Otolaryngology Cases

The University of Cincinnati Clinical Portfolio

Myles L. Pensak

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This book is dedicated to the caring support and professional staff who we, the physician members of the Department of Otolaryngology–Head and Neck Surgery, are fortunate to work beside in the daily care of our patients at UC Health, Cincinnati Children's Hospital, and the VA Medical Center.

Contents

Foreword

I am quite honored to have been asked by Dr. Myles Pensak to write a foreword for his text, *Otolaryngology Cases: The University of Cincinnati Clinical Portfolio*. We have been blessed by the University of Cincinnati for educating several of our finest clinical faculty, three of whom have endowed professorships here at the University of North Carolina at Chapel Hill. Dr. William Shockley did his residency at the University of Cincinnati, Dr. Mark Weissler did his fellowship in Head and Neck Oncology there, and Dr. Amelia Drake did her Pediatric Otolaryngology fellowship at Cincinnati also. All three are fabulous teachers, outstanding clinicians, and wonderful investigators.

Keeping that in mind, I reviewed the chapters in this book and have found the style of each presentation to be outstanding. In every area of otolaryngology, a clinical pathology is presented in a patient-based scenario. The material is very easy for the reader to follow, and the differential diagnoses and management options are very current. Any reader can understand the rationale for therapy and the different modalities that are used in planning for diagnosis and management. The bibliography at the end of each chapter allows the reader to probe further into the background information that would lead to management options for each clinical scenario.

This is the kind of organized approach to patient care that one finds in the minds of the best clinicians in medicine. Dr. Pensak and his colleagues at the University of Cincinnati have done an outstanding job of conveying their expertise to the reader in a way that allows any clinician to understand the nuances of state-of-the-art diagnosis and therapy. I feel that this book has an outstanding capacity to elevate the level of care in our field to that which would be available in one of the leading institutions in our country, the University of Cincinnati School of Medicine!

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Preface

For health care providers confronted with an ever-expanding universe of basic science research and clinical information, the challenge is to determine what resources will optimally provide a contemporary vantage detailing best-practice assessment and management for clinical situations. Moreover, recent discoveries in molecular, immunobiologic, and electrophysiologic sciences form the basis for understanding disease process, but complex and detailed studies frequently take significant time to translate from the contemporary literature to the conventional and traditional textbook. Despite changes in this electronic information age, textbooks remain the cornerstone and foundation for information transmission in our discipline, but rarely are they a convenient resource for the practitioner wishing to integrate diagnostic, assessment, and therapeutic strategies for specific disease entities.

Otolaryngology Cases: The University of Cincinnati Clinical Portfolio is a portfolio of case studies selectively chosen by the editors and faculty of the Department of Otolaryngology–Head and Neck Surgery of the University of Cincinnati Academic Health Center to represent a broad and diverse collection of common clinical entities encountered by the general otorhinolaryngologist. Furthermore, each case presentation is formatted to provide a robust differential diagnosis and a cost-effective assessment algorithm, as well as options for best-practice clinical management. A series of questions about each case optimizes the learning experience; answers to these questions are provided in the back of the book. Bibliographic references are provided for readers seeking additional information.

The text has been divided into areas of subspecialization, including facial plastic and reconstructive surgery, head and neck oncologic surgery, pediatric otolaryngology, general otolaryngology, and otology/neurotology. It is the hope of the authors that this text will provide both students and the experienced practitioner with a clearly articulated, well-organized, state-of-the-art studies profile to enhance and optimize the care given to patients.

Readers are encouraged to visit the deparment's web site, www.ent.uc.edu, for further patient and professional information, as well as related interdisciplinary links.

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Pediatric Otology

1

Congenital Hearing Loss

Evan J. Propst, Jay Paul Willging, and Robin T. Cotton

History

A 22-month-old girl was referred for hearing evaluation after failing a newborn hearing screening at birth and realizing a plateau in function with hearing aids and delayed speech. She was the product of a full-term pregnancy and normal delivery, complicated by influenza at 6 months' gestation. The patient's mother received promethazine hydrochloride (Phenergan), a histamine type 1 receptor antagonist, during pregnancy for nausea. Family history included a cousin with Usher syndrome. The patient failed the newborn hearing screening with distortion product otoacoustic emissions (DPOAEs) absent for the 2- to 6-kHz range (**Fig. 1.1**). Auditory brainstem response (ABR) testing via air and bone conduction was consistent with severe to profound bilateral sensorineural hearing loss (SNHL) (**Fig. 1.2**). Genetic testing was negative for GJB2 (Connexin 26) and Usher syndrome. She received sequential cochlear implants and is making great strides in her speech and listening.

Differential Diagnosis—Key Points

About 6 in 1000 children in the United States are born annually with mild to moderate hearing loss, and 1 in 1000 are born with severe to profound SNHL. Fifty percent of these are due to environmental causes such as maternal infection (toxoplasmosis, rubella, cytomegalovirus, rubella), prematurity, postnatal viral or bacterial infections, or ototoxic drugs. The other 50% are due to genetic causes. Most genetic hearing loss is nonsyndromic (70%). Most nonsyndromic hearing loss is autosomal recessive (80%), with autosomal dominant (17%), X-linked (3%), and mitochondrial (<1%) hearing losses making up the remainder of the group. The remainder of genetic hearing loss is syndromic (30%), meaning that these individuals have other features consistently related to their hearing loss.

Autosomal Recessive Nonsyndromic Hearing Loss

Connexins

Mutations in the gap junction β-2 gene (*GJB2*) encoding the connexin 26 protein (Cx26) on the long arm of chromosome 13 (13q11) account for 50% of autosomal recessive nonsyndromic hearing loss (ARNSHL). The most common *GJB2* mutation is 35delG. Cx26 is expressed in the stria vascularis, spiral ligament, spiral limbus, and supporting cells of the cochlea, forming intercellular channels called *gap junctions*. Gap junctions allow potassium to be recycled from

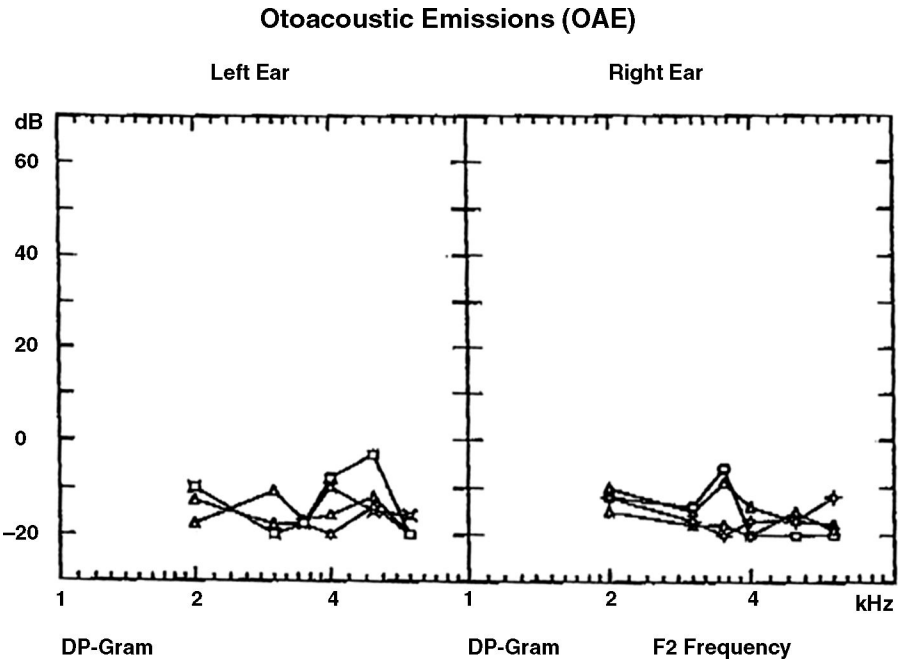


Fig. 1.1 Distortion product otoacoustic emissions were absent bilaterally for the 2- to 6-kHz range.

the endolymph to the stria vascularis to maintain the endolymphatic potential. Most *GJB2* mutations confer severe to profound hearing loss. A 342-kb deletion in the *GJB6* gene encoding the connexin 30 protein (Cx30) upstream from *GJB2* is responsible for 3% of heterozygotes (deaf individuals with a *GJB2* mutation on only one of two alleles) in the United States.

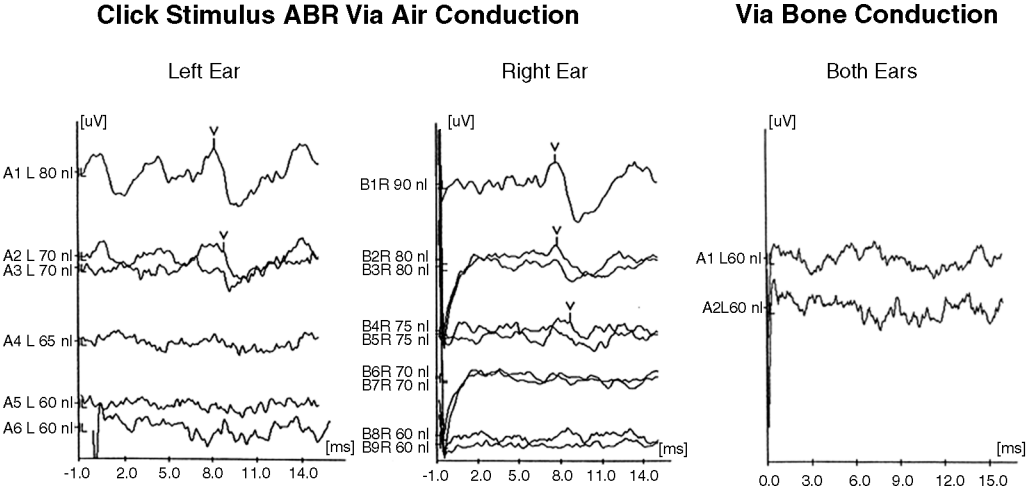


Fig. 1.2 Auditory brainstem response (ABR) testing via air and bone conduction was consistent with severe to profound bilateral sensorineural hearing loss.

Higher rates have been identified in Spanish populations (66%).

Otoferlin

The *OTOF* gene encodes otoferlin, a membrane-anchored, calcium-binding protein that plays a role in the exocytosis of synaptic vesicles at the auditory inner hair-cell ribbon synapse. It is associated with prelingual profound SNHL and inherited auditory neuropathy–dyssynchrony (AN-AD), which is characterized by the presence of otoacoustic emissions (OAEs) and a cochlear microphonic on ABR (one early cochlear wave with no waves from the auditory pathway). Treatment includes frequency modulation (FM) devices and hearing aids for mild to moderate hearing loss and cochlear implantation for severe SNHL.

Autosomal Dominant Nonsyndromic Hearing Loss

Connexins

Dominant mutations in the *GJB2* and *GJB6* genes have been reported but occur less frequently than recessive mutations.

Enlarged Vestibular Aqueduct Syndrome

Patients with enlarged vestibular aqueduct syndrome (EVAS) have large vestibular aqueducts, hearing loss, and incomplete cochlear partitioning (Mondini malformation) similar to that seen in Pendred syndrome, without thyroid abnormalities. The hearing loss ranges from mild to profound, the audiogram is flat or down-sloping, and the loss is sensorineural or mixed. SNHL can be progressive or sudden following head trauma. The *SLC26A4* mutation seen in Pendred syndrome is identified in 25% of patients with EVAS.

WFS1 Gene

Mutations in the *WFS1* gene confer SNHL affecting frequencies below 2000 Hz and worsening over time. Patients retain excellent understanding of speech, and many choose not to wear hearing aids.

COCH Gene

Mutations in the *COCH* gene are associated with high-frequency SNHL that presents in the second to fourth decade of life and progresses to profound SNHL by the fifth decade. Many experience recurrent bouts of vertigo similar to Ménière disease.

Mitochondrial

Mitochondrial proteins interact with chromosomal proteins to facilitate energy production. All mitochondrial inheritance comes from the maternal egg. Mitochondrial mutations are present in 1 to 2% of newborns, with the 12S rRNA *A1555G* mutation being the most commonly reported. This mutation is structurally similar to bacterial 16S rRNA, and aminoglycosides aimed at bacterial rRNA can erroneously attack mitochondrial rRNA, resulting in ototoxicity. Fifteen percent of people in the United States and most people in Mongolia with aminoglycoside ototoxicity have the *A1555G* mutation. Family members may have milder hearing loss without being exposed to ototoxic medications.

X-Linked

Stapes Fixation with Perilymphatic Gusher

Mutations in *POU3F4* can lead to profound SNHL, mixed hearing loss, an enlarged internal auditory canal with a thin or absent lamina cribrosa (bony plate between the auditory canal and the cochlea), and stapes fixation with perilymphatic gusher during stapes surgery. Young males with a family history of X-linked hearing loss are most commonly affected; however, females can have mild expression of this gene.

Autosomal Recessive Syndromic Hearing Loss

Usher Syndrome

The incidence of Usher syndrome is 1 in 25,000, affecting 2 to 4% of the deaf population and 50% of the deaf and blind population.

Table 1.1 Usher Syndrome

Type	Common Gene	% of Patients	Hearing	Vestibular Function	Visual	Clinical
1	*MYO7A*	40%	Congenital bilateral severe to profound SNHL	Abnormal	Early onset RP by age 10 yr	Amplification ineffective Manual communication Late motor milestones
2	*USH2A*	60%	Moderate SNHL (sloping audiogram)	Normal	RP onset in second decade	Hearing aids effective Oral communication
3	*USH3*A	3%	Progressive SNHL	Variable or progressive	Variable onset of RP	Variable

Abbreviations: RP, retinitis pigmentosa; SNHL, sensorineural heaing loss.

Individuals have SNHL, retinitis pigmentosa, and vestibular dysfunction. Three types of Usher syndrome are recognized (**Table 1.1**). Electroretinography can identify photoreceptor abnormalities in children 2 years of age, which is earlier than can be done using funduscopic examination. Rehabilitation with hearing aids, cochlear implantation, and educational and vocational planning is essential.

Pendred Syndrome

Pendred syndrome accounts for 1 to 2% of profound congenital deafness. Mutations in the *SLC26A4* (PDS) gene and pendrin protein affect iodine and chloride ion transport. The syndrome is characterized by severe to profound or progressive SNHL, cochlear abnormalities (Mondini malformation, large vestibular aqueduct, malformed vestibular canals), vestibular dysfunction, and euthyroid goiter in early puberty or adulthood. Genetic testing is used to make the diagnosis because the perchlorate discharge test that was historically used is neither sensitive nor specific. Thyroid function tests should be obtained and exogenous thyroid hormone used rather than thyroid surgery.

Jervell and Lange-Nielsen Syndrome

Jervell and Lange-Nielsen syndrome accounts for 0.1% of profound deafness. It is characterized by severe to profound SNHL, QT-interval prolongation, large T waves, and torsade de pointes on electrocardiography (ECG), with ventricular arrhythmia, syncope, seizure, or sudden death in the second or third decade of life. There may be a family history of syncope, seizure, sudden infant death syndrome (SIDS), or sudden death, and first-degree relatives may have cardiac irregularities without SNHL. Mutations in *KVLQT1* and *KCNE1* that regulate potassium channels in the ear and heart have been identified in some families. Patients and family members should receive an ECG and be referred to cardiology for beta-blockade.

Biotinidase Deficiency

Biotin is a water-soluble B vitamin essential for gluconeogenesis, fatty acid synthesis, and amino acid catabolism. About 1 in 60,000 children are born without biotinidase and cannot recycle biotin. Failure to add biotin to the diet will result in irreversible SNHL, optic atrophy, seizures, hypertonia, developmental delay, ataxia, skin rash, alopecia, and conjunctivitis. Consider this disorder when a child presents with SNHL and ataxia.

Refsum Disease

This rare disorder, caused by faulty phytanic acid metabolism, manifests as severe progressive SNHL and retinitis pigmentosa. Patients have a high serum concentration of phytanic acid and

can be treated with dietary modification and plasmapharesis.

Autosomal Dominant Syndromic Hearing Loss

Waardenburg Syndrome

Waardenburg syndrome accounts for 2% of profound deafness. Abnormalities in tissues of neural crest origin lead to SNHL (unilateral or bilateral), enlarged vestibular aqueduct, pigmentary changes such as a white forelock (varying age of first appearance), early graying of the hair, vitiligo, synophrys (eyebrows grow together), blue eyes or heterochromia iridis (different colored eyes), dystopia canthorum (lateral displacement of inner canthi), pinched nose, craniofacial abnormalities, neural tube defects, gastrointestinal dyskinesia, and limb defects. Four types are recognized, the most common being type 1 (**Table 1.2**).

CHARGE Syndrome

The CHARGE acronym stands for eye **c**oloboma, **h**eart defects, choanal **a**tresia, **r**etarded development, and **g**enitourinary and **e**ar anomalies. The incidence is 0.1 to 1.2 per 10,000, and more than 75% of patients have mutations in the *CHD7* gene. A definite clinical diagnosis includes all four major characteristics: (1) coloboma of the iris, retina, choroid, or disc; (2) choanal atresia (bilateral or left-sided); (3) cranial nerve (CN) dysfunction (CNI, CNVII, CNVIII, CNIX, CNX); (4) characteristic ear (short, wide, asymmetric protruding pinna with little lobe, ossicular and Mondini malformations, hypoplastic or absent semicircular canal). A probable diagnosis includes one major plus several minor characteristics, which include (1) developmental delay and hypotonia; (2) growth deficiency; (3) square face, prominent forehead and nasal bridge, and flat midface; (4) cleft lip, palate, or both; (5) cardiovascular malformations, (6) tracheoesophageal fistula; and (7) delayed puberty. Patients should be evaluated for respiratory distress, aspiration, swallowing, and hearing and also should be referred for neurologic, visual, cardiac, and urological workup.

Branchio-oto-renal Syndrome (Melnick-Fraser Syndrome)

The incidence of branchio-oto-renal (BOR) is 1 in 40,000. *EYA1* gene mutations on chromosome 8q13 are identified in 40% of individuals with the BOR phenotype, which is inherited with high penetrance and variable expressivity. The clinical diagnosis is based on three major criteria, two major and two minor criteria, or one major criterion and an affected first-degree relative (**Table 1.3**). The combination of a hypoplastic cochlea, facial nerve deviated to the medial side of the cochlea, funnel-shaped internal auditory canal, patulous eustachian tube, and kissing carotid arteries are pathognomonic for BOR.

Stickler Syndrome

The incidence of Stickler syndrome is 1 in 8000; however, only 40% have SNHL. Stickler syndrome is a connective tissue disorder characterized by ocular findings (myopia, cataract,

Table 1.2 Waardenburg Syndrome

Type	Name	Common Gene	Differentiating Clinical Feature(s)
1	Waardenburg	*PAX3*	Dystopia canthorum Hearing loss in 20%
2	Tietz syndrome	*MITF*	Dystopia canthorum absent Hearing loss in 50%
3	Klein-Waardenburg	*PAX3*	Features of WS1 plus upper limb defects
4	Shah-Waardenburg	*SOX10, EDNRB, EDN3*	Features of WS2 plus Hirschsprung disease

Table 1.3 Branchio-oto-renal Syndrome

Major	Minor
Hearing loss (conductive, sensorineural, mixed)	External ear
Prehelical pits	Middle ear
Branchial cleft cyst/fistula	Inner ear
Renal dysplasia/malformed calyces, aplasia, polycystic kidney	Tags
	Other: facial asymmetry, palate abnormalities

retinal detachment), hearing loss (high-frequency conductive loss or progressive SNHL), flat "scooped-out" midface, micrognathia, cleft palate, Robin sequence, spondyloepiphyseal dysplasia, short stature, precocious arthritis, marfanoid habitus, and mitral valve prolapse. Evaluation of hearing, vision, the airway, and heart is required. Contact sports should be avoided because of the risk of retinal detachment, and family members should be screened for visual or hearing impairment (**Table 1.4**).

Neurofibromatosis Type 2

Mutations in the tumor suppressor gene schwannomin on chromosome 22q11 lead to neurofibromatosis type 2 (NF-2). Diagnostic criteria include (1) bilateral acoustic neuromas; or (2) unilateral acoustic neuroma or two of neurofibroma, meningioma, glioma, schwannoma, juvenile lenticular opacity, and a first-degree relative with NF-2. Symptoms include unilateral hearing loss, tinnitus, imbalance, and vertigo in the second decade of life, with contralateral symptoms around 2 years later. Diagnosis requires magnetic resonance imaging (MRI) with gadolinium. Treatment includes observation, gamma knife, or surgery. Family members should receive genetic testing.

Treacher Collins Syndrome (Mandibulofacial Dysostosis)

Mutations in the *TCOF1* gene and treacle protein lead to bilateral ear anomalies (microtia, aural atresia, conductive hearing loss, SNHL, vestibular dysfunction), down-sloping palpebral fissures, coloboma of the outer lower eyelid, absent eyelashes, hypoplastic zygomatic arch, micrognathia, cleft palate, parotid hypoplasia, and normal intelligence. Evaluation of hearing and sleep apnea is required. In contrast, individuals with Goldenhar syndrome have unilateral facial anomalies and upper eyelid coloboma.

Apert-Crouzon Syndrome

The incidence of Apert syndrome is 1 in 65,000. *FGFR2* mutations that result from increased paternal age lead to coronal suture craniosynostosis (prominent forehead, flat occiput), large fontanelles, mental retardation, hearing loss (fixed stapes footplate, wide cochlear aqueduct, absent internal auditory canal), down-sloping palpebral fissures, proptosis, exposure keratitis, optic nerve anomalies, midface hypoplasia, down-sloping corners of the mouth, narrow "byzantine" palate with cleft, dental anomalies, fused cervical vertebrae, tracheal anomalies,

Table 1.4 Stickler Syndrome

Type	Common Gene	Differentiating Clinical Feature(s)
1	*COL2A1*	Classic Stickler syndrome, see description in text
2	*COL11A1*	More severe hearing loss and thick bundles throughout vitreous cavity of eye
3	*COL11A2*	No ocular abnormalities

cardiac changes, genitourinary and gastrointestinal anomalies, and syndactyly of the hands and feet (digits 2, 3, and 4 joined in a middigital mass). Features of Crouzon syndrome are similar but without syndactyly. Evaluation of hearing, the airway, vision, and heart is required.

Connexins

KID (keratoderma-ichthyosis-deafness) syndrome, Vohwinkel syndrome (mutilating keratoderma), and palmoplantar hyperkeratosis are due to *GJB2* mutations.

X-Linked

Alport Syndrome

The incidence of Alport syndrome is 1 in 10,000. *COL4A5* mutations lead to high-frequency, progressive SNHL in the second decade, retinal flecks, lens protrusion, spherophakia, congenital cataract, and progressive glomerulonephritis. Symptoms are worse in males. Audiologic evaluation and early management of renal disease are essential.

Mohr-Tranebjaerg Syndrome
(Deafness Dystonia Optic Atrophy)

Mutations in *TIMM8A* cause progressive SNHL, dystonia, myopia, visual-field reduction, cortical blindness, and mental deterioration.

Norrie Disease

Mutations in the *NDP* gene cause congenital rapidly progressive blindness, ocular degeneration, microphthalmia, progressive SNHL in the second decade, and progressive mental deterioration.

Otopalatodigital Syndrome

Mutations in the *FLNA* gene lead to otopalatodigital (OPD) type 1, mild form) and OPD type 2 (severe form, death in first year). Features include conductive, SNHL, or mixed hearing loss, cleft palate, oligohypodontia, short proximal thumbs, hypoplastic distal phalanges and great toe with large sandal gap, bowed limbs, joint restriction, prominent supraorbital ridges, down-slanting palpebral fissures, hypertelorism, broad nasal bridge and tip, and normal intelligence.

Wildervanck Syndrome

Wildervanck syndrome affects women and its features are a combination of Klippel-Feil anomaly (SNHL or mixed hearing loss, facial asymmetry, fused cervical vertebrae, torticollis, renal and cardiovascular anomalies) and Duane syndrome (lateral rectus innervated by CNIII).

Mitochondrial

MELAS

MELAS stands for mitochondrial encephalopathy, lactic acidosis, and stroke. Hearing loss is also present.

MIDD

MIDD is maternally inherited diabetes and deafness.

Kearns-Sayre Syndrome

Kearns-Sayre syndrome comprises SNHL, ataxia, short stature, delayed puberty, ophthalmoplegia, and retinopathy.

MERRF

MERRF (myoclonic epilepsy with ragged red fibers) includes SNHL, ataxia, epilepsy, and optic atrophy.

Chromosomal Hearing Loss

Down Syndrome

The incidence is 1 in 1500 for mothers under age 30 and 1 in 50 for mothers over age 40. The most common cause is trisomy 21. The clinical diagnosis includes 4 of 10 features: (1) flat face, (2) slanted palpebral fissures,

(3) anomalous auricles, (4) excess skin on the back of the neck, (5) hypotonia, (6) hyperflexible joints, (7) simian crease, (8) fifth finger midphalanx dysplasia, (9) pelvic dysplasia, (10) poor startle reflex. Atlantoaxial instability is common, and a shoulder roll should not be used. Examination of hearing, the airway, heart, and pelvis is required.

Turner Syndrome

Females with Turner syndrome (monosomic X chromosome) have hearing loss (conductive, SNHL, mixed), webbed neck, short stature, and gonadal dysgenesis.

◆ Test Interpretation

1. Historically only children at risk underwent hearing evaluation. Risk factors include family history of SNHL, in utero infection (toxoplasmosis, rubella, cytomegalovirus [CMV], herpes, syphilis), prematurity, syndromes, craniofacial anomalies, birth weight <1500 g, Apgar scores less than 3 at 5 minutes or less than 6 at 10 minutes, respiratory distress, mechanical ventilation longer than 10 days, hyperbilirubinemia requiring exchange transfusion, bacterial meningitis, ototoxic medication, head trauma, otitis media with effusion lasting 3 months, temporal bone fracture, neurodegenerative or demyelinating disease, or parental concern. Less than 50% of infants with hearing loss were detected, leading to implementation of the infant hearing screening program. Newborns at risk who pass the screen should be monitored for changes in hearing every 6 months for 3 years.
2. Universal newborn screening has decreased the age of detection from 2.5 years to less than 4 months, resulting in decreased rates of speech and language delay. Automated OAEs (AOAEs) measure the outer cochlear hair cell response to acoustic stimuli presented through a probe in the external auditory canal. Transient evoked (TEOAEs) and distortion product (DPOAEs) assess frequency-specific function from 500 to 6000 Hz as long as there is no contamination by ambient noise, vernix, or middle ear pathology. Infants at risk for SNHL should undergo automated ABR (AABR) by presenting a click stimulus at 35 dB hearing loss to the auditory canal and recording from forehead surface electrodes. Screening AABR cannot differentiate the type or degree of hearing loss. This process has a sensitivity of 100%, specificity of 99.7%, referral rate of 2%, and positive predictive value of 83.3% (Hall, 2004). Infants who do not pass should have both ears retested within 1 month using AOAE and AABR. Failure to pass retesting requires diagnostic OAE and ABR by an audiologist to determine hearing thresholds. Click-evoked ABR tests the 1000 to 4000 Hz range, and tone burst ABR and auditory steady-state response (ASSR) testing provide frequency-specific information. The test battery should include air- and bone-conduction ABR, high-frequency tympanometry, OAE, stapedial reflexes, tone-burst ABR or ASSR or both, and behavioral audiometry. Reversing polarity during air-conduction ABR ensures that the response is neural and not the cochlear microphonic. Amplification should be provided within 1 month, visual acuity assessed, and families offered genetic counseling.
3. Children older than 5 months should undergo audiometric testing to establish the degree and nature of hearing loss for each ear.

   Behavioral observational audiometry (BOA)

   BOA relies on a reflexive change of state in response to a sound. Imprecision, rapid habituation, and examiner bias make it useful only for children age 5 months or younger. The diagnosis of hearing loss should never rely on BOA alone.

   Visual reinforcement audiometry (VRA)

   Children older than 5 months can be tested by reinforcement with a visual stimulus after turning toward an auditory stimulus. VRA can be used in a sound-field environment or with insert headphones and can suggest ear differences at various frequencies.

   Conditioned play audiometry (CPA)

   Children 2 years of age can be taught to perform an activity (i.e., drop a

block in a bucket) in response to an auditory stimulus. Standard techniques can be used, and normal adult threshold levels are expected.

Standard adult testing

Children 5 years of age can undergo adult testing. Concerning results should be followed by OAE, ABR, and/or ASSR testing.

4. Syndromic children with SNHL should be tested for known associated features. A sequential diagnostic paradigm has been proposed for the workup of idiopathic ARNSHL for a more efficient and cost-effective approach (Preciado et al, 2005, **Fig. 1.3**). This does not obviate the need for specific testing when dictated by clinical examination or imaging when planning surgical intervention. Syphilis testing (Venereal Disease Research Laboratory and fluorescein treponema antibody test) should be performed when results are not available.

◆ Diagnosis

Bilateral SNHL

◆ Medical Management

Hearing aids should be fitted as soon as the hearing loss is identified. Genetic counseling regarding the cause of the hearing loss, additional deficits, vocational planning, and the implications for siblings or future offspring should be offered. Appropriate referrals should be made.

◆ Surgical Management

Indications for bone-anchored hearing aids include the following: (1) conductive or mixed hearing loss where conventional hearing aids cannot be used (e.g., anomaly of external or

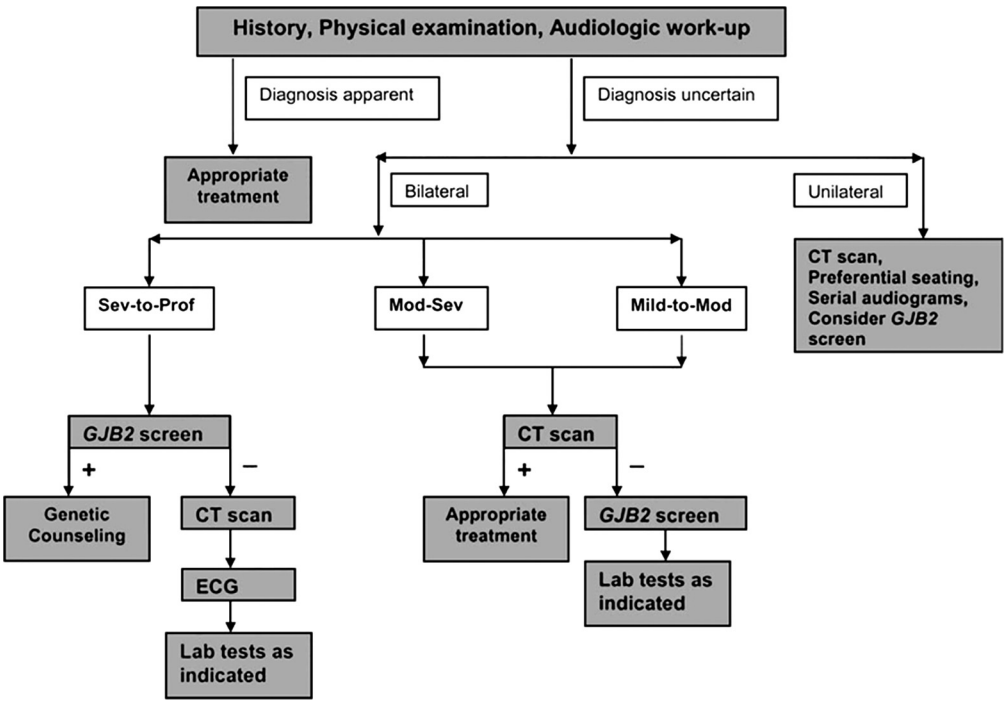


Fig. 1.3 Sequential diagnostic paradigm for the workup of idiopathic autosomal recessive nonsyndromic hearing loss. CT, computed tomography; ECG, electrocardiography; Mod, moderate; Sev, severe. (From Preciado DA, Lawson L, Madden C, et al. Improved diagnostic effectiveness with a sequential diagnostic paradigm in idiopathic pediatric sensorineural hearing loss. Otol Neurotol 2005;26(4):610–661.)

middle ear, chronic infection, unmanageable feedback), (2) unilateral conductive or mixed hearing loss, (3) unilateral severe to profound SNHL. Indications for cochlear implantation include (1) bilateral severe to profound SNHL, (2) aided open-set sentence recognition (e.g., HINT) score of 40% for Medicare, 60% for private payers, (3) when the patient does not benefit from amplification and aural rehabilitation, (4) implantable cochlea with nerve present, (5) motivated patient and family.

◆ Rehabilitation and Follow-up

Patients should have an audiologic reevaluation every 3 months during the first year and every 6 months thereafter. Hearing aids should be calibrated on a regular basis and new molds fitted when required. Periodic audiometric reevaluation is necessary to rule out fluctuation or progression of hearing loss. Speech and language therapy is imperative for the promotion of language and communication. Patients should see an otologist at least on an annual basis to reassess the accuracy of the initial diagnosis, inspect for diseases of the external and middle ear, evaluate for progression of hearing loss and suitability of the patient's educational program, and as needed if changes are noted in hearing. Avoidance of ototoxic medications and loud noise exposure by using hearing protection is essential. Children with cochlear implants require extensive rehabilitation involving specially trained audiologists, speech and language therapists, and auditory verbal therapists on a long-term basis to achieve maximal results. Parents must be made aware of this before implantation. With proper amplification, speech and language therapy, and educational programs, patients with congenital hearing loss can usually fully participate in social activities, school, and work.

◆ Questions

1. Most nonsyndromic hearing loss is
- **A.** X-linked recessive
- **B.** X-linked dominant
- **C.** Autosomal recessive
- **D.** Autosomal dominant
- **E.** Mitochondrial

2. Most autosomal recessive nonsyndromic hearing loss is due to
- **A.** Mutations in the *GJB6* gene
- **B.** Mutations in the *EYA1* gene
- **C.** Mutations that affect gap junctions in the stria vascularis
- **D.** Mutations in the *WFS1* gene
- **E.** Mitochondrial non-nuclear DNA

3. Universal newborn hearing screening is based on the premise that
- **A.** The auditory brainstem response measures action potentials from outer cochlear hair cells
- **B.** Visual reinforcement audiometry is best used in children younger than 2 months
- **C.** Outer hair cells in the cochlea emit signals spontaneously and following stimulation
- **D.** Syndromic hearing loss is often difficult to diagnose in early childhood
- **E.** Early intervention with a cochlear implant can help children with sensorineural hearing loss and a pure-tone average of 50 dB or worse.

Suggested Readings

Ballana E, Ventayol M, Rabionet R, Gasparini P, Estivill X. Connexins and deafness homepage. www.crg.es/deafness. February 9, 2009

Demichele AM, Ruth RA. Newborn Hearing Screening. eMedicine. February 1, 2008

Hall JW III, Smith SD, Popelka GR. Newborn hearing screening with combined otoacoustic emissions and auditory brainstem responses. J Am Acad Audiol 2004;15(6):414–425

Isaacson B, Lee KH, Kutz JW, et al. Cochlear Implants, Indications. eMedicine. July 8, 2008

Johnson R, Greinwald J. Genetic hearing loss. In: Bailey BJ, Johnson JT, eds. Head & Neck Surgery—Otolaryngology. Lippincott Williams & Wilkins, 2006: Chapter 94

Preciado DA, Lawson L, Madden C, et al. Improved diagnostic effectiveness with a sequential diagnostic paradigm in idiopathic pediatric sensorineural hearing loss. Otol Neurotol 2005;26(4):610–615

Propst EJ, Blaser S, Gordon KA, Harrison RV, Papsin BC. Temporal bone findings on computed tomography imaging in branchio-oto-renal syndrome. Laryngoscope 2005;115(10):1855–1862

Smith RJ, Van Camp G. Deafness and hereditary hearing loss overview. GeneReviews. www.geneclinics.org/profiles/. January 2007

2

Otitis Media

J. Matthew Dickson and Robin T. Cotton

History

A 19-month-old boy arrived in the emergency department with a 3-day history of left ear pain and a 1-day history of left facial paralysis. He had been seen by his primary care physician the day before and started on amoxicillin (40 mg/kg/day) for acute otitis media. His facial function was normal at that time. The patient had a history of recurrent otitis media requiring four to five courses of antibiotics per year since birth. Physical examination revealed an alert boy with a temperature of 39° C. Auricular examination was unremarkable. The left external auditory canal was occluded with granulation tissue. Cranial nerve examination demonstrated complete left-sided facial paralysis classified as a House-Brackmann VI paralysis (**Table 2.1**). The remainder of the head and neck examination revealed no abnormality. A computed tomography (CT) scan was then ordered.

Differential Diagnosis—Key Points

1. Complications of otitis media can be classified according to extracranial and intracranial involvement (**Table 2.2**). The differential diagnosis of extracranial complications of acute otitis media can be divided into two groups: intratemporal and extratemporal. Intracranial complications include extradural granulation tissue, sigmoid sinus thrombosis, brain or subdural abscess, otitic hydrocephalus, and meningitis. An accurate history, complete otolaryngologic and neurologic examination, and radiographic imaging are needed to differentiate the various complications.
2. All cases of acute otitis media involve inflammation of the mastoid air cells. However, clinically significant acute mastoiditis is a clinical diagnosis based on the findings of suppurative otitis media, postauricular swelling with loss of postauricular crease, and protrusion of the auricle. Coalescent mastoiditis is a specific radiographic diagnosis based on CT and is differentiated from acute mastoiditis by radiographic evidence of loss of the bony septations.
3. Suppurative labyrinthitis occurs when bacterial invasion penetrates the otic capsule, usually via the round window or oval window. The classic presentation is rapid onset of vertigo, sensorineural hearing loss, nausea, and vomiting during an episode of acute otitis media. In the absence of associated meningitis, the cerebrospinal fluid pressure and analysis are normal. Suppurative acute petrositis occurs when there is extension of the middle-ear infection into the petrous

Table 2.1 House-Brackman Facial Nerve Grading System

Grade	Gross	At Rest	Motion
I. Normal	Normal	Normal tone and symmetry	Normal
II. Mild dysfunction	Slight weakness on close inspection, slight synkinesis	Normal tone and symmetry	Forehead: good to moderate movement Eye: complete closure with minimum effort Mouth: slight asymmetry
III. Moderate dysfunction	Obvious but not disfiguring facial asymmetry. Synkinesis is noticeable but not severe. May have hemifacial spasm or contracture	Normal tone and symmetry	Forehead: slight to moderate movement Eye: complete closure with effort Mouth: slight weakness with maximum effort
IV. Moderately severe dysfunction	Asymmetry is disfiguring or obvious facial weakness	Normal tone and symmetry	Forehead: no movement Eye: incomplete eye closure Mouth: asymmetric with maximum effort
V. Severe dysfunction	Only slight, barely noticeable, movement	Asymmetry	Forehead: no movement Eye: incomplete closure Mouth: slight movement
VI. Total paralysis	No movement	No movement	No movement

Source: House JW, Brackman DE. Facial nerve grading system. *Otolaryngol Head Neck Surg.* 1985;93:146–147.

Table 2.2 Complications of Acute Otitis Media

I. Extracranial
- A. Intratemporal
  1. Acute mastoiditis
  2. Coalescent mastoiditis
  3. Labyrinthitis
  4. Petrositis
  5. Facial nerve dysfunction
  6. Tympanic membrane perforation
- B. Extratemporal
  1. Subperiosteal abscess
  2. Bezold abscess

II. Intracranial
- A. Extradural granulation tissue and/or abscess
- B. Sigmoid sinus thrombosis
- C. Brain abscess
- D. Otic hydrocephalus
- E. Meningitis
- F. Subdural abscess

apex, resulting in symptoms of retro-orbital pain, persistent otorrhea, and sixth cranial nerve palsy. This symptom complex is known as the Gradenigo triad. Facial paralysis, usually unilateral, can occur during an episode of acute otitis media either secondary to direct inflammation through a bony dehiscence in the tympanic segment of the facial nerve or secondary to osteitis involving the bony fallopian canal.

4. Extratemporal complications occur when infection progresses to involve the cortical bone surrounding the mastoid air cells. Osteitis of the lateral cortex can result in the development of a subperiosteal abscess. The patient usually presents with more pronounced auricular protrusion, loss of the postauricular crease, and fluctuance over the mastoid. Osteitis of the medial or inferior mastoid cortex can result in the development of a deep neck space infection known as Bezold abscess.
5. Acute mastoiditis can be accompanied by a significant amount of inflammation of the cartilaginous external auditory canal or

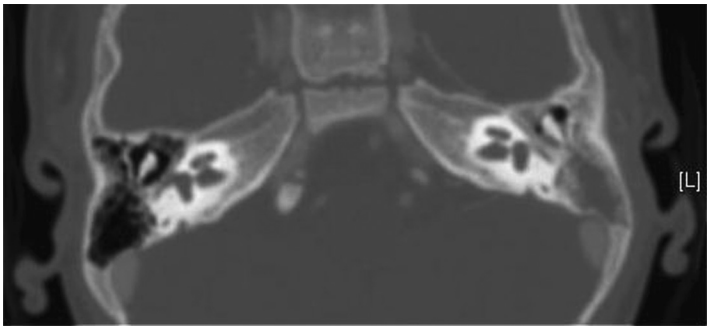


Fig. 2.1 Axial computed tomography scan of temporal bones: left mastoiditis.

granulation tissue, making visualization of the tympanic membrane difficult or impossible in the awake child. This can clinically mimic acute external otitis without middle-ear or mastoid involvement. The physical examination can help differentiate the two entities. Manipulation of the external auditory canal by pulling on the tragus is extremely painful in acute external otitis, but not in mastoiditis. Occasionally, initial response to intravenous (IV) and ototopical treatments is needed to make an accurate diagnosis.

6. Intracranial complications of acute otitis media manifest with a broad range of signs and symptoms. Presentation can range from headache and lethargy to seizures and focal neurologic signs. Sigmoid sinus thrombosis classically presents with "picket-fence" fevers, toxemia, septic embolization, and torticollis. However, further complications can occur with thrombus propagation, including jugular foramen syndrome, otic hydrocephalus, coma, and even death.
7. Brain abscesses can be difficult to diagnose because the signs and symptoms are often subtle. Presentation depends on the stage of the abscess. The first stage, cerebritis, is accompanied by generalized symptoms of headache, malaise, fever, and drowsiness, followed by a quiescent or latent phase that can last for weeks. In the third phase, abscess formation occurs with focal neurologic signs. The final phase, termination, results in rupture of the abscess, leading to rapid deterioration and death.
8. Meningitis presents with classic findings of headache, fever, and neck rigidity. Kernig and Brudzinski signs are positive. Spread of infection to the meninges is by hematogenous dissemination, inner ear malformations (incomplete partitioning), or direct spread (middle fossa dehiscence, meningoencephalocele).

◆ Test Interpretation

The CT scan revealed opacification of the left middle ear and mastoid with normal mastoid trabeculae. There was no bony dehiscence of the tympanic segment of the facial nerve course. The lateral mastoid cortex was intact (**Fig. 2.1**). CT is the appropriate modality to assess the mastoid and temporal bone. If intracranial involvement is suspected, contrast-enhanced magnetic resonance imaging is a superior investigation. It is more sensitive for the detection of an early brain abscess, sigmoid sinus thrombosis, subdural abscess, and extradural granulation tissue.

◆ Diagnosis

Acute otitis media with facial nerve paralysis

◆ Medical Management

Empirical IV antimicrobial coverage should include coverage for the most common organisms that cause acute otitis media, including *Streptococcus pneumoniae, Haemophilus influenzae,* and *Moraxella catarrhalis*. Once culture results are obtained, coverage can be narrowed. Following surgical drainage, symptoms usually dissipate quickly, and oral antibiotic therapy can be instituted. Ciprofloxacin-dexamethasone otic drops should be instilled after placement of tympanostomy tubes. This allows ototopical

treatment of the infection and inflammation. Oral steroid therapy should also be started to address inflammation associated with the facial nerve paralysis.

Surgical Management

On the day of presentation, this child underwent bilateral myringotomy and pressure-equalizing tube placement. He was started on IV cefuroxime at the time of surgery in addition to ciprofloxacin-dexamethasone otic drops. Cultures taken at the time of surgery grew penicillin-sensitive *S. pneumoniae*. By postoperative day 3, facial function had improved, but a significant weakness was still present (House-Brackmann IV). The ear was no longer draining, and the middle ear was dry. The patient was sent home on amoxicillin, otic drops, and a tapering course of oral steroids on postoperative day 4.

Most surgeons believe that a facial nerve paralysis complicating acute otitis media requires urgent surgical drainage of the middle-ear space. The middle ear can be drained using a wide-field myringotomy or pressure-equalizing tube. Whether to address the mastoid is more controversial. When a coalescent mastoiditis is present in a child with extracranial complications of acute otitis media, a cortical mastoidectomy is advised. When mastoid septae are intact, tympanostomy tube placement, medical treatment, and close observation constitute a reasonable initial approach. Any deterioration of the child warrants a mastoidectomy.

Rehabilitation and Follow-up

The patient was seen in clinic 1 week later, and facial function had fully recovered. At his last follow-up appointment 6 months after surgery, this child's pressure-equalizing tubes had extruded, the tympanic membranes were intact, and hearing was normal. He has had one episode of otitis media since this complication.

Questions

1. Facial nerve dysfunction characterized by an obvious, but not disfiguring, difference between the two sides, complete eye closure with effort, and normal tone at rest is classified as a House-Brackman:
   - **A.** Grade II
   - **B.** Grade III
   - **C.** Grade IV
   - **D.** Grade V
2. The Gradenigo triad, the symptom complex associated with petrositis, is characterized by all of the following except:
   - **A.** Cranial nerve VII palsy
   - **B.** Retro-orbital pain
   - **C.** Persistent otorrhea
   - **D.** Cranial nerve VI palsy
3. Which of the following is the most common organism causing acute otitis media?
   - **A.** *Haemophilus influenzae*
   - **B.** *Moraxella catarrhalis*
   - **C.** *Streptococcus pneumoniae*
   - **D.** *Staphylococcus aureus*

Suggested Readings

Bluestone CD, Klein JO. Intratemporal complications and sequelae of otitis media. In: Bluestone CD, Stool SE, Alper CU, et al, eds. Pediatric Otolaryngology, 4th ed. Philadelphia, PA: WB Saunders; 2003:687–763

Gliklich RE, Eavey RD, Iannuzzi RA, Camacho AE. A contemporary analysis of acute mastoiditis. Arch Otolaryngol Head Neck Surg 1996;122(2):135–139

Lim JW, Mukesh P, Selesnick SH. Complications of otitis media. In: Hughes GB, Pensak ML, eds. Clinical Otology. 3rd ed. New York, NY: Thieme; 2007:250–257

Nadol JB Jr, Eavey RD. Acute and chronic mastoiditis: clinical presentation, diagnosis, and management. Curr Clin Top Infect Dis 1995;15:204–229

3

Congenital Facial Paralysis

Christopher T. Wootten and Robin T. Cotton

History

A 1-month-old infant was brought to the outpatient otolaryngology clinic on referral from an outside institution. He had a history of stridor at birth requiring tracheotomy, micrognathia, cleft palate, and a suspicion of facial paralysis. The child was born vaginally without forceps at full term, weighing 2890 g. After birth the child demonstrated stridor and retractions requiring intubation. A tracheotomy was performed for bilateral vocal-fold paralysis. The maternal history was not significant for any illness during this pregnancy, and labor lasted around 4 hours. There was no family history of congenital facial paralysis. No facial asymmetry was noted at rest, but facial expression was masklike despite agitation. The patient did not follow a penlight with his eyes. Otherwise, the physical examination did not reveal any evidence of craniofacial, otologic, neurologic, or systemic abnormalities.

Differential Diagnosis—Key Points

1. Congenital facial paralysis can be broadly divided into developmental and traumatic causes.
2. The differential may be reduced to a few salient diagnoses by employing a careful history and physical examination, a neurotologic evaluation, and electrophysiologic testing. Imaging plays a secondary role, but high-resolution computed tomography (CT) scanning will emphasize temporal bone anomalies or injury, and magnetic resonance imaging (MRI) of the brain may define concomitant central developmental anomalies.
3. In some series, traumatic facial nerve injuries accounted for most cases of congenital facial paralysis. For most infants, the mechanism is either positioning of the infant's head against the mother's sacrum during prolonged labor or through the use of forceps extraction. By either mechanism, the thin fallopian canal may become indented, compressing the nerve. Risk factors include primiparity, birth weight greater than 3500 g, and forceps extraction.
4. Developmental facial paralysis is usually accompanied by other malformations of the first and second branchial arches. Cleft palate, hypoplastic maxilla, duplication of the palate, Pierre-Robin sequence, pinna anomalies, microtia, and atresia are all commonly reported with congenital facial paralysis. In addition, sensorineural hearing loss that accompanies congenital facial paralysis, isolated lower-lip or upper-facial palsy, or other co-occurring cranial neuropathies suggest a developmental cause. Some of these anatomic and neurologic

findings that accompany congenital facial paralysis are grouped into Möbius syndrome, congenital unilateral lower-lip palsy (or asymmetric crying facies), and the oculoauriculovertebral syndromes (including Goldenhar syndrome).

Test Interpretation

In addition to masklike facies and abducens diplegia, the physical examination demonstrates Pierre-Robin sequence, drooling, and a prominent upper lip. The infant does not tear with crying, and the surface of the tongue is somewhat smooth. The right hand is smaller than the left. Middle-ear effusions are present bilaterally. A sound-field audiogram demonstrates moderate rising to mild hearing loss. Electromyography (EMG) showed no fibrillations or voluntary facial activity. A noncontrasted MRI of the brain demonstrated concave contour of the upper dorsal medulla, most likely related to an absence of the facial colliculus.

Diagnosis

Möbius syndrome: Although the cause of Möbius syndrome remains elusive, agenesis of cranial nuclei, agenesis of target muscles, vascular compromise, and retrograde atrophy of facial and abducens nuclei have all been postulated. Möbius syndrome has been associated with first-trimester insults; misoprostol and thalidomide ingestion; and abnormalities of chromosomes 3, 8, 10, and 13 in certain familial cases (which account for ~2% of all cases of Möbius syndrome).

Medical Management

Möbius syndrome is nonprogressive, and occasionally patients are able to compensate partially for some of their cranial neuropathies. The medical management of Möbius syndrome is guided by the severity of the patient's symptoms, which in turn are based on the nerves affected. Eighty percent of patients have bilateral facial palsy; 75% have abducens paralysis, and about 25% have hypoglossal involvement. Usually VIII is spared, but involvement of IX, X, and XI is not uncommon. In addition, there is a high occurrence of musculoskeletal anomalies in patients with Möbius syndrome (up to a third). About 15% of patients with Möbius syndrome have absence of certain thoracic muscles that are developing in the early first trimester (Poland anomaly).

Electrophysiologic testing allows the clinician to offer prognosis in congenital facial paralysis. A useful battery of tests includes maximal stimulation testing, evoked electromyography-electroneuronography (EEMG/ENoG), and EMG. When the diagnosis is unclear based on history and physical examination, electrophysiologic testing should be performed as soon as possible after birth to select the rare infant requiring surgical intervention and to predict the timing of recovery. Patients with complete facial nerve paralysis (House-Brackmann 6/6) are divided into four groups based on electrophysiologic testing:

- Group 1—Patients seen within 3 days of birth who can be stimulated by EEMG. Testing should be repeated every 3 to 5 days until clinical return of function.
- Group 2—Patients seen within 3 days who can initially be stimulated by EEMG but progress to a nonstimulatable state on repeat testing
- Group 3—Patients seen within 3 days of birth but not stimulatable by EEMG
- Group 4—Patients not seen until more than 3 days after birth (when degeneration distal to a crush or transection prevents stimulation). EEMG in these patients may yield confusing results.

Group 2 and 3 patients should undergo follow-up EMG after 2 to 3 weeks. The presence of polyphasic potentials or voluntary action potentials predicts recovery, whereas fibrillation potentials indicate denervation. Electrical silence indicates the absence of facial musculature. EMG in Möbius syndrome may demonstrate poor recruitment of polyphasic action potentials. However, a patient with bilateral congenital facial and abducens palsies accompanied by limb anomalies will gain little by extensive electrophysiologic testing.

The early morbidity of facial diplegia centers on the inability to protect the eyes through decreased tearing and incomplete closure. Although some patients produce tears, the Bell phenomenon is absent in these patients, and

the clinician must be aware of the risk of exposure keratitis. Artificial tears and ophthalmic ointments are useful. A formal consultation with a pediatric ophthalmologist allows for a corneal evaluation and the assessment and treatment of ophthalmoplegia (some patients with bilateral abducens palsy develop marked internal strabismus). Similarly, incomplete oral closure facilitates drooling in the neonate and oral incompetence with food lodging in the buccal vestibule in infants and children. Referral to a speech therapist for oral incompetence is helpful to optimize feeding and articulation.

Lower cranial neuropathies are associated with dysphagia, dysarthria, vocal-cord paralysis, or hypomobility and aspiration. A flexible endoscopic examination of swallowing is useful to note pooling of secretions, aspiration, and vocal-fold mobility issues. A fluoroscopic swallowing examination is complementary. Patients with confirmed dysphagia and aspiration are at risk for recurrent pulmonary infections. Symptomatic vocal-fold paralysis may require formal microlaryngoscopy and bronchoscopy to exclude concomitant airway lesions.

◆ Surgical Management

The surgical management of congenital facial paralysis relates to its cause and prognosis. Complete facial paralysis from birth trauma carries a spontaneous recovery rate close to 90%. Conversely facial paralysis associated with Möbius syndrome does not recover. The degree of disability observed in infancy may evolve as the patient ages and the infantile fat pads diminish. Although early gold weight insertion may benefit patients with insufficient lid closure, attempts at facial reanimation are usually delayed until the elementary school age or older (when peer pressure affects the child's perceived disability).

Because of facial motor nerve atresia, the facial reanimation ladder is limited in Möbius syndrome. Direct facial nerve anastomosis and interposition grafting have no role in the management of facial paralysis in Möbius syndrome. Likewise, nerve transposition, which relies on connecting a functional motor nerve (e.g., contralateral VII or ipsilateral XII) to a nonfunctioning VII distal to a lesion, will not reanimate a patient with Möbius syndrome. Instead, pedicled temporalis or masseter transposition (cranial nerve V is spared in Möbius syndrome) or microneurovascular free muscle transfer (utilizing cranial nerve V or an unaffected cranial nerve XI) are the typical means of reanimation in this population. Static fascial slings are a less desirable option.

Other deficits commonly associated with Möbius syndrome are managed using a multidisciplinary approach. Craniofacial anomalies are corrected surgically by a craniofacial team composed of otolaryngology, plastic surgery, ophthalmology, and oral surgery. Limb anomalies are managed by orthopedics.

◆ Rehabilitation and Follow-up

The prognosis for recovery in congenital facial paralysis secondary to a developmental anomaly such as Möbius syndrome is poor. Alongside ongoing speech and feeding therapy and ophthalmologic management, there is a nearly uniform need for reanimation-suspension procedures later in life. Because most patients with Möbius syndrome achieve normal intelligence and because autism occurs more commonly in patients with Möbius syndrome, neuropsychological follow-up is necessary as well.

◆ Questions

1. Most cases of traumatic congenital facial paralysis are thought to be caused by:
- **A.** Fracture of the petrous temporal bone
- **B.** Facial lacerations severing terminal branches of VII
- **C.** Indentation of VII in the fallopian canal as it exits the stylomastoid foramen
- **D.** Fracture of the styloid process

2. From most common to least common, the cranial nerves affected in Möbius syndrome are:
- **A.** VI, XII, V
- **B.** VII, XII, VI
- **C.** VI,VII, VIII
- **D.** VII, VI, XII

3. Which of the following facial reanimation techniques is commonly employed in Möbius syndrome?
A. XII-VII anastomosis
B. Microneurovascular free muscle transfer
C. VII-VII cross-facial graft
D. Direct anastomosis of severed portion of VII

Suggested Readings

Carr MM, Ross DA, Zuker RM. Cranial nerve defects in congenital facial palsy. J Otolaryngol 1997;26(2):80–87

Cattaneo L, Chierici E, Bianchi B, Sesenna E, Pavesi G. The localization of facial motor impairment in sporadic Möbius syndrome. Neurology 2006;66(12):1907–1912

Lifchez SD, Matloub HS, Gosain AK. Cortical adaptation to restoration of smiling after free muscle transfer innervated by the nerve to the masseter. Plast Reconstr Surg 2005;115(6):1472–1479

May M. Facial paralysis at birth: medicolegal and clinical implications. Am J Otol 1995;16(6):711–712

Palmer CA. Möbius syndrome. http://emedicine.medscape.com/article/1180822. April 30, 2007.

Pedraza S, Gámez J, Rovira A, et al. MRI findings in Möbius syndrome: correlation with clinical features. Neurology 2000;55(7):1058–1060

Shapiro AM, Shaitkin BM, May M. Facial paralysis in children. In: Bluestone CD, Stool SE, Alper CM, eds. Pediatric Otolaryngology, 4th ed. Philadelphia, PA: WB Saunders; 2003:369-387

Sjögreen L, Andersson-Norinder J, Jacobsson C. Development of speech, feeding, eating, and facial expression in Möbius sequence. Int J Pediatr Otorhinolaryngol 2001;60(3):197–204

Strömland K, Sjögreen L, Miller M, et al. Mobius sequence: a Swedish multidiscipline study. Eur J Paediatr Neurol 2002;6(1):35–45

Verzijl HT, van Es N, Berger HJ, Padberg GW, van Spaendonck KP. Cognitive evaluation in adult patients with Möbius syndrome. J Neurol 2005;252(2):202–207

Verzijl HT, Padberg GW, Zwarts MJ. The spectrum of Möbius syndrome: an electrophysiological study. Brain 2005;128(Pt 7):1728–1736

Zuker RM, Goldberg CS, Manktelow RT. Facial animation in children with Möbius syndrome after segmental gracilis muscle transplant. Plast Reconstr Surg 2000;106(1):1–8

4

External Auditory Canal Atresia

Jay Paul Willging and David L. Walner

History

A 5-year-old girl was referred from her pediatrician for evaluation of a right-ear deformity. The pinna was abnormally small, and the tympanic membrane could not be visualized. She had failed a hearing test in the right ear. She was the product of a normal gestation and delivery and had no history of trauma. An audiogram had demonstrated a maximal conductive hearing loss on the right, with normal hearing on the left.

The physical examination revealed a small but grossly normally shaped pinna. No external auditory canal was present. The left pinna, external auditory canal, and tympanic membrane appeared normal. Balance function appeared normal. The child had no problem with physical activity or ambulation, and she was able to ride a bike without difficulty. Facial nerve function was normal and symmetric bilaterally. There was no facial asymmetry. There was no mandibular cant, suggesting equal vertical ramus height. No other head and neck abnormalities were identified.

Differential Diagnosis—Key Points

1. A thorough history and physical examination are essential when evaluating children with external ear deformities. The history should include any maternal drug use or toxic exposure during pregnancy to identify any possible teratogens. A family history of ear deformities, craniofacial anomalies, or hearing loss should be elicited. The physical examination should focus on craniofacial development. Syndromes associated with ear canal and pinna abnormalities should be explored (Treacher Collins, Goldenhar, Crouzon, Alpert, Pfeiffer, Klippel-Feil, branchio-otorenal, 18q− chromosome, hemifacial microsomia). It is estimated that 45% of patients with aural atresia or stenosis can have an underlying syndrome. The functioning of the facial nerve must be documented.
2. The frequency of moderate to severe forms of congenital aural atresia has been estimated at 1 in 8000 live births in the United States. Unilateral atresia is six times more common than bilateral forms of the disorder. Unilateral atresia more commonly affects the right side. A positive family history of external auditory canal atresia may be elicited in 14% of cases.
3. A multidisciplinary team should be involved with the evaluation and management of children with external auditory canal stenosis, in addition to the otolaryngologist. Associated abnormalities may lead a geneticist to the diagnosis of a syndrome. Many children have associated midface and mandibular abnormalities to

warrant additional reconstructions by plastic or craniofacial surgeons. Dental and orthodontic problems are also commonly associated with this disorder. Audiologists and speech pathologists are essential for the rehabilitative processes that may be required.

4. The literature includes various classification systems for microtia. One system reflects the ease with which the ear may be reconstructed. The more severe the grade, the more extensive the reconstruction required and the less successful the cosmetic outcome.
   Grade I: A slightly small ear with identifiable structures with a small but present external ear canal
   Grade II: A partial external ear with a stenotic external ear canal producing a conductive hearing loss
   Grade III: Absence of the external ear with a small vestigial structure composed of rudimentary cartilage and fibrofatty tissue and an absent external ear canal and eardrum, creating a maximal conductive hearing loss
   Grade IV: Total absence of the pinna
5. In 1992, Jahrsdoerfer et al developed a 10-point grading system to determine surgical candidacy based on key features from the computed tomography (CT) scan and appearance of the external ear. A score of 8 or above in experienced hands is associated with postoperative speech-reception thresholds of less than 30 dB HL in 80% of cases.
   The following is a summary of this 10-point grading system:

   - Stapes present = 2 points
   - Oval window open = 1 point
   - Middle ear space well developed = 1 point
   - Facial nerve identified in a normal position = 1 point
   - Malleus-incus complex present = 1 point
   - Mastoid pneumatization well developed = 1 point
   - Incus-stapes connection present = 1 point
   - Round window present = 1 point
   - External ear well formed = 1 point

6. It is possible for a canal cholesteatoma (epithelial inclusion cyst) to develop medial in a stenotic external auditory canal. Canal cholesteatomas may be suspected clinically when squamous debris is found in the ear canal remnant, and is identified radiographically as widening of the EAC.

Test Interpretation

An audiologic assessment is necessary in children affected by aural atresia. If bilateral atresia is present, a masking dilemma will exist that needs special evaluation techniques. Aural atresia is most commonly associated with a maximal conductive hearing loss. In cases of unilateral atresia, audiometric evaluation of the opposite ear is important in the rehabilitative planning of the child.

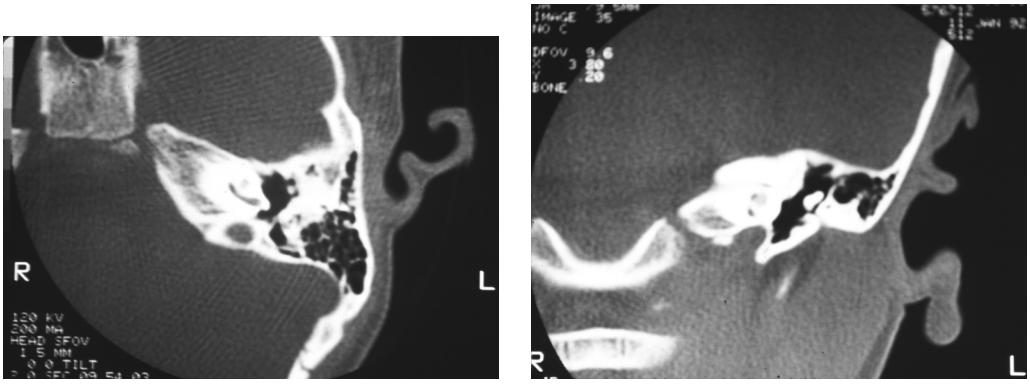
CT of the temporal bone is necessary for preoperative planning (**Fig. 4.1**). This should be a high-resolution scan using 1.5-mm cuts. It may be obtained at any time before the surgical repair, but it is customary to wait until a time proximate to the planned repair. Surgical repair is often performed before the child's entrance into school. The CT scan is used to assess the integrity of the tympanic bone, the extent of mastoid and middle-ear pneumatization, the presence of ossicles, and the course of the facial nerve and also to determine whether any abnormalities of the inner ear are present.

Diagnosis

1. Grade 1 deformity of the right pinna
2. External auditory canal atresia

Medical Management

Adequate auditory stimulation should be provided to an infant as soon as reasonable. In cases of bilateral atresia, this can be accomplished with bone-conduction hearing aids when the child is 4 to 6 months old. Implantable bone-conduction aids are generally not recommended until the skull thickness is sufficient to hold the implant, which is generally around the age of 3 years. Bone-conduction hearing aids or bone-anchored hearing aids



A,B

Fig. 4.1 (A,B) Computed tomography images of aural atresia with well-aerated middle ear and mastoid air cell system, with the presence of ossicles and a facial nerve in the normal location.

(BAHAs) provide serviceable hearing to patients. No additional reconstruction is necessary if CT examinations show a temporal bone with features unlikely to yield serviceable hearing after reconstruction. It is ideal to provide binaural hearing in children for hearing in noisy environments and to provide directionality of sound localization.

◆ Surgical Management

Preoperative assessment is important to determine surgical candidacy. A functioning and normal inner ear should be present. A Jahrsdoerfer score of 8 or greater is associated with good success in obtaining serviceable hearing after the surgical reconstruction. Scores of 6 or lower are generally better served without canal reconstruction unless to eradicate developing canal cholesteatomas. The risk of facial nerve paralysis must be discussed with families, and vigilance must be practiced to protect the nerve.

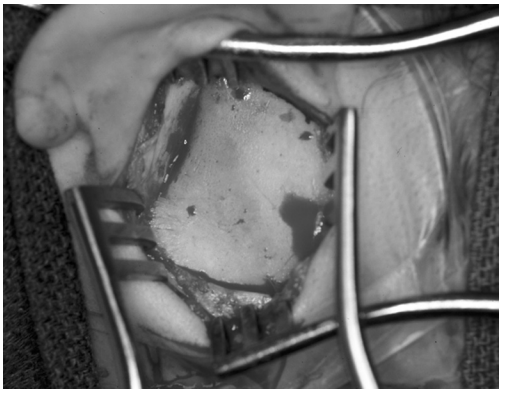
The timing of surgery and the expectations of family are key. Bilateral atresia cases should be operated on before the child enters school, around the age of 5. The better hearing or more normal radiographically appearing atretic ear should undergo repair. Surgery for unilateral cases is controversial. Some think surgery should be deferred until age 18 years, when the patient is able to participate in the decision process. Others think the advantage of binaural hearing outweighs the surgical risks and the surgery should be done at a young age.

Microtia repair is classically performed before construction of the external auditory canal. This sequence is not uniformly accepted, however. It is best to approach this issue of timing of each repair with the family and the surgeons performing each part of the procedure.

Surgical repair of the pinna must be tailored to the specific anatomic problem. The first stage of the microtia repair involves the construction and implantation of the auricular framework created from autologous rib cartilage. The second stage involves transposing preexisting auricular remnants to create the lobule. The third stage involves correction of the aural atresia and alignment of the newly created pinna to the newly created meatus of the external auditory canal. The fourth and fifth stages of the repair involve creating a tragus and releasing the auricular framework from the side of the skull. Three-month intervals between stages are recommended.

The aural atresia repair must be carefully performed to protect the facial nerve, which may be located anteriorly from its normal location in the mastoid. Intraoperative monitoring of the facial nerve is advocated. The repair involves creating an auditory canal, removing the atretic plate, restoring the sound-conducting mechanism, creating a new external auditory meatus, and lining the newly created

A,B



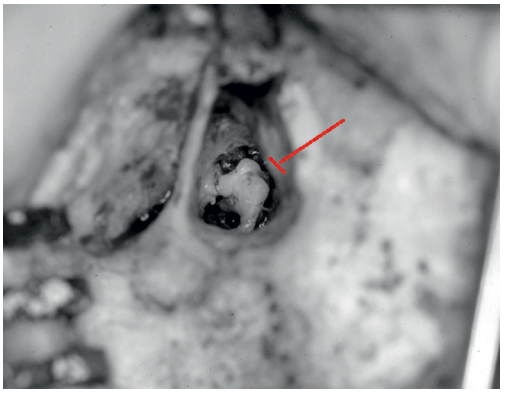


Fig. 4.2 (A,B) Surgical view of the operative site, with tegmen, temporal mandibular joint, and cortex covering mastoid air cell system defined. The ossicular mass is abnormal and will be located centrally in the reconstructed tympanic membrane.

canal with a split thickness skin graft. The canal is generally centered over the ossicular mass and bounded anteriorly by the temporal mandibular joint, superiorly by the tegmen, and posteriorly by the mastoid air cell system (**Fig. 4.2**).

A 30% revision rate is seen with long-term follow-up. The external auditory canal has a tendency to restenosis over time. This may encroach on the tympanic membrane and interfere with hearing thresholds as well.

Implantable bone abutments are available to secure posts to allow prosthetic devices to be attached to the skull. Some families prefer to use autograft materials to correct microtia. Others prefer the simplicity of a prosthetic device. The quality of the cosmetic result is generally better with prosthetic ears compared with that obtained with the use of autologous cartilage. The same abutments are used for the BAHA device. The implantable abutments have revolutionized the treatment of patients with severe deformities that made them less than ideal surgical candidates.

◆ Rehabilitation and Follow-up

Continued aural rehabilitation is essential for all hearing-impaired children. Regular otologic examinations should be performed twice annually. In addition, regular visits with a speech therapist may be beneficial.

◆ Questions

1. Which of the following statements is incorrect with regard to aural atresia?
- **A.** Unliateral atresia more commonly affects the right side.
- **B.** Unilateral atresia is more commonly encountered than bilateral conditions.
- **C.** A family history of aural atresia is identified in 50% of cases.
- **D.** It is estimated that aural atresia has a frequency in the United States of 1:8000 live births.

2. Regarding the surgical management of congenital aural atresia:
- **A.** Jahrsdoerfer's rating system for middle-ear structures correlates poorly with surgical success for hearing restoration.
- **B.** A magnetic resonance imaging scan provides the ideal assessment of structures related to reconstruction.
- **C.** Surgical intervention may be undertaken at any age.

D. Postoperative speech-reception thresholds of less than 30 dB of hearing loss can be achieved in properly selected cases

3. Syndromes associated with ear canal and pinna abnormalities do not include:
A. Crouzon and Stickler
B. Treacher Collins and hemifacial microsomia
C. Goldenhar and Alpert
D. Klippel-Feil and 18q− chromosome
E. Branchio-otorenal and Pfeiffer

Suggested Readings

Brent B. Auricular repair with autogenous rib cartilage grafts: two decades of experience with 600 cases. Plast Reconstr Surg 1992;90(3):355–374

Brent B. The team approach to treating the microtia atresia patient. Otolaryngol Clin North Am 2000;33(6): 1353–1365

Calzolari F, Garani G, Sensi A, Martini A. Clinical and radiological evaluation in children with microtia. Br J Audiol 1999;33(5):303–312

Chang SO, Lee JH, Choi BY, Song JJ. Long term results of postoperative canal stenosis in congenital aural atresia surgery. Acta Otolaryngol Suppl 2007;558:15–21

De la Cruz A, Teufert KB. Congenital aural atresia surgery: long-term results. Otolaryngol Head Neck Surg 2003; 129(1):121–127

Jahrsdoerfer RA, Yeakley JW, Aguilar EA, Cole RR, Gray LC. Grading system for the selection of patients with congenital aural atresia. Am J Otol 1992;13(1):6–12

Siegert R, Mattheis S, Kasic J. Fully implantable hearing aids in patients with congenital auricular atresia. Laryngoscope 2007;117(2):336–340

Teufert KB, De la Cruz A. Advances in congenital aural atresia surgery: effects on outcome. Otolaryngol Head Neck Surg 2004;131(3):263–270

II

Adult Otology

5

Cholesteatoma

Catherine K. Hart and Ravi N. Samy

History

A 52-year-old woman with a 1-year history of progressive left-sided hearing loss and otorrhea also complains of tinnitus with aural fullness. She had a tympanostomy tube placed 6 months ago. She also describes intermittent imbalance but no true vertigo.

The patient has no history of trauma, nor does she have a history of recurrent ear infections during childhood. The patient's father had a history of multiple otologic surgeries. Otomicroscopic examination of the right ear reveals a normal external auditory canal with a mildly atelectatic tympanic membrane. Examination of the left ear canal demonstrates a tympanostomy tube in place. There is a pars flaccida retraction pocket with squamous debris surrounded by granulation tissue. Weber lateralizes to the left ear, and Rinne is positive bilaterally. No nystagmus is noted. Cranial nerve function is intact. Gait and Romberg are normal.

Differential Diagnosis—Key Points

A *cholesteatoma* is a lesion of the temporal bone that is lined with keratinizing squamous epithelium. Cholesteatoma may be localized to the epitympanum, or it may follow preformed pathways to invade the mastoid, middle ear, or petrous apex. Cholesteatoma may be acquired (most common), congenital, or iatrogenic in origin.

1. *Primary cholesteatoma* is due to longstanding negative middle-ear pressure and retraction of the tympanic membrane resulting from chronic eustachian tube dysfunction (most common).
2. *Secondary cholesteatoma* results from the growth of squamous epithelium through a tympanic membrane perforation into the middle ear. The perforation may be the result of infection or trauma.
3. *Congenital cholesteatoma* is thought to be an embryonic rest of epithelial tissue that fails to involute during middle-ear development.
4. *Iatrogenic cholesteatoma* can occur as a result of implantation of squamous epithelium into the middle ear as a result of trauma or a surgical procedure.
5. The differential diagnosis of cholesteatoma includes chronic otitis media with or without suppuration, keratosis obturans, tuberculous otitis, osteoradionecrosis, squamous cell or basal cell carcinoma, foreign-body reaction, and granulomatous disease.

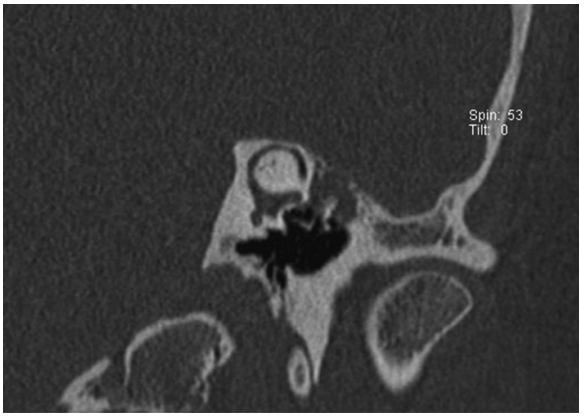


Fig. 5.1 Computed tomography scan with soft tissue mass in epitympanum eroding tegmen and incus.

Test Interpretation

A baseline audiogram should be obtained for all patients with cholesteatoma. In cases of associated dizziness, electronystagmography is warranted. Computed tomography scanning with 1-mm high-resolution cuts in the axial and coronal planes allows visualization of the cholesteatoma and any resultant osseous erosion of the bony labyrinth, tegmen–posterior fossa dural plate, and fallopian canal. **Figure 5.1** demonstrates the typical findings of an acquired cholesteatoma with a soft tissue mass in the epitympanum and bony erosion of the tegmen tympani and incus.

An audiogram (**Fig. 5.2**) reveals the typical conductive hearing loss associated with cholesteatoma.

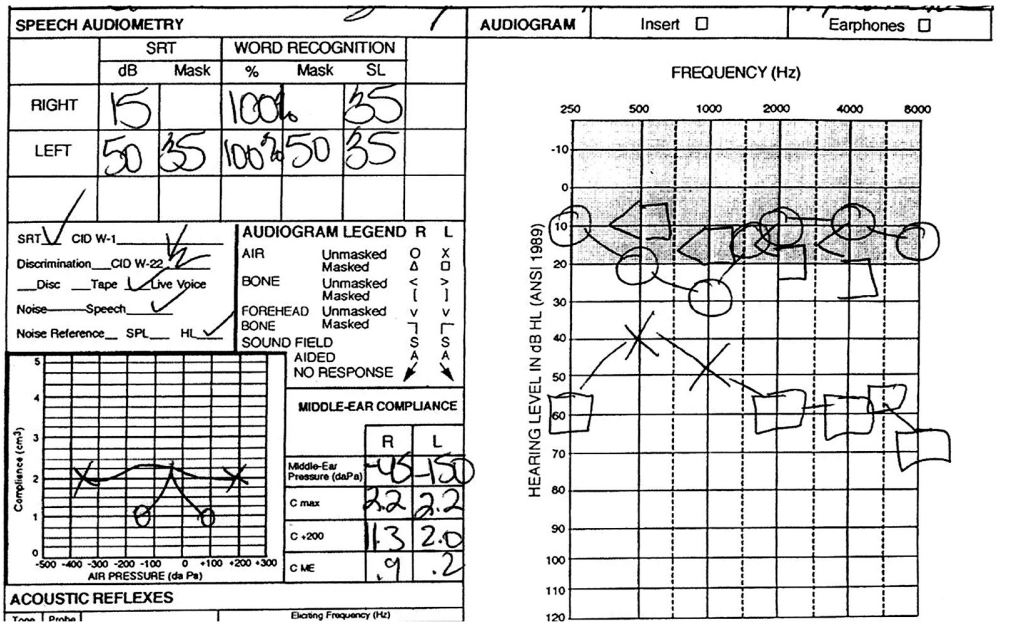


Fig. 5.2 An audiogram demonstrates the typical conductive hearing loss associated with cholesteatoma. The tympanogram is type B (flat) due to the presence of a tympanostomy tube. Patient has good word recognition bilaterally.

◆ Diagnosis

Acquired cholesteatoma

◆ Medical Management

The medical management of cholesteatoma includes in-office debridement and treatment of acute inflammatory changes. Dry ear precautions (e.g., with cotton ball and petroleum jelly) are mandatory. Ototopical antibiotics with steroids are used to reduce secondary infection and reduce granulation tissue formation. Systemic antibiotic therapy can also be used for concomitant infection. However, definitive treatment of cholesteatoma requires surgical intervention.

◆ Surgical Management

The options for management of cholesteatoma include canal wall-up or canal wall-down tympanomastoidectomy. If a canal wall-up technique is used, a facial recess (posterior tympanotomy) approach is needed to assess the sinus tympani (the region of the temporal bone with the highest rate of recurrent or residual disease). A canal wall-down approach is the gold standard for treatment of cholesteatoma, particularly when an impending complication is present (e.g., cochlear fistula). The location of cholesteatoma, the extent of bone destruction, and the amount of mucosal inflammation will dictate whether reconstruction during the initial procedure is possible. Staging of the ear (also known as a second-look procedure) is often used to assess for recurrent or residual cholesteatoma and is typically performed 6 to 12 months after the initial procedure. At the time of the second look, an ossicular chain reconstruction can be performed.

◆ Rehabilitation and Follow-up

A high index of suspicion is needed to assess for recurrence of disease over the life of the individual patient. If successful ossicular reconstruction is not achieved, consideration should be given to a bone-anchored hearing aid or referral for traditional hearing aid evaluation.

◆ Questions

1. What is the most common site of recurrence for cholesteatoma?
2. What procedure is considered the gold standard for treatment of cholesteatoma?
3. What is the most common type of cholesteatoma?

Suggested Readings

Glasscock ME III, Haynes DS, Storper IS, Bohrer PS. Surgery for chronic ear disease. In: Hughes G, Pensak M, eds. Clinical Otology, 3rd ed. New York, NY: Thieme; 2007

Persaud R, Hajioff D, Trinidade A, et al. Evidence-based review of aetiopathogenic theories of congenital and acquired cholesteatoma. J Laryngol Otol 2007;121(11): 1013–1019

Smith JA, Danner CJ. Complications of chronic otitis media and cholesteatoma. Otolaryngol Clin North Am 2006; 39(6):1237–1255

6

Otosclerosis

Collin M. Burkart and Ravi N. Samy

History

An otherwise healthy 40-year-old woman complains of bilateral hearing loss. The patient describes a gradual decline in her ability to hear over the past 10 years. She has tried a traditional hearing aid on the left side only (her worse hearing ear). However, she is tired of using the hearing aid and is seeking an alternative. She has no previous history of otologic or neurotologic disease nor symptoms other than hearing loss.

Physical examination demonstrates normal-appearing external auditory canals. Under binocular microscopy, the patient's tympanic membranes appear to be normal with normal pneumatic otoscopy and a negative fistula test. The Weber, at 512 Hz, was midline and the Rinne was negative (bone conduction greater than air) bilaterally at 512 Hz. The remaining head and neck examination was within normal limits.

Differential Diagnosis—Key Points

1. Based on history and physical examination, this patient appears to have a conductive hearing loss bilaterally. It will be important to further the evaluation with audiometric studies.
2. The differential diagnosis for conductive or possibly mixed hearing loss is expansive, and history provides invaluable information in clarifying the cause. Otosclerosis should always be considered. If a history of recurrent ear infections is present, then ossicular discontinuity resulting from incus erosion is a possibility, as is ossicular fixation from tympanosclerosis. A history of trauma may also lead toward a diagnosis of ossicular discontinuity. Middle-ear masses or space-occupying lesions such as glomus tumors or cholesteatoma must also be contemplated. Paget disease (osteitis deformans) is another possibility, as is osteogenesis imperfecta (van der Hoeve–de Kleyn syndrome).
3. Otosclerosis, a common histologic finding, is present in 10% of the white population. However, only around 10% of those with histologic findings have clinical symptoms. Therefore, about 1% of white patients will consult their physician with this disease. It is less common in other ethnic groups. It occurs more commonly in females, with a 2:1 ratio, and when a patient has the disease, 80% of the time the contralateral ear has histologic changes as well. The most common age for diagnosis is in the early to mid 30s, with reports in the past indicating that the disease seems to progress more rapidly during pregnancy, but more recent data indicate that progression during pregnancy is not as significant as previously believed.

4. Otosclerosis may manifest as conductive hearing loss, mixed hearing loss, or, rarely, with only a sensorineural hearing loss (cochlear otosclerosis). Most commonly, patients have a conductive or mixed loss with good word discrimination. Vestibular symptoms are rare but can be present. Tinnitus is also a frequent finding associated with the disease.

◆ Test Interpretation

Audiogram

The audiometric evaluation in this patient demonstrates a bilateral moderate conductive hearing loss with speech reception threshold being symmetric at 50 dB on the right and 55 dB on the left. Her word recognition was 100% bilaterally. She demonstrated a mild sensorineural hearing loss at 2000 Hz consistent with a Carhart's notch. Tympanometry was normal bilaterally.

Radiography

This patient did have a high-resolution temporal bone computed tomography (CT) scan performed. Although not essential to the determination of the diagnosis, it can be helpful in certain instances (e.g., atypical cases or cases in which superior canal dehiscence is a possibility). The CT scan can also be helpful in determining whether there is cochlear involvement. A halo sign can be seen because of demineralization of bone around the otic capsule.

◆ Diagnosis

Otosclerosis

◆ Medical Management

It is thought that all patients with a diagnosis of otosclerosis need to be presented with the option of medical management. The options for medical management include oral sodium or calcium fluoride. This medication is taken daily; it can demonstrate prevention of progression of symptoms in 80% of patients. This is especially helpful in cochlear otosclerosis. Sodium fluoride can be used regardless of whether the patient intends to undergo surgery, has undergone surgery, or is not planning on pursuing surgical treatment.

An alternative to surgery that still provides improvement in hearing is amplification. This is always a viable option, without the operative risk. Many patients will choose amplification with traditional hearing aids. In recent years, the training in otolaryngology programs has changed such that residents are getting less experience with performing stapedectomy or stapedotomy. Therefore, evidence indicates that younger practicing general otolaryngologists are operating less on patients with otosclerosis and recommending hearing aids more often.

◆ Surgical Management

Surgical management is available for treatment of the conductive hearing loss of otosclerosis; it is very effective. In the properly selected patient, the results can be satisfying for the patient. However, the patient needs to be reminded that the procedure is elective. The risks and benefits need to be reviewed carefully. Although the risks are rare, potential serious complications of stapes surgery (stapedotomy, stapedectomy) include sensorineural hearing loss of varying degrees (anacusis 1% chance), facial nerve injury, tympanic membrane perforation, perilymphatic fistula, vertigo, and dysgeusia, can result from injury of the chorda tympani nerve. Absolute contraindications to the procedure include surgery on the only hearing ear, active infection, or X-linked conductive hearing loss (risk of cerebrospinal fluid gusher). Patients with multiple medical co-morbidities should also be managed with caution. The procedure can be performed with the patient under local or general anesthesia. Stapes surgery is often performed with the use of a CO_2 or argon laser or direct microsurgical dissection with or without the use of an otologic drill. Often the piston of the prosthesis is packed with fascia or tissue of the surgeon's choice (e.g., vein, perichondrium, loose areolar tissue).

This patient underwent a right transcanal tympanotomy and stapedotomy with the CO_2 laser under local anesthesia. She did well and was able to demonstrate improved hearing

on the table. At her postoperative visit, she reported improvement in hearing with a positive Rinne at 512 Hz on the right side.

An alternative surgical option is a bone-conducting hearing aid. The current option available in the United States is a bone-anchored hearing aid, which provides good results for patients with otosclerosis without many of the risks of stapedectomy or stapedotomy and is technically less challenging. However, the negatives of wound healing and care as well as the need to continue to wear an external hearing aid persist. All options should be presented to the patient so that an informed decision can be made.

Rehabilitation and Follow-up

In the immediate postoperative period, while the patient is healing from the stapedectomy or stapedotomy and the fenestration is sealing, the patient must avoid straining or strenuous activity. It is recommended that the patient avoid heavy lifting for about a month postoperatively. The patient should also be on dry-ear precautions. Many otologic surgeons will prescribe a week of postoperative antibiotics. The patient should avoid nose blowing, and sneezing with an open mouth should be encouraged. Activities that involve changes in barometric pressure, such as SCUBA diving or flying in nonpressurized cabins, are also risky for the healing fenestration and should be avoided. If the surgery is successful and the patient has bilateral disease, the second side can be considered for operative repair around 6 months after the first (because of the risk of delayed hearing loss after stapes surgery). The patient should have long-term follow-up with serial audiograms to monitor for recurrence, dislodgment of the prosthesis, or development of cochlear involvement.

Questions

1. A patient arrives at your office with known otosclerosis in the right ear. He desires improvement for his hearing loss. He states that he is a chef; therefore, his sense of taste is very important to him. What are his options?
2. Carhart's notch occurs at what frequency?
3. A 40-year-old man has right-sided hearing loss. He has a history of recurrent ear infections. Tuning fork tests and audiometric studies confirm a conductive hearing loss of around 25 dB. Otomicroscopic examination reveals a mobile but thickened tympanic membrane. What test might be useful to diagnose the cause of the conductive hearing loss?

Suggested Readings

Fritsch MH, Naumann IC. Phylogeny of the stapes prosthesis. Otol Neurotol 2008;29(3):407–415

Ruckenstein MJ, Staab JP. Who is performing stapedectomy surgery? Implications for residency and fellowship training. Laryngoscope 2008;118(7):1224–1227

Shine NP, Rodrigues S, Miller S, Packer P. Bilateral stapedectomy: association between first- and second-ear surgical findings and their effects on the second-ear outcome. Ann Otol Rhinol Laryngol 2008; 117(3): 207–211

7

Glomus Jugulare

Todd M. Wine and Ravi N. Samy

History

A 45-year-old woman's chief complaint is tinnitus in the left ear. The tinnitus is pulsatile and has been present for years. She has noticed hearing loss and aural fullness. She denies headaches, dizziness, or other neurotologic symptoms. Her medical history is significant only for allergies; she has no surgical history.

Physical examination revealed a midline Weber and positive Rinne bilaterally. Examination of the pinnae and external auditory canals bilaterally is normal. Otomicroscopic examination is normal on the right. However, on the left, there is an anterior, inferior mesotympanic purplish blue mass. Pneumatic otoscopy revealed blanching of the mass (Brown sign). Cranial nerve examination reveals no abnormalities.

Differential Diagnosis—Key Points

1. Paragangliomas of the temporal bone include glomus tympanicum and glomus jugulare. They arise from neural crest cells residing in the region near the jugular bulb or Arnold and Jacobsen nerves. Although these are often slow growing tumors, they can be quite extensive by the time they become symptomatic. Histologically, these tumors are made of type I chief cells arranged in organoid-nested pattern with type II sustenacular cells (also known as zellballen or balls of cells). These tumors can present with a violaceous middle-ear mass noted on otomicrosopic examination. Pulsatile tinnitus is the most common symptom followed by hearing loss. The pulsatile tinnitus reflects the vascular nature of the tumor and is due to turbulent blood flow. Glomus jugulare is differentiated from glomus tympanicum by extension into the hypotympanum and jugular bulb. As its location is near the jugular foramen, glomus jugulare can cause symptoms related to dysfunction of cranial nerves IX through XII, including dysphagia, hoarseness, speech difficulties, and shoulder weakness.
2. Vascular anomalies of the middle ear most often manifest with pulsatile tinnitus. These anomalies include high-riding jugular bulb, aberrant internal carotid artery, and petrous carotid artery aneurysms. These anomalies are readily identified using high-resolution computed tomography (CT) scans of the temporal bone. High-riding jugular bulb (6% of patients) is most common and occurs when the jugular bulb extends superior to the inferior tympanic annulus. The aberrant carotid artery has CT findings that include an enlarged inferior tympanic canaliculus, enhancing mass in

the hypotympanum, absence of the vertical segment of the internal carotid artery, and absence of the carotid bony canal.

Test Interpretation

Finding the cause and determining treatment plans regarding otologic problems often start with obtaining an audiometric evaluation that includes pure tones, speech reception threshold and discrimination, tympanometry, and acoustic reflexes. Typically, the audiogram will show conductive hearing loss when vascular anomalies or tumors of the middle ear are present. Sensorineural hearing loss can be present if there is extension into the otic capsule, temporal bone, or internal auditory canal.

The cornerstone of the diagnosis of a vascular abnormality of the middle ear requires imaging. Both high-resolution CT scan of the temporal bones and magnetic resonance imaging (MRI) provide useful information and limit the differential diagnosis. The high-resolution CT scan with axial and coronal views provides excellent detail of the temporal bone, allowing the middle ear, petrous carotid, and jugular foramen to be evaluated. This should allow the differentiation of vascular anomalies of the temporal bone, glomus tympanicum, and glomus jugulare. Glomus jugulare tumors often have erosion of the caroticojugular spine.

Multiplanar MRI is especially critical for glomus jugulare tumors. The coronal views allow assessment of the inferior extension of the tumor through the jugular foramen and into the neck. The axial views allow the relationship to the petrous carotid to be visualized while assessing any intracranial extension. T1-weighted images will have intermediate signal intensity, whereas T2 imaging has high signal intensity. Flow voids are present in both (salt and pepper pattern), and the tumor will strongly enhance with contrast. MRI is also helpful for determining the presence of synchronous paragangliomas of the head and neck (e.g., contralateral carotid body tumor).

Carotid and vertebral angiography (four-vessel angiography) is typically not needed for diagnostic purposes for glomus tumors. However, angiography with preoperative embolization is performed before the surgical extirpation of the glomus jugulare tumor to reduce the amount of intraoperative blood loss and the subsequent transfusion of blood products.

Laboratory evaluation must include urine and serum catecholamines because around 4% of paragangliomas are hormonally active. It should be noted that if screening reveals excess catecholamines, adrenal imaging is important to rule out a concomitant pheochromocytoma.

This patient's CT scan revealed a rather large mass arising from the jugular foramen with intracranial and cervical extensions with erosion of the temporal bone. MRI revealed an enhancing left jugular foramen mass with extension into the neck inferiorly and to the seventh and eighth cranial nerve complexes intracranially (**Figs. 7.1, 7.2, 7.3**).

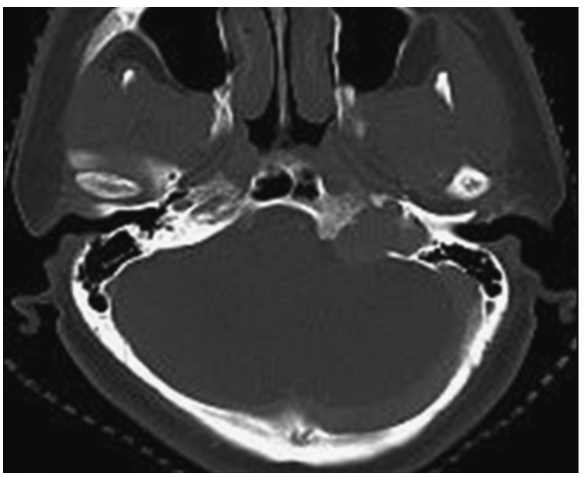


Fig. 7.1 Axial computed tomography scan of temporal bone demonstrating jugular foramen erosion.

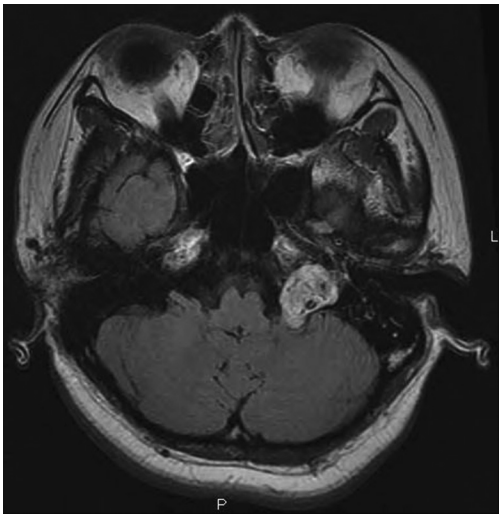


Fig. 7.2 Axial T1-weighted magnetic resonance image revealing enhancement of left jugular foramen mass as well as intracranial extension. Notice flow-void patterns in the mass.

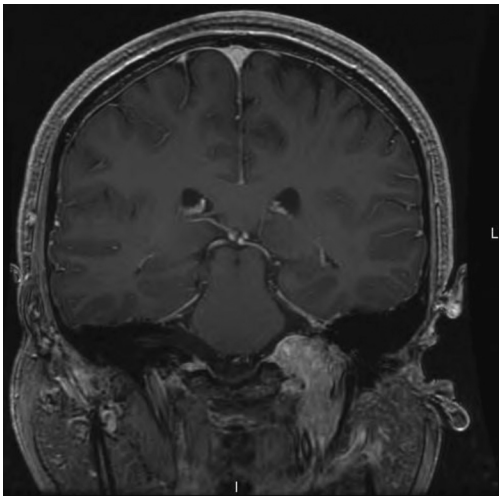


Fig. 7.3 Coronal T1-weighted magnetic resonance image revealing left jugular foramen mass with cervical extension.

◆ Diagnosis

Glomus jugulare

◆ Medical Management

The diagnosis of a paraganglioma necessitates investigation for the presence of excess catecholamines. As stated earlier, only around 4% of head and neck paragangliomas are secreting. If the catecholamines are elevated, it is vitally important to ensure that blood pressure is being properly controlled. This is critical in the perioperative period because surgical manipulation can cause a dangerous release of catecholamines. Thus, perioperative α and β blockade should be performed.

Factors such as patient age, medical co-morbidities, tumor size, involvement of critical structures, and the patient's wishes dramatically affect treatment decisions.

Although surgery is the mainstay of treatment, nonsurgical treatment options include observation (typically used for older or frail patients) and radiotherapy. Radiotherapy is aimed at preventing further tumor growth and therefore progression of symptoms. Radiotherapy for glomus jugulare can be performed via external-beam radiation (for larger tumors) or stereotactic radiation therapy. In the short term, radiation therapy carries a decreased risk of cranial nerve deficit compared with surgical treatment options, and radiation has proven effective at aborting further growth in 70 to 90% of cases. However, there is a chance of malignant transformation. In addition, regrowth rates of up to 20% have been reported with radiotherapy.

◆ Surgical Management

Surgical management is the only curative treatment option for glomus jugulare. The extent of the tumor is vitally important and dictates the best surgical approach for each individual. All surgical approaches are based on having adequate control of vital structures in close approximation to the tumor, such as the lower cranial nerves and the internal carotid artery and internal jugular vein.

The facial nerve lies in close proximity to the jugular bulb. This necessitates either the fallopian bridge technique (working around a suspended, immobilized mastoid segment of the facial nerve) or facial-nerve mobilization techniques for adequate access. The carotid artery must be controlled proximally in the neck. In larger tumors, an infratemporal fossa approach is necessary to control the petrous carotid artery.

Cranial nerves IX, X, XI, and XII must be assessed during the tumor resection as well. If they are uninvolved, it might be possible to spare their function. In addition to these critical structures, preoperative assessment of intracranial involvement is important; cerebrospinal fluid (CSF) leaks and posterior fossa hemorrhage can occur though rarely.

Rehabilitation and Follow-up

As expected with these operations, cranial nerve deficits are the most common morbidities. Cranial nerve VII is often weak postoperatively whenever mobilized. Postoperative attention to eye protection is critical so that ocular morbidities can be avoided. Injury to the lower cranial nerves will affect speech and swallowing to varying degrees. Rehabilitation with speech therapy is critical in achieving adequate speech and swallowing and can be assisted by vocal cord medialization. Ultimately the rehabilitative goal after surgical extirpation of a glomus jugulare is to have a patient with ocular protection, auditory rehabilitation with hearing aids, and speech and swallowing rehabilitation.

Questions

1. What is the only curative treatment option for the management of glomus jugulare tumors?
2. What cranial nerves are at risk with glomus jugulare tumors (treated or untreated)?
3. What percentage of glomus jugulare tumors are hormonally active?

Suggested Readings

Brackmann DE, Arriaga MA. Surgery for glomus and jugular foramen tumors. In: Brackmann DE, Shelton C, Arriaga MA, eds. Otologic Surgery. Philadelphia, PA: W.B. Saunders Company; 2001:478–492

Foote RL, Pollock BE, Gorman DA, et al. Glomus jugulare tumor: tumor control and complications after stereotactic radiosurgery. Head Neck 2002;24(4):332–338

Kaylie DM, O'Malley M, Aulino JM, Jackson CG. Neurotologic surgery for glomus tumors. Otolaryngol Clin North Am 2007;40(3):625–649

Krych AJ, Foote RL, Brown PD, Garces YI, Link MJ. Long-term results of irradiation for paraganglioma. Int J Radiat Oncol Biol Phys 2006;65(4):1063–1066

Li G, Chang S, Adler JR Jr, Lim M. Irradiation of glomus jugulare tumors: a historical perspective. Neurosurg Focus 2007;23(6):E13

Ramina R, Maniglia JJ, Fernandes YB, Paschoal JR, Pfeilsticker LN, Coelho Neto M. Tumors of the jugular foramen: diagnosis and management. Neurosurgery 2005; 57(1, Suppl)59–68

8

Sudden Sensorineural Hearing Loss

Catherine K. Hart and Ravi N. Samy

History

A 75-year-old man presents with right-sided hearing loss 2 days before presentation. Although there was no history of trauma preceding the hearing loss, the patient states that he was recovering from a cold. He also describes concomitant, right-sided tinnitus and aural fullness. He denies otorrhea, aural fullness, headache, or vision changes. He denies previous ear infections. He does have a history of left profound sudden hearing loss 10 years ago. His medical history includes hypertension, type 2 diabetes mellitus, coronary artery disease, and hypercholesterolemia. His family history is negative for autoimmune disease or hearing loss.

Neuro-otologic examination is significant for Weber and Rinne tests that the patient could not hear. Microscopic examination of his external auditory canals and tympanic membranes reveals normal anatomy with no evidence of acute infection or sequelae of previous otitis media. His cranial nerves are intact, and there is no spontaneous, gaze-evoked, or positional nystagmus. His cerebellar examination is unremarkable, and his gait and Romberg are normal.

Differential Diagnosis—Key Points

Sudden sensorineural hearing loss (SNHL) is defined as a 30-dB or greater sensorineural hearing loss occurring in at least three contiguous frequencies within 3 days or less. The possible causes of sudden SNHL are numerous. Often, the potential causes can be narrowed by the patient's history and physical examination. It is helpful to consider the possible causes in categories, such as infectious, inflammatory, vascular, neoplastic, traumatic, toxic, and idiopathic.

1. *Infectious.* Many infectious processes can affect the inner ear either unilaterally or bilaterally. Viral infections are commonly implicated, including cytomegalovirus, mumps, influenza, herpes simplex, measles, rubella, and human immunodeficiency virus. An association between upper respiratory tract infections (i.e., viral URIs) and sudden SNHL. Bacterial meningitis or labyrinthitis, syphilis, and mycobacterial or fungal infections may also be implicated. Infections are probably the most common cause of sudden SNHL.

2. *Inflammatory.* Several inflammatory processes can cause sudden SHNL, including autoimmune inner ear disease, Cogan syndrome, Wegener granulomatosis, polyarteritis nodosa, rheumatoid arthritis, systemic lupus erythematosus, and other rheumatologic disorders. These immune-mediated etiologies can be isolated to the inner ear (primary) or may be due to systemic autoimmune disease (secondary).
3. *Vascular.* Occlusion of the labyrinthine artery may result in sudden SNHL. Other vascular diseases—including leukemia, sickle cell disease, vasculitis, and embolization during coronary bypass surgery—can cause acute SNHL.
4. *Neoplastic.* Multiple tumors may directly or indirectly affect the inner ear and result in SNHL. About 13% of patients with acoustic neuromas present with sudden SNHL. Other tumors that can cause SNHL include meningioma and other cerebellopontine angle tumors, facial neuromas, squamous cell carcinoma of the external auditory canal or middle ear, glomus tumors, and papillary adenocarcinoma of the endolymphatic sac.
5. *Traumatic.* Penetrating or blunt trauma to the temporal bone can result in SNHL through direct damage to the labyrinth or internal auditory canal, subluxation of the stapes, or concussive injury.
6. *Toxics.* Several ototoxic medications can result in SNHL. The most commonly encountered toxins include aminoglycosides, loop diuretics, and cisplatin. These agents commonly cause bilateral SNHL, but unilateral loss may occur.
7. *Miscellaneous.* Other possible causes of sudden SNHL include diabetes mellitus, perilymphatic fistula, cochlear hydrops, congenital inner-ear deformities, and operative complications of otologic surgery.

◆ Test Interpretation

Investigation into the cause of this hearing loss should include serologic testing with complete blood count, erythrocyte sedimentation rate, C-reative protein, coagulation studies, thyroid function studies, lipid profile, and fluorescent treponemal antibody absorption. Additional testing can include rheumatoid factor, antinuclear antibody, heat-shock protein 70 antibody, and Lyme titers.

Audiometric testing, including pure-tone thresholds, speech reception and discrimination, and acoustic reflexes, is mandatory. In this case, the audiogram reveals profound bilateral sensorineural loss (**Fig. 8.1**). Magnetic resonance imaging with the internal auditory canal–cerebellopontine angle with contrast is required to rule out a retrocochlear lesion, such as an acoustic neuroma.

◆ Diagnosis

Unilateral right-sided SNHL in a patient with a history of prior contralateral profound, sudden SNHL

◆ Medical Management

Although the natural history is variable, many patients with sudden SNHL experience spontaneous improvement within the first 2 weeks. The treatment regimens are numerous. However, the most widely accepted treatment is corticosteroids, administered systemically (orally or intravenously) or transtympanically. If systemic steroid therapy is undertaken, close attention must be paid to systemic side effects, such as elevated blood sugars (worsening diabetes management), elevated blood pressure, and aseptic femoral head necrosis; prophylaxis for gastric ulcers must be administered.

Additional treatment modalities have been used less commonly, including vasodilators, inhaled carbogen, anticoagulants, and antivirals.

◆ Surgical Management

Surgical management may be indicated in the setting of a retrocochlear mass lesion. Cochlear implantation may be offered in the setting of bilateral profound hearing loss.

◆ Rehabilitation and Follow-up

This patient will require close monitoring of blood glucose levels and medical status during the period of oral steroid use. Follow-up

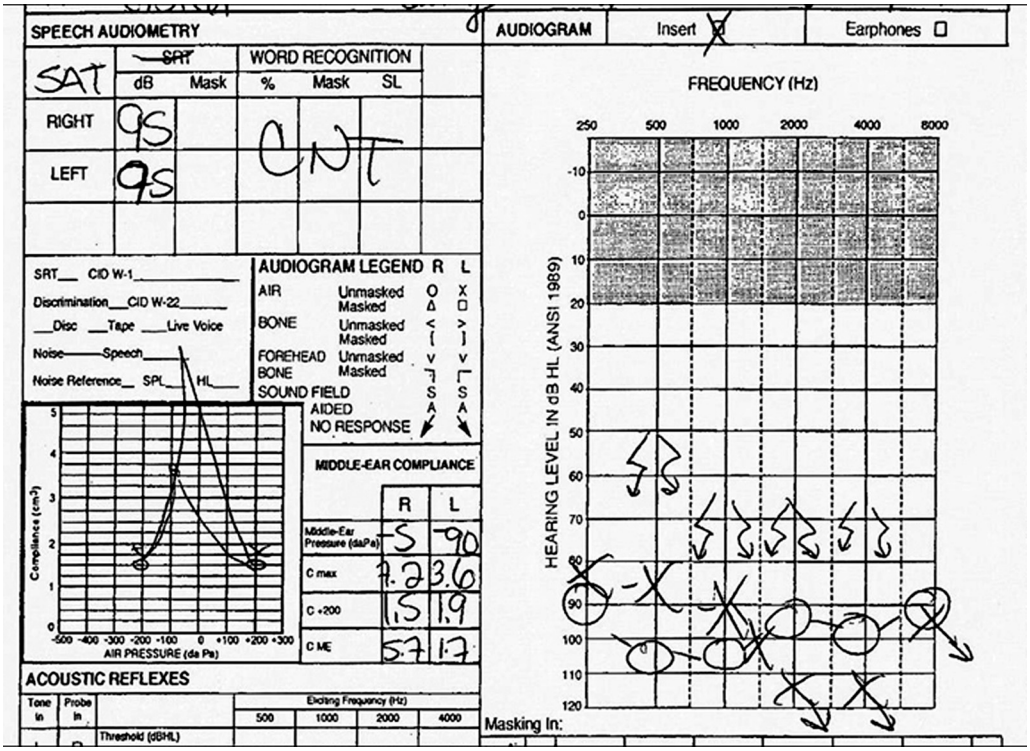


Fig. 8.1 Profound bilateral sensorineural hearing loss.

audiometry as well as testing to exclude retrocochlear pathology must be obtained. If this patient has no return of hearing, cochlear implantation would be appropriate.

◆ Questions

1. What is the most common cause of sudden sensorineural hearing loss?
2. What study is required of all patients with sudden sensorineural hearing loss to rule out a retrocochlear process?
3. By what two routes have steroids been administered for sudden SNHL?

Suggested Readings

Monsell EM, Teixido MT, Wilson MD, Hughes GB. Nonhereditary hearing loss. In: Hughes G, Pensak M, eds. Clinical Otology, 3rd ed. New York, NY: Thieme; 2007

O'Malley MR, Haynes DS. Sudden hearing loss. Otolaryngol Clin North Am 2008;41(3):633–649

Rauch SD. Clinical practice. Idiopathic sudden sensorineural hearing loss. N Engl J Med 2008;359(8): 833–840

9

Ménière Disease

Sarmad Sabour and Ravi N. Samy

History

A 46-year-old man with a chief complaint of dizziness has had four attacks of vertigo over the past 5 months. The episodes last for 1 to 2 hours and are associated with nausea and vomiting. He also complains of left aural fullness, hearing loss, and tinnitus. He has no history of ear surgery or family history of ear disease. He has no history of noise exposure, use of ototoxic medication, or history of head trauma. His medical history is significant for hypertension and allergies. Physical examination reveals Weber to lateralize to the right; Rinne is positive bilaterally. Otomicroscopic examination reveals no effusion, and a fistula test is negative. The remainder of his head and neck and neurotologic examinations are normal.

Differential Diagnosis—Key Points

1. Ménière disease is characterized by episodic vertigo, fluctuating sensorineural hearing loss, tinnitus, and aural fullness. There is variability in the symptoms and progression of the disease, although it primarily affects white individuals; peak incidence is at age 40 to 60 years. Ménière disease can also occur in children but less often than in adults.
2. Differential diagnosis includes perilymphatic fistula, dizziness associated with otitis media, benign positional vertigo of childhood, tumors of the skull base (such as meningiomas and acoustic neuromas), and autoimmune inner-ear disease. An autoimmune inner-ear disease workup must be considered in patients with bilateral Ménière disease. Migraine-associated vestibulopathy (MAV) must also be considered in the differential. Definite migrainous vertigo consists of episodic vertigo, migraine headache based on the International Headache Society criteria, and a migraine symptom during two episodes of vertigo. However, some patients can have MAV without headaches being present. Spontaneous or positional vertigo can last up to several days, and other aura symptoms may exist. Hearing loss can be present but is generally mild and nonprogressive. MAV is a diagnosis of exclusion but may be the most common entity confused with Ménière disease.
3. Pathogenesis stems from endolymphatic hydrops, most likely from insufficient absorption through the endolymphatic sac, either from an anatomic or functional standpoint. One theory considers microruptures of the membranous labyrinth

with leakage of potassium-rich endolymph into the perilymphatic space. There may also exist autoimmune, viral, or ischemic factors.

4. Allergy can play a role in Ménière exacerbations. It is theorized that antigenic exposure can lead to fluid changes in the endolymphatic sac, ion disturbances, and vestibulocochlear neurotoxicity. Accumulation of immune complexes may affect the sac's filtering ability. Endolymphatic hydrops results in saccular distention, which can then abut the stapes footplate with a resultant Hennebert sign. As a result of this phenomenon, stapes procedures are contraindicated in active Ménière disease.
5. Ménière disease is the idiopathic presentation of the syndrome of hearing loss, tinnitus, vertigo, and aural fullness. Clinically, attacks generally last 20 minutes to hours. Recurrent vestibulopathy or atypical Ménière disease may be confounding diagnoses. A rare presentation is the drop attacks of Tumarkin, characterized by sudden falls secondary to spatial disorientation but without loss of consciousness. The Lermoyez presentation is that of improving hearing loss and tinnitus with onset of vertigo. Acute attacks show horizontal nystagmus. Hearing loss is typically low frequency and fluctuating in the early period but can progress to a moderate sensorineural hearing loss in all frequencies.

◆ Test Interpretation

Standard testing consists of an audiogram with a typical low-frequency hearing loss (**Fig. 9.1**). Electronystagmography for evaluation of vestibular function and electrocochleography to assess for resultant increased SP:AP ratios are additional tests to consider. For suspicion of retrocochlear pathology, magnetic resonance imaging with contrast is undertaken.

◆ Diagnosis

Left-sided Ménière disease

◆ Medical Management

Medical treatment consists of lifestyle changes; salt restriction and diuretics are used to reduce endolymph accumulation (hydrops). Additionally, patients are told to stop smoking and limit alcohol and caffeine consumption. Stress reduction is important. Symptomatic treatment for acute episodes incorporates sedatives or antiemetics. There may be a role for positive pressure application to the middle ear as a means for redistributing inner-ear fluids. Meniett device use may provide benefit for vertigo reduction in select patients. Disequilibrium between vertigo attacks may persist. Vestibular rehabilitation is of benefit between episodes of vertigo to minimize disequilibrium and risk of falls. Intratympanic gentamicin (aminoglycoside therapy) can reduce vestibular attacks but with a greater than 30% chance of associated hearing loss.

◆ Surgical Management

For about 10% of patients, medical therapy fails and surgical therapy is required. Despite ablative procedures, central adaptation is still needed. Hearing-conservation procedures include endolymphatic sac decompression with or without mastoid shunt placement and vestibular neurectomy (via retrosigmoid, retrolabyrinthine, or middle cranial fossa routes). Intratympanic steroids can be used as a nonablative office procedure for their anti-inflammatory, immunosuppressive, and possible antioxidant effects (although the effects are considered temporary). Ablative treatment is done with a labyrinthectomy or vestibular nerve section procedure.

◆ Rehabilitation and Follow-up

Ménière disease affects an individual's quality of life because of changes in the cochleovestibular system. The patient will need long-term follow-up with serial audiograms to assess for progression of symptoms, including those of tinnitus, hearing loss, and

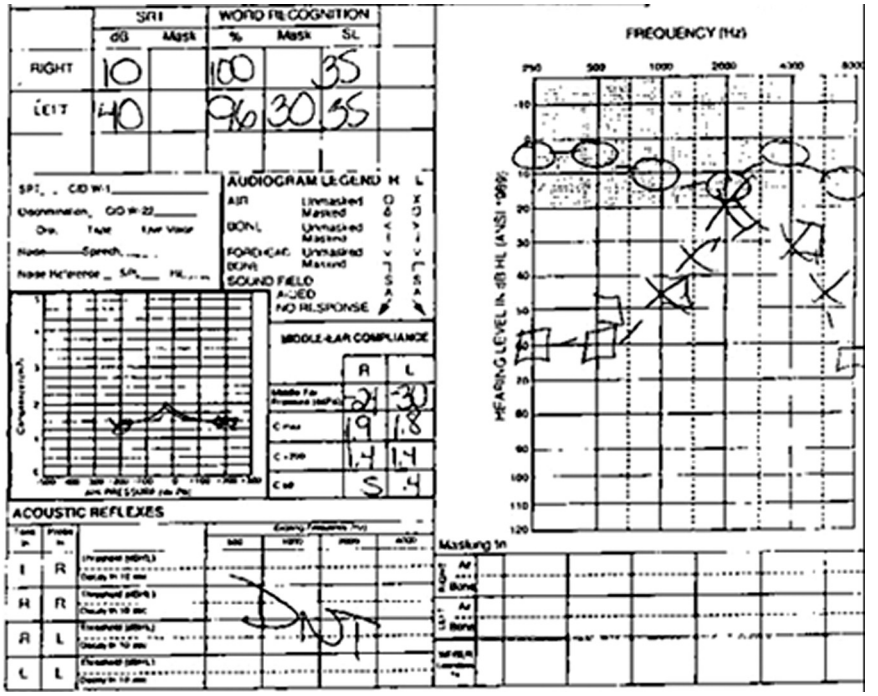


Fig. 9.1 Asymmetric sensorineural hearing loss of the left ear with good word recognition score (low-frequency hearing loss is common in patients with Ménière disease).

continued dizziness. Patients can also develop symptoms in the contralateral ear. Tinnitus and hearing loss can often be improved with use of a hearing aid.

◆ Questions

1. Name three different treatments for Ménière disease that are ablative.
2. What disease is most often confused with Ménière disease?
3. Ménière disease typically affects which hearing frequency range initially (low, mid, or high)?

Suggested Readings

Bodmer D, Morong S, Stewart C, Alexander A, Chen JM, Nedzelski JM. Long-term vertigo control in patients after intratympanic gentamicin instillation for Ménière's disease. Otol Neurotol 2007;28(8):1140–1144

Brinson GM, Chen DA, Arriaga MA. Endolymphatic mastoid shunt versus endolymphatic sac decompression for Ménière's disease. Otolaryngol Head Neck Surg 2007;136(3):415–421

De La Cruz A, Borne Teufert K, Berliner KI. Transmastoid labyrinthectomy versus translabyrinthine vestibular nerve section: does cutting the vestibular nerve make a difference in outcome? Otol Neurotol 2007;28(6):801–808

Gates GA, Verrall A, Green JD Jr, Tucci DL, Telian SA. Meniett clinical trial: long-term follow-up. Arch Otolaryngol Head Neck Surg 2006;132(12):1311–1316

Honaker J, Samy RN. Migraine-associated vestibulopathy. Curr Opin Otolaryngol Head Neck Surg 2008;16(5):412–415

Lee L, Pensak ML. Contemporary role of endolymphatic mastoid shunt surgery in the era of transtympanic perfusion strategies. Ann Otol Rhinol Laryngol 2008;117(12):871–875

Peterson WM, Isaacson JE. Current management of Ménière's disease in an only hearing ear. Otol Neurotol 2007;28(5):696–699

10

Benign Paroxysmal Positional Vertigo

Maria V. Suurna and Myles L. Pensak

History

A 45-year-old man complains of dizziness following a motor vehicle collision. He describes the symptoms as a spinning sensation associated with mild nausea that mostly occurs when he turns to the left while lying in bed or when he quickly gets up. Most recently he experienced a similar episode when he tilted his head back while standing on a ladder. The symptoms usually last less than a minute and improve if he stays still. The patient denies any associated hearing loss, tinnitus, or aural fullness. He has not experienced any recent infections and has never been exposed to any ototoxic agents. Other than mild hypertension, he is in good health.

A physical examination reveals a well-appearing man in no distress. The cranial nerve examination demonstrates no abnormalities. No spontaneous nystagmus is noted. The Weber is midline and the Rinne is positive bilaterally. The external auditory canals and tympanic membranes are normal. The Romberg and Fukuda tests are negative. When a Dix-Hallpike maneuver is performed, the patient experiences severe vertigo in the left-ear-down position. He is noted to have a down-beating rotary nystagmus, which lasts around 15 seconds. On the repeat maneuver he again experiences vertigo; however, the latency and fatigability of the nystagmus become apparent.

Differential Diagnosis—Key Points

1. Benign paroxysmal positional vertigo (BPPV) is attributed to cupulolithiasis or canalithiasis (or both) and usually affects a posterior semicircular canal. It is characterized by onset of vertigo in association with a head movement, particularly when turning in a supine position. Vertigo is usually noted after 5 to 10 seconds following the head movement and lasts about 15 to 60 seconds. It is often associated with nausea.
2. Vestibular neuronitis is the most common cause of acute vertigo. It often manifests with an acute onset of vertigo with associated nausea and vomiting. Severe vertigo usually lasts 24 to 48 hours. The symptoms gradually subside over a course of days to weeks. Viral infections and rarely ischemia of the labyrinth or brainstem are thought to be likely causes of this condition.
3. Ménière disease is characterized by severe episodes of vertigo often associated with fluctuation in hearing, aural fullness, and tinnitus. These episodes tend to be

recurrent, and duration can range from hours to days.

4. Oval and round window perilymph fistulae usually occur as a result of head trauma or barotrauma. Patients often present with fluctuant or progressive sensorineural hearing loss and vertigo immediately following a traumatic event. Coughing, straining, or lifting weights frequently heightens the symptoms.
5. Superior semicircular canal dehiscence syndrome is characterized by vertigo and oscillopsia induced by a loud sound or by a change in the middle ear or intracranial pressure.
6. Acute or chronic otitis media can manifest with associated vertigo caused by an inflammatory or infectious process directly affecting the labyrinth.
7. Eustachian tube dysfunction resulting in a myringostapediopexy or a myringoincudopexy can cause episodic vertigo.
8. Acoustic neuroma is the most common tumor affecting the eighth cranial nerve. Because of slow growth of the tumor, patients rarely show symptoms of vertigo. Hearing loss and tinnitus are more commonly the presenting symptoms, associated with mild balance dysfunction.
9. Other cerebellopontine angle lesions such as meningiomas, cholesterol granulomas, paragangliomas, arachnoid cysts, and metastatic tumors should be considered in patients presenting with vertigo.
10. Autoimmune inner-ear disease is an autoimmune disorder that usually affects only the inner ear. Patients usually present with fluctuant or progressive unilateral or bilateral sensorineural hearing loss that can be associated with vertigo. The symptoms may improve with high-dose corticosteroid treatment.
11. Syphilitic labyrinthitis is rare but can present with sensorineural hearing loss and vertigo.
12. Migraine-associated vertigo can be diagnosed in patients with episodic vertigo, lightheadedness, and disequilibrium associated with migraine headaches.
13. Demyelinating disorder such as multiple sclerosis can present with episodic vertigo.
14. Microvascular disease, vertebrobasilar insufficiency, or a stroke may lead to symptoms of vertigo and disequilibrium and should be considered in the differential diagnosis, particularly in older patients.
15. Many medications are commonly associated with lightheadedness. Rarely is true vertigo a potential side effect.
16. Psychiatric illnesses can be associated with range of symptoms, including unsteadiness and vertigo.
17. Mal de debarquement syndrome is a persistent rocking sensation experienced following disembarking from a boat after a long journey.

◆ Test Interpretation

A Dix-Hallpike maneuver is used to diagnose BBPV. Audiometric testing, radiographic imaging, and vestibular testing should be used when a diagnosis of BBPV is not obvious and other causes of vertigo are suspected.

In this case, the Dix-Hallpike test was positive on the left. The patient developed provoked geotropic fatigable nystagmus with the left side of the head in the down position (**Fig. 10.1**). Caloric testing revealed no abnormalities (**Fig. 10.2**).

◆ Diagnosis

Benign paroxysmal positional vertigo

◆ Medical Management

In most cases the clinical course of BPPV is self-limited, and symptoms resolve within weeks to months. Cawthorne positional exercises, Semont liberatory maneuver, and Epley's canalith repositioning procedure can be used to treat BPPV. More than 80% of patients experience resolution of their symptoms after initial treatment with a canalith repositioning procedure; and with repeated treatment more than 90% of patients have resolution of BPPV. Canalith repositioning maneuvers are the recommended modality for treatment of BPPV. Treatment of patients with vestibular suppressant medications such as antihistamines or benzodiazepines is not recommended.

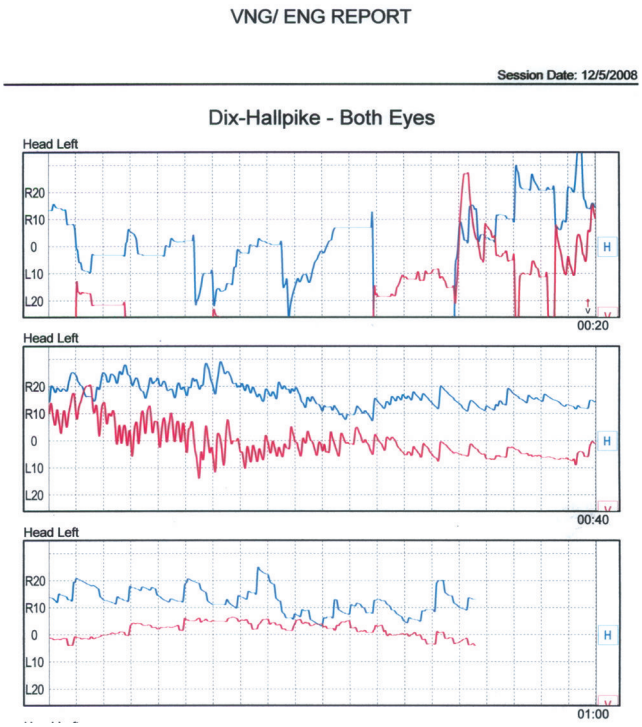


Fig. 10.1 Videonystagmography tracing shows nystagmus with left side of the head in the down position on Dix-Hallpike test.

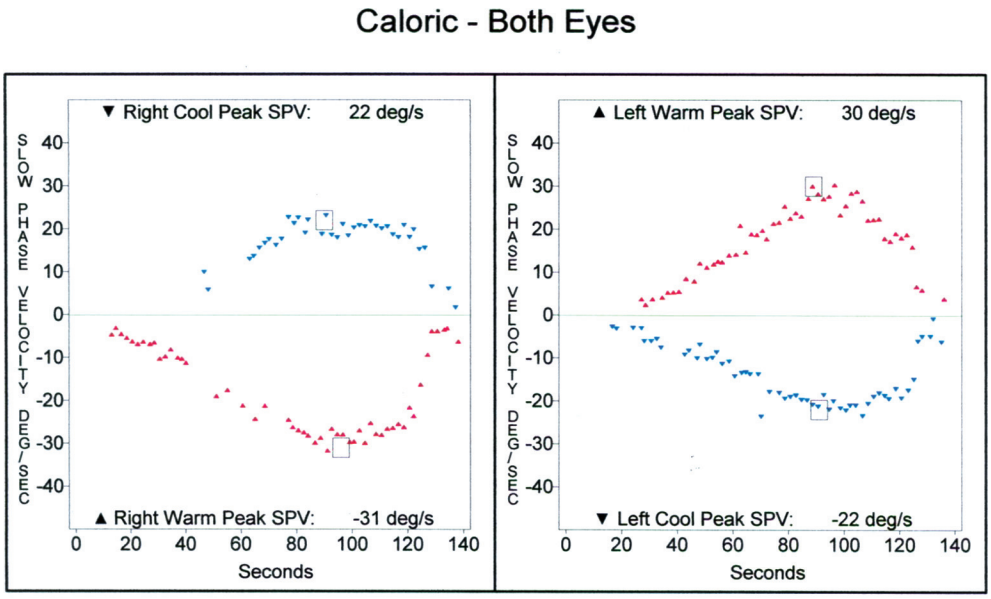


Fig. 10.2 Videonystagmography. Caloric testing. Demonstrates a normal response to warm (red) and cold (blue) irrigation. SPV, slow phase velocity.

Surgical Management

Surgical intervention is usually reserved for patients with disabling symptoms of BPPV lasting longer than a year and who did not respond to canalith repositioning procedures, medical treatment, and physical therapy. Singular neurectomy and posterior semicircular canal occlusion have been successful in management of intractable BPPV. The procedures are considered technically demanding and bear a risk of hearing loss.

A singular neurectomy technique involves a transcanal approach. The singular nerve, also known as a *posterior ampullary nerve*, is exposed inferior and medial to the round window niche and transected.

A posterior semicircular canal ampullary occlusion technique requires a mastoidectomy approach. The posterior semicircular canal is identified. It is skeletonized, and the membranous portion of the labyrinthine canal is exposed. Bone wax, fascia, or bone chips can be used to plug the posterior canal.

Rehabilitation and Follow-up

Most patients with BPPV have complete resolution of symptoms with canalolith repositioning procedures. The symptoms, however, can recur, and it is essential for patients to maintain adequate follow-up. Patients should be reassessed 1 month after treatment to check for resolution of symptoms. It is important to counsel patients about the risk for falls associated with BPPV. In cases of recurrence, patients may require a repeated canalith repositioning treatment or vestibular rehabilitation. Surgical approaches could be considered and discussed in debilitating cases of BPPV. Other peripheral or central causes of vertigo need to be considered in cases of persistent BPPV, and more testing might be required.

Questions

1. A patient complains of recurrent episodes of dizziness. The episodes are described as sudden loss of balance, usually lasting for less than a minute, and are precipitated by rapid movement. Neurotologic examination is benign except for an occasional facial twitching. Dix-Hallpike maneuvers are performed and are negative. What test should be performed next?
   A. Audiogram with acoustic reflexes
   B. Electronystagmogram
   C. Computed tomography scan of temporal bone
   D. Magnetic resonance imaging

2. A patient undergoes canalith repositioning maneuver for a diagnosis of benign paroxysmal vertigo leading to improvement in symptoms. The patient returns a few weeks later complaining of short-lasting vertigo with head movements. Dix-Hallpike maneuver is performed, and no nystagmus is provoked on either side. The supine roll test is then performed, and the patient is noted to have geotropic nystagmus on the right. What is the likely cause of vertigo?
   A. Perilymph fistula
   B. Posterior canalithiasis
   C. Lateral canalithiasis
   D. Superior canalithiasis

3. A patient has sudden onset of vertigo and hearing loss. The patient denies any trauma. Other than right beating nystagmus, the neurotologic examination is unremarkable. The audiogram confirms decreased sensorineural hearing on the right. A contrasted magnetic resonance imaging is ordered. What would be an appropriate immediate next step?
   A. Order an electronystagmogram
   B. Prescribe high-dose oral corticosteroid treatment
   C. Perform an Epley maneuver
   D. Reassure and have the patient return to clinic in 1 week

4. Immediately after a car accident a patient experiences an onset of episodic vertigo and fluctuation of hearing. He presents to the ear, nose, and throat office the following day. He notes exacerbation of symptoms by sneezing. During pneumatic otoscopy evaluation of the right ear, the patient experiences a short-lasting episode of vertigo. The remainder of the examination is unremarkable. Audiogram performed in the office shows

normal hearing. What would be a recommended treatment?

A. High dose of corticosteroids
B. Bed rest
C. Middle-ear exploration
D. Epley maneuver

Suggested Readings

Bhattacharyya N, Baugh RF, Orvidas L, et al; American Academy of Otolaryngology-Head and Neck Surgery Foundation. Clinical practice guideline: benign paroxysmal positional vertigo. Otolaryngol Head Neck Surg 2008;139(5, Suppl 4)S47–S81

Gacek RR. Pathophysiology and management of cupulolithiasis. Am J Otolaryngol 1985;6(2):66–74

Leveque M, Labrousse M, Seidermann L, Chays A. Surgical therapy in intractable benign paroxysmal positional vertigo. Otolaryngol Head Neck Surg 2007;136(5): 693–698

Schuknecht HF. Cupulolithiasis. Arch Otolaryngol 1969; 90(6):765–778

11

Perilymphatic Fistula

Collin M. Burkart and Ravi N. Samy

History

The patient, an 18-year-old white man, was admitted to the hospital because of a skiing accident in which he hit a tree. The impact was mostly to the back and left side of his head (the occiput and left parietal region). The patient was taken to the hospital emergency room via helicopter and was evaluated and admitted by the trauma service. In the course of the workup, he was found to have a mild subdural hematoma that was not expanding on serial evaluations and did not require treatment. He also had a left temporal bone fracture on a head computed tomography (CT) scan. The otolaryngology service was consulted.

On evaluation by the otolaryngology service, the patient was awake and pleasant, complaining of a mild headache, extreme dizziness, and decreased hearing in his left ear. He reported that his dizziness was a true spinning sensation; he felt nauseous when his eyes were open. There was blood and crusting in the left external auditory canal; the canal was lacerated; the tympanic membrane could not be visualized clearly. The patient had tenderness in the postauricular region.

On neurotologic examination, the patient demonstrated rightward beating nystagmus, and cranial nerves I through XII were intact except for left cranial nerve VIII. The patient had a Weber at 512 Hz that went to the right and was responsive to the Rinne at 512 Hz bilaterally with air conduction greater than bone conduction on the right side but bone conduction greater than air conduction on the left side. A Dix-Hallpike test was negative. His cerebellar function was normal on examination (finger to nose). The rest of his neurologic examination was normal and nonfocal. A high-resolution thin-cut (1.5-mm sections) CT scan of the temporal bones was ordered.

Differential Diagnosis—Key Points

1. At this point, based on the history and physical examination, there are a few possibilities about the cause of the patient's symptoms. There is a known temporal bone fracture on the left, but the extent is not well described on a head CT. Therefore, the patient may have a fracture through the labyrinth or internal auditory canal, causing the apparent sensorineural hearing loss as well as vertigo. The patient also apparently has a bloody middle-ear effusion, which could be contributing to the hearing loss. There also needs to be concern for the possibility of ossicular chain disruption. Another possibility is a perilymphatic

fistula, either isolated or in combination with the above.

2. A high-resolution thin-cut (1.5-mm sections) CT scan of the temporal bone is needed to evaluate thoroughly and precisely the lines of fracture to help in determining the diagnosis as well as the proper course of evaluation and treatment.
3. Temporal-bone fractures can be classified by a traditional anatomic system in which the fracture is designated as longitudinal, transverse, or mixed. These designations are based on the fracture line relative to the long axis of the petrous ridge of the temporal bone. Historically it has been shown that most commonly the fractures are longitudinal (~80% of the time), with the remainder being either transverse or mixed. Blows to the side of the head often result in longitudinal fractures (as with this patient), whereas blows to the back of the head often result in transverse fractures. An accepted teaching has been that longitudinal fractures are more likely to manifest with hemotympanum, rupture of the tympanic membrane, laceration of the external auditory canal, and ossicular injury resulting in a conductive hearing loss. Conversely, it has been an accepted teaching that transverse fractures are more likely to result in violation of the otic capsule resulting in sensorineural hearing loss. A more recent classification scheme has been developed based on whether the fracture does or does not invade the otic capsule. The new scheme is designed to provide improved functional and clinical prognosis. Fractures that disrupt the otic capsule will nearly uniformly result in some degree of sensorineural hearing loss, whereas fractures that do not disrupt the otic capsule are most likely to be associated with a conductive hearing loss. Also, if the otic capsule is involved, the facial nerve is much more likely to be injured. The fracture line needs to be evaluated for structures that are affected, such as the petrous internal carotid artery.
4. The management of perilymphatic fistula (particularly spontaneous ones) is one of the most controversial topics in otolaryngology. This controversy is due in large part to the nature of the disease process and the inherent difficulty and subjectivity of diagnosis. Most authorities agree on the existence of traumatic or postsurgical perilymphatic fistula, with the most controversial topics related to spontaneous development of sudden sensorineural hearing loss associated with the formation of a perilymphatic fistula. The theorized pathogenesis of perilymphatic fistula is that an abnormal communication between the inner and middle ear is formed. This can be due to traumatic fracture of the temporal bone, disruption or subluxation of the stapes footplate (more common in children due to congenital disorders), or barotrauma (either implosive phenomena where elevation of the atmospheric pressure causes inward displacement of the membranes of the inner ear and communication between the middle and inner ear or explosive phenomena where increases in intracranial pressure cause abnormal communication between the inner and middle ear). Patients with a stapes subluxation often have a mixed hearing loss, usually with a severe to profound sensorineural component and intense vertigo. The difficulty in studying perilymphatic fistula is due to the nature of determining the diagnosis. The only way a diagnosis can be determined is by exploratory tympanotomy allowing visualization of the perilymphatic fistula, which even when present may be difficult to confirm in the operating room. The natural history of a perilymphatic fistula is not known because there has not been a report of one being seen on tympanotomy and not repaired. It is thought that common locations for the development of perilymphatic fistula are the fissula antefenestram or a fissure identified between the ampulla of the posterior canal and the round window niche. Therefore, in the case of exploratory tympanotomy, these areas deserve thorough evaluation.

◆ Test Interpretation

Further testing in this scenario is warranted because of the patient's constellation of symptoms. The concerning factor here is that the patient on clinical examination is demonstrating

an apparent mixed hearing loss; however, the intense vertigo raises suspicion that this process involves more than just the middle ear.

- *Audiology*: Audiologic testing demonstrated normal hearing on the right side, with a speech reception threshold at 10 dB and a speech discrimination score of 100%. On the left side, a high-frequency severe mixed loss was found, with some preservation of the bone line (cochlear function) in the lower frequencies. Below 1500 Hz the bone line was near normal with a moderate to severe conductive loss. Overall, his speech reception threshold on the left was 65 dB, and the speech discrimination score was 64%.
- *Electronystagmography-videonystagmography*: These tests demonstrated spontaneous beating nystagmus. Calorics were not performed because of the nature of the injury to the tympanic membrane and external auditory canal.
- *Radiography*: As mentioned, a high-resolution CT scan of the temporal bones, with 1.5-mm sections and axial and coronal cuts, was performed. This revealed an anatomically described longitudinal fracture of the temporal bone that traversed anterior to the otic capsule and appeared to spare the otic capsule. The stapes and oval window niche could not be adequately visualized to determine whether subluxation had occurred as a result of soft tissue opacity in the middle ear, but there was a small air bubble in the vestibule, called a *pneumolabyrinth*. This finding was highly suggestive of stapes subluxation.

Diagnosis

Traumatic temporal bone fracture with formation of perilymphatic fistula resulting from stapes subluxation

Medical Management

The patient was treated with antiemetics and vestibulosuppressants. These medications controlled the vertiginous symptoms well enough to bring comfort to the patient. Some experts also advocate the use of systemic steroids to prevent further deterioration in sensorineural hearing. There is debate in the literature concerning the appropriate time to intervene surgically for a perilymphatic fistula. Many argue that with the clinical picture described the patient should be taken immediately to the operating room for exploration and repair. This is in an effort to save as much hearing and vestibular function as possible. Also, in this case, there is obvious concern for stapes subluxation resulting from the clinical and radiographic findings. In a situation with less clarity, it may be appropriate to follow up with the patient clinically to monitor for improvement. Vestibular suppressants should be discontinued as soon as tolerated by the patient to improve and expedite central adaptation. If during the course of monitoring, the patient has persistent vestibular symptoms or any evidence of worsening hearing loss, most authorities would agree that exploratory surgery would be warranted at that time.

Surgical Management

This patient was taken urgently to the operating room. A transcanal exploratory tympanotomy with repair of the traumatic perilymphatic fistula was performed. A posteriorly displaced incus was found, as was a posterior subluxation of the stapes footplate with perilymph leaking from the anterior portion of the footplate. The incus was removed, and the stapes footplate was gently repositioned. Once in place, temporalis fascia was harvested and placed around the stapes footplate. This fascia was cut into thin strips and then applied. Pressed Gelfoam soaked in antibiotic irrigation was placed in the middle ear.

Postoperatively, the patient demonstrated maximum conductive hearing loss across all frequencies, but low-frequency sensorineural hearing was maintained. The high-frequency loss did not improve. The vertigo resolved completely in a few days, and there were no operative complications.

Rehabilitation and Follow-up

This patient has several options concerning improving his hearing in the future. He would benefit from reexploration and placement

of an incus prosthesis or a partial ossicular reconstructive prosthesis. An alternative option would be the placement of a bone-conductive hearing aid, such as a bone-anchored hearing aid. The patient would also benefit from amplification with a traditional hearing aid. It may also benefit the patient to undergo a course of vestibular rehabilitation therapy to improve and optimize his balance functioning.

◆ Questions

1. All suspected perilymphatic fistulae (PLF) must be explored urgently. True or false?
2. Which type of scan is most commonly used to assess the presence of a fracture line or other abnormality that could cause a PLF (CT or magnetic resonance imaging)?
3. Surgical repair of PLF is guaranteed to improve the patient's hearing. True or false?

Suggested Readings

Albera R, Canale A, Lacilla M, Cavalot AL, Ferrero V. Delayed vertigo after stapes surgery. Laryngoscope 2004;114(5):860–862

Minor LB. Labyrinthine fistulae: pathobiology and management. Curr Opin Otolaryngol Head Neck Surg 2003;11(5):340–346

Sim RJ, Jardine AH, Beckenham EJ. Long-term outcome of children undergoing surgery for suspected perilymph fistula. J Laryngol Otol 2009;123(3):298–302

12

Cochlear Implant

Jedidiah J. Grisel and Ravi N. Samy

History

A 29-year-old white woman presented with hearing loss in both ears since birth. She has used hearing aids since 3 years of age. Since then, her hearing loss has slowly and progressively worsened. Even with the hearing aids, she can converse only intermittently on the telephone, primarily with family members. The patient also complains of bilateral tinnitus but denies otalgia, otorrhea, aural fullness, and headaches. There is no previous history of ototoxic medication use, ear or head trauma, noise exposure, or family history of hearing loss.

Physical examination revealed a normal head and neck examination, including otomicroscopic examination of the ear canals and tympanic membranes. The patient could not hear the Weber tuning fork examination; Rinne was positive bilaterally. The remainder of the otoneurotologic examination was within normal limits.

Differential Diagnosis—Key Points

1. This physical examination is consistent with bilateral sensorineural hearing loss (congenital). Audiometric testing is needed to define the nature and extent of this hearing loss.
2. The differential diagnosis of bilateral sensorineural hearing loss is lengthy and includes a variety of conditions: (1) hearing loss associated with systemic manifestations (infectious, immunologic, metabolic), (2) syndromic and nonsyndromic congenital hearing loss, (3) ototoxic medications, (4) autoimmune inner-ear disease, and (5) idiopathic.
3. A thorough history, physical examination, audiometric and serologic testing, and imaging studies are useful in identifying a specific cause. Although the precise cause for deafness cannot always be determined, it is helpful to classify patients into three categories of prognostic significance: (1) postlingually deafened (deaf at or after age 5 years), (2) congenitally deafened children (prelingual deafness), and (3) congenitally deafened adolescents and adults.
4. Congenital anatomic conditions that preclude successful cochlear implantation are *Michel deformity* (congenital cochlear agenesis), cochlear ossification (such as labyrinthitis ossificans after pneumococcal meningitis), and *small internal auditory canal syndrome*, in which the cochlear nerve may be congenitally deficient or absent (also known as *cochlear hypoplasia* or *aplasia*, respectively). Acquired cochlear nerve trauma (such as with temporal-bone fractures) or resected cochlear nerves-cochlear

blood supply (as with acoustic neuroma resection) are also contraindications for cochlear implant placement.

◆ Test Interpretation

For this patient (nonsyndromic, congenital hearing loss now with postlingual deafness), audiometric testing in the best-aided condition reveals a speech reception threshold (SRT) of 90 dB with discrimination of 30% and 35% in the left and right ears, respectively. A profound sensorineural hearing loss is identified bilaterally. Tympanometry reveals type A tympanograms, and acoustic reflexes are absent bilaterally. Magnetic resonance imaging (MRI) with and without contrast reveals normal intracranial anatomy with no retrocochlear lesions (such as acoustic neuromas), normal cochlear nerves, and good fluid signal in the cochleas.

◆ Diagnosis

Bilateral profound sensorineural hearing loss

◆ Medical Management

Many patients with mild to moderate sensorineural hearing loss will respond well to conventional amplification with hearing aids. In fact, criteria for cochlear implant candidacy (see below) require that hearing aids be attempted before implantation. When cochlear implantation is being considered, it is important that no active infection (either otitis externa or otitis media) is present.

◆ Surgical Management

Cochlear implantation has been clinically approved by the U.S. Food and Drug Administration (FDA) for both prelingually and postlingually deafened children and adults. Adult candidacy includes patients who are 18 years or older with bilateral severe to profound hearing loss with minimal benefit from hearing aids (sentence recognition score $<60\%$ in best-aided condition) and no medical contraindications. Pediatric candidacy includes patients 12 months or older with bilateral profound hearing loss, lack of development of auditory skills with minimal hearing aid benefit, no medical contraindications, and enrollment in an education program that stresses auditory development. For children older than 25 months, word recognition scores should generally be less than 30%. Cochlear implantation can be performed in children younger than 12 months when labyrinthitis ossificans is at risk of occurring after trauma or meningitis.

Preoperative assessment includes a thorough history and physical examination, auditory testing, radiologic evaluation, and psychosocial and educational assessment. Generally, auditory testing includes pure-tone and speech-reception thresholds, speech discrimination, and specialized audiologic testing for potential cochlear implant candidates (such as the hearing in noise test, or HINT). Radiologic evaluation includes a computed tomography (CT) scan or MRI of the temporal bone to determine mastoid pneumatization, inner-ear patency, and potential ear anomalies. From a psychosocial standpoint, it is important to determine the patient's motivation for success as well as family support. For children, it is important to formalize a multidisciplinary education plan before surgery.

A cochlear implant device consists of an external portion (sound processor) and an implanted portion (internal receiver-stimulator). The sound processor contains a microphone and a magnet with associated radiofrequency coil that connects to the implanted portion and stimulates the active electrode placed through the cochleostomy into the scala tympani of the cochlea. The commercially available implants in the United States are the Advanced Bionics (Sylmar, CA), Med-El (Innsbruck, Austria), and Cochlear Nucleus (Sydney, Australia) implant systems.

The most common method of implantation involves a postauricular mastoidectomy with facial recess to gain access to the round window. A well is prepared in the squamosa for placement of the receiver stimulator. A cochleostomy is then created anterior/inferior to the round window with placement into the scala tympani. Careful insertion of the active electrode is then performed. Meticulous attention to detail in wound closure and avoidance of infection are important because wound-related problems are the most common complications.

Additional complications of cochlear implantation include bleeding, infection, facial nerve injury, dizziness, taste changes, tinnitus, cerebrospinal fluid leaks, meningitis, and false insertion of the electrode into the hypotympanic cells. Late complications can include device failure, infection, migration, and extrusion, which may require device removal. It is recommended that pneumococcal vaccination be given to all cochlear implant recipients to reduce the risk of meningitis.

◆ Rehabilitation and Follow-up

After cochlear implantation, around 4 weeks of healing time is allowed before activating the device. When activation occurs, computer software programs are used to identify appropriate settings for the patient (also known as *mapping*). These settings are adjusted to create a map that customizes the device to the patient's specific hearing needs. Close follow-up with the audiologist is needed throughout the first year to achieve this customization. Oral rehabilitation and educational programs are crucial to successful implantation in children.

Hearing outcomes depend on many factors, including the type of deafness (prelingual or postlingual), duration of deafness, age at implantation (for children), mode of communication, socioeconomic status, motivation, family support, and others.

◆ Questions

1. Into what portion of the cochlea should the active electrode be inserted?
2. What is the most common complication of cochlear implant surgery?
3. Meningitis can cause what condition that precludes cochlear implantation?

Suggested Readings

Miyamoto RT, Kirk KI. Cochlear implants and other implantable auditory prostheses. In: Bailey BJ, Johnson JT, et al, eds. Head & Neck Surgery—Otolaryngology, 4th ed, vol 2. Philadelphia: Lippincott Williams & Wilkins; 2006:2265–2275

Nadol JB Jr, Eddington DK. Histopathology of the inner ear relevant to cochlear implantation. Adv Otorhinolaryngol. 2006;64:31–49

NIH Consensus Conference. Cochlear implants in adults and children. JAMA. 1995;274:1955–1961

III

Neurotology and the Skull Base

13

Cerebrospinal Fluid Otorrhea

Todd M. Wine and Ravi N. Samy

History

A 27-year-old woman consulted an otolaryngologist for history of recurrent right-sided otitis media not responding to antibiotics. She complained of reduced hearing in the right ear. She had mild tinnitus and aural fullness but no vertigo. She had a longstanding history of headaches that had not changed in character recently. She was morbidly obese and had type 2 diabetes mellitus. She has a surgical history of adenotonsillectomy and bilateral tympanostomy tubes as a child.

Physical examination was significant for right middle-ear effusion; Weber lateralized to the right ear at 512 Hz, Rinne + AU 512 Hz. Audiometric testing revealed mild conductive hearing loss of the right ear with 100% speech discrimination.

She underwent successful myringotomy and insertion of a pressure equalization tube (PET) in her right tympanic membrane. Subsequently, she had clear otorrhea that was treated with topical antibiotics and steroids without improvement.

She was referred to a neurotologist for probable cerebrospinal fluid (CSF) otorrhea. Examination revealed pulsating otorrhea from the right PET. Beta-2 transferrin testing was positive. The patient was taken into surgery for repair of the middle fossa encephalocele with cranial bone graft, PET removal, and fat-graft myringoplasty. Postoperatively, a lumbar drain was kept in place for five days.

Differential Diagnosis—Key Points

1. Chronic otitis media with or without cholesteatoma can cause osteitis of tegmen and progressive thinning or dehiscence resulting in CSF otorrhea (if the tympanic membrane is not intact) or middle-ear effusion.
2. CSF otorrhea is common in patients with a history of head trauma resulting in skull-base fractures. CSF otorrhea occurs in about 1 to 6% of all skull-base fractures. CSF otorrhea could present immediately at the time of injury, and when it does, resolution in 10 to 14 days is most common when bed rest, head of bed elevation, and a stool softener are used. If a patient has effusion after a temporal-bone fracture, the suspicion of CSF effusion should be high; a computed tomography (CT) scan of the temporal bones should be performed to evaluate for defects of the tegmen. If this is diagnosed after the initial 10 to 14 days, operative intervention is likely required for resolution.
3. Spontaneous CSF otorrhea or rhinorrhea can occur and is diagnosed when no specific temporal-bone abnormality or disease

precedes the otorrhea. This diagnosis has several potential causes. One congenital theory suggests that small communications exist through the tegmen as a result of aberrant embryologic development. The arachnoid granulation model suggests that some arachnoid granulations do not terminate in venous structures and blindly end within the temporal bone. Over time, the continuous pulsations of CSF in these granulations cause pressure-induced erosion of the tegmen, eventually resulting in a communication into the mastoid or middle ear. Third, another congenital theory suggests that excessive resorption of bone during pneumatization can lead to tegmen dehiscence, encephalocele formation, and subsequent CSF leakage.

4. In addition to the above-listed causes, further medical problems are increasingly recognized as being associated with CSF otorrhea and rhinorrhea. Benign intracranial hypertension should be considered with CSF otorrhea-effusion and is specifically characterized by papilledema, headache, diplopia, and pulsatile tinnitus. Radiographically, benign intracranial hypertension can be suspected when there is an empty sella, and this too has been associated with CSF otorrhea and rhinorrhea. Additionally, morbid obesity and obstructive sleep apnea have been associated with an increased incidence of CSF otorrhea.

◆ Test Interpretation

Diagnosis of CSF otorrhea is primarily by history and physical examination. Persistent clear fluid emanating from the ear is considered to be CSF until proven otherwise. Definitive diagnosis is made with laboratory and radiographic studies. Laboratory studies include immunoelectrophoretic identification of beta-2 transferrin, which is pathognomonic for CSF. This test can be invaluable, but it is not readily available in many laboratories. Glucose testing has been of little clinical value because it has a high false-positive rate.

Radiographic examination greatly helps in achieving diagnosis. A thin-cut CT scan of the temporal bones without contrast with axial and coronal views demonstrates the bony anatomy of the temporal bone, including the ossicles, tegmen, and labyrinth. It also reveals soft tissue opacification in either the mastoid or middle ear. Magnetic resonance imaging (MRI) allows differentiation of the brain tissue from other common causes of opacification, namely, cholesteatoma. CT scan with metrizamide contrast injected intrathecally can demonstrate the site of CSF leak. This can be falsely negative with either small or intermittent leaks.

Specifically, the CT scan of this patient reveals epitympanic opacification and a 5-mm defect in the tegmen tympani. Additionally, there is opacification of the right mastoid, and the mastoid cavities bilaterally reveal sclerosis and underpneumatization (**Fig. 13.1**).

MRI (not pictured) revealed opacification of the right middle ear and mastoid, as well as a partial empty sella and enlarged CSF space around the optic nerves bilaterally.

◆ Diagnosis

1. CSF otorrhea
2. Meningoencephalocele

◆ Medical Management

Although some debate exists in the literature regarding prophylactic antibiotics to prevent bacterial meningitis, most neurotologists think antibiotics do not need to be administered unless meningitis has actually occurred. Prophylactic antibiotics may select for resistant organisms if meningitis occurs. However, patients should receive a *Streptococcus pneumoniae* vaccine to reduce the risk of pneumococcal meningitis.

◆ Surgical Management

The surgical approach to a defect in the tegmen of the temporal bone may consist of either the middle fossa or transmastoid approach or a combination of the two. The location and size of the defect, patient's age, co-morbidities, and desires of patient must be considered while planning the operation. An isolated tegmen mastoideum defect can allow for a

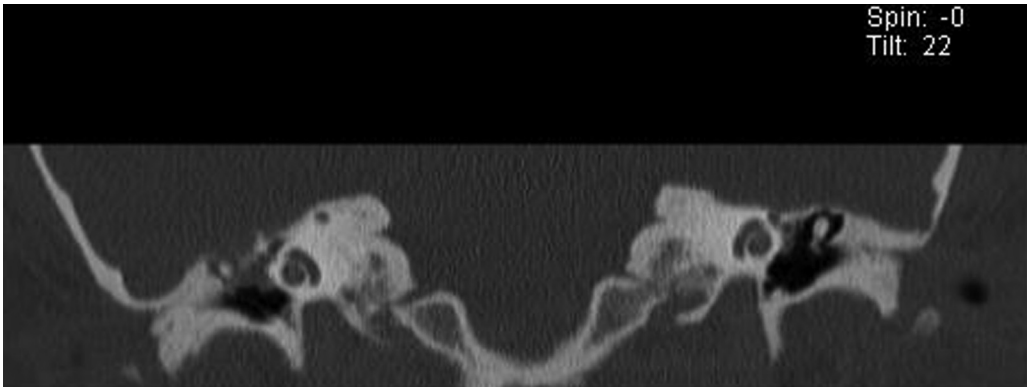


Fig. 13.1 Computed tomography of the coronal temporal bone revealing right middle-ear opacification and dehiscence of the tegmen tympani.

transmastoid approach (if the defect is single and smaller than 2 cm). Typically the larger and more numerous the defects, the more beneficial middle fossa craniotomy becomes, either alone or in combination with mastoidectomy, particularly with defects in the tegmen tympani.

After reducing the encephalocele with bipolar cautery (and resecting the nonfunctional brain tissue as needed), a multilayered closure offers the best success rate. During this approach, any encephalocele is bipolared and resected from the mastoid or middle ear. Layered closure from the middle fossa is then performed using a variety of materials, including bone, temporalis muscle (as a free graft or rotation flap), fascia, and abdominal fat. A typical repair would consist of a sandwich of fascia-bone-fascia using temporalis fascia and outer-table calvarial bone graft. Fibrin glue can be used to reinforce the repair.

If it is medically contraindicated to perform a craniotomy, a useful technique for large or multiple defects is to obliterate the mastoid, middle ear, and eustachian tube with abdominal fat. A lumbar drain is typically placed at the time of surgery and continued for up to 5 days postoperatively.

◆ Rehabilitation and Follow-up

Postoperatively, the patient should initially be monitored in the hospital for any evidence of CSF leak (via the ear, skin incision, or nose) as well as for signs and symptoms of meningitis. The head of bed should be elevated, and the patient should have an adequate bowel regimen preventing straining. At discharge, the patient should understand that decreased activity and avoidance of straining are critical for operative success. The patient can resolve normal activity 1 month postoperatively.

◆ Questions

1. What protein test can be obtained to assess for the presence of a cerebrospinal fluid leak and is considered pathognomonic?
2. What radiologic test is most commonly used to assess for the size and location of skull-base or tegmental defects?
3. True or false: The brain tissue present in an encephalocele is functional and must not be injured.

Suggested Readings

Brown NE, Grundfast KM, Jabre A, Megerian CA, O'Malley BW Jr, Rosenberg SI. Diagnosis and management of spontaneous cerebrospinal fluid-middle ear effusion and otorrhea. Laryngoscope 2004;114(5):800–805

Graham MD, Lundy LB. Dural herniation and cerebrospinal fluid leaks. In: Brackmann DE, Shelton C, Arriaga MA, eds. Otologic Surgery. Philadelphia, PA: WB Saunders Company; 2001:216–225

Gubbels SP, Selden NR, Delashaw JB Jr, McMenomey SO. Spontaneous middle fossa encephalocele and cerebrospinal fluid leakage: diagnosis and management. Otol Neurotol 2007;28(8):1131–1139

LeVay AJ, Kveton JF. Relationship between obesity, obstructive sleep apnea, and spontaneous cerebrospinal fluid otorrhea. Laryngoscope 2008;118(2):275–278

Nahas Z, Tatlipinar A, Limb CJ, Francis HW. Spontaneous meningoencephalocele of the temporal bone: clinical spectrum and presentation. Arch Otolaryngol Head Neck Surg 2008;134(5):509–518

Prichard CN, Isaacson B, Oghalai JS, Coker NJ, Vrabec JT. Adult spontaneous CSF otorrhea: correlation with radiographic empty sella. Otolaryngol Head Neck Surg 2006;134(5):767–771

Rao AK, Merenda DM, Wetmore SJ. Diagnosis and management of spontaneous cerebrospinal fluid otorrhea. Otol Neurotol 2005;26(6):1171–1175

Savva A, Taylor MJ, Beatty CW. Management of cerebrospinal fluid leaks involving the temporal bone: report on 92 patients. Laryngoscope 2003;113(1):50–56

14

Pulsatile Tinnitus

Melissa McCarty Statham and Ravi N. Samy

History

A 63-year-old white man seeks a second opinion with a complaint of constant left-sided pulsatile tinnitus for about 1 year. He states that it is progressive and is 10 of 10 in severity. He also reports rare left-sided otalgia. Otherwise, he denies vertigo, aural fullness, hearing loss, or otorrhea. His otologic history is significant for a history of left-sided auricular trauma during his childhood, although he cannot remember the incident well because of his young age at the time. He denies any significant history of otitis media. He reports a history of noise exposure from riding motorcycles. His family history is significant for his father dying of a ruptured intracranial aneurysm.

On neuro-otologic examination, Weber examination is midline, Rinne examination is positive bilaterally, examination of the pinnae is normal, and microscopic examination reveals normal mobile tympanic membranes bilaterally. Compression of the ipsilateral jugular vein and provocative head movement fail to abate the patient's tinnitus. Auscultation with a Toynbee tube fails to demonstrate audible pulsatile tinnitus. Examination of the cranial nerves (CNs) reveals normal function of CN II though XII.

Audiometric testing reveals normal hearing acuity and normal tympanometrics. Outside high-resolution computed tomography (CT) of the temporal bone shows no intratemporal tumors or other abnormalities. A carotid artery duplex study demonstrates mild irregularities of bilateral carotid systems without any hemodynamically significant stenosis.

Differential Diagnosis—Key Points

1. Vascular anomalies of the middle ear: Vascular anomalies in the middle ear often present with pulsatile tinnitus. An aberrant or laterally displaced petrous carotid artery, a dehiscent or high-riding jugular bulb, a persistent stapedial artery, and an intratympanic carotid aneurysm represent the most common types of vascular anomalies.

   To diagnose an aberrant carotid artery, high-resolution CT reveals the petrous carotid artery entering the middle ear through an enlarged inferior tympanic canaliculus, an enhancing hypotympanic "mass," and the carotid foramen and vertical segment of the petrous internal carotid artery are absent. Accurate radiographic diagnosis is imperative because this "mass" should never be biopsied. A persistent stapedial artery is usually noted with an enlarged anterior tympanic segment of the fallopian canal and an absent foramen spinosum on high-resolution CT scan. This entity may be

associated with an aberrant petrous carotid artery.

A high-riding jugular bulb is the most common vascular anomaly of the middle ear and is easily seen on CT scan of the temporal bone. It is defined as a jugular bulb extending above the inferior tympanic annulus. On otoscopic examination, this is seen as a blue-hued mass protruding from the hypotympanum. High-resolution CT scan reveals a focal absence of the sigmoid plate, and the superolateral jugular bulb appears to enter the middle ear as a mass. Accurate radiographic diagnosis is also imperative because this "lesion" should never be biopsied.

2. Idiopathic intracranial hypertension: Idiopathic intracranial hypertension (IIH; or *pseudotumor cerebri* or *benign intracranial hypertension*) often presents with retroocular and pulsatile headache, pulsatile tinnitus, papilledema, and elevated intracranial pressure. This process is most prevalent in obese women of childbearing age. If left untreated, IIH may progress to permanent visual field deficits and blindness. The associated pulsatile tinnitus is thought to derive from turbulent venous flow in the transverse and sigmoid sinuses. Diagnosis is confirmed with elevated cerebrospinal fluid pressure (demonstrated with lumbar puncture) and radiographic imaging.
3. Venous etiology: Venous hum tinnitus can be demonstrated in patients with a tortuous sigmoid sinus or a previous history of sigmoid sinus thrombosis and subsequent recanalization. Diagnosis is confirmed with elimination of the tinnitus with gentle jugular-vein compression or by turning the patient's head to the contralateral side.
4. Palatal myoclonus: Palatal myoclonus is caused by contraction of tensor palatini, levator veli palatini, tensor tympani, salpinopharyngeal, or superior constrictor muscles. It is described as a clicking sound that is rapid (up to 60 to 200 beats per minute), repetitive, and intermittent. The tinnitus does not dissipate while the patient is asleep. The muscle spasms may be seen either transorally or transnasally. Because of the rapidity of the clicking, this entity does not typically have a "heartbeat" or pulsatile nature.
5. Jugulotympanic paraganglioma: These represent a common cause of pulsatile tinnitus. Glomus bodies are neural crest derivatives that reside along the course of the Jacobson and Arnold nerves and in the region of the jugular bulb. Paragangliomas are thought to arise from these glomus bodies and migrate in close association with sympathetic ganglia. These highly vascular masses belong to the diffuse neuroendocrine system and are composed of chief cells and oxyphil cells. Paragangliomas usually grow slowly and may be present for several years before causing symptoms.

   In addition to pulsatile tinnitus, associated symptoms are usually based on location and nearby involved anatomic structures. Involvement of the middle ear can lead to conductive hearing loss, aural bleeding, and Brown sign (blanching of the lesion with positive pressure). Symptoms suggestive of tumor extension into associated structures include vertigo, sensorineural hearing loss, otalgia, and lower cranial neuropathy (CNs IX–XII).
6. Dural arteriovenous malformation (AVM): Dural AVM, or dural arteriovenous fistula, is one of the most common anomalies identified in patients with pulsatile tinnitus. In these vascular anomalies, an arteriovenous shunt is contained within the leaflets of the dura mater. The arterial supply is exclusively supplied by branches of the carotid or vertebral arteries before they penetrate the dura mater. A dural AVM may represent a true fistula, which is defined as a single or multiple dilated arterioles that connect directly to a vein without a nidus, and they are high-pressure and high-flow. Most commonly, these are found adjacent to a dural venous sinus, with a slight left-sided predominance. The junction of the transverse and sigmoid sinuses is the most common location.

   Evidence suggests that dural AVMs of the transverse–sigmoid junction are acquired, not congenital, lesions that result from collateral revascularization after a previous thrombosis of the involved dural venous sinus. This is most commonly seen from sigmoid sinus thrombosis secondary to infection or trauma. In most cases, the occipital artery is the dominant arterial feeding vessel (**Table 14.1**).

Table 14.1 Clinical Symptoms Reported in 27 Cases of Dural Arteriovenous Malformations

Symptom	n (%)
Pulsate tinnitus	25 (92%)
Occipital bruit	24 (89%)
Headache	11 (41%)
Visual impairment	9 (33%)
Papilledema	7 (26%)

Source: From Sundt TM Jr, Piepgras DG. The surgical approach to arteriovenous malformations of the lateral and sigmoid dural sinuses. J Neurosurg 1983;59:32–39.

Depending on the direction of flow, these lesions may be life-threatening, and complete evaluation for pulsatile tinnitus is warranted. Physical examination may reveal a loud bruit that may be auscultated over the mastoid or occiput. Dural AVMs are most commonly diagnosed via a combination of contrast-enhanced magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA).

◆ Test Interpretation

Investigating the cause of pulsatile tinnitus is necessary because potentially life-threatening pathology may be present. Pure-tone audiometry and speech audiometry are necessary tests, and tympanometry may objectively demonstrate pulsations. Pulsatile tinnitus is frequently associated with a low-frequency hearing loss, which is thought to be artifactual. Otic capsule and temporal bone invasion by paraganglioma will likely lead to a sensorineural hearing loss. If paraganglioma is suspected, evaluation of catecholamine secretion is imperative.

Complete blood count with differential: Anemia with an associated increased cardiac output can result in a transmitted pulsatile tinnitus.

Thyroid function studies: Hyperthyroid states may present with pulsatile tinnitus, along with the other constellation of symptoms associated with the disease state, which is resultant from increased cardiac output.

Radiographic evaluation: High-resolution CT scan of the temporal bone demonstrates bony anatomy of the cranial base foramina and will delineate vascular anomalies of the middle ear and cranial base (otosclerosis and superior canal dehiscence have also been rarely reported to cause pulsatile tinnitus). MRI, with and without gadolinium, and MRA are useful together to identify paraganglioma and dural AVMs.

4-Vessel angiography: This study is useful in delineating dural AVMs and for definitive or preoperative embolization of these lesions or paraganglioma (**Fig. 14.1**).

◆ Diagnosis

Dural AVM

◆ Medical Management

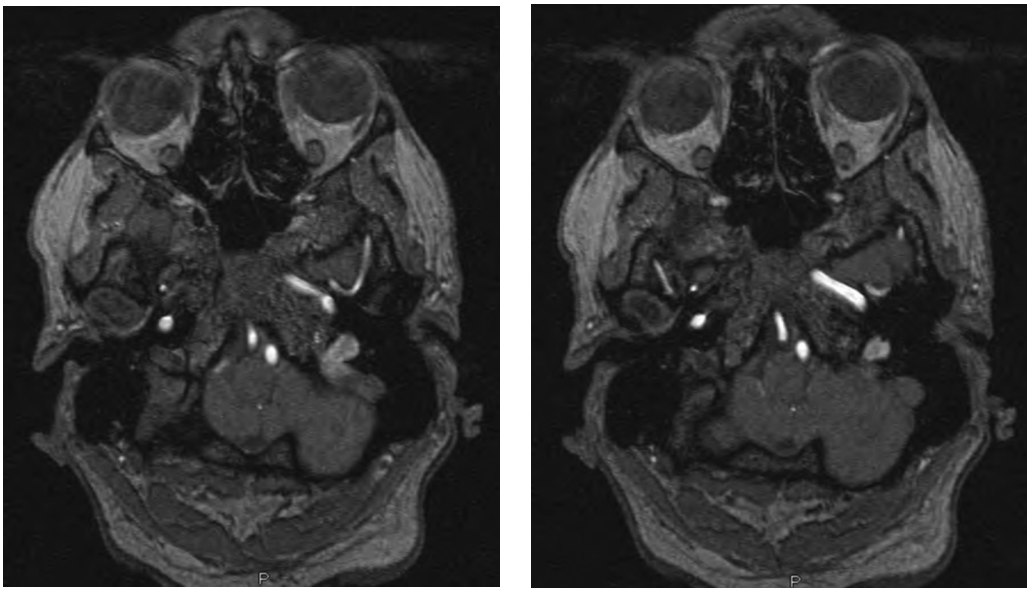
Dural AVMs may be observed if they are small and the patient's symptoms are amenable. Therapy for dural AVMs is usually endovascular embolization with or without microneurosurgical resection, but stereotactic radiosurgery has been described as a viable treatment in patients with benign AVM that has failed other treatments. Radiosurgery is contraindicated in large dural AVMs or in those at risk of hemorrhage.

◆ Surgical Management

Indications for treatment of a dural AVM include neurologic dysfunction, bleeding from the lesion, or intractable symptoms. Referral to neurosurgical colleagues is warranted for evaluation for possible treatment or expectant management. Endovascular embolization may be performed via transarterial or transvenous access. Preoperative embolization usually facilitates surgical intervention because blood loss can be rapid and extensive.

◆ Rehabilitation and Follow-up

Patients with dural AVM will need to be followed up if expectant management is chosen. In addition, serial imaging will be required of patients posttreatment to assess for recurrence or need for further treatment.



A B

Fig. 14.1 (A,B) Axial T1-weighted postcontrast magnetic resonance images demonstrate a high-flow left dural arteriovenous fistula at the level of the distal sigmoid sinus-jugular bulb region, with arterial supply from transosseous arterial vessels in the region of the left jugular tubercle and occipital condyle. Note the high-contrast uptake in the left jugular bulb and sigmoid sinus junction compared with the right side.

◆ Questions

1. True or false? The best way to diagnose a vascular middle-ear lesion is by biopsy.
2. Idiopathic intracranial hypertension is most commonly seen in what group of individuals?
3. Name the two methods to treat dural arteriovenous malformations.

Suggested Readings

Greenberg SF. Handbook of Neurosurgery, 6th ed. New York, NY: Thieme; 2006:843–844

Shiley SG, Folmer RL, McMenomey SO. Tinnitus and hyperacusis. In: Cummings CW, ed. Otolaryngology—Head and Neck Surgery, 4th ed. Philadelphia, PA: Elsevier Mosby; 2005

Sundt TM Jr, Piepgras DG. The surgical approach to arteriovenous malformations of the lateral and sigmoid dural sinuses. J Neurosurg 1983;59(1):32–39

15

Acoustic Neuroma

Randal Leung and Ravi N. Samy

History

A 57-year-old man describes a 12-month history of slowly progressive right-sided hearing loss. Associated symptoms include unilateral right tinnitus, mild sense of aural fullness, disequilibrium without vertigo, and occasional morning headaches. The remaining history is noncontributory.

Physical examination reveals a well-developed, well-nourished man in no acute distress. Both external auditory canals and tympanic membranes are normal. Facial nerve examination demonstrates House-Brackman grade I to VI function bilaterally. Tuning fork testing reveals a Weber lateralizing to the left side with bilaterally positive Rinne tests. Head thrust test was positive for a corrective saccade on the right; however, Romberg and Fukuda tests were normal. Corneal hypesthesia was noted with use of a cotton-tipped applicator on the right (i.e., diminished corneal reflex). The remaining otoneurologic examination was normal.

Differential Diagnosis—Key Points

1. Differential diagnosis would include endolymphatic hydrops, an episode of labyrinthitis, or a cerebellopontine angle (CPA) lesion (such as an acoustic neuroma, meningioma, or facial neuroma); a malignancy causing such symptoms would be rare. Many times, after an exhaustive search and workup, the cause of such symptoms remains unknown or idiopathic. If bilateral symptoms had occurred, autoimmune inner-ear disease would also be included in the differential diagnosis.
2. Endolymphatic hydrops typically presents with symptoms of fluctuating hearing loss, tinnitus, aural fullness, and episodes of vertigo lasting 20 minutes to hours. However, patients may present initially without the classic history; only after observing the evolution of symptoms over time, the patient's response to initial medical therapy, and excluding CPA lesions may the diagnosis become evident.
3. The head-thrust test is a useful and simple bedside test of the vestibular ocular reflex in the horizontal plane and is positive in instances of a unilateral vestibular weakness. Small differences between sides might not be seen clinically when observing for the corrective saccade after a rapid lateral head turn. However, if measured by electronystagmography, even small differences will be noted with this test.
4. Vestibular function tests are useful to determine the residual peripheral vestibular

function on the affected side. This is helpful when counseling patients regarding their expected vestibular recovery and prognosis following treatment. Other than differentiating a pure peripheral or central vestibular insult, it does not give any further differentiation in terms of diagnosis. The use of vestibular function tests and vestibular evoked myogenic potentials may provide an indication as to whether an acoustic neuroma is arising from the superior or inferior vestibular nerve; however, this is of little practical importance.

5. Audiometric testing in cases of retrocochlear lesions will typically display speech rollover if a speech audiogram is performed.

Test Interpretation

An audiogram is performed and reveals a mild high-frequency sensorineural hearing loss in the left ear and a mild to severe sensorineural hearing loss in the right ear. Acoustic reflexes were absent in both ears. Speech discrimination was 100% at 25 dB on the left; however, there was no word recognition on the right at either the expected speech reception threshold or a suprathreshold presentation level. Gadolinium-enhanced magnetic resonance imaging scanning is then undertaken and reveals an enhancing right CPA mass that extends into the internal auditory canal and is producing a mild mass effect on the adjacent cerebellum and pons (**Fig. 15.1**).

Diagnosis

Acoustic neuroma

Medical Management

Management options for acoustic neuroma include observation, radiation therapy, and surgical extirpation. When counseling patients on treatment options and recommendations, factors that come into consideration include the patient's overall health and co-morbidities, age, residual hearing level, initial tumor size on presentation, location of tumor, presence of brainstem compression, treatment risks, and patient wishes.

Observation of acoustic neuromas is generally not recommended for large tumors already displaying brainstem compression or those with more progressive and acute presentations that suggest rapid growth. A history of malignancy also raises the possibility that the CPA lesion may represent a metastasis. Observation protocols are somewhat controversial but all involve serial MRIs and close clinical follow-up.

Stereotactic radiosurgery is aimed at halting growth of the tumor. It is a valid option for smaller tumors (those without brainstem compression), older patients, and those unable to tolerate surgery; patients should be adequately counseled with respect to the risks and side effects of this treatment, including consultation with a radiation oncologist, if the patient desires. Specific mention should

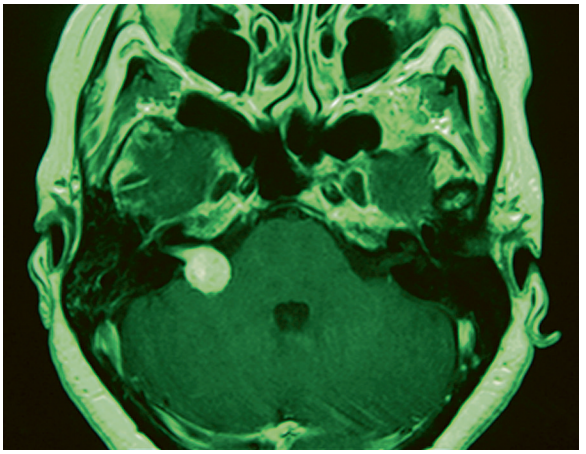


Fig. 15.1 Right cerebellopontine-angle tumor with internal auditory canal extension and mass effect on the cerebellum and pons.

be made of hearing loss, vertigo, facial nerve dysfunction, delayed tumor growth, radiation-induced malignancy, radiation failures, and a greater risk of facial nerve paralysis and hearing loss if surgical intervention is required in the future for radiation failures. If the presenting tumor is large, has large cystic components, or has evidence of brainstem compression, then it is generally not recommended because of acute postradiotherapy swelling worsening any existing compression.

◆ Surgical Management

Surgery is the mainstay of treatment for acoustic neuromas, particularly with younger, healthy patients and large tumors. Surgical approach options for acoustic neuromas include retrosigmoid, translabyrinthine, and middle fossa routes. The translabyrinthine approach is the most commonly used approach for resection of CPA tumors. However, the retrosigmoid and middle fossa routes are the only ones capable of hearing preservation. The choice between these is based primarily on tumor size, location, and residual hearing function. The general health of the patient and surgical expertise of the treating team are also of importance.

If the patient elects for surgical intervention, then because of the significantly lowered sensorineural thresholds, together with absent speech discrimination, a translabyrinthine approach is warranted to reduce any retraction on the cerebellum as well as to allow for early identification of the facial nerve in large tumors.

A translabyrinthine dissection is undertaken, and the initial mastoidectomy identifies the sigmoid sinus, sinodural angle, tegmen, facial nerve, and semicircular canals. Complete removal of bone overlying the sigmoid sinus, posterior fossa, and middle fossa is undertaken; the superior petrosal sinus is identified. Labyrinthectomy is then performed, and the completion of bone removal delineates the internal auditory canal from the fundus to porus acousticus. Active stimulation of the facial nerve in the lateral part of the internal auditory canal confirms the visual verification of the nerve and allows dissection of the tumor from the lateral portion of the internal auditory canal with sectioning of the vestibular and auditory nerves. Dissection of the tumor progresses to the porus, with attention then turned to the CPA with opening of the dura. Meticulous dissection of the tumor from the facial nerve and adjacent cranial nerve is undertaken, facilitated by active stimulation of the facial nerve to identify the nerve as its course deviates over the tumor surface. Total tumor removal is accomplished, and the facial nerve is preserved.

◆ Rehabilitation and Follow-up

Strict eye care is mandatory following any treatment for acoustic neuroma. If there is evidence of poor eye closure and corneal exposure, eye lubricants, ointments, and moisture chambers are used to prevent drying and inadvertent injury to the cornea. Temporary measures to facilitate eye closure include gold weight or lateral tarsorrhapy while waiting for future return of function. Future measures for facial reanimation may include static or dynamic procedures as indicated in the individual case.

◆ Questions

1. What surgical approach is most commonly used to resect cerebellopontine angle tumors, including acoustic neuromas?
2. What surgical approaches can be used for hearing preservation in acoustic neuromas, depending on size and location of the tumor?
3. Acoustic neuromas arise from cranial nerve VIII. What cranial nerve is the second most commonly affected cranial nerve with these tumors (preoperatively)?

Suggested Readings

Battaglia A, Mastrodimos B, Cueva R. Comparison of growth patterns of acoustic neuromas with and without radiosurgery. Otol Neurotol 2006;27(5):705–712

Bennett M, Haynes DS. Surgical approaches and complications in the removal of vestibular schwannomas. Otolaryngol Clin North Am 2007;40(3):589–609, ix–x

Doherty JK, Friedman RA. Controversies in building a management algorithm for vestibular schwannomas. Curr Opin Otolaryngol Head Neck Surg 2006;14(5):305–313

16

Petroclival Meningioma

Maria V. Suurna and Myles L. Pensak

History

A 42-year-old woman is referred to the office with a 6-month history of progressive gait and postural instability. The patient complains of intermittent left facial hyperesthesia and pain. She recently developed left occipital headaches. There is no history of head trauma or recent infections. The remainder of her medical history is unremarkable.

Her examination reveals a well-appearing, slightly anxious woman in no distress. The external auditory canals and tympanic membranes are normal. The head and neck examination shows no masses or lesions. On neurologic examination she is noted to have marked unsteadiness; an ataxic gait; and an inability to perform rapid axial, rotary, or translational movements. The patient has a positive Fukuda test; however, the Dix-Hallpike maneuver with Frenzel glasses reveals no evidence of spontaneous or positional nystagmus. Gross motor and sensory function is intact. The cranial nerve examination reveals reduced sensation in the V2 and V3 distribution on the left side.

Differential Diagnosis—Key Points

Skull-base lesions can manifest with a variety of symptoms, depending on their size and location. Patients with lesions involving the petrous apex may manifest with any combination of symptoms, the most common being decreased hearing, tinnitus, vestibular dysfunction, balance problems, headache, facial paralysis, facial pain, and facial paresthesia or hyperesthesia. Cranial nerves V, VII, and VIII are most commonly involved.

1. After acoustic neuroma, meningioma is the second most common tumor in the cerebellopontine angle (CPA). Meningiomas are locally invasive benign neoplasms. Patients with petroclival meningiomas most commonly manifest with complaints of facial pain, headache, balance disturbance, and decreased hearing.
2. Acoustic neuroma is the most common neoplasm of the CPA. This is a benign tumor that originates most commonly on the vestibular portion of the eighth nerve. Depending on the size of the tumor, the patients may have hearing loss, balance dysfunction, tinnitus, and signs of brainstem compression.
3. Other neoplasms of CPA, such as trigeminal neuroma, paraganglioma, chordoma, chondrosarcoma, endolymphatic sac tumor, nasopharyngeal carcinoma, or metastatic lesion, should be considered.
4. Metastatic neoplasms should be in the differential diagnosis for older patients, with the petrous apex being the most common

site for metastasis within the temporal bone. Breast, lung, prostate, melanoma, and malignant neoplasms of the kidney were the most common to metastasize to the temporal bone.

5. Cholesterol granuloma is the most common petrous apex lesion and is thought to form as a result of either hemorrhage into the air cells of the petrous apex and a subsequent breakdown of the red blood cells and inflammatory reaction or inspissation of trapped apical air-cell mucus.
6. Cholesteatoma (epidermoid cyst) of the petrous apex can be acquired or congenital. Acquired cholesteatoma usually results from the middle-ear disease in which epithelial cells gain access to the petrous apex. With cholesteatoma expansion, bone erosion and injury to the surrounding structures may occur.
7. Mucocele forms as a result of obstruction of mucus-secreting cells within a pneumatized petrous apex. The expansion of this lesion may lead to bone erosion.
8. Effusion or trapped fluid of the petrous apex is thought to result from infection of the middle ear or mastoid.
9. Petrous apisitis results from the spread of infection toward the petrous apex in patients with acute otitis media. With wide use of antibiotics this pathology is rarely encountered. In severe cases patients may show a combination of symptoms known as Gradenigo syndrome, which includes retro-orbital pain, otorrhea, and lateral gaze palsy resulting from inflammation of cranial nerve VI.
10. Arachnoid cysts arise from the arachnoid membrane and are filled with cerebrospinal fluid. The cysts may lead to erosion of petrous apex and cause compression of the cranial nerves, brainstem, or cerebellum.

◆ Test Interpretation

The initial evaluation of the patient with petroclival meningioma will include obtaining a baseline audiogram with tympanometry and acoustic reflexes. An electronystagmogram (ENG) allows for better assessment of vestibular function. An auditory brain response test can be useful for an initial evaluation of suspected retrocochlear pathology.

Imaging is very useful in evaluating and differentiating the petrous apex pathology. Magnetic resonance imaging (MRI) with gadolinium is the preferred imaging study when evaluating meningiomas. Meningiomas are usually isointense or slightly hypointense to the adjacent gray matter on T1-weighted images and have variable intensity on T2-weighted images. They enhance on gadolinium-enhanced T1-weighted images. The lesions tend to have a broad-based shape and presence of a dural tail.

Computed tomography (CT) scan images of the temporal bone allow for evaluation of the extension of the petrous apex lesions and are useful in assessment of invasion of the surrounding bony structures. It is difficult to differentiate the petrous apex lesions based on a CT study. The CT scan can be used in conjunction with MRI

In this case, on audiogram the patient has normal hearing and tympanometry. The ENG demonstrates right beating positional nystagmus. The bithermal caloric testing showed a 32% left-sided weakness. The auditory brain response test reveals a prolongation of a wave V on the left side. MRI demonstrates a CPA mass. It is isointense to the adjacent gray matter on T1- and T2-weighted images, and postcontrast imaging reveals intense homogeneous enhancement of the lesion (**Fig. 16.1**).

◆ Diagnosis

Petroclival meningioma

◆ Medical Management

The management plan should be based on the patient's medical condition, age, symptoms, and the location and size of the tumor. In cases of small tumors, observation is generally recommended. Radiation therapy may be used as a primary intervention or supplemental in cases of surgical subtotal cytoreductive treatment. Stereotactic radiosurgery has replaced conventional radiation. This treatment is generally not used for large tumors.

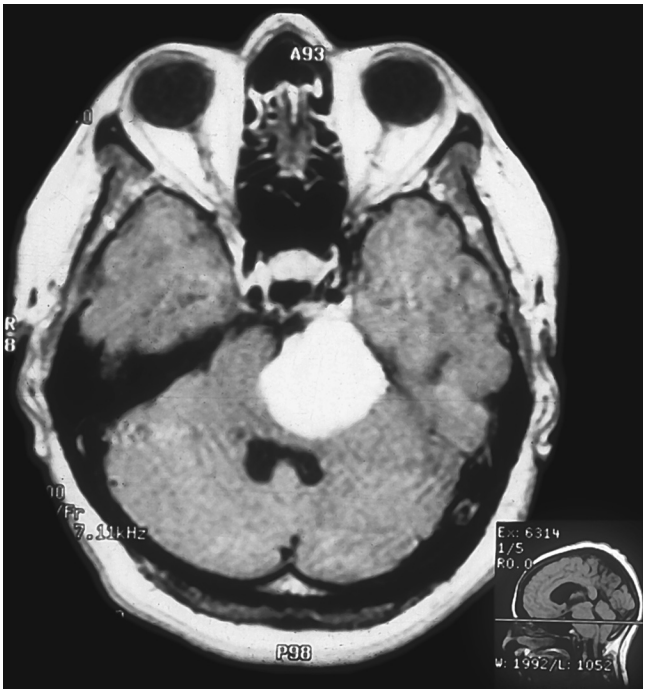


Fig. 16.1 Petroclival meningioma shown on postcontrast T1-weighted image

Surgical Management

Currently several surgical approaches to the petroclival region allow for optimal complete or subtotal excision of petroclival meningiomas. Suboccipital, extended middle fossa, translabyrinthine, transcochlear, or transpetrosal approaches that allow access for removal of petroclival meningiomas. The choice of the approach is guided by the location, size of the tumor, and the surgical team's experience.

In this case the patient underwent petroclival meningioma excision using a transpetrosal approach. She recovered well from the procedure and had no neurotologic complications. Her gait and postural stability have significantly improved following the procedure.

Rehabilitation and Follow-Up

Meningiomas are classified into three World Health Organization grades based on the lesion's histopathology and likelihood of recurrence, with grade I lesions having a low rate of recurrence after complete excision and grade III lesions having the highest recurrence rate. Meningiomas are slow growing tumors, and a long-term follow-up with repeat MRI imaging is necessary to surveillance for a recurrence. In cases of residual tumor, radiation treatment might be considered. Depending on the neurologic impairment of the patient resulting from the disease or treatment, audiologic, facial nerve, vestibular, or ophthalmologic rehabilitation may be necessary.

Questions

1. On magnetic resonance imaging (MRI) the patient was found to have a lesion in the petrous apex. The lesion was found to be hyperintense on T1- and T2-weighted images. No postcontrast enhancement of the lesion was seen. What is the most likely diagnosis?

A. Acoustic neuroma
B. Cholesteatoma
C. Cholesterol granuloma
D. Meningioma

2. A patient has progressive symptoms of facial pain and recent onset of disequilibrium. What is the best study to differentiation of the underlying pathology?
   A. Auditory brainstem response
   B. Audiogram
   C. Computed tomography with contrast
   D. MRI with gadolinium
3. Following meningioma removal using a translabyrinthine approach, what is the most common immediate postoperative complication?
   A. Cerebrospinal fluid leak
   B. Facial nerve paralysis
   C. Hematoma
   D. Vertigo
4. What cerebellopontine-angle neoplasm is likely to manifest with an isolated unilateral hearing loss?
   A. Acoustic neuroma
   B. Metastatic neoplasm
   C. Paraganglioma
   D. Petroclival meningioma

Suggested Readings

Abdel Aziz KM, Sanan A, van Loveren HR, Tew JM Jr, Keller JT, Pensak ML. Petroclival meningiomas: predictive parameters for transpetrosal approaches. Neurosurgery 2000;47(1):139–150

Isaacson B, Kutz JW, Roland PS. Lesions of the petrous apex: diagnosis and management. Otolaryngol Clin North Am 2007;40(3):479–519

Park CK, Jung HW, Kim JE, Paek SH, Kim DG. The selection of the optimal therapeutic strategy for petroclival meningiomas. Surg Neurol 2006;66(2):160–165

Pensak ML, Van Loveren H, Tew JM Jr, Keith RW. Transpetrosal access to meningiomas juxtaposing the temporal bone. Laryngoscope 1994;104(7):814–820

17

Temporal Bone Fracture

Abdul-Aleem A. Kadar and Ravi N. Samy

History

A 21-year-old skier who was not wearing a helmet struck a tree and suffered a loss of consciousness. On arrival to the emergency department, the patient was neurologically intact but reported bilateral hearing loss, vertigo, tinnitus, and a headache. Physical examination reveals serosanguineous otorrhea from the right ear with a small, posterior perforation and left hemotympanum. Mastoid ecchymoses are noted bilaterally. There was no response to tuning forks; the patient was unable to communicate except via writing. The facial nerve was graded as a House-Brackmann grade I/VI bilaterally. His neurologic examination was nonfocal except for right-beating nystagmus. The remainder of his head and neck examination was unremarkable.

Differential Diagnosis—Key Points

1. Temporal bone fracture (TBF) is a fracture of the skull base that accounts for 75% of skull-base fractures in adults. It is usually a part of multisystem trauma and often requires multispecialty attention, particularly from a neurologic standpoint evaluating for epidural or subdural hematomas, temporal-lobe contusions, or other facial injuries.
2. Traditionally TBF is classified as longitudinal, transverse, or mixed (a fracture with significant comminution or fracture lines running in both longitudinal and transverse directions is termed a *mixed fracture*). Although this system is still widely used, it suffers descriptive limitations and may not accurately predict the presence of sequelae, such as profound hearing loss or cerebrospinal fluid (CSF) leakage. One alternative method of classifying TBF is the otic capsule–based system (which describes the fracture as involving or sparing the otic capsule).
3. Using the traditional classification system, longitudinal fractures are the most common (80%) and are frequently caused by a lateral blow to the skull in the parietal or temporal regions. The fracture typically runs in a coronal plane and extends from the squamous portion of the temporal bone along the anterior portion of the petrous apex to the region of the foramen lacerum and foramen ovale, passing through the posterior-superior aspect of the external auditory canal. Because of its laterality and direction, the otic capsule is preserved, as is the internal auditory canal. The hearing loss is usually conductive and results from tympanic membrane perforation, hemotympanum, or disruption of the ossicular chain. Sensorineural

hearing loss can occur but typically is not profound or complete.

4. Transverse fractures are less common (20%) and are caused by intense blows to the occipital region or by a direct frontal injury. The injury usually runs perpendicular to the petrous ridge from the foramen magnum across the petrous pyramid, often including the internal auditory canal and into the foramen spinosum. Fractures across the internal auditory canal and labyrinth result in profound anacusis and immediate debilitating vertigo. The incidence of facial paralysis may be as high as 50%.
5. Patients who have suffered TBF warrant a thorough otologic and neurotologic assessment. The external auditory canals are assessed for skin lacerations and fracture lines (with attention to prevention of stenosis occurring). Any CSF leak should be closely looked for and noted. The patient should also be assessed for CSF rhinorrhea. Ophthalmologic evaluation examines for full ocular movement, visual acuity, and the presence of nystagmus. Early and full assessment of facial nerve function, although problematic in obtunded patients, is of paramount importance.
6. After full neurologic assessment and once the patient is stable, hearing assessment should be done to obtain an audiometric profile and establish bone-conduction threshold levels. Electronystagmography and rotation-chair assessment may help in assessing labyrinthine function but have no clinical relevance in acute labyrinthine injury.

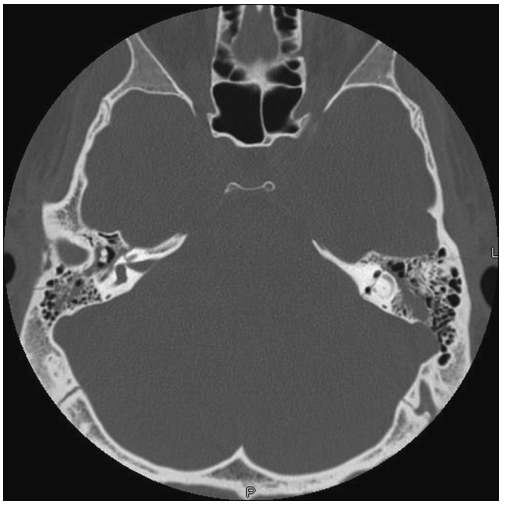


Fig. 17.1 Noncontrast axial computed tomography scan of the temporal bones. Blood is noted in the mastoids bilaterally. The fracture on the right involves the mastoid. The fracture on the left involves the superior semicircular canal with air in the canal.

Test Interpretation

1. The pure-tone audiogram shows a complete absence of response bilaterally.
2. Computed tomography scan shows TBFs extending through the otic capsules bilaterally (**Figs. 17.1 and 17.2**); pneumolabyrinth is noted on the left.

Diagnosis

Bilateral TBFs (mixed, otic capsule involving)

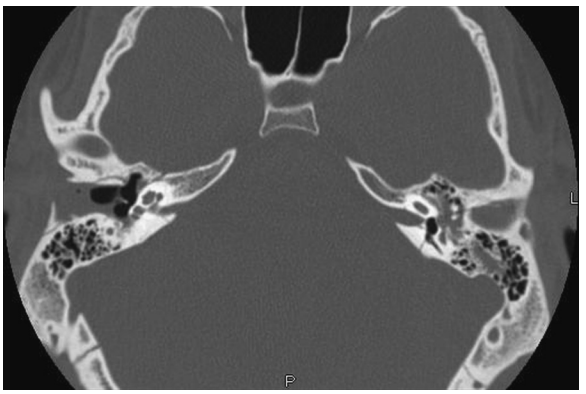


Fig. 17.2 Computed tomography image inferior to Figure 17.1. The right side shows an anterior external auditory canal wall fracture as well as a fracture posterior to the internal auditory canal. On the left side, the prior fracture and pneumolabyrinth noted on Figure 17.1 are shown to be continuing inferiorly to the level of the vestibule and posterior semicircular canal.

Medical Management

1. Conservative treatment is used for most patients with TBF; however, neurologic complications associated with TBF, such as epidural and subdural hematomas, petrous internal carotid artery injury, and hydrocephalus, need to be addressed urgently.
2. Bleeding from the external auditory canal is a usual feature; packing with a wick and using dry-ear precautions and otic drops are important to reduce the chance of stenosis.
3. Acute vestibulopathy in TBF patients may be due to direct otic capsule injury, labyrinthine concussion, brainstem or nuclei injury, or perilymphatic fistula. The symptoms usually improve as activity resumes and may be treated with vestibular suppressant medications; rehabilitation exercises may help in early recovery of balance. The prognosis is excellent for most patients.
4. CSF otorrhea and rhinorrhea will often resolve spontaneously (>90%) and may be helped with head elevation, stool softener, and bed rest.
5. Importance of early and thorough assessment of facial nerve function cannot be overemphasized.
6. The patient should be monitored for delayed complications of TBF, such as cholesteatoma formation from implantation of external auditory canal skin.

Surgical Management

Temporal bone fractures can cause both immediate and delayed complications, as listed below. Surgical management and rehabilitation are predicated on the type of complications that occur and are related to hearing loss, vertigo, or facial nerve dysfunction.

1. Conductive deafness is due mostly to blood in the external canal, laceration of the tympanic membrane, or hemotympanum. If the conductive loss persists for 6 months or longer, then tympanoplasty for associated ossicular damage should be considered. Hemotympanum typically resolves within 90 days.
2. Severity of sensorineural hearing loss usually correlates with the magnitude of otic capsule–internal auditory canal damage and can affect intelligibility and understanding of speech as well. Although high-dose steroids can be tried in the hopes of reversing this loss, a hearing aid or cochlear implant may be needed (this patient underwent simultaneous bilateral cochlear implant placement).
3. A high degree of suspicion for perilymph fistulae will necessitate an exploratory tympanometry for temporal treatment of either an oval window or round window fistulae.
4. Immediate-onset facial paralysis requires a facial nerve exploration or decompression.

Questions

1. What is the most common type of temporal bone fracture when using the traditional classification system?
2. What percentage of temporal bone fracture–associated cerebrospinal fluid leaks resolve on their own?
3. Name the mechanism by which cholesteatoma formation can occur after temporal bone fractures.

Suggested Readings

Conoyer JM, Kaylie DM, Jackson CG. Otologic surgery following ear trauma. Otolaryngol Head Neck Surg 2007;137(5):757–761

Johnson F, Semaan MT, Megerian CA. Temporal bone fracture: evaluation and management in the modern era. Otolaryngol Clin North Am 2008;41(3):597–618, x

Wu KC, Shu MT, Yang CC. Autologous ossiculoplasty following temporal bone fracture. J Laryngol Otol 2009;123(1):50–53

IV

The Facial Nerve

18

Facial Reanimation

David B. Hom

History

A 65-year-old man 3 months previously had undergone a parotidectomy for mucoepidermoid carcinoma requiring sacrifice of the facial nerve at the facial nerve stump 3 cm distal to the stylomastoid foramen. He complains of the inability to close his eyes and drooling when drinking.

He denies significant medical problems and reports no drug allergies or prescribed medications. He is also a nonsmoker and nondrinker. Physical examination revealed a well-nourished, well-developed white man in mild distress. His facial photos are shown in **Fig. 18.1**.

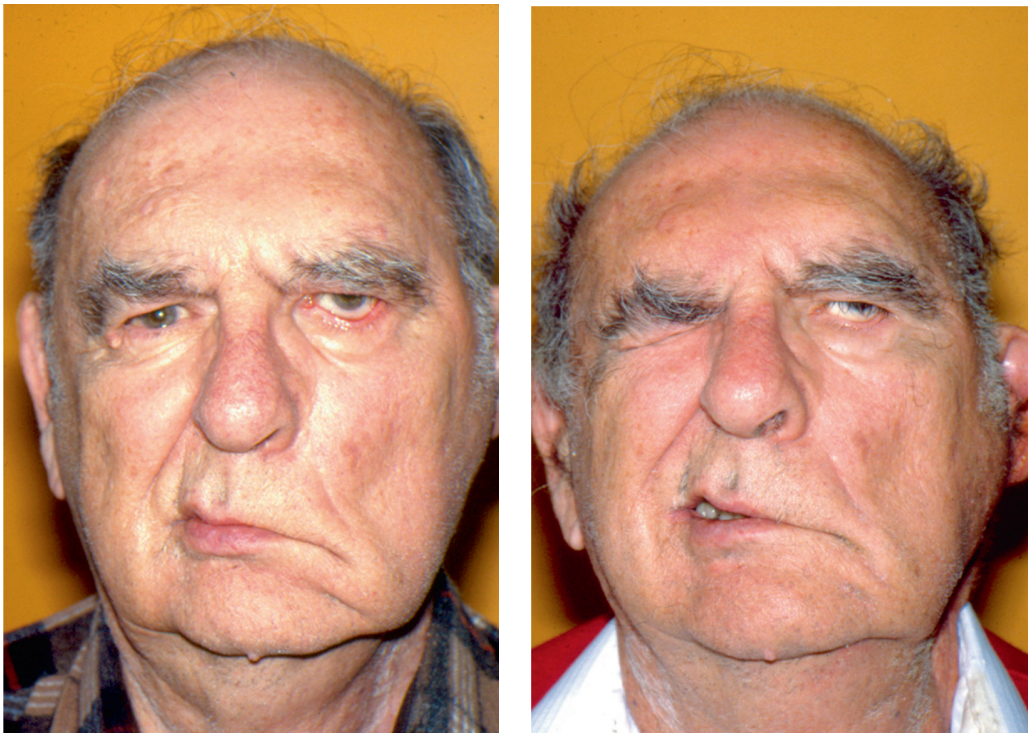
He has good Bell phenomena, extraocular movements intact, PERRLA (pupils equal, reactive to light and accommodation), sclera noninjected. Head and neck examination reveals a well-healed facial incision with obvious full facial paralysis on the left side. He has a ptotic eyebrow, paralytic lagophthalmos, paralytic ectropion, lack of tone on his lower face, and drooping of his lateral commissure. His other cranial nerves are fully intact; his remaining head and neck examination is unremarkable.

Differential Diagnosis—Key Points

1. Facial nerve paralysis is a significant functional and disfiguring deficit. It results from trauma, infection, tumor ablation, and Bell palsy. The facial nerve is one of the most commonly injured cranial nerves because of its long anatomic path from the brainstem to the facial musculature. The site of injury along the nerve's path determines the type of neurologic deficit seen clinically.
2. When the possibility of facial nerve injury has occurred, its major distal motor branches should be checked on physical examination by having the patient lift the eyebrows (frontal branch), squint (temporal zygomatic branch), wrinkle the nose and smile (buccal branch), and frown with puckering lips (mandibular branch).

Test Interpretation

Facial movement of each facial region correlates with the functional integrity of the facial nerve proximal to that region. If voluntary facial movement is present, there is no need



A B

Fig. 18.1 (A, B) Photos of a patient with facial paralysis in the resting and grimacing state.

for nerve excitability testing. The Schirmer test for tearing can be helpful to more fully determine whether the nerve injury has occurred proximal to the geniculate ganglion in the temporal bone.

When a patient presents with an acute facial paralysis lasting longer than 1 month, tumors along the facial nerve pathway should be ruled out. A computed tomography scan or magnetic resonance imaging evaluating the brainstem and parotid gland is helpful in this evaluation.

◆ Diagnosis

Facial nerve paralysis

◆ Medical Management

Facial nerve paralysis can be classified by the severity of nerve damage. This can be helpful to predict its prognostic recovery. When the facial nerve is paralyzed, the terms *neurapraxia*, *axonotmesis*, and *neurotmesis* are useful to describe its injured state. Neurapraxia is often described as "a bruised nerve." It represents the least injured state of a paralyzed nerve. It occurs from a physiologic nerve-conduction blockade from myelin sheath injury. The nerve can still be electrically stimulated externally because the structural integrity of the nerve remains intact. Prognosis for recovery of neurapraxia is usually good, with recovery occurring within 3 to 4 weeks.

Axonotmesis is the state in which the nerve axon is damaged, leading to distal axon degeneration. Nerve regeneration involves the proximal axon to grow again distally within its previous endoneurial sheath to reestablish itself. Recovery can be spontaneous but might not be totally complete.

In neurotmesis, complete injury or transection of the nerve has occurred. Because the epineurium (outer nerve sheath) is damaged, the regenerating proximal nerves sprout outside the nerve sheath, forming neuromas and aberrant nerve circuitry and resulting in abnormal facial movement. A permanent neurologic deficit often results.

After many months, if a regenerating nerve reaches a different facial-muscle motor endplate, abnormal innervation has occurred, resulting in uncoordinated facial movement (*synkinesis*). An example of this state is when a patient attempts to close his eyes, his mouth also moves. In this instance, facial nerve rehabilitation and botulinum injection can be helpful in assisting facial nerve coordination and function.

Eye protective care is crucial in patients with facial paralysis. If the cornea becomes scratched, it can lead to keratitis and later to blindness if not managed. Eye protective measures include an eye ointment at night and nonpreservative eye drops every 2 to 3 hours during the day.

Eye-patch chamber shields with an elastic strap add comfort to the affected dry eye. Sunglasses that curve around the temples or with side protector are helpful when it is windy outside.

For Bell's palsy for which no cause is found for the facial paralysis and other causes have been ruled out, the prognosis is a 94% chance of full recovery if partial facial paralysis is present. If complete facial paralysis is evident, there is a 60% chance of full recovery. Possible medical treatment includes steroids and antiviral medications, which may help patients with poor recovery.

◆ Surgical Management

Surgical goals to restore facial function from facial paralysis are to achieve the following:

1. Facial symmetry at rest
2. Facial symmetry with voluntary motion
3. Separate and selective function of individual muscle groups
4. Spontaneous facial motion during emotional expression

Presently no surgical procedure can achieve all these goals. Also, no single surgical procedure is suitable for all patients. Thus the surgical management of facial paralysis is tailored to match the goals of the patient.

Surgical treatment for facial nerve paralysis is based on the length of time of nerve paralysis and the site of injury. The timing for surgical repair of facial paralysis can be classified as *immediate* (0–3 weeks), *delayed* (3 weeks to 2 years), and *late* (over 2 years). Depending on these time periods, the optimal method to reanimate the facial paralysis can be determined. The importance of classifying such injuries is that procedures, which involve nerve-to-nerve reattachment, are most successful if performed within 2 years of injury. This is because after 2 years denervation muscular atrophy has likely occurred, preventing future innervation. In this circumstance other surgical techniques, such as muscular slings, are required for facial movement.

Immediate and Delayed Repair

The optimal method to repair a transected facial nerve is to connect its severed ends directly as soon as possible. Doing so within 3 days will make it eaiser to find the distal end of the nerve since it will only be electrically stimulated up to this time interval. Once the nerve has been sutured together, the regenerating proximal nerve travels along its attached distal segment at 1 to 3 mm each day. Because the cell body of the facial nerve is located at the brainstem, the more proximal the facial nerve injury, the more time is needed for the regenerating nerve to reinnervate its facial musculature.

If a significant distance between severed nerve ends exists (i.e., resulting from nerve resection from tumor removal or trauma), this gap is bridged with nerve grafts taken from the greater auricular nerve from the neck or the sural nerve from the lower leg.

In the instance in which the nerve cannot be surgically reattached because of its location (i.e., facial nerve transection near the brainstem from an intracranial injury or tumor), other motor cranial nerves can be connected to the facial nerve distally. In this case the hypoglossal nerve is commonly used to attach to the distal facial nerve. For facial movement, when such a patient wants to smile, he learns to push his tongue against his incisors. By connecting the hypoglossal nerve to the facial nerve, 90% of patients achieve good resting facial muscle tone, preventing a sagging face. Disadvantages are unilateral weakness of the tongue. Thus this procedure should not be done when there is a deficit of cranial nerve IX, X, or XII.

Late Repair

Two years after the onset of facial paralysis, denervation atrophy of the facial musculature has likely occurred. To determine the condition of the distal facial nerve, electromyography (EMG) is helpful to determine its physiologic state. If the EMG shows electrical silence, the facial muscles have undergone denervation atrophy. Under these circumstances, nerve grafting or nerve reattachment using other cranial nerves will not be effective. In this instance, temporalis or masseter muscle slings can be rotated to give facial movement to the upper and lower face. These dynamic muscle slings are innervated by the fifth cranial nerve. After this surgery, the patient is able to move his face by biting down.

In cases when a patient requires facial support and cannot undergo a muscle sling procedure, static sling procedures are performed. Slings made from autogenous and artificial materials can be used to suspend sagging tissues of the lower eyelid, cheek, and lips.

Restoring Eyelid Closure

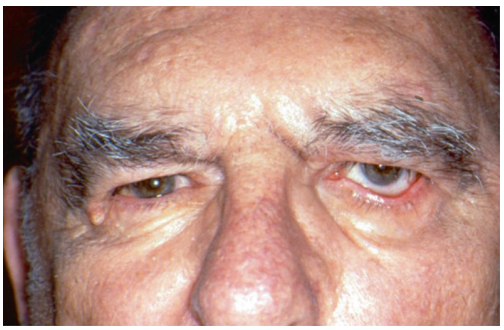
A patient with facial nerve paralysis is frequently unable to close the eyelids because of paralysis of the orbicularis oculi muscle. In addition, lower eyelid sagging (paralytic ectropion) is often apparent because of the lack of muscular tone of the lower eyelid. With the absence of tearing, these conditions make the cornea very vulnerable to exposure and keratitis. The traditional method to prevent exposure keratitis from facial paralysis has been a tarsorrhaphy. However, disadvantages of a tarsorrhaphy include restriction of the lateral visual field, the inability to cover the cornea completely, and unacceptable appearance of patients.

Over the past 15 years, the placement of gold weight implants to the upper eyelid has helped many patients to improve functional eyelid closure. By having a tailored gold weight inserted within the upper eyelid, a patient is able to open and close the eyes spontaneously while being upright.

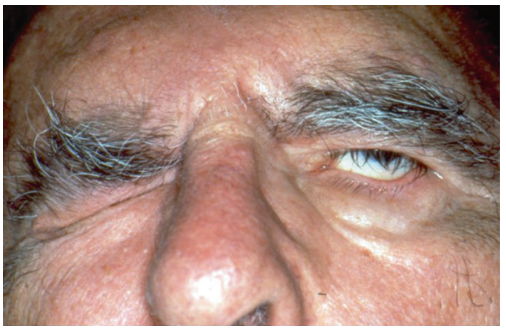
These implants work by giving additional leverage to the upper eyelid, thus allowing the eyelid to close by gravity spontaneously. Eyelid opening is not impaired since the third cranial nerve and the sympathetic nerves innervate eyelid levator function. In addition, procedures to tighten the lax lower eyelid can optimize functional eyelid closure and corneal protection (**Fig. 18.2**). Because gold weight implants are easily removed, gold weight placement can be useful in patients who have an anticipated prolonged temporary facial paralysis.

◆ Rehabilitation and Follow-up

Rehabilitation with facial exercises can be beneficial in strengthening and retraining the facial muscle with a physical therapist, speech pathologist, or occupational therapist that is interested in treating patients with facial paralysis.

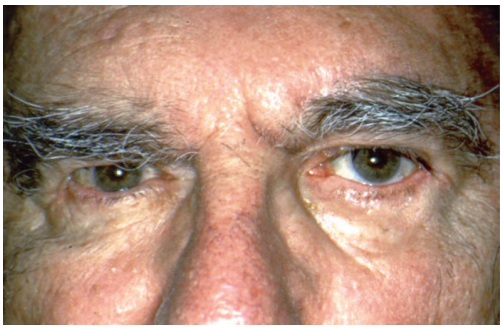


A

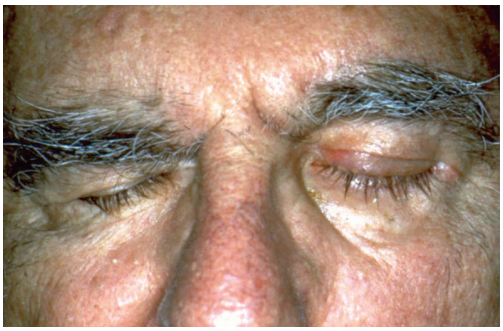


B

Fig. 18.2 Photos of a patient with facial paralysis before surgery **(A)** in the relaxed state and **(B)** attempting eye closure. *(Continues)*



C



D

Fig. 18.2 (*Continued*) **(C and D)** Photos after gold weight implantation to the upper eyelid, lower eyelid tightening, and a direct browlift.

◆ Questions

1. In facial nerve paralysis, what nerve state injury best represents a physiologic nerve-conduction blockade, which can still be electrically excitable from an external stimulus?
- **A.** Neurapraxia
- **B.** Axonotmesis
- **C.** Neurotmesis
- **D.** Wallerian degeneration
- **E.** Neurotization

2. A 40-year old woman had complete transaction of her facial nerve from a transverse temporal fracture 2 years previously. What test would be most helpful to determine the best treatment for facial reanimation?
- **A.** Temporal bone computed tomography scan with contrast
- **B.** Electroneuronography
- **C.** Electromyography (EMG)
- **D.** Hilger stimulation test
- **E.** Schirmer test

3. A 55-year-old woman has had complete facial paralysis for 2 years and would like movement of her lower face. EMG shows electrical silence. What would be the best treatment for facial reanimation?
- **A.** Temporalis muscle sling
- **B.** Gracilis free flap
- **C.** Hypoglossal nerve transposition to facial nerve
- **D.** Facial sling with tensor fascia lata
- **E.** Greater auricular cable nerve graft

4. An 80-year-old woman has had complete facial paralysis for 2 years and would like support to her lower lip to prevent drooling. EMG shows electrical silence. What would be the best treatment for facial reconstruction?
- **A.** Temporalis muscle sling
- **B.** Gracilis free flap
- **C.** Hypoglossal nerve transposition to facial nerve
- **D.** Facial sling with tensor fascia lata
- **E.** Greater auricular cable nerve graft

Suggested Readings

Bascom DA, Schaitkin BM, May M, Klein S. Facial nerve repair: a retrospective review. Facial Plast Surg 2000;16(4):309–313

Hadlock TA, Greenfield LJ, Wernick-Robinson M, Cheney ML. Multimodality approach to management of the paralyzed face. Laryngoscope 2006;116(8):1385–1389

May M, Schaitkin BM. The Facial Nerve, 2nd ed. New York, NY: Thieme; 2000

Tate JR, Tollefson TT. Advances in facial reanimation. Curr Opin Otolaryngol Head Neck Surg 2006;14(4): 242–248

19

Bell Palsy

Melissa McCarty Statham and Ravi N. Samy

History

A 34-year-old white woman with a 12-day history of a rapidly progressive, complete left-sided facial paralysis reports a history of an upper respiratory infection, which began presumably virally caused but progressed to bronchitis, for which she received antibiotic therapy. One day after beginning the course of systemic antibiotics, she noted an ulcerative lesion in her oral cavity. Four days after the appearance of this lesion, she began experiencing rapidly progressive loss of function of the left side of her face.

The patient denies any history of antecedent otologic or neuro-otologic infection or trauma. She complains of right-sided otalgia, aural fullness, and hyperacusis in her left ear. Her medical history is significant only for migraine headaches. The patient denies tinnitus, vertigo, or otorrhea. She has not traveled to any deer tick–infested areas.

Physical examination reveals a well-developed woman in no apparent distress. The facial nerve function on the left side is House-Brackmann grade VI/VI (i.e., complete absence of volitional function). The contralateral facial mimetic function is normal. The pinnae and external auditory canals are normal, and microscopic examination reveals normal mobile tympanic membranes bilaterally. In particular, no vesicular lesions are noted on physical examination. The head and neck examination, as well as the remainder of her cranial nerve examination, are within normal limits.

Differential Diagnosis—Key Points

The differential diagnosis for the cause of paroxysmal facial paralysis can be organized in the following manner:

Congenital

Möbius syndrome, also known as congenital facial diplegia, is the congenital agenesis of bilateral facial and abducens nerves. These patients usually present as infants with complete lack of bilateral mimetic function and bilateral abducens paralysis. Although it occurs less commonly, cranial nerves III and IV, as well as the lower cranial nerves, can be additionally affected.

Hemifacial microsomia is a progressive bony and soft tissue deformity in which half of the lower face fails to form secondary to a developmental malformation of the first and second branchial arches. Facial paralysis is present in a

significant portion of these patients, who are diagnosed in early childhood.

Inflammatory

Neurosarcoidosis is an inflammatory condition of unknown cause in which noncaseating granulomas involve multiple organs, most commonly lung, as well as the nervous system. Most commonly, this manifests as cranial neuropathy, with paroxysmal facial paralysis the most common presenting symptom. Cerebrospinal fluid (CSF) or serum should be analyzed for angiotensin-converting enzyme to confirm the diagnosis. Further evaluation for systemic sarcoidosis should include, but not be limited to, chest radiograph, bronchoscopy with biopsy of mediastinal lymph nodes, and contrast-enhanced magnetic resonance imaging (MRI) of the brain. Involvement of the facial nerve may be unilateral or bilateral and relapsing-recurring. Even in the absence of concomitant rheumatologic or pulmonary disease, investigation for neurosarcoidosis should be considered as the cause of facial paralysis in patients with a suspicious history.

Melkersson-Rosenthal syndrome is a rare disorder characterized by recurring facial nerve paralysis, facial and lip swelling, and the development of a furrowed tongue. The facial paralysis may be unilateral or bilateral. Onset is usually in late childhood or adolescence. Noncaseating granulomas may be seen in lip biopsies of some of these patients, but this is not seen in all patients with the syndrome. The cause is currently unknown, but there may be a genetic predisposition to the disorder.

Infectious

Lyme disease is caused by an infection with the organism *Burrelia burgdorferi*, which is carried by the deer tick, *Ixodes scapularis*, as a vector. The disease presentation varies widely, but classically patients exhibit erythema migrans and a flulike illness in the initial stages of the disease. Musculoskeletal, cardiac, psychiatric, and neurologic manifestations, including facial paralysis, may follow. Lyme disease should be considered in the differential diagnosis in patients with a history compatible with potential exposure to a tick bite. Although rare, Lyme disease may be a cause of bilateral facial paralysis. Diagnosis is confirmed with serology studies to the aforementioned organism.

Herpes zoster oticus, also known as *Ramsay Hunt syndrome*, is a viral infection caused by reactivation of varicella zoster virus, affecting the geniculate ganglion and causing facial nerve inflammation, edema, and compression within the fallopian canal. Typically patients have severe otalgia and vesicular lesions of the ipsilateral ear and face. Varicella zoster infections without concomitant skin lesions, termed *zoster sine herpete*, can occur and is sometimes mistaken for Bell palsy.

Although it is the most common cause of facial palsy, Bell palsy remains a diagnosis of exclusion. The presentation is that of a rapidly progressive facial paralysis, although patients may exhibit variable degrees of weakness. The pathophysiology is thought to derive from reactivation of latent herpes simplex virus, causing inflammation, edema, and compression of the facial nerve in the fallopian canal.

Neoplastic (Benign and Malignant)

Facial neuroma: See the case presentation on facial neuroma for more detail.

Chronic otitis media with and without cholesteatoma may manifest with facial paralysis because erosion of the fallopian canal may allow the inflammatory response (e.g., hypertrophic mucosa and granulation tissue) to involve the facial nerve. Rarely facial paralysis is the sole presenting symptom of a patient with cholesteatoma.

Cerebellopontine angle masses, such as vestibular schwannomas, meningiomas, lipomas, or unusually large lesions associated with the trigeminal nerve or the lower cranial nerves, may manifest with facial paralysis. Typically the symptoms associated with other involved structures are also present because it is extremely rare for such lesions to have facial paralysis as the only presenting symptom. Head and neck processes (e.g., squamous cell carcinoma and benign parotid gland neoplasms) may also present with an acute onset of facial paralysis.

Test Interpretation

Electroneuronography (ENoG), also known as *evoked electromyography*, may be obtained after 72 hours after the initial onset of the facial palsy to follow the degree of degeneration quantitatively. In general, patients exhibiting degeneration of less than 90% will have an excellent spontaneous recovery of their facial paralysis. In the setting of degeneration greater than 90%, the degree of recovery is much less predictable.

Electromyography (EMG) may establish the presence or absence of voluntary motor units. This is a complementary study to use along with the ENoG because the presence of voluntary motor units in the setting of greater than 90% degeneration is suggestive of good spontaneous recovery of facial function. Lack of voluntary motor units in the same setting is a negative prognostic indicator of recovery of facial nerve function. Both ENoG and EMG have supplanted the use of minimal nerve excitability testing or maximal stimulation testing.

Computed tomography (CT) scan may be obtained if a neoplastic process is suspected or in the setting of preoperative evaluation of anatomic structures in patients who are to undergo surgical management of the facial paralysis.

The use of MRI in the initial diagnosis of the patient with Bell palsy is of limited clinical utility. In cases of Bell palsy and neurosarcoidosis, enhancement along the course of the facial nerve has been demonstrated to a variable degree. Patients who do not demonstrate any improvement by 6 months should undergo MRI with gadolinium to evaluate for possible neoplastic pathology. In the presence of other clinical findings, such as additional cranial neuropathy, or in the setting of recurrent or bilateral facial paralysis, MRI is certainly warranted to complete a thorough evaluation. Additionally, patients who elect to undergo operative intervention as treatment (see middle-fossa approach below) will undergo preoperative MRI to rule out a neoplastic cause.

Diagnosis

Bell palsy

Medical Management

Instituting high-dose systemic corticosteroid therapy early in the course of Bell palsy and herpes zoster oticus has been found to be efficacious (typically 1 mg/kg prednisone). Additional use of systemic antiviral agents has been generally considered to be efficacious (typically 1 g three times a day of valacyclovir); yet data from randomized prospective clinical trials are less clear, with some trials suggesting no benefit from antiviral therapy, whereas others establish a statistically significant improvement in patient outcomes. Antiviral therapy, in addition to systemic corticosteroids, has certainly been established as efficacious in the setting of herpes zoster oticus. Antiviral therapy should always be implemented in the immunocompromised patient with Bell palsy or herpes zoster oticus.

Attention to eye care is imperative in the management of patients with Bell palsy. Ocular lubricants and sodium chloride ophthalmologic drops are applied periodically. In addition, the eye should be securely patched or covered while the patient is sleeping, and adequate eye coverage should be used during the daytime, when there is a possibility of dust or wind causing corneal irritation (e.g., with glasses or bubble patch).

Surgical Management

The best surgical management for Bell palsy remains controversial. Many centers perform serial ENoG with complementary EMG to determine patients whose facial paralysis is unlikely to resolve spontaneously with good return of function, generally accepted as a House-Brackmann grade I–II. In patients in whom degeneration is found to be greater than 90% on ENoG and no volitional motor units on EMG, surgical decompression is offered. It has generally been accepted that patients in whom facial paralysis has been present longer than 14 days are unlikely to benefit from surgical decompression, and as such, these patients are managed expectantly.

To date only one multicenter prospective clinical trial examining surgical decompression has been performed. Patients with complete facial

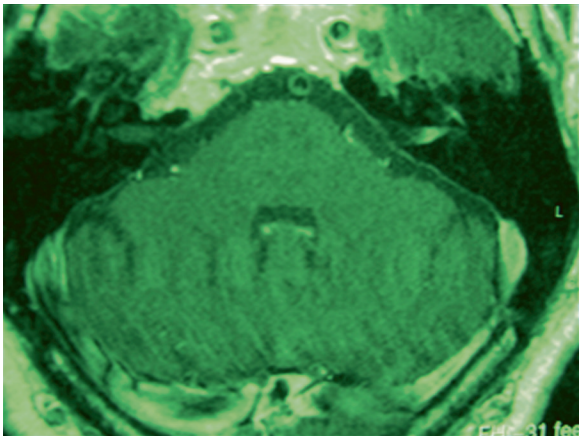


Fig. 19.1 Axial magnetic resonance image with contrast of patient with Bell palsy shows enhancement of left facial nerve in fundus of internal auditory canal, labyrinthine segment, and portion of tympanic segment.

nerve paralysis, greater than 90% degeneration on ENoG, and no evidence of intact voluntary motor units on EMG testing were candidates for surgical decompression within 2 weeks of onset of paralysis. Patients who underwent decompression within 2 weeks of onset of paralysis showed a 91% chance of a House-Brackmann scale grade I or II facial nerve function versus a 42% chance of obtaining a House-Brackmann scale grade I or II if treated with steroids only. Even taking these findings into account, the vast majority of patients with Bell palsy improve, and a small percentage, though improved, will attain a poor return of facial nerve function.

For patients who elect to undergo facial nerve decompression, a middle cranial fossa approach is used to decompress the labyrinthine segment of the facial nerve (the site of pathology in Bell palsy) (**Fig. 19.1**). This is performed from the fundus of the internal auditory canal to the meatal foramen and to the proximal tympanic segment. The meatal foramen is the entrance to the labyrinthine segment and represents the anatomically narrowest portion of the fallopian canal. Risks of surgical decompression include anacusis, dizziness, and other potential complications associated with intracranial procedures (CSF leaks, seizures, stroke).

◆ Rehabilitation and Follow-up

Although adequate data are lacking, facial rehabilitation exercises are considered important, and these are instituted as early in the disease process as possible to attempt to keep synkinesis at a minimum. Vigilant eye care should be maintained to prevent drying of the cornea and abrasion. For patients in whom return of orbicularis oculi function is anticipated to take longer than 6 months, reversible surgical procedures (such as placement of an upper lid gold weight) may be performed to optimize corneal protection. In the event a patient develops synkinesis several months after Bell palsy, botulinum toxin injections may be performed to reduce hypertonicity and restore facial symmetry.

◆ Questions

1. True or false: Bell palsy most likely has a viral cause.
2. What portion of the facial nerve is involved in the pathophysiology of Bell palsy?
3. In Bell palsy, what is the most important complication to avoid?

Suggested Readings

Adour KK. Medical management of idiopathic (Bell's) palsy. Otolaryngol Clin North Am 1991;24(3): 663–673

Adour KK, Byl FM, Hilsinger RL Jr, Kahn ZM, Sheldon MI. The true nature of Bell's palsy: analysis of 1,000 consecutive patients. Laryngoscope 1978;88(5):787–801

Adour KK, Ruboyianes JM, Von Doersten PG, et al. Bell's palsy treatment with acyclovir and prednisone compared with prednisone alone: a double-blind, randomized, controlled trial. Ann Otol Rhinol Laryngol 1996; 105(5):371–378

Fisch U. Surgery for Bell's palsy. Arch Otolaryngol. 1981; 107(1):1–11

Gantz BJ, Rubinstein JT, Gidley P, Woodworth GG. Surgical management of Bell's palsy. Laryngoscope 1999; 109(8):1177–1188

Samy RN, Gantz BJ. Surgery of the facial nerve. In: Glasscock ME, Gulya AJ, eds. Surgery of the Ear, 5th ed. Hamilton, Ontario, Canada: BC Decker; 2003:615–639

Sullivan FM, Swan IR, Donnan PT, et al. Early treatment with prednisolone or acyclovir in Bell's palsy. N Engl J Med 2007;357(16):1598–1607

20

Iatrogenic Facial Palsy

Collin M. Burkart and Ravi N. Samy

History

A 40-year-old woman is taken to the operating room to undergo surgical management for right chronic otitis media with cholesteatoma. In the operating room, she undergoes an uneventful right tympanomastoidectomy with facial recess. A facial nerve monitor is used. The patient is injected preoperatively in the usual fashion with 1% lidocaine with 1:100,000 epinephrine for the postauricular and external auditory canal incisions. The extended facial recess is performed (drilled through the incus buttress and the chorda tympani nerve). No facial nerve dehiscence is noted throughout the case; the facial nerve is identified in the tympanic and mastoid segments. The nerve monitor does not indicate any nerve stimulation. The rest of the procedure is uneventful. However, on awakening, the patient has a facial nerve palsy of House-Brackmann grade III–VI.

Differential Diagnosis—Key Points

1. Iatrogenic facial nerve injury or palsy is a rare but serious complication of otologic surgery. Concerns in the immediate postoperative period include the effect of local anesthetic or packing. Mastoid dressing and packing should be immediately removed to see whether there is any benefit to the function of the facial nerve. It is also possible that the canal injections of lidocaine, especially if a tympanic membrane perforation was present, may be contributing to the facial palsy. The effect of the lidocaine may persist for several hours.
2. After several hours (enough time for the effect of the local anesthetic to subside), the patient should be reexamined. If there is no improvement and the patient has a dense facial palsy (House-Brackmann grade VI), the patient should be taken back to the operating room immediately for exploration of the facial nerve.
3. If the injury manifests as a partial facial nerve paralysis, the patient should be managed medically and expectantly as described below under medical management.
4. In this case, intraoperative facial nerve monitoring was used. Intraoperative facial nerve monitoring, which remains controversial (particularly from a medicolegal standpoint), has become standard of care in neuro-otologic surgery for vestibular schwanomma. In routine middle-ear or mastoid surgery, it is most often beneficial for revision cases, complex cases, and when residents or fellows are involved. The use of intraoperative facial nerve monitoring is not intended to replace surgical

experience and judgment but merely to augment the otologic surgeon's decision making. Although the monitor has proven helpful in identification of the facial nerve, it has not eliminated facial nerve injury in otologic surgery.

5. The site of injury in cases of iatrogenic facial palsy will vary based on the type and location of surgery being performed. In cases of surgery involving chronic otitis media, the tympanic segment is a common site of injury to the facial nerve. This is likely due to the fallopian canal being dehiscent in this area more often relative to other locations of the nerve secondary to congenital dehiscence, cholesteatoma, and granulation tissue or hypertrophic mucosa. The next most common site of injury is the mastoid segment of the facial nerve, especially at the location of the second genu, where the nerve changes from a horizontal to vertical trajectory. A third location that is of concern is the anterior epitympanic space just distal to the geniculate ganglion and first genu. Disease in this area, combined with decreased visualization and exposure, place this area of the nerve at risk, but removing portions of the ossicles and cog will aid in exposure. Another possible location of injury, which is the suspected location in this case, is at the attachment of the chorda tympani nerve and facial nerve. If the chorda tympani nerve is transected with the otologic drill, it is theorized that it can cause a small hematoma formation compressing the facial nerve in the fallopian canal. Alternatively heat-induced neuropraxia (from the otologic drill) can cause such a palsy as well.

Test Interpretation

None

Diagnosis

Postoperative or iatrogenic facial paresis or paralysis. The diagnosis of postoperative facial nerve paralysis or paresis is made by clinical observation. A complete facial paralysis does not require any electrophysiologic or radiographic studies and should be immediately explored. A partial weakness (as in this case) or delayed onset should be followed up similarly to a Bell's palsy. If a delayed complete palsy occurs, electroneuronography (ENoG) and electromyography (EMG) are used by some surgeons to determine whether facial nerve exploration and decompression are needed (see below).

Medical Management

In a case of acute, complete postoperative facial nerve paralysis, there is no role for further testing or medical management and the situation warrants surgical management as described below.

For situations of partial or delayed facial nerve paralysis, as in this case, medical management and further testing are indicated. In this case, the patient had immediate onset of facial nerve paresis with a House-Brackmann grade III. Therefore, she was immediately placed on steroids. The patient was kept in house overnight postoperatively and intravenous dexamethasone was used. The patient had not demonstrated improvement at the time of discharge on postoperative day 1. She was discharged with a 1-week taper of steroids and had an early follow-up appointment scheduled. The patient was seen weekly during the immediate postoperative period while closely monitoring facial nerve function. Her facial nerve function never decreased below a House-Brackmann grade III and at 1 month postoperatively, her facial nerve function was demonstrating signs of improvement and was a House-Brackmann grade II. Facial nerve injury with only a partial decrease in function is expected to return to normal or near normal by 6 to 12 months postoperatively. Although the patient had good eye closure, she was counseled on the importance of eye care and using artificial tears and eye ointment as needed.

In a case in which the facial nerve had delayed onset of paralysis postoperatively (i.e., 3 days or longer), the situation should be managed differently. Steroids and antiviral agents should be used because often the cause is edema and inflammation secondary to viral reactivation. Recently, studies have questioned

the usefulness of antiviral medication in Bell's palsy. In fact, although steroids are effective in Bell's palsy, antiviral medications alone or combined with steroids demonstrate no effectiveness. At this time, it is the opinion of the authors that postoperative delayed-onset facial paralysis should be managed with steroids and antiviral medications. The patient should also be monitored serially with ENoG and EMG. If the nerve degenerates over 90% and no voluntary motor potentials are found, surgical exploration should be considered. Of note, ENoG will be useful around 3 to 21 days after onset of complete paralysis.

Surgical Management

If the need arises to return to the operating room, the patient and room should be prepared for an operation to expose the entire course of the facial nerve. The patient should have a bone-line audiogram performed before surgery to confirm the level of sensorineural hearing. To expose the geniculate region of the nerve, a middle cranial fossa approach may be warranted. If the patient has nonfunctional hearing, a translabyrinthine approach would provide excellent exposure of the nerve without subjecting the patient to a craniotomy.

The facial nerve would need to be identified and exposed in the vertical segment, and after opening up the facial recess, the full tympanic segment would need to be exposed. Also, if warranted, the geniculate region might need to be exposed via the middle cranial fossa approach. If the nerve appears only edematous and hyperemic but still intact, it should be decompressed along its course. If an area of injury is identified, the method of repair is determined by the extent of injury. A partial transaction of less than 50% warrants full decompression alone, but if over 50% of the nerve is transected, the edges should be freshened and the nerve reapproximated as well as decompressing the nerve. If the nerve is transected completely, the edges should be freshened, and the nerve should be freed, decompressed, and reapproximated with tension-free closure. In situations where a segment of nerve is missing and tension-free closure cannot be attained, an interposition or cable graft should be used.

Rehabilitation and Follow-up

As with all cases of facial palsy, ophthalmic care with appropriate coverage and ointment is important. Facial-muscle exercises can be initiated, and the patient may be referred to a physical therapist who specializes in facial paralysis.

Facial paralysis, while rare, is a recognized complication of otologic surgery that can be devastating for both the patient and the surgeon. It is crucial that the patient and the patient's family have a complete understanding of the risks, especially as they relate to facial nerve injury, before surgery. If an injury occurs, it is essential that the surgeon discuss the nature and prognosis of the injury with the patient or responsible family member as soon as possible. The options for management should also be addressed as quickly as possible. All information should be honest, direct, and clear. In these situations, a high level of stress and anxiety will be felt by both the patient and the surgeon. It is important, in discussing reexploration, to convey a clear understanding of the expectations of facial nerve restoration. In cases of facial nerve transaction, the best outcome to be expected is a House-Brackmann grade III. Although often the surgeon may wish to perform the reexploration, it is appropriate to request assistance from a more senior colleague or even to have the patient seen on referral to a regional or national expert.

Questions

1. If a postoperative facial palsy resolves within just a few hours after the case is completed, what is the most common reason?
2. If a delayed complete palsy occurs, what electrodiagnostic tests can be used to determine whether further surgical management is warranted?
3. True or false: Use of a facial nerve monitor is guaranteed to prevent injury to the facial nerve.

Suggested Readings

Danner CJ. Facial nerve paralysis. Otolaryngol Clin North Am 2008;41(3):619–632, x

Mancini F, Taibah AK, Falcioni M. Complications and their management in tympanomastoid surgery. Otolaryngol Clin North Am 1999;32(3):567–583

Weber PC. Iatrogenic complications from chronic ear surgery. Otolaryngol Clin North Am 2005;38(4): 711–722

V

The Oral Cavity and Pharynx

21

Field Cancerization

Ernest C. Manders, Eyad Khabbaz, and Keith M. Wilson

History

A 45-year-old woman presents with a raised tender area in the right posterior ventral tongue. Two years previously she had undergone transoral resection of squamous cell carcinoma of the anterior left ventral tongue. Pathology revealed in situ tumor only, without invasive component. These areas are anatomically separate from one another. She has no tobacco or significant alcohol history and is generally healthy.

Examination revealed a well-nourished individual without obvious lesion of the posterior tongue. The anterior area previously resected now has a pyogenic granuloma (confirmed by biopsy), and palpation of the right posterior ventral tongue reveals a slightly raised, firm area that is tender to the touch (**Fig. 21.1**). The neck does not have appreciable lymphadenopathy.

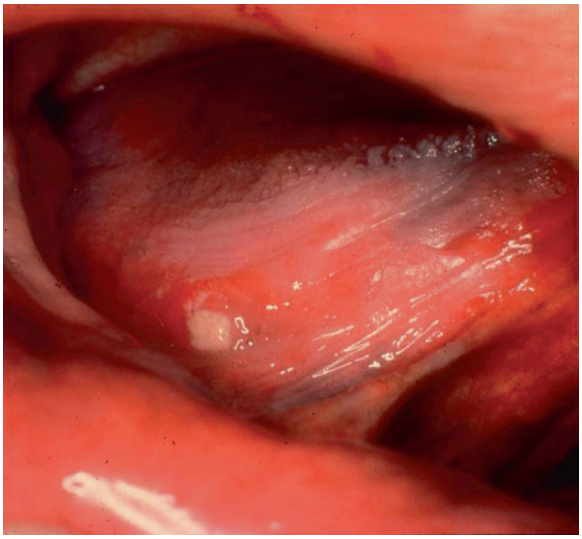


Fig. 21.1 Lesion of the right posterior ventral tongue. Raised white leukoplakic center surrounded by erythematous halo with distinct border.

Differential Diagnosis—Key Points

1. *Leukoplakia* is a white or gray area of mucosa that may represent a pathologic change that is neoplastic or traumatic in nature. It has a low incidence of transformation to carcinoma (about1%).
2. *Erythroplakia* is a red or pink plaquelike area of mucosa. This has a 90% incidence of being malignant or undergoing malignant transformation.
3. The clinician must evaluate lesions of the upper aerodigestive tract carefully because they can look fairly similar and are often described similarly, although their causes may be quite distinct. Trauma, burn, rheumatoid and connective tissue diseases (Crohn disease), and infectious causes (aphthous ulcers) can all cause plaquelike lesions of discoloration in the oral mucosa.
4. Suspicion of neoplasm must guide the examiner. Neoplasms rarely have a distinct border. Also, carcinogenic agents will contact many cells and therefore create areas of leukoplakia, carcinoma in situ, microinvasive carcinoma, and invasive carcinoma, all in the same anatomic region or in the same lesion. Pain is a key element of the history and can help guide the biopsy.
5. Agents such as tobacco, alcohol, and betel nuts have been well documented as carcinogens of the aerodigestive tract. More recently, human papillomavirus, especially types 16 and 18, has been implicated in squamous cell carcinoma in the patient without the traditional carcinogenic exposures.

Test Interpretation

1. Biopsy is critical to pathologic diagnosis of carcinoma. The clinician should be guided to biopsy areas that are painful, firm areas on palpation, or areas previously demonstrating carcinoma.
2. Staining with toluidine blue can be helpful to assess areas of condemned mucosa and can help guide the biopsies. More recently, techniques involving an oral rinse of dilute acetic acid solution or swab of toluidine blue and luminescence under specific wavelengths of light (ViziLight) may further help to direct biopsies.
3. Panendoscopy is often necessary to rule out a second primary.

Diagnosis

Microinvasive squamous cell carcinoma of the ventral tongue stage I

Medical Management

1. Vigilance and a low threshold to biopsy a suspicious site are the most important aspects of managing this disease. There is no chemopreventive agent that is yet widely accepted, although this has been the subject of clinical trials. Ingestion of antioxidants and the trace nutrients in black raspberries and blueberries have been examined for these ends.
2. Radiotherapy can be used for this disease, although it has the disadvantages of treating a diffuse area, with the possibility of recurrence in an adjacent site outside the radiation field. More commonly, radiotherapy is directed at a single distinct lesion.

Surgical Management

For palpable lesions, cold knife excision remains an excellent way to remove the lesion and obtain tissue for pathologic examination. CO_2 laser ablation may be a useful adjunct for destruction of a lesion while penetrating only superficially into the underlying tissues. The disadvantage of this technique is that it destroys any tissue for microscopic examination. Often combining these two modalities is the most useful course.

Rehabilitation and Follow-up

In this case, the lesion on the posterior tongue was resected under general anesthesia. This revealed microinvasive carcinoma. The patient underwent imaging and an ipsilateral selective neck dissection, revealing no

positive lymph nodes. She has since presented several more times over the following years, each time with a painful area on the left tongue. These areas are biopsied, often in the office, and if high-grade atypia or carcinoma in situ is found, CO_2 laser ablation with the patient under general anesthesia has been done. The neck must be clinically examined at these visits for lymphadenopathy. Communication between clinician and patient is very important in surveillance of the condemned mucosa.

◆ Questions

1. A 61-year-old white man who is a heavy smoker and occasional drinker with multiple painful oral cavity lesion consults you. What is the best way to diagnose these lesions and rule out malignancy?
   A. Incisional biopsies from all lesions in clinic
   B. Fine-needle aspiration of these lesions
   C. Panendoscopy, including laryngoscopy, bronchoscopy, and esophagoscopy with incisional or excisional biopsies if feasible
   D. Computed tomography scan of the head and neck with contrast.
   E. Magnetic resonance imaging of the head and neck

2. Which lesion of the oral cavity has the highest rate of malignant transformation?
   A. Leukoplakia
   B. Erythroplakia
   C. Aphthous ulcer
   D. Hyperkeratosis
   E. Lichen planus

3. Which of the following statements about oral cavity suprastaining with toluidine blue is incorrect?
   A. It is an easy technique to identify a suspicious mucosal lesion.
   B. It helps in guidance of the biopsy.
   C. Luminescence under specific wavelengths of light may help direct the biopsy.
   D. If the mucosa did not take the stains, this rules out malignancy.
   E. It can be done in clinic and in the operating room.

4. The risk factor(s) for squamous cell carcinoma of the oral cavity is (are):
   A. Use of tobacco
   B. Alcohol use
   C. Chewing betel nut
   D. Human papilloma virus type 16 and 18, especially tonsil cancer
   E. All of the above

5. The most common malignancy of the oropharyngeal mucosa is:
   A. Spindle cell carcinoma
   B. Squamous cell carcinoma
   C. Lymphoepithelial carcinoma
   D. Mucoepidermoid carcinoma
   E. Adenoid cystic carcinoma

Suggested Readings

DeVries N, Gluckman JL, eds. Multiple Primary Tumors of the Head and Neck. New York, NY: Thieme; 1990

Haddad RI, Shin DM. Recent advances in head and neck cancer. N Engl J Med 2008;359(11):1143–1154

Park EJ, Pezzuto JM. Botanicals in cancer chemoprevention. Cancer Metastasis Rev 2002;21(3-4):231–255

22

Nasopharyngeal Carcinoma

Eyad Khabbaz and Keith M. Wilson

History

A 56-year-old white woman has a 3-month history of intermittent blood-stained left nasal discharge. Five weeks ago she noticed an increase of the nasal discharge with left-sided ear pressure, discomfort, and mild hearing loss. She was treated with antibiotics by her primary care physician for acute rhinosinusitis and acute otitis media. After 2 weeks of treatment, the symptoms continued and the patient noticed a left neck lump. She comes here today for evaluation and treatment of her persistent symptoms. She denies otorrhea, otalgia, facial nerve weakness, vertigo, or tinnitus. She denies previous ear or nose surgery. She had worked as an accountant for almost 30 years. She has been retired for 5 years. She denies vision changes, sensation changes, dysphasia, voice changes, aspiration, or weight loss. She has smoked half a pack per day for almost 30 years and admits to drinking one or two beers daily. Her medical history is insignificant other than newly diagnosed diabetes mellitus.

On physical examination, the left tympanic membrane is retracted and amber-colored fluid fills the middle ear space. After adequate topical anesthesia and decongestion, rhinoscopy and nasopharyngoscopy were performed using a 30-degree Hopkins rod. This showed a lesion that fills the left nasopharynx and obliterates the fossa of Rosenmuller and obstructs the eustachian tube orifice. The lesion appears as friable and ulcerative without obstructing the choana. A biopsy was performed at this point using biopsy-cup forceps. Neck palpation reveals a 3 × 2 cm nontender, firm, mobile lymph node in the left upper jugular level. The right neck is negative for any palpable lymph nodes. Extraocular movements are normal. The remainder of the upper aerodigestive tract examination is normal.

Differential Diagnosis—Key Points

1. The finding of unilateral serous otitis media in an adult should make the clinician suspicious for a nasopharyngeal tumor, especially with palpable cervical adenopathy. These observations may lead the primary care physician to a presumptive diagnosis of acute otitis media and prescription of antibiotics without thorough evaluation of the nasopharynx. One or two courses of antibiotic therapy will lead to a delay in diagnosis, thus allowing further tumor growth. Skull-base invasion is often characterized by headache and extraocular muscle weakness.
2. Lymphoma is the second most common tumor to arise in the nasopharynx. Therefore

queries regarding fevers and night sweats, constitutional type B symptoms, should be made.

3. This patient does not complain of nasal obstruction or epistaxis. These symptoms, along with headache or head pressure, are often present with nasopharyngeal malignancy but are frequently dismissed as allergy or sinus problems
4. The differential diagnosis of a nasopharyngeal mass with evidence of adenopathy in the neck is a malignancy until proven otherwise. A brief list of possible malignancies include squamous cell carcinoma, lymphoma, adenocarcinoma, adenoid cystic carcinoma, mucoepidermoid carcinoma, plasma cell myeloma, rhabdomyosarcoma, malignant melanoma, fibrosarcoma, chondrosarcoma, and clival chordoma. The most important next step in the diagnosis of the mass is a biopsy and histopathologic examination. Fine-needle aspiration cytology is safe and helpful, especially in those patients whose primary tumor is hard to biopsy or unknown. Also it is important to prove metastasis in questionable cases.

◆ Test Interpretation

A patient with a nasopharyngeal mass, cervical adenopathy, and unilateral serous otitis media must undergo several diagnostic tests to be appropriately diagnosed and staged before treatment can be instituted.

1. *Computed tomography (CT) scan.* CT scan with contrast from the skull base to the clavicles will delineate the extent of the primary tumor and cervical metastases. Some surgeons advocate the importance of obtaining imaging before biopsy of the tumor. If it is convenient for the patient to undergo biopsy with minimum morbidity at the initial visit, it should be done. Imaging is obtained subsequently. Skull-base invasion occurs in 30% of the cases. CT scan can demonstrate soft tissue extension in the nasopharynx as well as the parapharyngeal space. It is sensitive in detecting any skull-base bone erosion. CT scan helps in identifying perineural spread. Tumor extension in the region between the foramen rotundum and foramen lacerum may involve the anterior group of cranial nerves (II through VI). Posterior cranial fossa extension may affect the lower cranial nerves (VII through XII).
2. *Magnetic resonance imaging (MRI).* In patients with obvious signs of intracranial extension, including cranial neuropathies or other signs suggestive of central nervous system invasion, MRI should be obtained to evaluate the extent of disease. MRI is better in differentiating tumor from inflammation. It is also more sensitive at evaluating retropharyngeal extension and deep cervical nodal metastasis. MRI shows bone marrow infiltration even without bone erosion, which may indicate increased risk for distant metastasis.
3. *Positron emission tomography (PET), especially if combined with CT scan (PET-CT scan)* is more sensitive at detecting persistent and recurrent nasopharyngeal carcinoma (NPC), both locally and in the neck. PET-CT scan may help in detecting distant metastasis. Suspected lesions may need further study with bone scan, CT of the chest, liver scintigraphy, or bone marrow biopsy.
4. *Biopsy.* In cooperative patients, a nasopharyngeal mass may be easily biopsied transnasally in the office after adequate topical anesthesia. This is done under endoscopic guidance. Minimal bleeding occurs that is easily controlled with a light pack of a hemostatic agent. Tumor biopsy specimens suspected to be lymphoma should be placed in saline for fresh specimen preparation. Often the tissue obtained by transnasal biopsy is enough for pathologists to diagnose lymphoma with special stains and immunotyping. In the case where the biopsy specimens are of questionable quality or if the patient is reluctant or unable (secondary to significant septal pathology) to undergo transnasal biopsy, a formal transoral nasopharyngeal biopsy is performed under general anesthesia in the operating room. Transoral biopsy is also needed in cases where a lesion appears to be completely submucosal. Deeper biopsies are often difficult to obtain in the office setting.
5. Histopathologically there are three distinct forms of NPC. An accurate diagnosis of the subtype is critical as prognosis may differ.

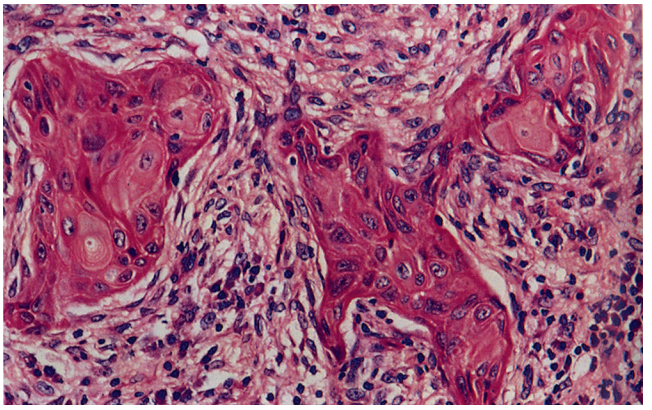


Fig. 22.1 On low-powered microscopy: Stratified polygonal cells, with keratinized islands.

The World Health Organization (WHO) classification is used herein.

- WHO type I (25% of total NPC cases in the United States): Keratinizing squamous cell carcinoma. These tumors show abundant intercellular bridges and keratin production. Histologically this is similar to other upper aerodigestive tract squamous cancers. It is rare in Southeast Asia. There are three grades in this type: well, moderately, and poorly differentiated.
- WHO type II (12%): Nonkeratinizing squamous cell carcinoma, with some maturation. Also known as *transitional cell carcinoma* because it resembles this form of bladder malignancy.
- WHO type III (63%): Undifferentiated carcinomas forming a diverse group of tumors, including lymphoepithelioma, anaplastic, clear cell, and spindle cell variants. This type represents the vast majority of cases (95%) in Southeast Asia.

A transnasal biopsy was performed in the patient in question, and histopathology revealed a keratinizing squamous cell carcinoma of the nasopharynx, WHO type I. (**Figs. 22.1 and 22.2**).

6. *Serology.* Latent Epstein-Barr virus (EBV) virus infection seems to be crucial in the pathogenesis of NPC. After initial infection, the virus remains latent in epithelial cells and B lymphocytes. In a patient with NPC, the immunoglobulin A (IgA) response to the early (EA) and late (VCA, capsid) antigens has been shown to be of diagnostic value. An elevated IgA anti-VCA titer is highly sensitive (95%), and an elevated IgA anti-EA is highly specific (>95%). The value as tumor marker in evaluating tumor eradication and detection of recurrence has not been established. The correlation is stronger in WHO types II and III. EBV DNA titers, on the other hand, seem to be an important index for prognostication. The implications

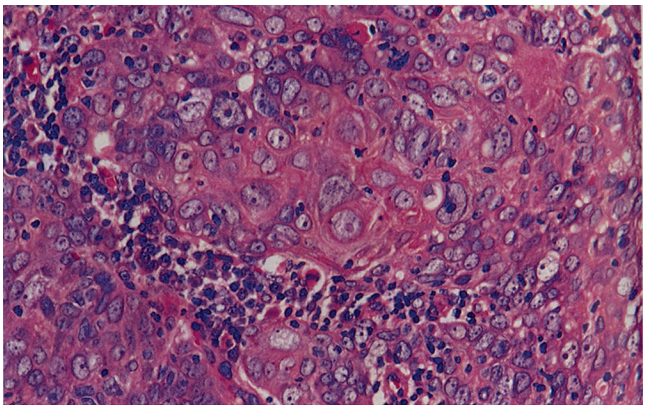


Fig. 22.2 On high-powered microscopy: Large pleomorphic nuclei with prominent nucleoli. The cell borders are distinct and separated by intercellular bridges.

of this test are in the screening of large populations in areas where nasopharyngeal carcinoma is endemic. In addition, in cases of an unknown primary with metastatic cervical adenopathy, EBV titers may suggest NPC and directed biopsies in the search for the primary tumor. EBV titers were not drawn on this patient.

◆ Diagnosis

1. Nasopharyngeal carcinoma, WHO type I, stage II B: T2b N1 M0.
2. Left serous otitis media

Staging of nasopharyngeal carcinoma (**Table 22.1**)

◆ Medical Management

Appropriate histopathologic diagnosis and staging are essential before any treatment is initiated.

Because of the location and anatomy and the good response, external-beam radiotherapy has been the primary treatment modality for NPC. The surrounding structures limit the dose of radiation. In general, the dose given to the primary tumor is in the range of 65 to

Table 22.1 Staging of Nasopharyngeal Carcinoma

Primary Tumor (T)	
TX	Primary tumor cannot be assessed
T0	Evidence of primary tumor
Tis	Carcinoma in situ
T1	Tumor confined to the nasopharynx
T2	Tumor extends to soft tissues
T2a	Tumor extends to the oropharynx and/or nasal cavity without parapharyngeal extension
T2b	Any tumor with parapharyngeal extension
T3	Tumor involves bony structure and/or paranasal sinuses
T4	Tumor with intracranial extension and/or involvement of cranial nerves, infratemporal fossa, hypopharynx, orbit, or masticator space
Regional Lymph Nodes (N)	
NX	Regional lymph node cannot be assessed
N0	No regional lymph node metastasis
N1	Unilateral metastasis in lymph node(s), 6 cm or smaller in greater dimension, above the supraclavicular fossa
N2	Bilateral metastasis in lymph node(s), 6 cm or smaller in greater dimension, above the supraclavicular fossa
N3	Metastasis in a lymph node(s) >6 cm and/or to supraclavicular fossa
N3a	>6 cm in dimension
N3b	Extension to the supraclavicular fossa
Distant Metastasis (M)	
MX	Distant metastasis cannot be assessed
M0	No distant metastasis
M1	Distant metastasis

(*Continued on page 104*)

Table 22.1 (*Continued*)

Stage Grouping			
0	Tis	N0	M0
I	T1	N0	M0
IIA	T2a	N0	M0
IIB	T1	N1	M0
	T2	N1	M0
	T2a	N1	M0
	T2b	N0	M0
	T2b	N1	M0
III	T1	N2	M0
	T2a	N2	M0
	T2b	N2	M0
	T3	N0	M0
	T3	N1	M0
	T3	N2	M0
IVA	T4	N0	M0
	T4	N1	M0
	T4	N2	M0
IVB	Any T	N3	M0
IVC	Any T	Any N	M1

75 Gy and to the neck, 65 to 70 Gy. In elective neck treatment, the dose will be only 50 to 60 Gy. Stereotactic radiosurgery boosts in the form of intracavity brachytherapy help control local rates in T1 and T2 disease and also may help T3 and T4 disease.

Chemotherapy used concurrently with radiotherapy is the treatment of choice in advanced locoregional disease. Combination of cisplatin and 5-fluorouracil is the most widely used and studied.

◆ Surgical Management

Surgical management is infrequent. Neck dissection may be required in patients whose primary tumor has been controlled and residual cervical metastases are palpable. Many surgical approaches to the nasopharynx were described. Transpalatal, transmaxillary, and transcervical are the most accepted. The anterolateral approach or the maxillary swing procedure has been used for salvage nasopharyngectomy.

A myringotomy with tube insertion should be performed on the patient with serous otitis. This will immediately relieve the symptoms of fullness and decreased hearing. Clinicians wishing to do so should obtain an audiogram before surgery on the ear, although microscopic examination in conjunction with Weber and Rinne tests with 512- and 1024-Hz tuning forks should suffice.

◆ Rehabilitation and Follow-up

The patient should be evaluated 10 weeks postradiation and chemotherapy for any persistent disease and then followed up with monthly examinations (with endoscopy) for 1 year after completion of treatment. Bimonthly examinations occur during the second year of follow-up, with trimonthly examinations during the third year. Yearly examinations are

performed after this time for the duration of the patient's life. PET-CT scan is of great value for detecting persistence and recurrence. Biopsy is helpful in suspected cases.

Long-term sequelae of radiotherapy may develop, including neuroendocrine, auditory, xerostomia, and soft tissue fibrosis.

◆ Questions

1. Nasopharyngeal carcinoma may present with all of the following except:
   A. Blood-stained nasal discharge and unilateral nasal obstruction
   B. Unilateral or bilateral cervical lymphadenopathy
   C. Unilateral sensorineural hearing loss
   D. Lower cranial nerve paralysis (XI, X, XI, and XII)
   E. Distant metastasis to the bone, lung, or liver

2. Epstein-Barr virus is correlated strongly with these types of nasopharyngeal carcinoma:
   A. Keratinizing squamous cell carcinoma of the nasopharynx, World Health Organization (WHO) type I
   B. Nonkeratinizing carcinoma of the nasopharynx, WHO type II
   C. Undifferentiated carcinoma of the nasopharynx, WHO type II
   D. Both B and C
   E. All of the above

3. A strong association has been established between nasopharyngeal carcinoma and Epstein-Barr virus (EBV). Which of the following is correct about EBV serologic study?
   A. EBV titers may be helpful in posttreatment surveillance.
   B. Elevated IgA anti-EA titer is highly specific (>95%).
   C. Elevated IgA anti-VCA titer is highly sensitive (95%).
   D. EBV DNA titer seems to be an important index for prognostication.
   E. All of the above are correct.

Suggested Readings

Chan AT, Teo PM, Leung TW, Johnson PJ. The role of chemotherapy in the management of nasopharyngeal carcinoma. Cancer 1998;82(6):1003–1012

Chu EA, Wu JM, Tunkel DE, Ishman SL. Nasopharyngeal carcinoma: the role of the Epstein-Barr virus. Medscape J Med 2008;10(7):165

Lin HS, Fee WE, Gosselin BJ, Sadeghi N, Slack CL, Meyers AD. Malignant nasopharyngeal tumors. eMedicine, Dec 2007

Paulino AC, Grupp SA, Gross S, Windle ML, Bergstrom SK, Chan HS, Arceci RJ. Nasopharyngeal Cancer. eMedicine, Oct 2008

Sivanandan R, Fee WE Jr. Benign and Malignant Tumors of the Nasopharynx. In: Cummings CW, Schuller DE, et al., eds. Otolaryngology—Head and Neck Surgery, 4th ed. Philadelphia, PA: Elsevier Mosby; 2005

Wei WI. Nasopharyngeal cancer. In: Bailey BJ, Johnson JT, et al., eds. Head and Neck Surgery—Otolaryngology. 4th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2006

23

Mandibular Reconstruction

Yash J. Patil

History

A 43-year-old white man is seen for evaluation of an ulcer in his anterior floor of mouth noted by his dentist. He states that this ulcer has been present for 5 months and is slowly growing. He notes that there is occasional bleeding when he eats. He has pain in his right ear when this area is irritated. The patient has a 40 pack-year history of tobacco use and drinks beer and liquor daily. On physical examination the ulcer extends directly over the anterior mandible, and no plane is palpable between the mandible and ulcer (**Fig. 23.1**). The mandible feels intact and is edentulous. Palpation deeply in the floor of mouth reveals submucosal extension that abuts the inner surface of the mandible and extends to the right of the midline. Overall this firm ulcerative lesion with indistinct borders and submucosal extension is greater than 3.5 cm.

Also, small (0.5 to 1.5 cm) submandibular (level 1) lymph nodes are palpable on the right. They are located in level one adjacent to the submandibular gland and are firm, nontender, and mobile. No other lymph nodes are palpable in the neck.

His dentist noted the ulcer 2 weeks ago and performed a biopsy. The patient's internist received a copy of the pathology report and ordered computed tomography (CT) scans of the neck and chest with intravenous contrast.

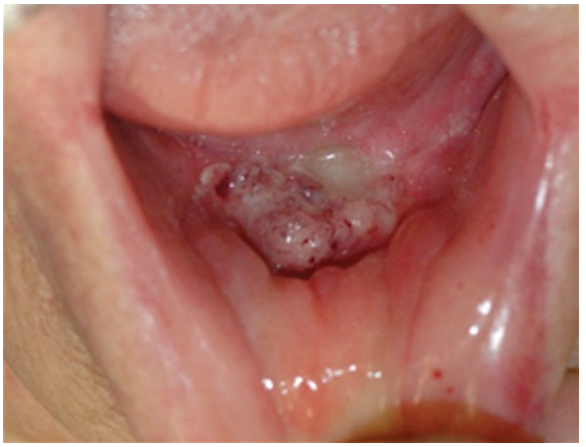
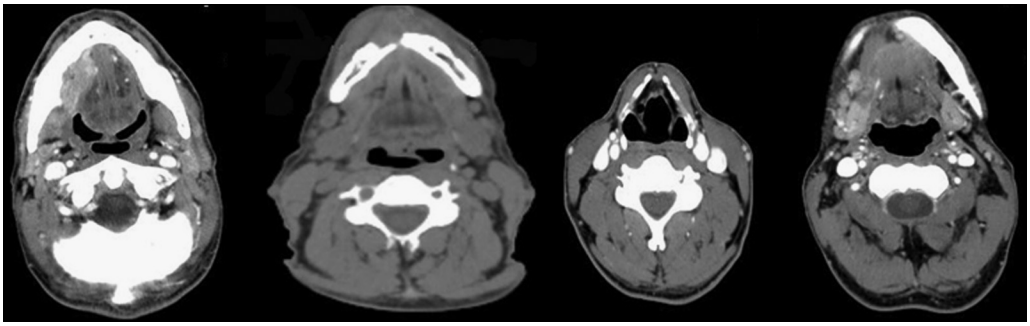


Fig. 23.1 Anterior floor of mouth.



A–D

Fig. 23.2 (A–D) Computed tomography of the neck with contrast.

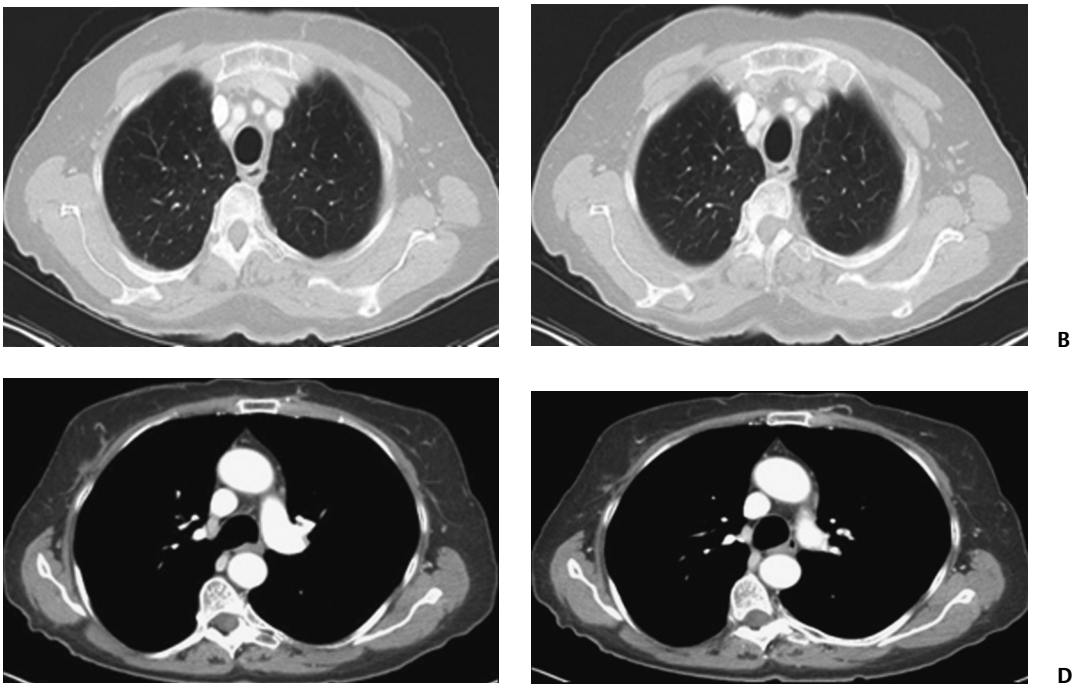
These tests are available for your review (**Figs. 23.2, 23.3, 23.4**).

◆ Differential Diagnosis—Key Points

The biopsy of the oral ulcer reveals well-differentiated squamous cell carcinoma (**Fig. 23.4**). You will note the presence of hyperchromatic cells forming keratin pearls beneath normal-appearing mucosa. Clearly there is invasion below the basement membrane and increased nuclear diameter-to-cytoplasm ratio in these atypical enlarged cells. This is characteristic of invasive squamous cell carcinoma.

◆ Test Interpretation

CT of the neck with contrast demonstrates a mass involving the floor of mouth, mandible, and some oral tongue. There is submucosal



A B C D

Fig. 23.3 Computed tomography of the chest: lung windows (**A,B**) and mediastinal windows (**C,D**).

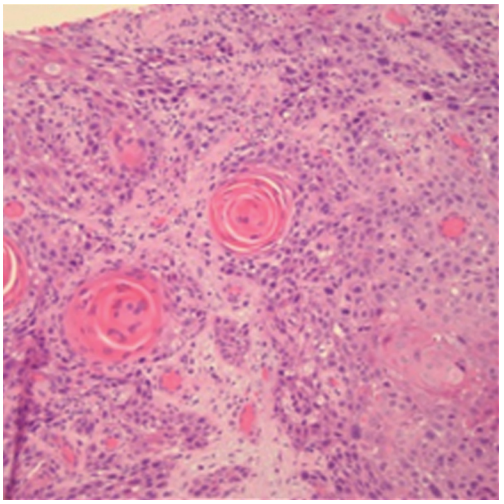


Fig. 23.4 Biopsy of the anterior floor of mouth ulcer.

extension along the right floor of mouth adjacent to the jaw. The mandible demonstrates erosion where the mass overlies the alveolar ridge. The right side of the neck demonstrates at least three lymph nodes adjacent to the submandibular gland.

CT of the chest demonstrates no pulmonary mass or lesion. Mediastinal windows demonstrate no enlarged lymph nodes.

◆ Diagnosis

T4N2bM0 stage IV squamous cell carcinoma of the anterior floor of mouth

◆ Medical Management

Treatment of oral cavity cancers must also take into account expected morbidity of treatment, whether surgical or nonsurgical. In general, primary radiation of oral cavity lesions carries the risk of osteoradionecrosis. For T1 and T2 lesions, surgery and radiation therapy have similar results. When patients are not suitable candidates for surgery or surgery might severely affect function, radiation is certainly an option. For advanced oral malignancies, surgery and radiotherapy in combination provide superior local control than either single modality. By choosing surgery as the initial modality, some effects of higher-dose radiotherapy can be avoided.

Neck observation, the "wait and see approach," is a poor choice for management of most early and advanced oral malignancies. The presence of occult nodal metastasis in a clinically negative neck is reported to be 20 to 44%. Diagnosis of these metastases is possible only by histologic evaluation after neck dissection. There is no uniform consensus on how much invasion at the primary site indicates a need for neck treatment or whether to measure invasion histologically or radiologically. Depth of invasion cutoffs range between 1.5 and 10 mm, and modalities such as ultrasound, CT, and magnetic resonance imaging (MRI) have all been studied. Furthermore, tumor characteristics such as differentiation, vascular invasion, perineural invasion, inflammatory reaction, and pattern of invasion have all been studied without consensus as to appropriate management of the neck. In general, observation of the neck should be considered in only the most superficial oral cancers as long as more aggressive features such as positive margins, lymphovascular invasion, perineural spread, and an aggressive invasion pattern are not noted after resection. The neck should be included when oral cavity cancers are treated with radiotherapy.

◆ Surgical Management

Transoral, composite resection, and pull-through approaches have all been described for resection of oral cancers. For early oral cancers, a transoral approach will generally suffice. When there is extension of tumor onto or close to the mandible, marginal or segmental mandibulectomy must be considered. Historically floor of mouth lymphatics were thought to drain through the mandibular periosteum and bone. As such, en bloc resection of the mandible was thought necessary to control lymphatic spread. Mandibular invasion in fact occurs through direct invasion, which makes preservation of the mandible oncologically feasible in those patients without invasion, even when lymphatic metastasis is present. Numerous investigators have studied the ability of physical examination to predict

mandibular invasion. Upward of 30% of patients with histologically proven mandibular invasion are clinically missed on examination. Although both CT and MRI in some studies have demonstrated high sensitivity and specificity for bone invasion, other studies clearly demonstrate high rates of mandible involvement histologically with no radiographic evidence of bone invasion. Although there is no foolproof method of determining mandibular invasion, segmental mandibulectomy should be considered in irradiated patients with tumor adjacent to the mandible and when clinical examination or radiographs suggest mandibular invasion. Furthermore, segmental mandibulectomy is indicated when there is significant soft tissue disease along the lingual and buccal cortices of the mandible or evidence of spread along the inferior alveolar nerve is seen on CT or MRI. Lastly, segmental mandibulectomy is indicated in older edentulous patients with significant loss of vertical mandibular height because marginal mandibulectomy would leave insufficient bone. The practice of "stripping" malignancy off the mandible and then sampling periosteum should generally be avoided. It must be emphasized that en bloc resection is a central theme in the surgical treatment of oral cancers.

Segmental mandibulectomy defects that extend to include the symphysis must be reconstructed for cosmetic, functional, and oncologic safety. Oncologically, advanced oral malignancies that necessitate segmental resection of the anterior mandible will generally require postoperative treatment with radiation or chemoradiation. Poor reconstruction will result in poor healing. Exposed bone edges, infection, and orocutaneous fistula will delay timely delivery of radiation. Mandibular plates spanning the symphysis extrude up to 80% of the time when unsupported by vascularized bone. Loss of anterior mandibular projection causes loss of support of the soft tissues around the chin. This cosmetic result is simply not acceptable by current reconstructive standards. Lastly, these patients will be dependent on gastrostomy and tracheostomy tubes if they are not reconstructed appropriately.

The most common alloplastic materials for mandibular reconstruction are titanium plates. These plates maintain normal occlusion of the remaining mandible. Frequently a regional flap such as a pectoralis flap or a soft tissue free flap is "wrapped" around the reconstruction plate to help prevent plate exposure. With these techniques, titanium plate reconstructions of the lateral and anterior mandible extrude around 30 to 80% of the time, respectively. In comparison, vascularized bone-containing flaps reconstruct the mandible with a greater than 90% success rate. The use of nonvascularized bone grafts also fail, especially when exposed to radiation therapy. Free tissue reconstruction is the gold standard for mandibular reconstruction.

Anterior mandible defects are best reconstructed using microvascular free tissue. Fibula, scapula, iliac crest, and radial forearm osteocutaneous free flaps may be used to reconstruct most bone and soft tissue defects of the oral cavity. Both fibula and iliac crest provide sufficient bone to support dental implants. The bone that is harvested using radial forearm osteocutaneous free flap is generally sufficient only for small bony defects that do not involve the anterior mandible. Also, the donor site requires plating to support the radius bone lest it fracture. Scapula free flap can be harvested with a considerable soft tissue component. When facial skin is resected in addition to oral mucosa, tongue, and mandible, the scapula donor site provides an excellent source of skin, soft tissue, and bone. The appropriate bone-containing free flap used to reconstruct oromandibular defects is determined by vascular anatomy, body habitus, patient and surgeon preference, and characteristics of the defect.

◆ Rehabilitation and Follow-up

Although one might argue that early detection of a recurrence or second primary cancer might not correlate with improved survival, surveillance should continue for no less than 2 years. Risk of second primary tumor (2 to 4% per year) continues to be a significant source of morbidity for these patients. In general, in the first year after treatment, patients should be seen every 1 to 3 months. In the second year, patients should be seen every 2 to 4 months. In the third year, follow-up should be scheduled at 3- to 6-month intervals.

In the fourth and fifth years after treatment, appointments should be made every 4 to 6 months. After 5 years, yearly appointments are acceptable. Also, at minimum, yearly chest radiographs and liver function tests should be obtained. Thyroid function tests should be checked yearly in patients who received radiation to the low neck. Lastly, symptoms should warrant further workup during this surveillance period.

Head and neck oncologic and reconstructive surgery can cause impaired speech, deglutition, and cranial nerve function. Effective rehabilitation is best accomplished with experienced speech, swallowing, and physical therapists.

◆ Questions

1. Donor sites used for the harvest of vascularized bone-containing flaps for mandibular reconstruction include all of the following except:
- **A.** Radial forearm flaps
- **B.** Pectoralis major flaps
- **C.** Fibula flaps
- **D.** Scapula flaps
- **E.** Iliac crest flaps

2. Loss of anterior mandibular projection causes all of the following except:
- **A.** Severe dysphagia likely necessitating gastrostomy tube
- **B.** Severe respiratory changes likely necessitating trachestomy
- **C.** Severe cosmetic changes
- **D.** Severe articulation changes
- **E.** Loss of sensation over the mentum

3. Resection of the mandible is required in the treatment of oral cancer when:
- **A.** There are obvious lymphatic metastases
- **B.** The tumor invades deeply close to the anterior mandible
- **C.** The patient is edentulous
- **D.** Lingual nerve paralysis is evident
- **E.** The inner cortex of the mandible is involved

Suggested Readings

Burkey BB, Coleman JR Jr. Current concepts in oromandibular reconstruction. Otolaryngol Clin North Am 1997;30(4):607–630

Mehta RP, Deschler DG. Mandibular reconstruction in 2004: an analysis of different techniques. Curr Opin Otolaryngol Head Neck Surg 2004;12(4):288–293

Shah JP. The role of marginal mandibulectomy in the surgical management of oral cancer. Arch Otolaryngol Head Neck Surg 2002;128(5):604–605

Wax MK, Bascom DA, Myers LL. Marginal mandibulectomy vs segmental mandibulectomy: indications and controversies. Arch Otolaryngol Head Neck Surg 2002; 128(5):600–603

24

Carotid Artery Rupture

Yash J. Patil and Judith C. McCaffrey

History

Profuse bleeding is noted from the right neck of a 55-year-old white man who underwent total laryngectomy, partial right pharyngectomy, and bilateral neck dissection 2 weeks ago, with adequate margins of resection. Radiation and chemotherapy were given 6 months ago as part of a laryngeal preservation protocol. Surgical resection was scheduled after tumor persistence was diagnosed on endoscopy 1 month ago. The patient noted worsening dysphagia, weight loss, and right otalgia since the completion of chemoradiotherapy. The postoperative course was complicated by right lower-lobe pneumonia and a pharyngocutaneous fistula. The fistula is located at the inferior aspect of the apron flap 4 cm to the left of the stoma. It has been treated conservatively with packing four times daily during the past week. The fistula extends superiorly for 3 to 4 cm. The packing was changed the night before, at which time the carotid artery was noted to be visible in the open wound (**Fig. 24.1**).

On arrival to the patient's room, there is a large amount of blood covering the patient's gown, bed, and several towels. The patient's blood pressure is 90/50 with a pulse of 120. There is active hemorrhage coming from the patient's neck wound.

Differential Diagnosis—Key Points

Bleeding from the neck in the postoperative setting is a potentially life-threatening situation. The differential diagnosis for such bleeding includes bleeding from the internal, common, or external carotid artery; jugular vein rupture; tracheoinnominate fistula; or, rarely, upper

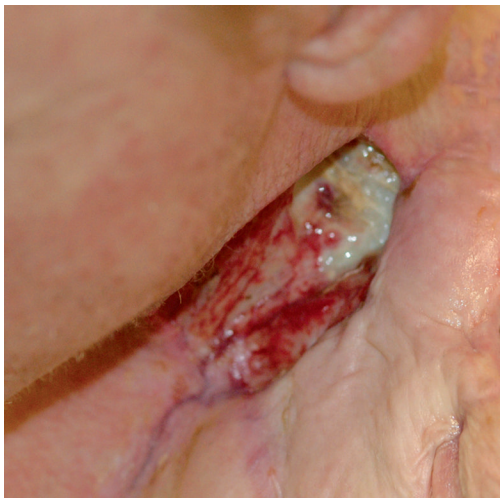


Fig. 24.1 Exposed cartoid artery and fistula

gastrointestinal bleed. Occasionally bleeding from granulation tissue or the transverse cervical system can create bleeding, but it is typically less dramatic.

Carotid artery rupture is potentially a life-threatening emergency. Reported mortality varies between 3 and 50%. Severe neurologic deficits are seen in 16 to 50% of survivors. Risk factors for carotid artery rupture include pharyngocutaneous salivary fistula, previous radiation therapy to the head and neck, head and neck ablative surgery, incompletely resected tumor in proximity to the carotid, and local infection. Most patients will have a "herald" bleed hours before carotid artery rupture. Local wound infection plays an important role in carotid artery rupture. Organisms that have been implicated include *Pseudomonas* spp., *Klebsiella* spp., *Staphylococcus aureus*, *Enterococci*, and anaerobes. When a pharyngocutaneous fistula is noted after tumor ablation in a radiated patient, every step should be taken to exclude and cover the vessels from the stream of saliva, prevent desiccation, and prevent local trauma from wound care.

The prevention of carotid artery rupture begins in the preoperative period. Often the size of the primary tumor and neck metastases necessitates extensive resection and neck dissection. Tumors of this nature typically affect oral intake. To maintain an adequate nutritional status, evaluation by a nutritionist, supplementation, and occasionally placement of a feeding tube preoperatively may be used.

Radiotherapy affects postoperative wound healing. Fibrosis, devascularization, and impaired wound healing are commonly seen in the radiated neck and contribute to carotid artery rupture.

Skin-flap viability is also compromised in radiated patients. Smoking cessation preoperatively and certainly postoperatively may decrease the risk of skin-flap loss. Careful consideration should be given to the placement of surgical incisions. Incision should not overlie the carotid artery and cross the vessel only once. Trifurcations over the carotid are to be avoided. Typically an apron flap will suffice. If it is necessary to "drop a limb," avoid placing it over the carotid.

Several intraoperative steps may aid in prevention of carotid artery rupture. When neck dissection is performed, the surgeon should avoid stripping the adventitia from the carotid artery. The carotid artery derives 80% of its blood supply from the adventitia, and its removal leads to ischemia. Coverage of the carotid artery with dermal grafts, regional flaps, or free flaps should be performed routinely in cases where the carotid is exposed in continuity with oral cavity or oropharyngeal defects. Watertight mucosal closure is imperative in preventing pharyngocutaneous fistula and wound infection. Once again, regional or free tissue reconstruction should be used to create a tension-free closure.

A sentinel bleed is noted before most carotid artery ruptures. This may be seen as blood-tinged oral secretions, blood clot in the mouth, or bleeding during a dressing change. Typically carotid artery rupture will occur within hours of the sentinel bleed. Once a sentinel bleed is noted, appropriate steps should be taken. The patient should be monitored in a setting where nursing staff are familiar with the care of head and neck patients. Blood should be available in a timely fashion, and the patient should be instructed not to strain or move about unnecessarily. Plans for immediate carotid artery exploration and coverage or endovascular intervention should be made.

◆ Test Interpretation

Elective cerebral angiogram before carotid rupture may provide insight into collateral circulation across an intact circle of Willis. Patients with no option for coverage of an at-risk carotid should undergo angiography and a carotid stent should be considered. Use of a stent, even in the presence of tumor, may decrease the incidence of carotid artery blowout and death. Stenting might not decrease the incidence of stroke, especially in patients with recurrent tumor, chronic wounds, and local wound infection. In this setting, the stent may extrude. Despite this, retrospective studies seem to indicate that rupture is much less frequent. If balloon-occlusion studies demonstrate collateralization, exclusion of the at-risk carotid may be considered, especially if further wound breakdown is noted. If collateral flow is not present, depending on the patient's prognosis, surgical reconstruction may be considered.

Cerebral angiography has little role in the management of acute carotid artery rupture because brain ischemia cannot be corrected in a timely fashion. Revascularization procedures involving the cervical carotid or as part of an intracranial-extracranial bypass may potentiate a hemorrhagic component to a preexisting ischemic stroke. Once rupture has occurred, control of bleeding is of paramount importance.

◆ Diagnosis

Rupture of the common carotid artery

◆ Medical Management

Immediate response by physicians and nursing staff is crucial in this life-threatening emergency. Airway, breathing, and circulation should be addressed in a coordinated and calm fashion. It is helpful to have a "crash cart" and tracheostomy tray brought to the room.

Digital pressure should be applied in the wound to control bleeding. Attempting to clamp the carotid artery in this setting is not advisable. Friable tissue and lack of visualization may worsen the bleeding. In general, direct pressure until transfer to the operating room will suffice.

Control of the airway by qualified staff is of paramount importance. A suction setup should be present and functioning. If a tracheotomy is present, a cuffed tracheostomy tube should be used to control and exclude the airway. In laryngectomy patients, the stoma should also be controlled with a cuffed tube. In the case of intraoral rupture, bleeding can be controlled with a tonsil sponge as external digital pressure is applied. If this is not successful, the airway should be kept clear of blood and clot with suction. Oxygen therapy with pulse oximetry should be initiated. In the case of respiratory arrest, the airway is secured with orotracheal intubation or tracheostomy with ambu-bag ventilation.

Simultaneously, large-bore IV access, 14- or 16-gauge peripheral intravenous (IV) catheters, or central venous access should be secured. As this is done, complete blood count and chemistries are sent. A type and cross for 4 units of blood should be drawn at this time. Balanced saline should be rapidly infused to stabilize blood pressure and pulse. Blood products are requested when the operating room is notified. These steps should be performed by physicians and nursing staff simultaneously. Commonly the patient maintains consciousness throughout the ordeal.

◆ Surgical Management

This patient with acute carotid artery rupture should be transported immediately to the operating room with a physician in attendance. The patient is placed under general anesthesia as fluid resuscitation continues. Hypotension and subsequent cerebral hypoperfusion should be avoided during both the intraoperative and the postoperative period. IV access is finalized and the patient is prepped and draped with continuous digital pressure still controlling the bleeding source. The goal of surgical exploration in this setting is to ligate the carotid and exclude the proximal and distal stumps from the infected wound and salivary fistula. The carotid artery should be isolated and dissected proximally and distally until suitable sites for ligation are identified. Ligation with a vascular stapling device or running prolene is simple and minimally traumatic. Once ligation is accomplished, it is prudent to cover the arterial ends with a regional flap and attempt closure of the pharyngocutaneous fistula. A pectoralis flap is an excellent source of regional tissue that may be harvested for this purpose. The use of a controlled fistula to direct saliva away from the stumps of the carotid artery should also be considered.

◆ Rehabilitation and Follow-up

Even after successful management of an acute carotid artery rupture, the patient may require long-term care for neurologic sequelae. In patients with recurrent unresectable malignancy, palliative care, including hospice, should be initiated. When carotid artery rupture occurs after ablative surgery or secondary to soft tissue necrosis after radiotherapy, routine cancer treatment and rehabilitation should be

initiated after the patient recovers. In summary, carotid artery rupture is a dreaded complication of head and neck surgery. In general, carotid artery rupture can be prevented with the steps outlined above. When faced with carotid artery rupture, coordinated teamwork directed toward timely surgical intervention can be lifesaving.

◆ Questions

1. Initial management of carotid artery rupture includes:
   - **A.** Applying direct pressure at the bleeding site
   - **B.** Securing the airway
   - **C.** Obtaining large-bore intravenous access
   - **D.** Notifying the operating room staff
   - **E.** All of the above
2. Which of the following is not a risk factor for carotid artery rupture?
   - **A.** Pharyngocutaneous fistula
   - **B.** Local wound infection
   - **C.** Previous radiation therapy
   - **D.** Tracheostomy
   - **E.** Neck dissection
3. Which of the following steps may help prevent carotid artery rupture?
   - **A.** Avoidance of trifurcating neck incisions
   - **B.** Maintaining a fascial covering over the carotid artery during neck dissection
   - **C.** Providing adequate nutrition in the perioperative period
   - **D.** Covering the carotid with vascularized tissue or dermal grafts
   - **E.** All the above

Suggested Readings

Cohen J, Rad I. Contemporary management of carotid blowout. Curr Opin Otolaryngol Head Neck Surg 2004;12(2):110–115

Simental A, Johnson JT, Horowitz M. Delayed complications of endovascular stenting for carotid blowout. Am J Otolaryngol 2003;24(6):417–419

Upile T, Triaridis S, Kirkland P, et al. The management of carotid artery rupture. Eur Arch Otorhinolaryngol 2005;262(7):555–560

25

Tonsillar Carcinoma

Alex Senchenkov and Keith M. Wilson

History

A 62-year-old carpenter arrives for evaluation of progressive and persistent sore throat, foul breath, odynophagia, and right-sided otalgia for 3 months. He reports unintentional weight loss of 25 pounds. He has a 40 pack-year smoking history and drinks socially. He is otherwise healthy. On physical examination, he has a friable, ill-defined, ulcerated mass in the right tonsillar area that extends into the soft palate and base of tongue mucosa (**Fig. 25.1**). The tumor is 4.5 cm in greatest dimension and mobile in relevance to the ramus of the mandible. There is no trismus. He has a 3-cm single, enlarged, firm upper jugular lymph node (level II). His examination is otherwise normal.

Differential Diagnosis—Key Points

1. This patient's overall presentation is highly suggestive of oropharyngeal malignancy:
   - **A.** Primary tumors cause progressive compression and invasion of surrounding structures and manifest as sore throat, otalgia, and odynophagia.
   - **B.** Ulceration frequently implies malignant nature of the disease process whereby tumor tissue outgrows its blood supply and necroses.
   - **C.** Cervical adenopathy is suggestive of regional metastatic disease.
2. *Histologic verification of diagnosis* is a crucial part of the oncologic workup of a primary tumor. Although squamous cell carcinoma accounts for more than 90% of oropharyngeal tumors, with the tonsil being the most frequent primary locale, other uncommon tumor types of the tonsil may have important treatment implications, including malignant melanoma, minor

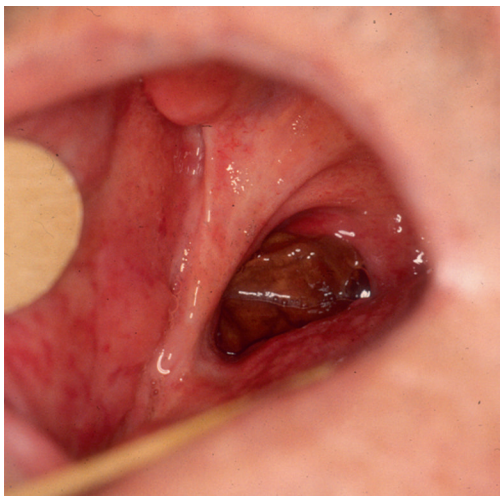


Fig. 25.1 Irregular mass situated in the right tonsil. The tumor extends into the base of tongue.

salivary gland tumor, soft tissue sarcoma, and, importantly, lymphoma. Incisional biopsy should be performed as a part of initial evaluation.

3. *Fine-needle aspiration (FNA) biopsy* with image guidance of (office) ultrasound is an effective way to secure tissue diagnosis of regional adenopathy.
4. Evaluation of the patient's overall condition is focused on the following:
   - **A.** *General health* of a head and neck cancer patient is frequently compromised by a long history of smoking and alcohol use. Careful history is taken to assess the severity of these habits as well as related pulmonary, cardiovascular, and gastrointestinal co-morbidities. The "CAGE questionnaire" (need to cut down on drinking, annoyance, guilt about drinking, need for an eye-opener) should be asked to identify patients at risk for *delirium tremens* to ensure timely prophylaxis. The *Karnofsky score* reflects global assessment of patient's well-being.
   - **B.** *Nutritional status* is often undermined by alcoholism, malignancy, and lack of social support. Unintentional weight loss as well as weight measurement during every visit should be documented. Complete blood cell count, serum albumin and prealbumin, basic metabolic panel, and liver function tests are assessed to determine overall nutritional status.
   - **C.** *Thyroid* screening of a head and neck cancer patient is recommended if he or she has received radiation therapy to the neck or if prior surgery was performed on or near the thyroid gland. Thyroid-stimulating hormone, free triiodothyronine, and free thyroxine levels would be adequate to evaluate thyroid function.
   - **D.** *Dental evaluation* is critical because radiation therapy is frequently a part of the treatment program. Appropriate dental care and extraction need to be arranged before initiation of radiation therapy.
   - **E.** *Social functioning and support* are frequently issues in head and neck cancer patients, and therefore involvement of a social worker in the head and neck team is critical. Patient's home, family, insurance situation, anticipated need for skilled-nursing facility placement, and home visiting nursing care all fall in the social worker's area of expertise.
5. Imaging plays an important role in clinical evaluation of the tumor spread, that is, *clinical staging*. A computed tomography (CT) scan of the head and neck with intravenous contrast is a common first-line study. A chest radiograph needs to be performed as a minimal workup to evaluate for potential lung cancer or metastasis. CT scanning of the chest would be a better screening test to identify pulmonary metastases or primary lung pathology. Magnetic resonance imaging (MRI) is useful in evaluating skull-base, skeletal, and neurovascular involvement. Positron emission tomography (PET), particularly in conjunction with CT (PET-CT), proved to be an expensive but effective study of regional and distal tumor spread. PET-CT should be considered for patients with advanced regional disease who have higher risk of distant metastasis.
6. Panendoscopy under general anesthesia allows evaluation of the upper aerodigestive tract for second primary tumors. If the patient is to be treated surgically, panendoscopy can be performed at the time of the resection. However, if the patient is a candidate for chemoradiation treatment, panendoscopy can be effectively combined with dental extraction and gastrostomy tube placement and a central venous access port. Likewise, additional biopsy should be performed if needed.

◆ Test Interpretation

1. Biopsy of the primary tumor in the office was consistent with moderately differentiated keratinizing squamous cell carcinoma. Likewise, squamous cell carcinoma cell clusters were identified in the FNA biopsy of the ipsilateral cervical lymph nodes. This histologically confirms the nature of the primary disease process and regional adenopathy.
2. CT scan revealed a $4.0 \times 4.5 \times 4.0$ cm mass occupying the right tonsillar fossa with an intact soft tissue plain between the tumor

and the mandible. Base of the tongue and soft palate are involved. There is also a 3.5-cm level 2 lymph node with central necrosis and multiple nodes smaller than 3 cm that are level 2 and 3. No retropharyngeal or parapharyngeal adenopathy is present. Chest radiograph is unremarkable.

◆ Diagnosis

Stage IV (T3N2bM0) squamous cell carcinoma of the right tonsillar fossa (**Table 25.1**)

◆ Medical Management

Combined modality therapy consisting of surgery and radiation therapy has been the mainstay of treatment of oropharyngeal cancers for several decades. There are two reasons for the limited popularity of surgical resection as an upfront treatment. First, wide surgical resection in this area is associated with a significant disturbance in swallowing function and possibly speech. Second, surgical resection, as a single-modality treatment, does not provide adequate clearance of the parapharyngeal and retropharyngeal lymphatic-bearing tissues to which oropharyngeal tumors can drain. Therefore, the optimal treatment strategy is individual and based on the extent of the tumor as well as patient preference.

Most tonsillar cancers are radiosensitive, especially tonsillar nonkeratinizing, poorly differentiated squamous cell carcinoma (lymphoepithelioma) that frequently presents with cystic neck metastases. Human papillomavirus (HPV; particularly subtypes 16 and 18) is associated with the development of tonsillar carcinoma. These HPV-related tonsillar cancers occur in younger individuals; in the absence of usual risk factors, these cancers are more susceptible to radiation therapy and generally carry a more favorable prognosis.

Early tonsillar cancer (T1N0M0, stage I) can be treated with radiotherapy as a single modality with durable results. Likewise, in intermediate stages such as T1N1, T2N0–1, and exophytic T3N0, certain radiotherapy techniques

Table 25.1 Staging of tonsillar carcinoma

Primary Tumor (T)	
T1	Primary tumor <2 cm in greatest dimension
T2	Primary tumor 2–4 cm in greatest dimension
T3	Primary tumor >4 cm in greatest dimension
T4	Primary tumor involving surrounding structures: bone, soft tissue of the neck, tongue
Regional Lymph Nodes (N)	
NX	Nodal status cannot be determined
N0	No evidence of nodal metastasis
N1	Metastasis to a single ipsilateral node ≤3 cm
N2a	Metastasis to a single ipsilateral node 3–6 cm in greatest dimension
N2b	Metastases to multiple ipsilateral nodes, none >6 cm
N2c	Metastases to bilateral or contralateral nodes, none >6 cm
N3	Metastasis to a lymph node >6 cm
Distant Metastasis (M)	
MX	Distant metastasis cannot be determined
M0	No distant metastasis
M1	Distant metastasis

can achieve 73 to 94% 5-year survival and locoregional control. Concurrent administration of radiation and chemotherapy aims to sensitize tumor cells with chemotherapy and thus make them more susceptible to radiotherapy improving locoregional antitumor effect. The unfortunate downside is a substantial increase in both acute and late morbidity of radiation that can be very troublesome to patients. Importantly, patients need to be medically fit to undergo concurrent chemoradiation therapy, which limits the number of patients eligible for this treatment plan.

◆ Surgical Management

Surgical resection is considered for patients with large-volume disease, particularly involving the mandible; patients who are not amenable or unwilling to undergo chemoradiation therapy; and patients who either had recurrence or failed to respond completely to radiation-based treatments. Two important concepts the surgeon needs to bear in mind are (1) *resectable tumor*, which implies that the extent of the tumor is such that it can be removed in its entirety with the goal of achieving negative margins; (2) *operable patient*, which means the patient's mental and physiologic reserve is sufficient to undergo the proposed surgical procedure.

Tumor resection must be performed with thorough understanding of three-dimensional anatomy of the oropharynx. Usually a 1-cm cuff of normal tissue around visible or palpable tumor is included in the resection with the goal of achieving tumor-free margins around the *entire* tumor bed, confirmed with intraoperative frozen section analysis. Preoperative imaging and surgical exposure are critical. Smaller, mobile tumors in the patients whose mouth can widely open are amenable to *transoral* approach. The resection can be facilitated by *transcervical* approach during the neck dissection whereby the vessels are controlled from below to allow wide resection of the tonsillar tumor. For larger tumors, tumors involving the mandible, and patients presenting with trismus, the resection should be done though mandibulotomy or segmental resection of the involved portion of the mandible. In general, mandibulotomy is the gold standard for comprehensive three-dimensional resection of oropharyngeal tumors.

Radial forearm free flap allows the transfer of thin and pliable skin and soft tissue of the forearm and has become the "workhorse" flap in pharyngeal reconstruction. Larger defects involving the mandible can be rebuilt with osteocutaneous free fibula flap if the patient has dentition and life expectancy greater than 6 months. Otherwise, the mandible is left unreconstructed and the defect is filled with a larger soft tissue flap such as anterolateral thigh or rectus abdominis musculocutaneous free flap.

Pectoralis major musculocutaneous flap (*pec flap*) is reliable, technically straightforward, faster to perform, and does not require microsurgical expertise. The downside is higher rate of leakage from suture line dehiscence caused by gravity and bulk of this flap. Although it is still a good reconstructive option, it is becoming the second-line flap in pharyngeal reconstruction.

Radical operations for oropharyngeal cancers frequently require tracheostomy for secretion and airway control and gastrostomy for nutritional access. Postoperative (adjuvant) radiation therapy is routinely added to treatment protocols unless the patient had undergone radiotherapy before the resection. Patients with unresectable disease can be treated with palliative chemotherapy and radiation therapy to slow the growth of the tumor and alleviate pain.

◆ Rehabilitation and Follow-up

The intensity of the follow-up is proportional to the patient's risk of recurrence. Most of the recurrences occur during the first 2 years following completion of treatment; therefore, patients are seen every 1 to 2 months for the first 2 years and every 3 to 6 months thereafter. In general, head and neck cancer patients should be followed up for a minimum of 5 years. Many recommend lifetime surveillance.

Follow-up care of patients treated with chemoradiation may present a clinical challenge. Radiation-induced fibrosis, scarring, contracture of the neck, limited range of the mandible, and chronic pain and edema all limit sensitivity and accuracy of physical examination. The head and neck surgeon needs to be

attuned to appearance or worsening in such symptoms as otalgia, dysphagia, odynophagia, loss of weight, and increasing fatigue, which can be early symptoms of recurrent tumor.

PET-CT becomes an important imaging modality in follow-up care of pharyngeal cancer patients after radiation-based treatments. Although some residual activity may be seen during the first posttreatment scan at 8 to 12 weeks, it should decrease with subsequent studies. Failure to do so should raise the concern of incomplete response that may require salvage surgery. Patients with advance neck disease (N2–3) are at increased risk to recur locally and distantly and should be followed regularly for persistent or recurrent disease.

Rehabilitation of head and neck cancer patients in general and tonsillar cancer patients in particular can frequently be a challenge that requires the approach of the specialty team, including social workers, head and neck nurses, dentists, speech and swallowing therapists, physical therapists, and nutritionists. However, the head and neck oncologic surgeon needs to recognize specific problems and refer the patient to the appropriate members of the rehabilitation team.

◆ Questions

1. Dental evaluation before radiation therapy for tonsillar cancer is necessary because
   A. Dental health is usually poor in head and neck cancer and patients can be lost to follow-up.
   B. Optimizing dentition will minimize dental decay from radiation.
   C. Dental extractions in irradiated patients may cause osteoradionecrosis.
   D. It is more important to address maxillary teeth because they are more likely to cause problems following radiation therapy.
   E. Diseased teeth that could be successfully treated and salvaged before radiation therapy never need to be extracted.

2. In a nonsmoking, nondrinking patient with tonsil cancer, what is the most likely cause of the cancer?
   A. Genetic predisposition
   B. Environmental exposure, such as household radon
   C. Viral infection, such as human papillomavirus (HPV)
   D. Vitamin D deficiency
   E. Chronic tonsillitis

3. The patient presented with T1N0M0 (stage 1) squamous cell carcinoma of the tonsil and was treated with radiation therapy. He initially responded but later relapsed and, despite the salvage treatment, developed first cervical and then distant metastases in the lungs, liver, and bones. His disease is now T0N2bM1 (stage IV).
   A. True
   B. False

Suggested Readings

Bradford CR, Futran N, Peters G. Management of tonsil cancer. Head Neck 1999;21(7):657–662

Jones AS, Fenton JE, Husband DJ. The treatment of squamous cell carcinoma of the tonsil with neck node metastases. Head Neck 2003;25(1):24–31

Myers EN, Suen JY, Myers JN, Hanna EYN. Cancer of the Head and Neck, 4th ed. Philadelphia, PA: WB Saunders, 2003

Silver CE, Rubin JS. Atlas of Head and Neck Surgery, 2nd ed. Philadelphia, PA: Churchill Livingstone, 1999

26

Odontogenic Tumors

Eyad Khabbaz and Keith M. Wilson

History

A 39-year-old white man has a history of progressive swelling of the left mandible. The swelling has enlarged over the past 2 to 3 months. Also, he noticed that his left lower teeth were becoming "crooked." His primary care physician ordered a panorex, which revealed a multilocular cyst expanding the left mandible with adjacent sclerosis. No bone erosion was noticed. He was then referred to the ENT outpatient clinic for further evaluation and management. On further questioning, he denies malocclusion or pain and numbness of the teeth or lips. However, he complains of discomfort in the left mandible and difficulty chewing on the left side and difficulty cleaning his teeth on the same side. He has no history of smoking, drinking, or previous exposure to radiotherapy.

On physical examination, swelling of the left mandible with external asymmetry of the lower face was noted. A firm 6 × 4 cm nontender mass is noted intraorally involving the left mandibular body and including both lingual and buccal surfaces. Teeth 17 through 21 are malpositioned. On palpation, crepitus (eggshell cracking) was noted. The overlying mucosa is intact with normal sensation. Externally, there is apparent deformity in the left lower face and submandibular region. Sensation of the lower lip is normal. The intraincisor opening is 45 mm with normal occlusion. Auscultation of the mass reveals no bruit. The remainder of the head and neck examination is within normal limits.

Differential Diagnosis—Key Points

1. Painless swelling of the jaw, either upper or lower, is a common presentation of jaw cysts and tumors. Dysesthesias, pain, and loose dentition suggest infection or malignancy. Another common finding that suggests malignancy is a nonhealing tooth socket after extraction. Osteogenic sarcoma may occur in the mandible or maxilla and may mimic an odontogenic tumor. Mucosal irregularity or ulceration is common in malignancy.
2. Vascular malformations (VMs) of the mandible may manifest as slow growing masses. These lesions are more common in children and are often associated with other physical findings. It is not uncommon for the VM to extend to the tongue with apparent lingual vein distention. A bruit heard on auscultation requires further evaluation with magnetic resonance imaging, magnetic resonance angiography, or angiography. If embolization is chosen

as definitive treatment or before surgical excision, angiography is the best study.

3. The clinical presentation of odontogenic cysts and tumors depends, to a certain extent, on the dimensions of the lesion. A small lesion without symptoms will be frequently missed on routine oral examination. However, evaluation of a devitalized or infected tooth will help diagnose these small cysts. Clinical absence of one or more teeth without a history of extraction may lead to the diagnosis.
4. The most useful radiologic evaluation of the maxilla and mandible is a panorex. It can assist with the differential diagnosis of jaw cysts and tumors; however, definitive diagnosis is made on biopsy and histopathologic examination. Slow growing cysts or benign tumors will have thin, sclerotic bony walls that are well defined. It can cause blunting of tooth roots and displacement of teeth or the inferior alveolar nerve. An aggressive, fast growing, or malignant lesion will show lysis and resorption of surrounding bone, without teeth displacement. It also has ill-defined margins.
5. Computed tomography (CT) is not cost-effective for routine evaluation. It is indicated in evaluating large lesions with significant anatomy distortion. Also, it is important to identify cortex erosion, especially when malignancy is suspected. CT evaluation is routine for pre-excision and reconstruction of advanced cases. CT is frequently necessary to evaluate the maxillary lesions fully.
6. Inflammatory and developmental cysts include periapical, dentigerous, globulomaxillary, and nasopalatine cysts. The most common is the periapical cyst; the dentigerous cyst, often associated with the crown of an unerupted molar, may rarely give rise to ameloblastoma, mucoepidermoid carcinoma, or squamous cell carcinoma. Odontogenic keratocyst (OKC) is considered a multicystic benign neoplasm with an increased propensity for recurrence after inadequate curettage. It is aggressive and can grow quite rapidly. Multiple keratocysts with recurrence may occur with basal cell nevus syndrome (BCNS). The diagnosis of BCNS must be entertained in patients with findings of bifid ribs, hypertelorism, widened nasal dorsum, multiple mandibular OKCs, and early development of basal cell carcinomas of the face and trunk. It is autosomal dominant disease with high penetration.
7. True odontogenic tumors arise from the cells of the enamel organ and include ameloblastoma, calcifying epithelial odontogenic tumor (Pindborg tumor), squamous odontogenic tumor, calcified odontogenic tumor (Gorlin cyst), odontogenic myxoma, cementoblastoma, ameloblastic fibroma, and odontoma. Ameloblastoma is considered the most primitive of the odontogenic tumors, and the odontoma is the most differentiated, showing histologic evidence of tooth formation. Other rarer tumors occur and are not mentioned here.
8. Tumors typically involving the mandible include ameloblastoma, OKC, calcifying epithelial odontogenic tumor, squamous odontogenic tumor, cementoblastoma, and ameloblastic fibroma. Tumors typically involving the maxilla include Gorlin cyst (also affects the mandible) and odontoma.
9. Metastatic tumor from breast carcinoma, renal cell carcinoma, and other primaries, as well as contiguous involvement of oral cavity and oropharyngeal squamous cell carcinoma, may occur in the jaws. A history of concurrent malignancy may suggest this in the differential diagnosis.
10. Ameloblastoma does not have a capsule and is extremely aggressive and infiltrative. Some advocate classifying it as low-grade or indolent malignancy. However, two rare variants exist, and each can metastasize: malignant *ameloblastoma* (benign histopathologically, the metastasis occur through implantation) and *ameloblastoma* carcinoma (with malignant cytologic features).

◆ Test Interpretation

Workup of a patient with a jaw mass includes a full head and neck examination, panorex, computed tomography (CT) scan, and biopsy.

1. Panorex. Multiloculated soap-bubble radiolucencies in the left body of the mandible

extending to the angle. Another possible appearance is a multiloculated lesion with internal septa and a honeycomb. Another less common finding is a single, well-demarcated, well-defined, unilocular, well-corticated, lucent lesion associated with the crown of impacted or unerupted teeth. In this case it is indistinguishable at radiography from odontogenic keratocysts and dentigerous cysts.

2. CT scan. Extensive inner cortical destruction with multiple areas of thinning external cortical bone by a multiloculated cystic mass.
3. Biopsy. Fine-needle aspiration biopsy of a solid jaw mass is difficult, if not impossible, to perform adequately. Therefore, an open biopsy should be performed. Biopsy is either complete enucleation in the case of small lesions or incisional in cases of suspected malignancy or larger tumors where preoperative counseling regarding treatment is necessary. Histopathologic examination of the biopsy is needed for diagnosis.

Biopsy of the tumor in the case presented shows an ameloblastoma arising from the epithelial lining of a dentigerous cyst surrounded by a fibrous capsule (**Fig. 26.1**).

On high-powered microscopy, islands of odontogenic epithelium are seen in a fibrous connective-tissue stroma. The columnar cell nuclei appear polarized away from the basement membrane (**Fig. 26.2**).

Diagnosis

Ameloblastoma left mandibular body

Medical Management

Ameloblastomas are relatively resistant to chemotherapy and radiation therapy. Thus there is no role for medical management of this medical problem.

Surgical Management

The various forms of odontogenic tumors are usually adequately treated with enucleation. In cases of OKC, there is a tendency for recurrence if simple enucleation is performed. A more aggressive approach, with extensive curettage and drilling of all bony loculations, should be performed. The surgeon should not be reluctant to perform a marginal mandibulectomy if there is extensive disease that cannot be adequately accessed with curettage and drilling. Recurrent OKC typically requires segmental mandibulectomy for adequate gross removal.

Opinions in the literature vary regarding the appropriate treatment of ameloblastoma. Although it is a benign disease histologically with rare metastases, ameloblastoma has a propensity for aggressive behavior with significant destruction of local structures. When inadequately treated with simple enucleation or curettage, ameloblastoma recurs in up to

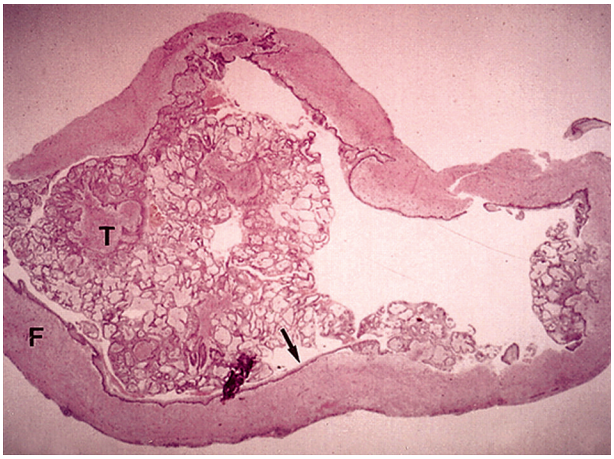


Fig. 26.1 Mural ameloblastoma. Low-power microscopy (hematoxylin-eosin stain) shows an ameloblastoma (*T*) arising from the epithelial lining (*arrow*) of a dentigerous cyst surrounded by a fibrous capsule (*F*).

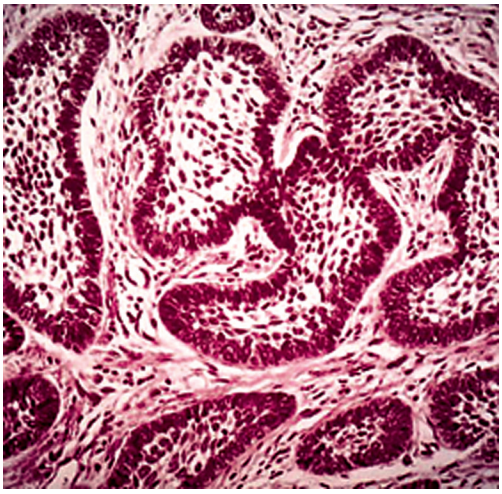


Fig. 26.2 On high-power microscopy, islands of odontogenic epithelium in a fibrous connective-tissue stroma. The columnar cell nuclei appear polarized away from the basement membrane.

50%. In the case of small and moderately sized tumors, a marginal mandibulectomy is performed, leaving a rim of inferior mandible intact. Large, destructive lesions with multiple loculations and obvious bony destruction of the cortices require segmental mandibulectomy for complete removal. One- to 1.5-cm bony margins and one uninvolved overlying anatomic barrier margin are advocated. Negative hard and soft tissue margins increase the cure rate to 98%. Reconstruction is best performed with a free tissue transfer. The most commonly used are free fibula and free scapula flaps. The role for radiotherapy in the management of odontogenic tumors is limited to malignancy, including carcinomas and sarcomas.

◆ Rehabilitation and Follow-up

Close follow-up is required, particularly in cases where enucleation was performed for OKC or ameloblastoma. Recurrence is the most common complication of these tumors, especially when the enamel organ is not entirely removed or wide surgical margins are not obtained. Malignant transformation may develop in the recurrence and, although rare, requires extensive revision surgery for treatment. A 5-year follow-up is mandatory, but a 10-year follow-up is prudent.

◆ Questions

1. Which of the following statements about ameloblastomas is incorrect?
- **A.** The central ameloblastoma can be unicystic, multicystic, or solid.
- **B.** The solid central form can give lung metastasis in rare cases.
- **C.** The peripheral ameloblastoma tends not to invade the alveolar bone.
- **D.** The unicystic form is the most aggressive among the central ameloblastomas.
- **E.** The most common histologic pattern is the follicular.

2. All of these are correct about benign odontogenic tumors except which one?
- **A.** Painless, slowly growing swelling of the jaw with normal mucosa
- **B.** On panorex, blunting teeth roots and displacement of teeth or inferior alveolar nerve
- **C.** Ameloblastoma requires 5 to 10 years of follow-up for the high potential of recurrence.
- **D.** A small lesion can be adequately treated with enucleation.
- **E.** It can cause dysesthesia, pain, and bone lysis and resorption with teeth loosening.

3. In a 43-year-old woman with a swollen jaw, a biopsy revealed ameloblastoma. What is the best treatment for these tumors?
- **A.** Simple enucleation or curettage
- **B.** Radiation therapy
- **C.** Radiation and chemotherapy
- **D.** Complete excision, including marginal, segmental, or partial mandibulectomy to obtain free surgical margins
- **E.** All the resections should be followed by radiation therapy.

Suggested Readings

Chung WL, Cox DP, Ochs MW. Odontogenic cysts, tumors, and related jaw lesions. In: Bailey BJ, Johnson JT, et al., eds. Head and Neck Surgery—Otolaryngology, 4th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2006

Dunfee BL, Sakai O, Pistey R, Gohel A. Radiologic and pathologic characteristics of benign and malignant lesions of the mandible, RSNA Annual Meeting, May 2006

Goldman KE, Tewfik TL, Talavera F, Calhoun KH, Slack CL, Meyers AD. Mandibular cysts and odontogenic tumors. eMedicine, March 2006

Larsen PL. Odontogenic cysts and tumors. In: Cummings CW, Schuller DE, et al., eds. Otolaryngology—Head and Neck Surgery, 4th ed. Philadelphia, PA: Elsevier Mosby; 2005

Sampson DE, Pogrel MA. Management of mandibular ameloblastoma: the clinical basis for a treatment algorithm. J Oral Maxillofac Surg. 1999;57(9):1074–1077

VI

The Larynx

27

Laryngeal Carcinoma

Collin M. Burkart, Alex Senchenkov, and Keith M. Wilson

History

A 62-year-old man presents for evaluation of persistent hoarseness that has lasted over the past 3 months. He reports mild dysphagia and odynophagia but no weight loss or otalgia. He has smoked one pack of cigarettes per day for 40 years and consumed alcohol socially. He is otherwise healthy and employed as a construction worker.

On physical examination, his larynx is mobile. On flexible fiberoptic laryngoscopy, he has a friable, ulcerated mass involving the laryngeal surface of the infrahyoid epiglottis and left false vocal fold. The true vocal folds (TVFs) are crisp and appear clear of tumor. The anterior and posterior commissures, arytenoids, and TVFs appear uninvolved by the tumor. The left TVF mobility is mildly impaired, whereas the right TVF is fully mobile and compensating with good glottic closure (**Fig. 27.1**). There is a left firm palpable enlarged upper jugular node less than 3 cm in diameter. There is no other adenopathy or abnormal masses in the head and neck region appreciated on examinatin. The patient's dentition is poor.

Differential Diagnosis—Key Points

1. This patient's presentation is consistent with a late-stage supraglottic carcinoma. The primary tumor involves the epiglottis and left false vocal fold with early impairment of mobility of the left TVF.
2. Histologic verification of diagnosis is a crucial part of oncologic workup of a primary tumor. Although squamous cell carcinoma is the most likely histologic diagnosis in this patient with increasing age, smoking, and alcohol use, other neoplastic and infectious processes, such as tumors of salivary gland origin (adenoid cystic, mucoepidermoid, and adenocarcinoma), neuroendocrine tumors, sarcomas, tuberculosis, and fungal infections must be ruled out. Incisional biopsy under general anesthesia should be performed as part of the clinical evaluation.
3. Fine-needle aspiration (FNA) biopsy with office ultrasound is an effective way to secure tissue diagnosis of regional adenopathy.
4. Evaluation of the patient's overall condition is focused on general health, nutritional

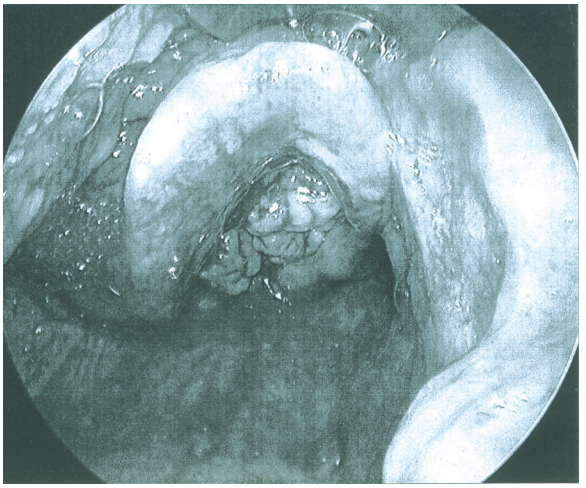


Fig. 27.1 Endoscopic view of the tumor of supraglottic larynx.

status, thyroid, dental evaluation, and social functioning and support.

Test Interpretation

1. Computed tomography (CT) scan of the head and neck with intravenous contrast is a common first-line study. This study revealed a 2.5 × 2.3 × 1.9 cm left supraglottic mass involving the epiglottis and left false vocal fold without involvement of the laryngeal cartilages. There is also a single 2.5 cm ipsilateral level 2 lymph node with central necrosis. No other adenopathy is present.
2. Panendoscopy. Evaluation of the upper aerodigestive tract is performed under topical anesthesia or, more commonly, under general anesthesia specifically to search for a synchronous cancer. If the patient is to be treated surgically, panendoscopy can be performed at the time of the resection. However, if the patient is a candidate for chemoradiation treatment, panendoscopy can be effectively combined with dental extraction and gastrostomy tube placement.
   a. Panendoscopy reveals no second primary. Bimanual palpation reveals fullness in the pre-epiglottic space but normal consistency of the base of the tongue. The laryngeal tumor biopsy obtained during direct laryngoscopy was consistent with moderately differentiated keratinizing squamous cell carcinoma.
3. FNA. FNA of the ipsilateral cervical lymph node reveals squamous cell carcinoma clusters.
4. Chest radiography. The chest radiograph is negative for metastatic foci. In general, the more advanced the stage of locoregional disease, the more vigilant one should be to identify distant metastases. A positron emission tomography (PET)/CT or CT of chest, abdomen, and pelvis could be considered. With only one cervical node, I would probably not go beyond a chest radiograph or a CT scan of the chest.

Diagnosis

Stage III (T2N1M0) squamous cell carcinoma of the supraglottic larynx

Medical Management

Radiation therapy is commonly used as a single-modality treatment for early laryngeal cancer (T1 and T2). Initial radiation therapy usually has a one-time application and often precludes future effective use of partial laryngectomy.

Chemoradiation treatment may include induction chemotherapy that is followed by radiation therapy for responders, as it was in the VAMC study, or concurrent chemotherapy and radiation where chemotherapy is given during the radiation treatment and serves as a

radiosensitizer that augments antitumor effect of radiation and unfortunately its toxicity.

Patients with clinically positive regional lymph nodes (N+) need to be treated, and management of a clinically negative neck (N0) is based on the risk of the metastatic spread. In early glottic cancers, it is safe to observe the neck because the risk of metastasis is low. Prophylactic treatment of the neck is warranted if the risk exceeds 20 to 25%. Generally, the neck in patients with supraglottic and subglottic tumors should be treated. The neck may be treated with radiation therapy as a single modality for N0 or N1 disease or chemoradiation for N2–N3 disease with neck dissection reserved for salvage cases.

Overall, quality of life for patients who retained their larynx is better than in those treated with total laryngectomy. Nevertheless, there is a higher rate of postoperative complications following a salvage laryngectomy among the patients for whom chemoradiation therapy failed. Moreover, despite the fact that most patients treated with chemoradiation therapy retained their larynges, several had poor laryngeal and swallowing functions that came as long-term sequelae of radiation damage exacerbated by chemosensitization.

◆ Surgical Management

Several surgical options are available as a single-modality treatment, including transoral endoscopic laser surgery, open partial laryngectomies (e.g., vertical partial laryngectomy, surpraglottic laryngectomy, and supracricoid laryngectomy), and total laryngectomy.

The main decision in the treatment of early laryngeal cancer is between radiation therapy and endoscopic excision–partial laryngectomy as a primary treatment modality. Overall, partial laryngectomy offers better initial local control, but it has higher cost and higher immediate postoperative morbidity. Transoral endoscopic laser surgery is less expensive than either radiation or partial laryngectomy and has a lower morbidity rate. The advantage of initial surgical treatment is higher immediate local control, fewer long-term sequelae, and that radiation could still be used in the future.

Advanced laryngeal cancer (T3 and T4) requires combined medical and surgical therapy consisting of either total laryngectomy followed by radiation therapy or chemoradiation with a possibility of salvage total laryngectomy. Historically, laryngectomy followed by radiation has been the mainstay of treatment for advanced laryngeal cancer.

Management of laryngeal cancer involves a dilemma of surgery- versus radiation-based treatment strategy in almost every case. The patient needs to understand the significant risks, complications, and side effects of the proposed treatment. Issues such as loss of larynx, postlaryngectomy speech rehabilitation, quality of voice, swallowing, quality of life after chemoradiation, as well as postoperative morbidity after primary versus salvage laryngectomy should be discussed in the initial treatment session with the laryngeal cancer patient.

Although radiation is an option to treat neck metastasis, surgical therapy is also a viable option. In cases of supraglottic and subglottic (as well as advanced T3–T4 glottic cancer), the N0 neck could be treated with a selective neck dissection. In N1 disease a selective neck or modified radical neck dissection could be used. In advanced neck disease (N2–N3), a modified radical neck or radical neck dissection would be the treatment of choice. The decision to use adjuvant radiation or chemoradiation is based on the extent of neck disease and the presence of extracapsular spread, perineural invasion, soft tissue invasion, or angioinvasion.

This patient was treated with open supraglottic laryngectomy, left modified radical neck dissection, and right selective neck dissection. Extracapsular spread was found on the neck specimen, and the patient was treated with postoperative radiation therapy.

◆ Rehabilitation and Follow-up

As with any oncologic condition, the intensity of the follow-up is proportional to the patient's risk of recurrence. Most of the recurrences occur during the first 2 years following completion of treatment; therefore, patients are seen every 1 to 2 months for the first 2 years and every 3 to 6 months for the next 3 years. At the 5-year mark, annual visits are encouraged.

The appearance or worsening in symptoms like otalgia, dysphagia, odynophagia, loss of weight, and increasing fatigue should alert the clinician as symptoms of recurrence. Further evaluation of these complaints should be pursued.

PET-CT plays a central role in follow-up care of the laryngeal cancer patients, especially after radiation-based treatments. Some residual activity in the tumor site may be seen at the first posttreatment scan at 10 to 12 weeks. If present, it should decrease with subsequent studies. Failure to do so should raise the concern of incomplete response that may require an operation for salvage. Most recurrences occur in the neck. Patients with advance neck disease (N2–N3) frequently recur and should be treated aggressively for persistent or recurrent disease.

Rehabilitation of head and neck cancer patients in general and laryngeal cancer patients in particular requires a team approach with input from speech and swallowing therapists, radiation and medical oncologists, radiologists, and social workers.

◆ Questions

1. A 57-year-old woman is being evaluated with a PET/CT at 2 months postoperatively for supraglottic laryngeal carcinoma for which she underwent an endoscopic laser procedure and bilateral neck dissections. There is mild positive uptake in the region of the larynx. What is the next best step in management?
- **A.** Operative biopsy of the area
- **B.** Magnetic resonance imaging
- **C.** Evaluate clinically followed by repeat imaging in 1 month
- **D.** Radiation therapy
- **E.** Open total or partial laryngectomy

2. A 63-year-old man with a longstanding history of tobacco and heavy alcohol use is diagnosed with supraglottic squamous cell carcinoma staged as a T2N0M0. Of the following, which is not an appropriate method of management?
- **A.** Endoscopic CO_2 laser excision of mass with bilateral selective neck dissection
- **B.** External beam radiation to supraglottis and neck
- **C.** Endoscopic CO_2 laser excision of tumor with radiation of bilateral neck
- **D.** Open partial laryngectomy with bilateral selective neck dissection
- **E.** Total laryngectomy

3. In which situation below does the patient not require treatment (either radiation or surgery) of the neck?
- **A.** T1 glottic squamous cell carcinoma
- **B.** T1 squamous cell carcinoma of the left aryepiglottic fold
- **C.** T2 squamous cell carcinoma of the subglottis
- **D.** Glottic squamous cell carcinoma involving the thyroid cartilage
- **E.** T1 base of tongue squamous cell carcinoma

Suggested Readings

Myers EN, Suen JY, Myers JN, Hanna EYN. Cancer of the Head and Neck, 4th ed. Philadelphia: WB Saunders; 2003

Silver CE, Ferlito A. Surgery for Cancer of the Larynx and Related Structures, 2nd ed. Philadelphia, PA: WB Saunders; 1996

Weinstein GS, Laccourreye O, Brasnu D, Laccourreye H. Organ Preservation Surgery for Laryngeal Cancer. San Diego, CA: Singular Publishing Group; 2000

28

Postradiation Laryngeal Framework Chondritis

Yash J. Patil

History

A 53-year-old white man is seen 2 years after receiving 7000 cGY of external-beam radiotherapy for a T4N0M0 squamous cell carcinoma of the glottis. On initial presentation, his glottic cancer extended to the anterior commissure with cartilage invasion visible on computed tomography (CT) scan. During therapy he had significant edema and pain. After radiotherapy, pain persisted but he did not require a feeding tube or tracheostomy. He failed to return after 1 year of follow-up and now notices increased hoarseness and difficulty breathing. He has continued to smoke but no longer drinks alcohol. His speech has been muffled for 1 week. He has decreased his oral intake because of pain on swallowing and has lost 5 pounds.

On physical examination the vital signs are normal. On deep inspiration, he is stridulous. There is woody induration of the entire neck with no palpable adenopathy. The larynx is tender to palpation. There is a fetid oral odor noted during the examination. Flexible laryngoscopy reveals poor true vocal cord motion with glottic edema and a 3- to 4-mm airway. The epiglottis is edematous. The airway is judged to be significantly compromised.

Differential Diagnosis—Key Points

1. The differential diagnosis should include recurrent cancer and chondronecrosis resulting from radiotherapy. Both diseases manifest with pain, edema, dysphagia, and airway compromise. Recurrence or persistence of disease after treatment is usually noted within the first year after treatment. Patients who develop postradiation chondronecrosis typically have pain and dysphagia throughout treatment, with symptoms worsening after the first year of treatment.
2. Almost 40% of patients who receive 7000 cGy of radiation therapy experience persistent laryngeal edema. Chandler proposed a grading system for laryngeal radionecrosis. Grades I and II are common and respond to humidification, voice restraint, discontinuation of smoking, and antibiotics. Grade III and IV reactions are more severe and do not respond well to conservative treatment (**Table 28.1**).
3. CT or CT with positron emission tomography (PET) scan may help to guide or avoid biopsy when there is suspicion of recurrent cancer. Biopsies may worsen mild chondronecrosis. On the other hand, tumor

Table 28.1 Chandler Laryngeal Radionecrosis Staging System

Grade	Symptoms	Signs	Treatment
1	Slight hoarseness and mucosal dryness	Slight edema, mobile vocal cords	Symptomatic care: control pain, control diabetes, humidity, antireflux therapy, smoking and alcohol cessation
2	Moderate hoarseness and mucosal dryness	Slight impairment of vocal cord mobility, moderate cord edema and erythema	Symptomatic care: control pain, control diabetes, humidity, antireflux therapy, smoking and alcohol cessation
3	Severe hoarseness with dyspnea, moderate odynophagia and dysphagia	Severe impairment of vocal cord mobility or fixation of 1 cord, marked edema, skin changes	Symptomatic care: control pain, control diabetes, humidity, antireflux therapy, smoking and alcohol cessation Consider steroids and antibiotics Consider imaging
4	Airway obstruction, severe pain, severe odynophagia, weight loss, dehydration	Fistula, foul odor, fixation of skin to larynx, immobile vocal cords, severe edema causing airway obstruction, fever	Control airway. If steroids and antibiotics do not alleviate obstruction, consider tracheostomy Symptomatic care: control pain, control diabetes, humidity, antireflux therapy, smoking and alcohol cessation Consider imaging

recurrence after radiotherapy can be largely submucosal and difficult to identify with endoscopy. In these situations, a CT/PET that is negative, or a CT scan that identifies suspicious areas, can be helpful.

◆ Test Interpretation

In general, head and neck cancer patients are followed up with routine clinical examination as well as chest radiography and liver function tests. Patients who have received radiation to the lower neck should also have thyroid function tests, especially when wound healing is problematic.

The chest radiograph and laboratory values were obtained and were within normal limits. CT scan of the neck revealed loss of laryngeal cartilage and fat stranding and fluid accumulation in the tissues around the larynx (**Fig. 28.1**). These findings are typically seen in chondronecrosis. The CT scan of the chest did not reveal any evidence of metastatic disease.

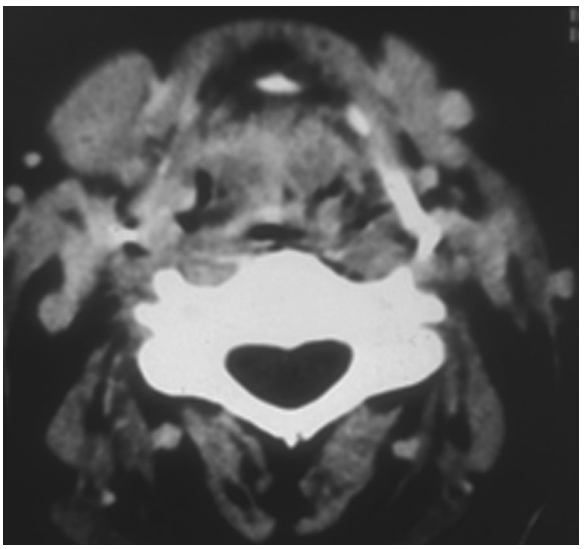


Fig. 28.1 Computed tomography scan of neck.

Direct laryngoscopy with biopsy is performed, and no residual malignancy is found.

Diagnosis

Postradiation laryngeal chondronecrosis, stage III

Medical Management

After organ-preservation therapy, patients with worsening symptoms such as loss of vocal cord function, pain, hemoptysis, or ulceration noted on examination must be evaluated for recurrence or persistence of cancer. Although a biopsy may worsen chondronecrosis, and interpretation of biopsies may be difficult in the radiated larynx, it is crucial to diagnose recurrent malignancy. If there is recurrent cancer within a radiated larynx, salvage laryngectomy is generally indicated.

Medical management of chondronecrosis of the larynx includes smoking and alcohol cessation, control of gastroesophageal reflux, and tight control of blood glucose in diabetic patients. Inhaled steam, corticosteroids, and antibiotics are all used in the management of early, Chandler stage I or II, chondronecrosis. Hyperbaric oxygen therapy may also provide some improvement in early stage chondronecrosis. Narcotics may be needed to alleviate pain.

Any fistula tract or tissue biopsy during the workup of chondronecrosis should be sent for culture and sensitivity. Other than culture-directed antibiotics, ciprofloxacin 750 mg, administered orally twice daily, is a good choice because it effectively treats *Staphylococcus aureus, Pseudomonas* spp., and enterococcal species. Surgical exploration with debridement of the necrotic tissue is recommended if no improvement is noted after at least 3 to 4 weeks of treatment.

Surgical Management

Surgical treatment of chondronecrosis is generally best accomplished with laryngectomy. Local debridement may be indicated when a specific site, such as the epiglottis or a portion of the hyoid bone or thyroid cartilage, is involved. **Figure 28.2** shows the greater cornu of the hyoid bone and the superior cornu of the thyroid cartilage that were removed endoscopically. In this instance, they had extruded into the pharynx of a radiated patient. Local debridement resulted in cessation of symptoms.

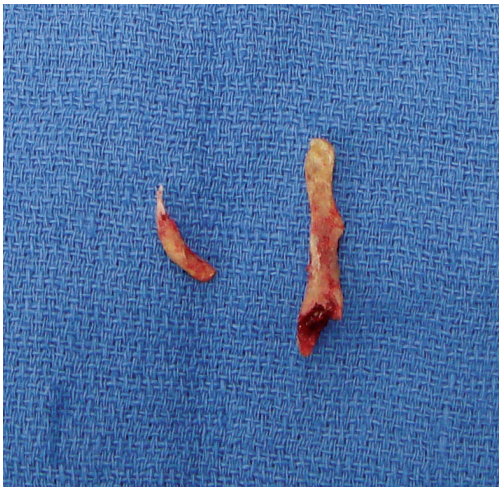


Fig. 28.2 Endoscopic debridement of exposed cartilage.

Advanced chondronecrosis, especially when there is dysfunction of the larynx with aspiration, is best treated with laryngectomy after demonstrated lack of improvement with maximal medical therapy. Laryngectomy will remove the infectious focus and prevent aspiration pneumonia. The patient's voice may be rehabilitated with tracheoesophageal puncture.

When recurrent or persistent cancer is found, only patients with small superficial recurrences in an otherwise functional larynx should be considered for treatment with open or endoscopic partial laryngectomy. Wound healing after radiation therapy is significantly impaired, and complications after laryngectomy, such as fistula, should be expected.

In this case, the patient had no response to humidification, antibiotics, and hyperbaric oxygen therapy. Increasing pain despite narcotic pain medication and worsening airway made it clear that surgical therapy would be required. The patient's airway remained patent, and tracheotomy was not required before definitive surgical treatment.

When total laryngectomy is planned for unresponsive advanced chondronecrosis, it is

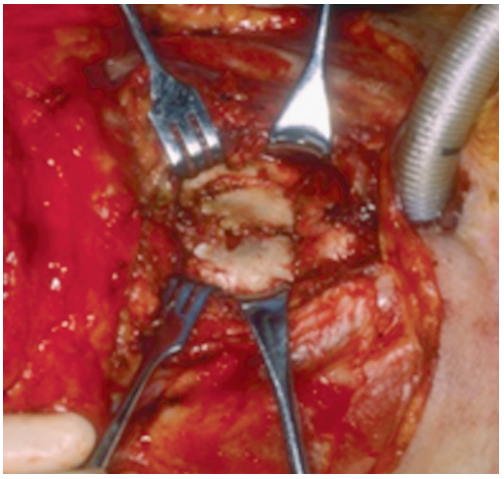


Fig. 28.3 Thyroid cartilage.

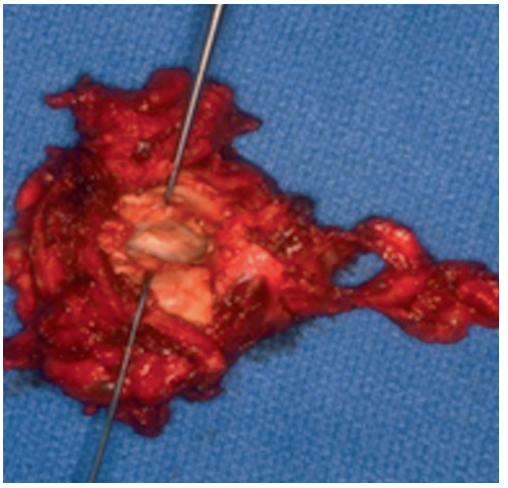


Fig. 28.4 Laryngectomy specimen.

prudent to biopsy the larynx after entry into the pharynx. If suspicion of malignancy is low and biopsies are negative, a narrow-field laryngectomy that preserves the strap muscles and piriform mucosa can be performed. This allows a tension-free closure with a layer of strap muscles to reinforce further the pharyngeal closure. In this case, note the mottled fractured appearance of the thyroid cartilage after separation of the strap muscles (**Fig. 28.3**). Examination of the specimen revealed white fibrotic mucosa (**Fig. 28.4**) with no areas suspicious for malignancy. The final pathology in this case revealed chondronecrosis and no residual tumor (**Figs. 28.5 and 28.6**). Note that the epithelium in **Fig. 28.5** shows no evidence of carcinoma. There is an inflammatory infiltrate in the deeper tissues and the cartilage appears acellular.

◆ Rehabilitation and Follow-up

Because postradiation surgery carries the potential for increased local complications, the wound should be observed for fistula or wound infection before oral feeding. Oral alimentation can usually be resumed after

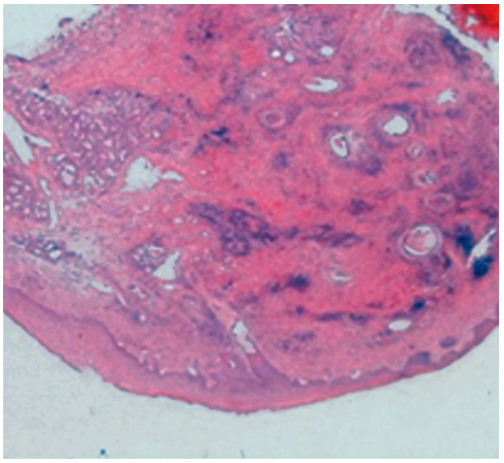


Fig. 28.5 Low-power hematoxylin-eosin stain.

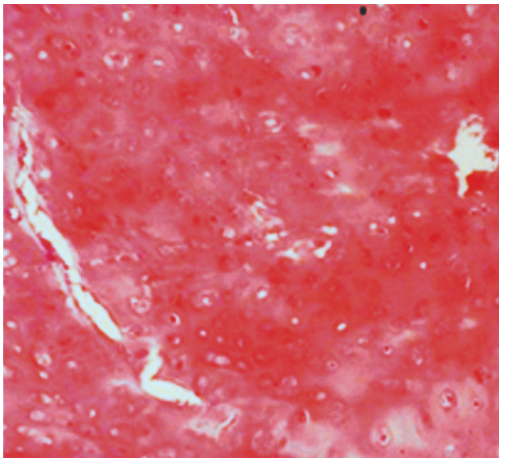


Fig. 28.6 High-power hematoxylin-eosin stain.

postoperative day 10. Speech can be initially rehabilitated with an electrolarynx and later with tracheoesophageal speech. Patients should be monitored for disease recurrence. Although one might argue that early detection of a recurrence or second primary cancer might not correlate with improved survival, surveillance should continue for no less than 2 years. Risk of second primary tumor (2–4% per year) continues to be a significant source of morbidity for these patients. In general, in the first year after treatment, patients should be seen every 1 to 3 months. In the second year, patients should be seen every 2 to 4 months. In the third year, follow-up should be scheduled at 3- to 6-month intervals. In the fourth and fifth years after treatment, appointments should be made every 4 to 6 months. After 5 years, yearly appointments are acceptable. Also, at minimum, yearly chest radiographs and liver function tests should be obtained. Thyroid function tests should be done yearly in patients who received radiation to the lower neck. Lastly, worsening symptoms should warrant further workup during this surveillance period.

◆ Questions

1. Typical symptoms seen in chondronecrosis of the larynx include all of the following except which one?
   A. Hoarseness
   B. Dysphagia
   C. Hemoptysis
   D. Neck-skin edema
   E. Dyspnea

2. Which of the following are useful treatments in the management of chondronecrosis?
   A. Antibiotics
   B. Humidification
   C. Control of gastroesophageal reflux
   D. Smoking cessation
   E. All of the above

3. Endoscopic features of laryngeal chondronecrosis include:
   A. Laryngeal edema
   B. Vocal cord fixation
   C. Cartilage exposure
   D. Mucosal dryness
   E. All of the above

Suggested Readings

Delanian S, Lefaix JL. Current management for late normal tissue injury: radiation-induced fibrosis and necrosis. Semin Radiat Oncol 2007;17(2):99–107

Fitzgerald PJ, Koch RJ. Delayed radionecrosis of the larynx. Am J Otolaryngol 1999;20(4):245–249

Hunter SE, Scher RL. Clinical implications of radionecrosis to the head and neck surgeon. *Curr* Opin Otolaryngol Head Neck Surg 2003;11(2):103–106

VII

The Thyroid

29

Papillary Carcinoma

David L. Steward

History

A 57-year-old man is referred for an incidental finding of a 2-cm thyroid mass seen on cervical spine magnetic resonance imaging (MRI). He denies any history of radiation exposure or family history of thyroid cancer. He denies hoarseness, weight change, fatigue, or palpitations.

Physical Examination

He has a palpable thyroid gland on the right larger than on the left with no discrete palpable nodule. He has no palpable lymphadenopathy. Indirect laryngoscopy reveals normal vocal function.

Differential Diagnosis—Key Points

This patient has an asymptomatic nonpalpable thyroid nodule found incidentally during cervical spine imaging. The risk of malignancy in thyroid nodules is about 5 to 10% in patients without significant risk factors. The risk is similar in incidental and palpable nodules, as well as apparently solitary and multinodular disease. Childhood radiation exposure or a strong family history of thyroid cancer would increase this risk. Ultrasound is the most sensitive and cost-effective imaging modality for thyroid nodules contained within the neck, and sensitivity for malignancy based on sonographic features exceeds 80%. Coupled with fine-needle biopsy and surgery if indicated, the sensitivity improves to 98 to 99%.

The patient has no symptoms to suggest hyperthyroidism or hypothyroidism but should undergo thyroid-stimulating hormone (TSH) measurement to assess thyroid function because the results may affect the medical management of the patient. For example, a solitary toxic adenoma has an extremely low risk (0.1%) of malignancy and likely will not require biopsy. In contrast, a nonfunctional nodule within a background of Graves disease requires biopsy to exclude malignancy before consideration for radioiodine therapy.

Test Interpretation

Screening TSH is indicated for all patients with nodular thyroid disease. If the screening is normal, no further thyroid function tests are necessary and the workup proceeds to exclude malignancy. If the TSH is high, suggestive of hypothyroidism, a free thyroxine (T_4) level should be obtained to confirm the diagnosis, and treatment with levothyroxine should be initiated and the workup proceeded to exclude

malignancy as in euthyroid patients. If the TSH is low, suggestive of hyperthyroidism, free thyronine (T_3) and free T_4 levels should both be obtained. In hyperthyroid patients with thyroid nodules, a radioiodine uptake and scan are helpful to determine the cause of the hyperthyroidism, which impacts management. The radioiodine uptake and scan are not indicated and are not helpful in the routine evaluation of euthyroid and hypothyroid patients.

Thyroglobulin, a precursor to thyroid hormone, is produced by benign thyroid follicular cells as well as by well-differentiated thyroid carcinomas (papillary, follicular, and Hürthle). It is a very sensitive postoperative and postablative marker for persistent or recurrent malignancy but is not helpful preoperatively to screen for malignancy and is not indicated.

Neck ultrasound is the best imaging modality for thyroid nodules contained within the neck or thoracic inlet. Sonographic features suggestive of malignancy include microcalcifications, irregular borders, extrathyroidal extension, and partially cystic or hypoechoic nodules. A combination of these features is more worrisome than any single feature. In the absence of suspicious sonographic features, the size of the nodule is used to determine the need for biopsy. Subcentimeter nodules do not require biopsy unless suspicious features are noted. Benign appearing nodules measuring 1 to 1.5 cm may be observed or biopsied at the preference of the patient and physician. Nodules greater than 1.5 cm should undergo biopsy to exclude malignancy in most cases. In the presence of suspicious sonographic features of the nodule or prior biopsy suspicious for malignancy, ultrasound should be performed to look for associated central and lateral nodal metastasis preoperatively so that these may be addressed surgically at the time of thyroidectomy.

Neck CT or MRI is rarely necessary unless there is clinical concern for invasive malignancy or in centers without experience in neck ultrasound for nodal disease in biopsy proven malignancy. Chest CT is indicated in cases of thyroid extension below the thoracic inlet for evaluation of inferior extent of the goiter and degree of tracheal deviation or compression. Chest CT is also indicated preoperatively or postoperatively to assess for pulmonary or mediastinal metastatic disease in biopsy proven malignancy.

Fine-needle biopsy is the best test to exclude malignancy in thyroid nodules with false-negative rates of 1 to 2% in experienced centers. Fine-needle biopsy sensitivity and specificity are significantly improved with ultrasound guidance to avoid sample error from inadvertent sampling of surrounding normal thyroid tissue with 10 to 15% false-negative rates. Palpation-guided fine-needle biopsy has insufficient negative predictive value and should not be performed unless ultrasound guidance is unavailable and the nodule is anteriorly located and discretely palpable. Ultrasound-guided fine-needle biopsy requires a minimum of two separate needle passes to sample the nodule. The nondiagnostic rate can be reduced by using three to four separate needles for each nodule.

Interpretation of the results of fine-needle biopsy requires an understanding of the limitations of cytologic evaluation. Nondiagnostic specimens contain fewer than six to eight follicular cell clusters and must be repeated to exclude malignancy. Repeatedly nondiagnostic biopsy may require surgery to exclude malignancy. Benign follicular and or Hürthle cells are the most common finding and do not require further biopsy or surgery but should be followed up with ultrasound to assess for significant interval growth, which, if present, may warrant repeat biopsy or surgery. Atypical, suspicious, or indeterminate follicular or Hürthle cells may be seen in follicular or Hürthle cell neoplasms (adenoma or carcinoma) or in follicular variant of papillary thyroid carcinoma and require surgery to exclude malignancy. Malignant cytology is most commonly seen in classic papillary thyroid carcinoma and requires surgery for definitive therapy rather than diagnostic purposes.

For this patient, screening TSH, diagnostic ultrasound, and ultrasound-guided fine-needle biopsy are necessary.

The TSH is normal and the patient biochemically euthyroid. Ultrasound demonstrated a $1.8 \times 2.1 \times 2.5$ cm hypoechoic, partially cystic right nodule with microcalcifications. Ultrasound-guided fine-needle biopsy was highly suspicious for papillary thyroid carcinoma. Repeat neck ultrasound revealed right level IV suspicious lymph node 1.2 cm, partially

cystic with microcalcifications. Ultrasound-guided fine-needle biopsy confirmed metastatic papillary thyroid carcinoma.

◆ Diagnosis

Metastatic papillary thyroid carcinoma

◆ Medical Management

Postoperative thyroid hormone suppressive therapy with levothyroxine to suppress TSH levels to subnormal levels has been shown to reduce recurrence and morbidity. The degree of TSH suppression is based on risk of recurrence. For low-risk patients, TSH suppression to 0.1 to 0.4 is recommended. Higher-risk patients or those with known persistent disease should have TSH suppression to less than 0.1. TSH suppression may result in atrial arrhythmia and osteoporosis, which can be reduced with less aggressive TSH suppression and adjunctive medical therapy when indicated.

Radioiodine thyroid remnant ablation may reduce risk of recurrence and mortality, especially in more advanced disease. Remnant ablation may facilitate long-term surveillance with thyroglobulin levels. Radioiodine treatment may increase the risk of second malignancy and may cause salivary or lacrimal gland dysfunction. Radioiodine is teratogenic and contraindicated in pregnancy.

◆ Surgical Management

Total thyroidectomy is indicated based on the cytologic diagnosis. Frozen section biopsy is less sensitive than cytology and is not indicated when a diagnosis of papillary carcinoma is made on cytology alone. Compared with hemithyroidectomy alone, total thyroidectomy has been shown to reduce the recurrence of thyroid cancer for tumors larger than 1 cm or in the presence of multicentricity. The recurrent and superior laryngeal nerves should be identified and preserved along with the parathyroid glands in situ with the blood supply intact. Parathyroid autotransplantation is performed if parathyroid devascularization occurs during dissection. Confirmation of parathyroid with biopsy and frozen section is recommended to avoid transplantation of tumor.

Because of the presence of lateral nodal metastasis, central (level VI) neck dissection and lateral (levels II–IV and possibly V) neck dissection are performed, with preservation of all vital structures not involved with cancer, including the internal jugular vein, sternocleidomastoid muscle, and spinal accessory nerve, and preferably with preservation of cervical sensory nerves as well.

Postoperative complications of thyroidectomy include transient or permanent recurrent or superior laryngeal nerve injury, with resultant dysphonia for unilateral injury and rare but possible airway obstruction from bilateral recurrent nerve injury. Hypoparathyroidism, transient or permanent, with resultant hypocalcemia can be diagnosed with rapid intact parathyroid hormone measurement in the recovery room ($<$8–15 pg/mL) or by serial calcium measurement in recovery, on the evening of surgery, and on the morning of postoperative day 1 ($<$8.0 mg/dL). Treatment of hypoparathyroidism includes frequent oral calcium supplementation (three or four times daily) along with early and aggressive 1,25 dihydroxy vitamin D supplementation (calcitriol).

Postoperative hematoma can result in airway obstruction requiring urgent opening of the wound to decompress the neck and return to the operating room for hematoma evacuation and control of bleeding. Use of neck drains does not prevent postoperative bleeding but may facilitate management if this complication occurs.

Postoperative hypothyroidism is an expected consequence of total thyroidectomy requiring thyroid hormone replacement therapy lifelong. Long-term therapy with levothyroxine (LT_4) sodium is initiated at 1.5 μg/kg/day with subsequent dose titration based on TSH levels obtained 5 to 6 weeks after initiation of therapy. Short-term therapy with liothyronine (LT_3) is an alternative in the early postoperative period when thyroid hormone withdrawal is planned for physiologic TSH elevation to facilitate radioiodine 4 to 6 weeks postoperatively. Liothyronine therapy at 25 μg daily in divided doses can be started 1 week postoperatively to limit the severity and duration of symptoms

from hypothyroidism during this period. Recombinant human TSH can be given intramuscularly an as alternative to physiologic TSH elevation from thyroid hormone withdrawal to prepare for radioiodine remnant ablation, avoiding the need for postoperative hypothyroidism or short-term liothyronine therapy.

Rehabilitation and Follow-up

Radioiodine whole-body scans had traditionally been the mainstay of scanning for recurrent or persistent cancer, but they have been shown to have low sensitivity (20%) in more recent studies. Serum thyroglobulin and neck ultrasound have been shown to have combined sensitivity greater than 90% in most studies and have become the mainstay of well-differentiated thyroid cancer surveillance. The presence of anti-thyroglobulin antibodies, common in thyroiditis and present in around 25% of thyroid cancer patients, severely limit the measurement and interpretation of serum thyroglobulin levels and should always be measured along with thyroglobulin. Further, the presence of persistent benign thyroid tissue following surgery without postoperative radioiodine ablation precludes discrimination of benign from malignant disease using thyroglobulin. TSH-stimulated thyroglobulin measurement with recombinant human TSH injection or thyroid hormone withdrawl improves the sensitivity of thyroglobulin to detect persistent or recurrent disease.

Questions

1. What is the most accurate test for the diagnosis of papillary thyroid carcinoma?
   - **A.** Ultrasound-guided fine-needle biopsy
   - **B.** Thyroglobulin
   - **C.** Computed tomography scan
   - **D.** Radioiodine scan
2. What is the most appropriate surgical treatment for cytologically diagnosed papillary thyroid carcinoma?
   - **A.** Hemithyroidectomy (lobectomy)
   - **B.** Subtotal thyroidectomy
   - **C.** Bilateral subtotal thyroidectomy
   - **D.** Total or near total thyroidectomy
3. What is the most appropriate thyroid-stimulating hormone target for levothyroxine therapy for a patient with recently diagnosed papillary thyroid carcinoma?
   - **A.** Subnormal
   - **B.** Normal
   - **C.** Supranormal

Suggested Reading

Cooper DS, Doherty GM, Haugen BR, et al, for the American Thyroid Association Guidelines Taskforce. Management guidelines for patients with thyroid nodules and differentiated thyroid cancer. Thyroid 2006;16(2):109–142

30

Medullary Thyroid Carcinoma

David L. Steward

History

A 55-year-old man has a palpable right thyroid mass discovered after his sister was diagnosed with medullary thyroid carcinoma. He denied hoarseness or symptoms of hypothyroidism or hyperthyroidism. He reported a history of medically controlled hypertension with occasional flushing spells. In addition to his sister with medullary thyroid carcinoma, his paternal uncle died of thyroid cancer of unknown origin.

The physical examination revealed a 2-cm right thyroid nodule without palpable lymphadenopathy. Vocal cord function is normal.

Differential Diagnosis—Key Points

The patient has a thyroid nodule with a family history of medullary thyroid cancer and is at high risk for familial medullary thyroid carcinoma, multiple endocrine neoplasia (MEN) IIa, or MEN-IIb. Genetic testing of the patient's sister and of the patient if the sister is positive or unavailable for testing is diagnostic and required for this autosomal dominant hereditary disease. If this patient is positive, all of his siblings and offspring require testing and therapeutic or prophylactic thyroidectomy if positive. MEN-IIa is associated with hyperparathyroidism and pheochromocytoma; MEN-IIb is associated with pheochromocytoma, mucosal neuromas, and pancreatic islet cell tumors. Biochemical screening for these associated disorders is important preoperatively to identify hyperparathyroidism, which should be addressed concomitantly at the time of thyroidectomy. Screening for pheochromocytoma is necessary preoperatively so that it can be treated before thyroidectomy is performed to minimize the risk of perioperative hypertensive crisis.

Diagnostic Tests

The workup of a thyroid nodule is reviewed in the previous chapter and includes screening thyroid stimulating hormone (TSH) ultrasound, and ultrasound-guided fine-needle biopsy, all of which are indicated and should be performed. The cytology specimen should be sent for immunohistochemical staining for calcitonin in addition to traditional hematoxylin and eosin staining.

Specific to this case is the family history of medullary thyroid carcinoma, which impacts the medical management of this patient. Calcitonin is produced by parafollicular c-cells within the thyroid, as well as their malignant counterpart medullary thyroid carcinoma. Serum calcitonin is a reasonable screening test in all patients with thyroid nodules because it has greater sensitivity than fine-needle

biopsy for the diagnosis of medullary carcinoma. In patients with a family history, it is mandatory. Slight elevation of serum calcitonin may be normal or may represent c-cell hyperplasia, a precursor to medullary carcinoma in hereditary patients. Larger elevations above 40 or 100 are more specific for medullary cancer.

Genetic testing for the *Ret* proto-oncogene is commercially available and indicated in all patients diagnosed with medullary carcinoma because 25% of apparently sporadic cases (no family history) are found to have the hereditary genetic alteration. If the index patient with medullary carcinoma is negative, then no further family members require genetic testing. If the index patient is positive, then all surviving parents, siblings, and offspring require testing. The specific codon affected will diagnose MEN-IIa versus MEN-IIb with important clinical implications regarding the aggressiveness of medullary thyroid carcinoma (codon 918 MEN-IIb generally more aggressive and earlier presentation) as well as associated endocrine neoplasias. *Ret*-positive family members have an approximate 90% risk of the development of medullary thyroid carcinoma and require biochemical workup and therapeutic or prophylactic thyroidectomy. Prophylactic removal of other endocrine glands is not recommended.

Pheochromocytomas are seen in about 50% of patients with MEN-IIa, but the risk may be codon specific. These benign adrenal tumors produce epinephrine, norepinephrine, and its precursors and metabolites, causing poorly controlled hypertension. All patients diagnosed with or suspected of having hereditary medullary thyroid carcinoma should have preoperative screening. Plasma and 24-hour urinary catecholamines and metanephrines are the most sensitive and specific tests. If these are abnormally elevated, imaging is indicated and removal of the pheochromocytoma before thyroidectomy is important.

Hyperparathyroidism, usually from multiple parathyroid adenomas or hyperplasia, is seen in around 20% of patients with MEN-IIa. Biochemical screening should include serum calcium and intact parathyroid hormone levels. If elevated, then concomitant parathyroidectomy at the time of thyroidectomy is indicated.

◆ Test Interpretation

Serum TSH is normal and the patient is euthyroid. Ultrasound reveals a 2-cm right-sided hypoechoic nodule with irregular borders and left-sided 1.3-cm hypoechoic nodule without sonographically suspicious lymph nodes. Ultrasound-guided fine-needle biopsy revealed cytologically suspicious cells that stain positive for calcitonin, diagnostic of medullary thyroid carcinoma bilaterally. Serum calcitonin is elevated at 570 pg/mL, suggesting possible metastatic disease. The *Ret* proto-oncogene test is positive for mutation in codon 609, confirming MEN-IIa syndrome. Plasma and 24-hour urinary catecholamines and metanephrines are normal. Calcium and parathyroid hormone levels are also normal. Neck MRI failed to demonstrate evidence of lateral cervical nodal metastasis.

◆ Diagnosis

MEN IIa syndrome with medullary thyroid carcinoma

◆ Medical Management

Postoperative thyroid hormone replacement therapy with levothyroxine sodium should be initiated immediately postoperatively at 1.5 μg/kg/day with titration of levothyroxine to keep TSH levels within the normal range, ideally between 1 and 3. Thyroid hormone suppression has no role in medullary thyroid carcinoma, nor does radioiodine therapy.

No chemotherapeutic agents are currently approved by the Food and Drug Administration for medullary thyroid carcinoma; however, several novel vascular endothelial growth factor inhibitors are in clinical trials and have shown some promise for partial response or stabilization of disease.

◆ Surgical Management

Total thyroidectomy with prophylactic comprehensive central compartment (level VI) dissection to include prelaryngeal (Delphian), pretracheal, and bilateral paratracheal lymph

nodes medial to the carotid arteries and extending inferiorly to the innominate artery in the superior mediastinum (level VII) is important to reduce recurrent or persistent disease. All efforts should be made to preserve grossly normal parathyroid glands in situ with blood supply intact in the absence of biochemical evidence of hyperparathyroidism. Prophylactic lateral neck dissection (levels II–V) might not be necessary in the absence of radiographically suspicious lymph nodes.

Rehabilitation and Follow-up

Serum calcitonin and carcinoembryonic antigen (CEA) levels are the mainstay of long-term medullary thyroid carcinoma surveillance. Undetectable calcitonin and CEA levels are suggestive of no persistent or recurrent disease. Semiannual to annual calcitonin and CEA levels and neck ultrasound are the mainstays of follow-up of these low-risk patients. Annual biochemical screening for associated endocrine neoplasias (hyperparathyroidism and pheochromocytoma) is important.

Persistently elevated calcitonin and CEA levels for more than 1 month postoperatively are consistent with persistent metastatic disease. Structural imaging of the neck, chest, liver, and brain using ultrasonography, computed tomography, and MRI to localize disease is performed. Bone metastasis is best localized with bone scintigraphy. Lower calcitonin levels are associated with regional disease, whereas higher levels are associated with distant metastatic disease. Radionuclide imaging using octreotide or fluorodeoxyglucose positron emission tomography may occasionally be helpful to localize disease if structural imaging is nonlocalizing.

Rapidly doubling calcitonin levels are worrisome for progressive disease. Doubling times less than 2 years are associated with reduced 10-year survival compared with greater than 2-year doubling times. Very rapid doubling times of less than 6 months are associated with poor 5- and 10-year survival.

Questions

1. Which of the following is not associated with medullary thyroid carcinoma?
- **A.** MEN I
- **B.** MEN IIa
- **C.** MEN IIb
- **D.** Sporadic medullary thyroid carcinoma

2. What is the appropriate surgical therapy for medullary thyroid carcinoma radiographically confined to the thyroid gland?
- **A.** Hemithyroidectomy
- **B.** Total thyroidectomy
- **C.** Total thyroidectomy with central node dissection
- **D.** Total thyroidectomy with bilateral modified radical neck dissection

3. What is the most specific tumor marker used in postoperative surveillance for medullary thyroid carcinoma?
- **A.** Thyroglobulin
- **B.** Thyroglobulin antibodies
- **C.** Calcitonin
- **D.** Parathyroid hormone

Suggested Reading

Cooper DS, Doherty GM, Haugen BR, et al, for the American Thyroid Association Guidelines Taskforce. Management guidelines for patients with thyroid nodules and differentiated thyroid cancer. Thyroid 2006;16(2):109–142

31

Graves Disease

David L. Steward

History

A 33-year-old woman has thyroid enlargement, palpitations, tremor, and weight loss. She denies visual changes. She is otherwise healthy and is on no medications. She reports a family history of a maternal aunt who had radioiodine therapy and her mother who is on thyroid hormone.

Physical examination reveals diffuse and doughy thyromegaly, mild asymmetric exophthalmos with bilateral lagophthalmos, tachycardia, and mild tremor. Extraocular motion is preserved.

Differential Diagnosis—Key Points

This patient has a fairly classic presentation for Graves disease with signs and symptoms of hyperthyroidism, diffuse thyroid enlargement, and ophthalmopathy. The family history suggests both hyperthyroidism and hypothyroidism on her mother's side. Current understanding of autoimmune thyroid disease suggests that Hashimoto thyroiditis and Graves disease are genetically related. Initial workup should focus on biochemical confirmation of the hyperthyroidism and amelioration of symptoms, with further workup focusing on the cause of the hyperthyroidism, which could include Graves, thyroiditis (Hashimoto or viral), or toxic adenoma or toxic multinodular goiter. The treatment and prognosis depend on the cause of the hyperthyroidism.

Test Interpretation

Biochemical diagnosis of hyperthyroidism includes thyroid-stimulating hormone (TSH), tri-iodothyronine (T_3), and free thyroxine (T_4) levels, with low TSH and high free T_3 or free T_4 levels being diagnostic. Subclinical hyperthyroidism is associated with low TSH with normal free T_3 and free T_4 levels.

The cause of hyperthyroidism can be evaluated using serum antithyroid antibody testing or with radioiodine uptake and scan. Thyroid ultrasound, especially if coupled with antibody testing, may be an alternative to the radioiodine scan in patients with Graves disease. Elevated anti-TSH receptor antibody levels are diagnostic of Graves disease. Elevated antithyroglobulin or antithyroid peroxidase (TPO) antibodies are consistent with chronic lymphocytic thyroiditis (Hashimoto). In the absence of positive antibody testing, elevated erythrocyte sedimentation rate (>60) is consistent with viral subacute thyroiditis (DeQuervan).

Radioiodine uptake and scanning will demonstrate marked elevated 24-hour iodine uptake (>35%) and a diffusely positive scan in Graves. The presence of a cold area within a background of Graves disease suggests a nodule

that requires workup to exclude malignancy. Radioiodine uptake will be low (<5–10%) in hyperthyroid patients with thyroiditis. Radioiodine uptake will be mild to moderately elevated in toxic adenoma or toxic multinodular goiter, with scanning demonstrating a solitary hot nodule or multiple hot nodules with the background thyroid suppressed.

Thyroid ultrasound will demonstrate characteristic diffuse thyroid enlargement with hypervascularity in Graves disease and occasionally will demonstrate nodular disease requiring further evaluation. Ultrasound will show a characteristic heterogeneously hypoechoic thyroid in thyroiditis, sometimes with infrathyroidal reactive lymph nodes. Toxic adenomas will appear as a solitary solid, possibly hypervascular, nodule but may require radioiodine scanning to assess the function of the nodule to differentiate Graves disease with a cold nodule from a hot nodule within a background of suppressed thyroid. Toxic multinodular goiter on ultrasound alone will be indistinguishable from nontoxic multinodular goiter and requires radioiodine scanning to identify the hyperfunctional nodule(s) and those that are hypofunctional, possibly requiring further workup (biopsy).

The TSH is undetectable with an elevated free T_3 and free T_4 consistent with hyperthyroidism. TSH receptor antibodies are elevated with normal TPO and thyroglobulin antibodies consistent with Graves disease. Ultrasound revealed a diffusely enlarged hypervascular thyroid without nodules, and radioiodine uptake was elevated with diffuse scanning consistent with Graves.

◆ Diagnosis

Hyperthyroidism and Graves disease

◆ Medical Management

Initial medical management includes use of a β-blocker to ameliorate the tachycardia, palpitations, and tremor and to protect the heart from the cardiovascular side effects of thyrotoxicosis. Subsequent medical management with thioamides (methimazole or propylthiouracil) will inhibit the synthesis of thyroid hormone and result in euthyroidism within 4 to 6 weeks in most patients. TSH, produced in the pituitary, may remain suppressed for several months, and evolution of effectiveness of thioamides relies on measurement of free T_3 and free T_4 levels. Long-term use of thioamides is associated with a low risk of marrow suppression and is less desirable than definitive treatment with radioiodine or surgery for most patients. Graves will spontaneously resolve in a minority of patients (10–30%), however, and thioamide weaning 1 to 2 years after initiation of therapy before definitive treatment is reasonable.

Corticosteroids are effective in medically refractory patients during acute toxic crisis or in preparation for surgery. The steroids inhibit the autoimmune stimulation of the TSH receptor by the anti–TSH receptor antibody and also inhibit peripheral conversion of T_4 to T_3.

Radioiodine therapy will render most patients with Graves hypothyroid but may take months for the effects to manifest. Radioiodine may need to be repeated in patients with very large thyroid glands or for whom a single treatment fails to resolve the hyperthyroidism. Radioiodine is associated with risk for worsening Graves orbitopathy and is a relative contraindication to this therapy. If planned, concomitant corticosteroids may reduce this risk. Radioiodine is teratogenic and absolutely contraindicated in pregnancy, and pregnancy should be deferred for 6 to 12 months following radioiodine treatment. It is not recommended during breastfeeding because of significant radioiodine uptake in lactating breast tissue. Radioiodine may be associated with a very small absolute risk of subsequent development of radiation-induced malignancy, which should be discussed with the patient.

Lagophthalmos will improve with β-blockers and rendering the patient euthyroid. Referral of the patient to an ophthalmologist is recommended for evaluation of possible corneal or other complications of ophthalmopathy. Adequate moisturization of the eye is important.

◆ Surgical Management

Total thyroidectomy is curative of hyperthyroidism. Thyroid surgery for Graves is more difficult than other cases in large part because of the hypervascular nature of the

glands. Preoperatively, patients should be rendered euthyroid with thioamide therapy and corticosteroids if necessary. Treatment with supersaturated potassium iodide 1 week preoperatively may further reduce thyroid vascularity and reduce the risk of thyroid storm. Perioperative β-blocker therapy is recommended to protect the heart. Postoperatively, thyroid hormone replacement with levothyroxine should be initiated. Attempts to perform subtotal thyroidectomy to avoid postoperative hypothyroidism are misguided and associated with risk for recurrence of hyperthyroidism.

Orbital decompression of the medial and inferior walls to allow decompression of the medial and inferior rectus muscles into the ethmoid and maxillary sinus can improve worsening exophthalmos but causes diplopia and is not indicated for mild orbitopathy. The eye disease may improve slowly 1 to 2 years following total thyroidectomy, presumably because removal of the thyroid reduces the overall antigen load presented to the immune system despite the fact that the antibodies reacting to the extraocular muscles are different from the TSH receptor antibodies causing hyperthyroidism.

◆ Rehabilitation and Follow-up

Long-term thyroid hormone replacement is necessary for patients treated with thyroidectomy or radioiodine therapy. Serial thyroid function testing and possibly complete blood counts should be performed for patients receiving thioamide therapy. This patient was treated with surgery because of the presence of ophthalmopathy and has been on levothyroxine postoperatively with stabilization and slight improvement in the eye disease.

◆ Questions

1. Which of the following thyroid tests are consistent with hyperthyroidism?
   - **A.** High TSH, low free tri-iodothyronine (FT_3), low free thyroxine (FT_4)
   - **B.** Low TSH, high FT_3, high FT_4
   - **C.** High TSH, high antithyroid peroxidase (TPO) antibodies (Ab), high antithyroglobulin Ab
   - **D.** Low TSH, low FT3, low FT4
2. Which of the following is consistent with Graves disease?
   - **A.** Low radioiodine uptake
   - **B.** Low thyroglobulin
   - **C.** High anti-TSH receptor Ab
   - **D.** High anti-TPO Ab
3. Which of the following is not an appropriate medical therapy for hyperthyroidism associated with Graves disease?
   - **A.** Methimazole
   - **B.** Levothyroxine
   - **C.** Propranolol
   - **D.** Radioiodine
   - **E.** Prednisone

Suggested Reading

Singer PA, Cooper DS, Levy EG, et al. Treatment guidelines for patients with hyperthyroidism and hypothyroidism. Standards of Care Committee, American Thyroid Association. JAMA 1995;273(10):808–812

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Hashimoto Thyroiditis

David L. Steward

History

A 42-year-old woman describes progressive fatigue, lethargy, and weight gain. She has no known medical problems. She takes no prescription medications but does take a multivitamin daily. She reports a family history of hypothyroidism but denies a family history of thyroid cancer. She denies a history of radiation exposure.

Physical examination reveals a firm thyroid gland bilaterally but is otherwise normal.

Differential Diagnosis—Key Points

This patient has symptoms of hypothyroidism, although these symptoms are not specific, and biochemical thyroid function testing is required to confirm the diagnosis. The most common cause of hypothyroidism in the United States is chronic lymphocytic thyroiditis, or Hashimoto thyroiditis, although antithyroid antibodies are rarely necessary for patient management. In older patients, hypothyroidism might not be the result of thyroiditis but rather a consequence of aging. Outside the United States, in iodine-deficient areas, hypothyroidism may be due to endemic iodine deficiency.

Test Interpretation

Thyroid-stimulating hormone (TSH) and free thyroxine (T_4) are indicated to confirm the suspected diagnosis of hypothyroidism, which is associated with a high TSH and low or normal free T_4. Free tri-iodothyronine (T_3) testing is generally not needed to confirm hypothyroidism, in contrast to hyperthyroidism. Antithyroid peroxidase (TPO) and thyroglobulin antibodies are rarely necessary but can confirm the cause as Hashimoto thyroiditis.

Because the thyroid is firm and palpable, ultrasound to rule out discrete nodule and possible cancer is indicated. Thyroiditis and hypothyroidism may increase the risk of malignancy. Ultrasound commonly reveals a hypoechoic heterogeneous thyroid gland in thyroiditis and occasionally benign-appearing infrathyroidal lymph nodes, especially in younger patients. Discrete nodules, if present within the thyroid, require fine-needle biopsy to exclude malignancy if sonographically suspicious or larger than 1.5 cm in diameter. Rapidly enlarging, diffuse enlargement should raise suspicion for thyroid lymphoma, which is rare but has a relatively increased risk in patients with thyroiditis.

This patient's TSH level is high at 33 (normal, 0.4–4.5) with a low free T_4 at 0.7 (normal, 0.9–1.8) confirming hypothyroidism. No further blood tests were performed. Neck ultrasound was consistent with thyroiditis without discrete nodule.

Diagnosis

Hypothyroidism with Hashimoto thyroiditis

Medical Management

Thyroid hormone replacement therapy with levothyroxine is instituted. Because of this patient's profound symptoms, high TSH level, and free T_4, this patient is started on full replacement dosing of 1.5 μg/kg/day. A TSH level is obtained 4 to 6 weeks later with titration of levothyroxine therapy to a target TSH of between 1 and 3. In older patients or in those with known coronary artery disease, initiation of therapy should be at lower dosing with slow incremental increases ultimately to the same target TSH level (i.e., 25 μg daily for 1 week, then 50 μg × 1 week, and then 75 μg, and so on).

Some studies have suggested improved cognitive function with combined levothyroxine (LT_4) and liothyronine (LT_3) therapy, but most studies have failed to demonstrate a benefit. Furthermore, LT_3 has a short half-life requiring twice or three times daily dosing and results in supraphysiologic T_3 levels transiently, potentially increasing the risk for cardiac and skeletal effects of hyperthyroidism. T_4 is peripherally converted to T_3, so LT_3 is not necessary for the vast majority of patients and is discouraged for routine use. For patients who continue to feel fatigued with target TSH between 1 and 2 and normal free T_4 levels, obtaining a free T_3 to ensure adequate peripheral conversion will usually prove normal, suggesting no need for LT_3 therapy.

Surgical Management

Surgical management is not indicated for hypothyroidism or thyroiditis unless there is suspicion for malignancy based on fine-needle biopsy of discrete nodules or because of rapid, progressive, diffuse enlargement refractory to thyroid hormone replacement therapy. Surgery is often more difficult in patients with thyroiditis as a result of fibrosis of the gland to surrounding structures. Further, Hashimoto thyroiditis is usually self-limited, with progressive gland atrophy precluding need for surgical intervention.

Rehabilitation and Follow-up

Semiannual to annual TSH testing to confirm adequacy of thyroid hormone replacement therapy is important. This patient was rendered euthyroid with levothyroxine with improvement in symptoms.

Questions

1. Which of the following tests are consistent with hypothyroidism?
   - **A.** Low thyroid stimulating hormone (TSH), low free thyroxine (FT_4)
   - **B.** High TSH, low FT_4
   - **C.** Low TSH, high FT_4
   - **D.** High TSH, high FT_4
2. Which of the following tests is most consistent with Hashimoto thyroiditis–associated hypothyroidism?
   - **A.** High antithyroid peroxidase (TPO) antibodies
   - **B.** High anti-TSH receptor antibodies (TRAb)
   - **C.** Low antithyroglobulin antibodies
   - **D.** Low antiphospholipid antibodies
3. Which of the following is the most appropriate medical management for hypothyroidism associated with Hashimoto thyroiditis?
   - **A.** Liothyronine
   - **B.** Levothyroxine
   - **C.** Methimazole
   - **D.** Propylthiourocil
   - **E.** Prednisone

Suggested Readings

Singer PA, Cooper DS, Levy EG, et al. Treatment guidelines for patients with hyperthyroidism and hypothyroidism. Standards of Care Committee, American Thyroid Association. JAMA 1995;273(10):808–812

33

Multinodular Goiter

David L. Steward

History

A 56-year-old woman with a longstanding palpable multinodular goiter is referred by her primary care physician for evaluation. The patient reports a family history of goiter and thyroid surgery but no family history of thyroid cancer. She denies history of radiation exposure. She denies dysphagia, dyspnea, or hoarseness. She denies symptoms of hypothyroidism or hyperthyroidism.

Physical examination reveals multinodular goiter with around 3-cm right-sided and 2-cm left-sided dominant nodules. Vocal cord function is normal. No other masses or evidence of adenopathy are noted.

Differential Diagnosis—Key Points

Multinodular goiter is common and often hereditary. Three main points come up when evaluating a patient with multinodular goiter: (1) Does this represent malignancy? (2) Is there endocrine dysfunction? (3) Is there evidence of compression of surrounding structures as a result of its size?

Test Interpretation

Ultrasound is the best imaging modality for thyroid nodules to assess the presence of nonpalpable nodules, for accurate sizing, to identify features suspicious for malignancy, and to guide fine-needle biopsy. Suspicious sonographic features of thyroid nodules have an 80% sensitivity for malignancy. Size alone is nondiscriminatory, but larger nodules (>1.5–2 cm) are often biopsied to exclude malignancy even in the absence of suspicious features. In the presence of multiple nodules, biopsy of suspicious nodules should be preferentially performed over benign-appearing larger ones. Often the largest on each side is biopsied along with any sonographically suspicious nodules, requiring biopsy of as many as four nodules to exclude malignancy within a large multinodular gland (see papillary thyroid carcinoma case for further description of ultrasound-guided fine-needle biopsy and cytologic findings).

Screening thyroid-stimulating hormone (TSH) should be done to exclude subclinical hyperthyroidism, which is common in multinodular goiter. If the TSH is low, then follow-up testing of free tri-iodothyronine (T_3) and free thyroxine (T_4) is necessary to confirm hyperthyroidism from toxic multinodular goiter.

Slowly growing multinodular goiters confined to the neck are often asymptomatic to the patient as the neck musculature and skin expand with the growth. Goiters that extend into the thoracic inlet and mediastinum more commonly cause tracheal deviation and compression requiring chest computed tomography for further evaluation. Pulmonary function tests in the sitting and supine position or with flexion and extension may be helpful to assess dyspnea. Barium swallow to evaluate dysphagia rarely demonstrates extrinsic compression but may be helpful to exclude other causes of dysphagia, such as esophageal dysmotility or strictures, especially in older individuals.

The TSH level is normal, suggesting a euthyroid state. Ultrasound confirms a multinodular goiter with solid dominant hypoechoic nodules bilaterally, 2.1 × 2.4 × 3.2 cm on the right and 1.8 × 1.5 × 2.5 cm on the left side. Numerous nonpalpable subcentimeter nodules are noted, along with a left superior pole hypoechoic nodule measuring 1.4 × 1.5 × 2.1 cm and partially cystic right inferior hypoechoic nodule measuring 1.3 × 1.4 × 2.1 cm. No evidence of nodal disease or extrathyroidal extension is seen. None of the nodules contains multiple suspicious features for malignancy.

Diagnosis

Multinodular goiter

Medical Management

Medical management generally involves fine-needle biopsy to exclude malignancy and observation for significant interval growth. This patient underwent fine-needle biopsy of all four nodules without evidence of malignancy and elected for observation.

Thyroid hormone replacement therapy to keep TSH levels in the low normal range between 1 and 3 is indicated for patients with hypothyroidism. Thyroid hormone suppression therapy with levothyroxine to suppress TSH to subnormal values may reduce goiter size for some patients but is associated with increased risk of cardiac arrhythmia and osteoporosis, and goiter recurs following discontinuation of suppressive therapy. Routine suppression is not recommended.

Radioiodine may be more effective than TSH suppression, but it requires higher doses of radioiodine for euthyroid multinodular goiter compared with patients with hyperthyroidism. Radioiodine may reduce volume size by 20 to 40% over a 1- to 2-year period but is associated with a small risk of malignancy and is not recommended except possibly for older or medically infirm patients with compressive symptoms who do not want to undergo surgery. TSH-stimulated radioiodine therapy may improve radioiodine uptake and subsequent volume reduction with lower radioiodine doses and should be considered if this mode of therapy is undertaken.

Surgical Management

Total thyroidectomy is curative for multinodular goiter that is bilateral. In unilateral nodular goiter, fine-needle biopsy to exclude malignancy followed by hemithyroidectomy will be sufficient for most patients but may require completion thyroidectomy if occult malignancy is discovered or subsequent enlarging nodular disease develops in the remaining lobe. Partial lobectomy, subtotal thyroidectomy, and bilateral subtotal thyroidectomy should be avoided because of the likelihood of recurrent nodular disease requiring reoperative surgery in a prior operative field with increased risk. Surgery should be reserved for cases suspicious for malignancy or for large or enlarging goiters, especially in younger patients. Involvement of the thoracic inlet or superior mediastinum or evidence of tracheal compression or significant deviation is indication for surgery.

Rehabilitation and Follow-up

Annual TSH testing and annual to biannual ultrasound are reasonable to evaluate for progression of nodular disease, development of new nodules, or development of thyroid dysfunction. This patient has demonstrated no significant disease progression or symptoms

in long-term follow-up. Repeat fine-needle biopsy to exclude malignancy in previously cytologically benign nodules without significant change on ultrasound is not necessary but should be considered if suspicious sonographic features appear or significant growth occurs.

Questions

1. What is the best imaging modality for a euthyroid patient suspected of having a thyroid nodule?
   - **A.** Ultrasound
   - **B.** CT scan
   - **C.** MRI scan
   - **D.** Radioiodine uptake and scan
   - **E.** PET scan
2. What biochemical screening test is indicated for a patient suspected of having a multinodular goiter?
   - **A.** Thyroglobulin
   - **B.** Calcitonin
   - **C.** Thyroid-stimulating hormone
   - **D.** Free thyroxine
   - **E.** Free thyronine
3. What is the best surgical procedure for management of an enlarging bilateral multinodular goiter with compressive symptoms?
   - **A.** Bilateral subtotal thyroidectomy
   - **B.** Unilateral subtotal thyroidectomy
   - **C.** Total thyroidectomy
   - **D.** Hemithyroidectomy (lobectomy)
   - **E.** Isthmusectomy

Suggested Reading

Cooper DS, Doherty GM, Haugen BR, et al, for the American Thyroid Association Guidelines Taskforce. Management guidelines for patients with thyroid nodules and differentiated thyroid cancer. Thyroid 2006;16(2):1–33

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Primary Hyperparathyroidism

David L. Steward

History

A 64-year-old woman has a history of osteoporosis and elevated serum calcium found on a routine renal panel. She denies a history of kidney stones or fragility fracture or a family history of hypercalcemia. She does report fatigue, which she had attributed to aging. She has no significant psychiatric history and denies gastrointestinal distress or bone pain. She is on a bisphosphonate for osteoporosis and a multivitamin. Her physical examination is unremarkable.

Differential Diagnosis—Key Points

Asymptomatic hypercalcemia associated with osteoporosis is most likely primary hyperparathyroidism. Confirmation with a repeat renal panel and simultaneous parathyroid hormone (PTH) level are indicated. More severe hypercalcemia can be seen with malignancy, either through tumor production of PTH-related peptide (PTHrP), which can be measured, and is seen in small cell lung cancer or through extensive bone metastases, such as with multiple myeloma. Hypercalcemia may also be seen with excess calcium or vitamin D ingestion and is termed milk–alkali syndrome. It may also be seen in granulomatous disease such as tuberculosis or sarcoidosis resulting from extrarenal vitamin D production in macrophages. Intact PTH levels will be low in these cases, essentially ruling out primary hyperparathyroidism. Familial hypocalciuric hypercalcemia can mimic primary hyperparathyroidism and is excluded with a 24-hour urinary calcium level below the reference range or a fractional excretion of calcium below 1%. Lithium may cause or mimic primary hyperparathyroidism via alteration in the calcium-sensing receptor. Thiazide diuretics may exacerbate or unmask primary hyperthyroidism and occasionally cause hypercalcemia.

Primary hyperparathyroidism is most often sporadic, with about 90% of patients having a solitary parathyroid adenoma and 10% having multigland disease, primarily four-gland hyperplasia or sometimes multiple adenomas. Familial primary hyperparathyroidism is seen with multiple endocrine neoplasia (MEN) type I and MEN-IIa. MEN-I is associated with a menin gene mutation resulting in parathyroid hyperplasia that is difficult to treat surgically. MEN-IIa is associated with a *ret* proto-oncogene mutation, and when accompanied by primary hyperparathyroidism this mutation is often associated with multiple parathyroid adenomas. Biochemical screening for medullary thyroid carcinoma and pheochromocytoma is indicated in MEN-IIa (see medullary thyroid carcinoma case).

Test Interpretation

A repeat measurement of calcium level with a simultaneous intact PTH level can confirm primary hyperparathyroidism if both are elevated. Obtaining ionized calcium may be more accurate than serum calcium but is more sensitive to pH change and may be less reliable in the outpatient setting. A renal panel routinely provides serum calcium, phosphorous, creatinine, and albumin levels, which aid greatly because the serum calcium can be corrected for albumin [corrected Ca = measured Ca + (4.0 − albumin) × (0.8)]. Further, serum phosphorous should be reduced in primary hyperparathyroidism. Creatinine or creatinine clearance can be an indication of renal function.

A 24-hour urinary calcium and creatinine measurement should accompany a renal panel to exclude familial hypocalciuric hypercalcemia. Patients on diuretics, especially thiazide diuretics, should discontinue use for about a month before testing.

In a patient with osteoporosis and suspected primary hyperparathyroidism, a 25-OH vitamin D level should be checked to look for associated secondary hyperparathyroidism, which if left untreated may exacerbate osteoporosis.

Bone densitometry to include the spine, hip, and radius should be performed if not already done to assess for severity of osteoporosis. The cortical long bones such as the radius may be preferentially affected in metabolic bone disease such as primary hyperparathyroidism.

This patient's serum calcium is elevated at 10.5 mg/dL (reference range, 8.4–10.4) with a normal albumin and simultaneously elevated intact PTH level of 78 pg/nL (reference range 20–65). Twenty-four-hour urine calcium was normal and 25-OH vitamin D level was low at 22 (range, 20–100). Bone density is reduced at all sites with T score below −2.5. This is consistent with primary hyperparathyroidism. It is important to understand that the diagnosis of primary hyperparathyroidism is biochemical and not based on imaging.

Diagnosis

Hypercalcemic and primary hyperparathyroidism with osteoporosis

Medical Management

Bisphosphonate therapy may improve or prevent progression of osteoporosis but has less effect on serum calcium. Vitamin D therapy may help to prevent progression of osteoporosis and improve hyperparathyroidism without significant worsening of hypercalcemia. Both are indicated in this patient. Calcium intake should not be restricted below 1200 mg daily because this will likely drive progression of osteoporosis. Dehydration can exacerbate hypercalcemia, and adequate fluid intake is advised and may help prevent kidney stones. Diuretics are controversial but may cause dehydration and are generally discouraged. Calcimimetics, such as cinacalcet, are calcium agonists used in the treatment of secondary hyperparathyroidism and may help to reduce serum calcium, but they are costly and are used primarily in patients for whom surgery has failed or who are too medically infirm to undergo surgery.

Surgical Management

Surgery is the most effective way to treat primary hyperparathyroidism. Historically, four gland explorations with removal of all enlarged parathyroid glands was the gold standard. Increasingly, unilateral or focused parathyroidectomy with removal of only an enlarged gland localized by imaging preoperatively is performed, often with intraoperative measurement of PTH levels to confirm resolution of the hyperparathyroidism. Post–adenoma removal levels that have fallen more than 50% from preincision baseline and within the reference range are considered consistent with successful resolution of hyperparathyroidism such that further surgical exploration is not warranted.

Parathyroid scintigraphy with technetium 99m sestamibi has consistently been the most sensitive parathyroid localization study but lacks anatomic detail and is less sensitive for smaller adenomas. This is often done with dual-phase technique, with early images showing thyroid and parathyroid tissue (along with salivary glands and heart) and delayed images after the thyroid washes out, showing the enlarged parathyroid. Single-photon emitted

computed tomography (SPECT) sestamibi and CT-fused sestamibi provide additional anatomic detail compared with standard anterior imaging alone. Subtraction of thyroid and parathyroid sestamibi image uptake with radioiodine imaging, which is taken up only by the thyroid gland, is an alternative strategy.

High-resolution neck ultrasound has been shown to provide equal or greater sensitivity for parathyroid adenoma localization in experienced centers at lower cost and with greater anatomic detail than sestamibi scanning. The two are complementary and are often both obtained.

CT and magnetic resonance imaging are generally less sensitive and are useful primarily for nonlocalized adenomas after previously failed surgery or for those suspected of being intrathoracic where ultrasound is not helpful.

Rehabilitation and Follow-up

This patient underwent high-resolution neck ultrasound with localization of a small right inferior parathyroid adenoma confirmed at surgery with intraoperative resolution of the hyperparathyroidism and postoperative resolution of the hypercalcemia. Adequate calcium and vitamin D intake is important in the management of the osteoporosis. Serum calcium should be measured 6 months postoperatively to ensure no recurrence and annually thereafter. Bone density testing should be repeated 2 to 3 years postoperatively.

Questions

1. Which of the following biochemical tests is consistent with primary hyperparathyroidism?
   - **A.** High calcium, low PTH
   - **B.** High calcium, high PTH
   - **C.** Low calcium, low PTH
   - **D.** Low calcium, high PTH
2. Which of the following bone density measurements is consistent with osteoporosis?
   - **A.** T score >2.5
   - **B.** T score >1.0 and <2.5
   - **C.** T score >-1.0 and <1.0
   - **D.** T score <-1.5 and >-2.5
   - **E.** T score <-2.5
3. During unilateral parathyroid exploration with removal of a single enlarged parathyroid gland, which of the following intraoperative PTH test results would suggest surgical resolution of primary hyperparathyroidism?
   - **A.** Postremoval PTH $>50\%$ of preremoval PTH
   - **B.** Postremoval PTH $<50\%$ of preremoval PTH

Suggested Readings

Bilezikian JP, Potts JT Jr, Fuleihan Gel-H, et al. Summary statement from a workshop on asymptomatic primary hyperparathyroidism: a perspective for the 21st century. J Clin Endocrinol Metab. 2002;87(12):5353–5361

Terris DJ, Gourin CG. Parathyroid diseases. In: Thyroid and Parathyroid Diseases: Medical and Surgical Management. New York, NY: Thieme; 2008

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Postthyroidectomy Hypocalcemia

David L. Steward

History

A 42-year-old man is seen in the emergency department complaining of perioral and extremity paresthesias and brachioradialis muscle spasm on postoperative day 3 after completion of a thyroidectomy for papillary thyroid carcinoma. He is taking calcium supplements 1 g three times daily after being discharged on postoperative day 1 with a serum calcium level of 7.8. On examination he has a positive Chvostek sign.

Differential Diagnosis—Key Points

Postthyroidectomy hypocalcemia resulting from hypoparathyroidism is a risk for any patient undergoing bilateral thyroid surgery, even if performed initially as a lobectomy-hemithyroidectomy with subsequent completion thyroidectomy for the contralateral lobe.

Postthyroidectomy hypoparathyroidism can be detected early in the recovery room with rapid intact parathyroid hormone (PTH) testing available at some institutions. PTH levels lower than 15 are associated with significantly increased risk for hypocalcemia. Serial calcium measurements performed in recovery on the evening of surgery and early morning postoperative day 1 can detect hypocalcemia. Corrected calcium levels below 8.0 mg/dL (reference range, 8.4–10.4) are diagnostic. Patients will often become symptomatic with corrected calcium levels below 7.5 to 8.0, with perioral numbness an early and specific complaint. Extremity paresthesias may be equally sensitive but less specific. Chvostek sign, facial twitching after tapping on the main trunk of the facial nerve, has a fairly high false-positive rate. Muscle spasm and rigidity are late and worrying signs of tetany and associated with corrected calcium levels below 7.0.

Oral calcium supplementation is the mainstay of therapy for hypocalcemia but requires concomitant vitamin D supplementation for calcium to be absorbed. PTH converts 25-OH vitamin D into 1,25-OH vitamin D, which is the active form. As such, patients with hypoparathyroidism require treatment with calcitriol (1,25-OH vitamin D). Many patients have vitamin D deficiency and may require load dosing with calcitriol to absorb calcium.

Test Interpretation

A stat calcium or renal panel is diagnostic for hypocalcemia. Ionized calcium may be more accurate but less reliable (see primary hyperparathyroidism case). In the postthyroidectomy setting, hypoparathyroidism is etiologic and

PTH testing is rarely needed for diagnosis once hypocalcemia develops.

Early rapid PTH testing in the operating room, or better in the recovery room, can diagnose hypoparathyroidism before development of hypocalcemia, allowing for early intervention and prevention of hypocalcemia.

Stat calcium was measured at 6.3 mg/dL (range, 8.4–10.4) in the emergency department. A renal panel showed a slightly low albumin at 3.8 with a corrected calcium of 6.4 and elevated serum phosphorus of 5.5 (range, 3.5–5.0). Intact PTH levels were sent but not available for several days. These were all confirmatory of hypocalcemia from postthyroidectomy hypoparathyroidism.

Diagnosis

Hypocalcemia from postthyroidectomy hypoparathyroidism

Medical Management

Severely symptomatic patients with critically low calcium levels such as this patient require intravenous calcium, preferably continuous infusion of 10 g calcium gluconate per 1 L of saline run over 10 to 12 hours until the calcium levels can be maintained within the low normal range. Intravenous boluses are an alternative to continuous infusion but may prolong the time to normalization. Oral supplementation with calcium should be instituted immediately with the frequency of dosing more important than the dose. Severe hypocalcemia may require 1 to 2 g calcium orally four to five times daily. Calcitriol should be given with a load dose of 1 to 2 μg and then 1 μg daily.

Surgical Management

No surgical treatment is currently available for postthyroidectomy hypocalcemia. An ounce of prevention is worth a pound of cure, which applies to parathyroid gland management during thyroidectomy. Parathyroid glands should be left in situ with the blood supply from the inferior thyroid artery left intact. When branches of the inferior thyroid artery are taken during thyroidectomy, they should be taken medial to the parathyroid glands. Parathyroid glands that are devascularized are minced and autotransplanted into the sternocleidomastoid or sternohyoid muscle. Before passing the thyroid specimen from the field, it is inspected to ensure that no parathyroid glands have been removed. If found, they are autotransplanted. Confirmation of parathyroid tissue with a frozen biopsy before autotransplantation may prevent transplantation of tumor in the presence of malignancy.

Rehabilitation and Follow-up

Oral supplementation with calcium and calcitriol is continued until the hypoparathyroidism resolves with target calcium between 8.0 and 9.0 mg/dL. If not resolved within 1 year, it is likely permanent. Most postthyroidectomy hypoparathyroidism is transient and will resolve within 1 month. Parathyroid glands that are autotransplanted may take longer to become functional.

This patient was admitted and treated with intravenous calcium until oral supplementation stabilized the serum calcium in the low normal range of 8.5 mg/dL. The patient was discharged on hospital day 2, on two Tums four times daily along with calcitriol 0.5 μg daily. The PTH level in the emergency department was less than 4 pg/nL (undetectable). Renal panel obtained 3 days after discharge showed calcium of 9.5 mg/dL with an elevated phosphorous consistent with adequately treated hypoparathyroidism. Weekly renal panels were obtained with a slow wean of calcium supplements from initial four times a day dosing to three times a day dosing, then twice daily, and then once daily tapered weekly based on the serum calcium levels. At 1 month, the patient's calcium was 8.5 mg/dL and intact PTH was 24 pg/nL, suggesting partial recovery of parathyroid function. By 3 months postoperatively, serum calcium, phosphorous, and PTH levels had normalized without supplements, suggesting resolution of the transient hypoparathyroidism.

Questions

1. Which of the following patient signs or symptoms is a reliable early indicator of postoperative hypocalcemia?
   A. Chvostek sign
   B. Extremity paresthesia
   C. Perioral paresthesia
   D. Brachioradialism muscle spasm
   E. Fatigue
2. What is the most important single biochemical test to order in a patient suspected of having postthyroidectomy hypocalcemia?
   A. 24-hour urinary calcium
   B. Calcitonin
   C. PTH
   D. Serum calcium
   E. Magnesium
3. What is the optimal medical management for a patient with postthyroidectomy hypocalcemia?
   A. Calcitonin
   B. PTH
   C. 25-OH vitamin D
   D. Calcitriol and calcium
   E. Levothyroxine

Suggested Reading

Jameson MJ, Levine PA. Complications of thyroid surgery. In: Terris D, Gourin C, eds. Thyroid and Parathyroid Diseases. New York, NY: Thieme; 2008

VIII

The Salivary Gland

36

Parapharyngeal Mass

Marci J. Neidich and David L. Steward

History

A 55-year-old woman with a history of three surgical resections for right recurrent pleomorphic adenoma is referred for evaluation of a neck mass. She originally had a parotid mass at the age of 25; this mass was resected, with pathology revealing pleomorphic adenoma. This recurred 1 year later, and a completion parotidectomy was performed. She had a long disease-free interval until 10 years ago, when she had another resection of mass in the parotid bed. She now has 4 months of an enlarging right neck mass, intermittent hoarseness, and right-sided otalgia. She has also noticed some mild dysphagia to solids over the past several weeks but no odynophagia. She has a history of tonsillectomy as a young child. She drinks no alcohol, has never smoked, and has no other symptoms or significant medical history.

Examination reveals a thin woman (body mass index 20) breathing quietly. She has a palpable right-sided level II mass on palpation of the neck. Intraoral examination reveals postsurgical absence of the tonsils and mild fullness of the right oropharyngeal wall. There is no erythema or exudate of the oropharynx. She is afebrile and has mild trismus. Ear examination is normal. On the right, she has an obvious facial droop at rest with incomplete eye closure consistent with a House-Brackman IV/VI. Otherwise, cranial nerves are grossly intact.

Differential Diagnosis—Key Points

1. Critical to the understanding of the diagnosis and management of these masses is knowledge of the anatomy and boundaries of the parapharyngeal space. The space is divided into two compartments through which various structures course. The anterolateral prestyloid space contains mostly fat, a variable portion of the deep lobe of the parotid, internal maxillary artery, inferior alveolar nerve, lingual nerve, and the auriculotemporal nerve. The posteromedial retrostyloid space contains the carotid, jugular vein, cranial nerves IX–XII, sympathetic chain, and lymph nodes. This knowledge will help narrow down the diagnosis once imaging delineates the location of the mass.
2. The differential diagnosis of masses that arise in this space is vast, with many case reports existing in the literature of rare presentations of various tumors. The most common diagnoses are presented below:
   A. Salivary gland tumors (40–50%): These may be from the parotid (benign or malignant, most commonly pleomorphic

adenoma) or minor salivary glands (arising either in the lateral pharyngeal wall or from ectopic tissue).

B. Neurogenic tumors (20–25%): This includes schwannoma, neurofibroma, glomus vagale, carotid body tumor.

C. Lymph node (15%): Lymph node enlargement may be primary (i.e., lymphoma) or metastatic disease from a pharyngeal cancer at any level.

D. One must keep in mind a variety of other miscellaneous tumors: these may be arising from muscle (rhabdomyoma, leiomyoma), connective tissue (lipoma, fibroma), or congenital (branchial cleft cyst, dermoid, lymphangioma). Another possibility is that of pseudotumors such as aneurysm, abscess, and myositis.

3. Patients with tumors in this space may have quite subtle symptoms despite a substantial sized mass. Often they are found incidentally when a scan is obtained for another purpose. Pain and cranial nerve palsies are red flags suggestive of malignancy, although these symptoms and signs may also occur in benign tumors. It is key to obtain a full review of systems and social history (i.e., tobacco and alcohol), which may raise suspicion of a primary pharyngeal cancer. Inquire about recent infections because the patient may present with an abscess once the primary tonsillitis has resolved. Some rare symptoms that may bring patients to attention include syncope, bradycardia, and hypertension from involvement of the carotid sinus.

4. The lesion is usually found on examination of the oropharynx. Further testing is usually needed for both consideration of the diagnosis and planning for the therapeutic approach.

◆ Test Interpretation

1. Computed tomography (CT) is a good first choice for evaluation of the parapharyngeal space. This modality can differentiate prestyloid from retrostyloid tumors; deep-lobe versus extraparotid tumors based on fat planes; and base of skull erosion. Relationship of the tumor to surrounding structures may give clues as to whether the tumor is benign or malignant. In this case the tumor was noted to be prestyloid.

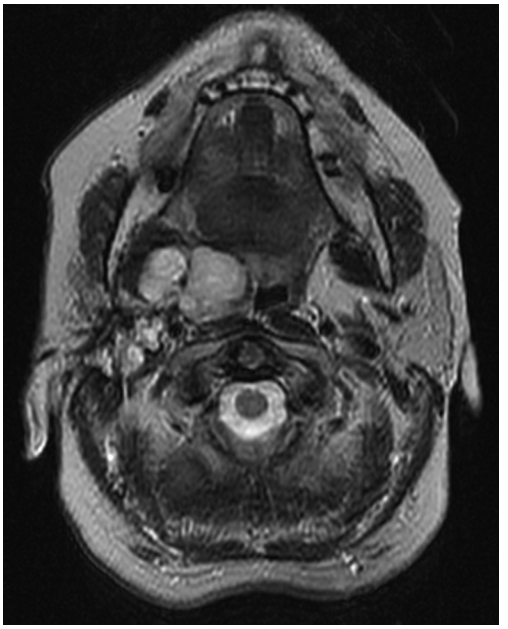

Fig. 36.1 T2-weighted magnetic resonance image of a parapharyngeal space mass.

2. Magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) may be valuable adjuncts, depending on the findings on the CT scan (**Fig. 36.1**). These studies can delineate intracranial extension and vascular involvement. Angiography may be needed if embolization is a consideration or for other preoperative planning.

3. Biopsy is not usually necessary preoperatively unless malignancy is suspected. If needed, biopsy should be done using fine-needle aspiration (FNA) with CT guidance, either by transoral or transcervical approach. Transoral open biopsy is dangerous, often unnecessary, and should be avoided because of the risks of tumor seeding and vascular injury.

4. If the diagnosis is suspected to be nodal metastases, panendoscopy should be performed. Additionally, if paraganglioma is suspected, obtain urinary catecholamines so that prophylaxis for hypertension and arrhythmias may be put into place during resection.

◆ Diagnosis

Recurrent pleomorphic adenoma involving the soft tissues of the neck

◆ Medical Management

In this specific case scenario, there is no need for preoperative medical management other than ensuring that there are no medical co-morbidities to surgery. The management of these masses is largely surgical. If an infectious cause is suspected, appropriate antibiotics should be started. Depending on the size of the lesion, airway observation is always a primary consideration. This patient did require a temporary tracheostomy placed at the time of her surgical resection for airway protection. One case report in the literature describes a lymphovenous malformation of the parapharyngeal space, which did regress with steroids and antibiotics alone.

◆ Surgical Management

Definitive diagnosis is usually made on surgical exploration. Because the exact procedure that is required may be unclear preoperatively, a thorough discussion must be had with the patient regarding possible approaches and risks of cranial nerve and vascular injury.

The two main options for approach are transoral and transcervical. Transoral approach is reserved only for very select cases, for the same risks noted in discussion of transoral biopsy. The vast majority of the time, a transcervical approach is used with a horizontal submandibular incision two finger breadths below the mandible. If the tumor arises from the parotid gland, this incision may be extended to perform superficial parotidectomy. This is unnecessary if the tumor is not connected to the parotid. If more access is needed, mandibular osteotomy may be performed, and this possibility should be included in discussions of informed consent with the patient. Vagal schwannomas are treated by subcapsular enucleation. Vagal paragangliomas may require preoperative embolization, and resection often requires sacrifice of the nerve. Grafting may be an option in this scenario.

In this specific case, the surgical resection involved an infratemporal preauricular approach as well as transmandibular approach to the mass, along with a selective neck dissection (levels I–IV). All lymph nodes examined were negative for disease.

◆ Rehabilitation and Follow-up

In most cases of tumors of minor and major salivary glands of the parapharyngeal space, recurrence is not a problem. Injury to cranial nerves in the region resulting from direct involvement of the tumor and need for sacrifice, or inadvertent injury during surgery, create the most problems postoperatively. This may lead to the need for further procedures such as gastrostomy and thyroplasty. Young patients with no other co-morbidities may compensate well and should be managed expectantly. This highlights the importance of examining and documenting cranial nerve function preoperatively.

Following surgery of the parapharyngeal space, there is documentation in the literature of the phenomenon known as “first-bite syndrome,” or pain that is felt in the region of the parotid following the first bite of a meal. This pain is thought to be due to disruption of sympathetic nerve function to the parotid.

The patient described in this case required tracheostomy and percutaneous endoscopic gastrostomy tube placement at the time of surgical resection; these tubes were eventually removed. She also received postoperative radiation therapy because of her history of multiple recurrences and invasion of soft tissue of the neck.

Questions

1. What is the most common tumor of the parapharyngeal space?
2. What compartment is involved in a tumor arising from the deep lobe of the parotid?
3. What is the best surgical approach for removal of a tumor of the parapharyngeal space?

Suggested Reading

Olsen KD. Tumors and surgery of the parapharyngeal space. Laryngoscope. 1994;104(5 Pt 2, Suppl 63):1–28

37

Recurrent Pleomorphic Adenoma

Collin M. Burkart and David L. Steward

History

A 38-year-old white man consults you about nodules on the right side of his face. He indicates that these nodules have been present for several years but seem to be growing larger and are becoming a concern for him. He reports no pain, no numbness or paresthesia of the face, and no facial weakness. These nodules are located on the right side of his face anterior and inferior to his ear near a scar that he reports is from a surgery to remove a benign tumor when he was in college (**Fig. 37.1**).

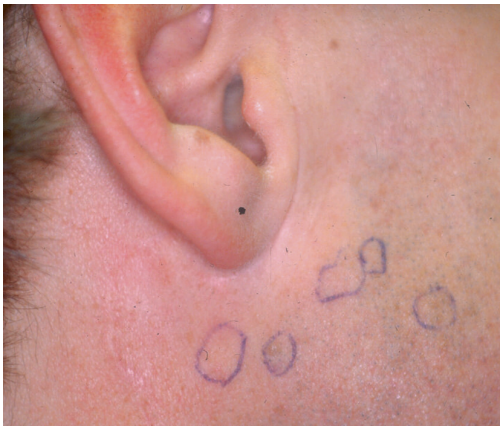

Fig. 37.1 Picture of a patient with recurrent pleomorphic adenoma. Lesions are outlined by a marking pen.

His main concern with these nodules is primarily cosmetic, although when asked, he admits to being worried about cancer. He is a nonsmoker and does not have a history of much sun exposure. He cannot remember the type of benign tumor he had removed; he does remember that his lower lip was weak on the right side for several months after the surgery but eventually returned to normal.

On physical examination, there is a well-healed preauricular scar consistent with a parotidectomy incision. Five palpable masses just anterior to this scar are subcutaneous and nonmobile. The largest of these masses is 1.1 cm in diameter; the remaining four are about 0.5 cm in diameter. They are nontender to palpation and located within a few millimeters of each other but clearly separate masses. Cranial nerve VII examination is normal bilaterally, and the intraoral examination is normal with no lateral pharyngeal bulge. The remainder of the head and neck examination is normal.

Differential Diagnosis—Key Points

1. In this setting of a preauricular mass, the differential diagnosis must include all parotid benign and malignant neoplasms and cutaneous neoplasms. However, with a history of having a benign mass removed

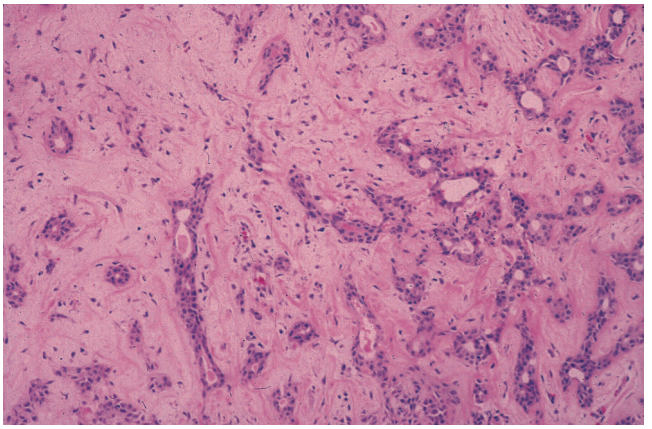

Fig. 37.2 Pathology demonstrating histology consistent with pleomorphic adenoma.

from the preauricular area, particular consideration must be given to the potential diagnosis of recurrent pleomorphic adenoma (**Fig. 37.2**).

2. In this setting it is important to determine as definitively as possible the nature of the previously excised mass. It is important to obtain previous medical records, and if possible histologic slides should be reviewed to confirm the previous pathologic diagnosis and its benign character.
3. A fine-needle aspiration with or without ultrasound guidance is an important diagnostic step in determining the nature of these preauricular masses. This is especially important if the previous pathology cannot be confirmed. This will aid in the management planning. The important distinction is to determine whether the lesions are benign or malignant.
4. Options for imaging include ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI). Some advocate using ultrasound routinely to follow up on patients who have had a pleomorphic adenoma treated as part of the follow-up clinic visit. In this situation, when disease is clearly present or in situations when the presence of disease is suspected, MRI is the best choice for imaging. MRI has been shown to be the best imaging modality to delineate the nature and extent of parotid lesions. In cases of recurrent pleomorphic adenoma, the palpable lesions may just be the "tip of the iceberg" and MRI has been shown to best demonstrate the nature of multiple lesions.
5. An important finding of the clinical examination is the function of the facial nerve. It is also important to ascertain whether the facial nerve function is abnormal and, if so, to what extent this is due to previous treatment. If, after the patient's initial therapy the facial nerve was normal but is now demonstrating weakness, concern for malignant transformation must be considered. Although recurrent pleomorphic adenoma has been reported to cause facial nerve weakness, especially when the nerve is encased in tumor or scar, this is not a common occurrence. It has been reported that malignant transformation more commonly presents in deep-lobe parotid pleomorphic adenoma as the disease tends to progress for a longer time asymptomatically. Malignant transformation is also a concern if other cranial nerve deficits are seen from involvement in the parapharyngeal space.

◆ Test Interpretation

1. Fine-needle aspiration: Positive for pleomorphic adenoma
2. Ultrasound: Multiple separate masses throughout the parotid bed with some not palpable
3. MRI: As indicated, MRI is the imaging modality of choice. It not only will demonstrate the extent of disease better than CT imaging, but it will also demonstrate the amount and location of residual parotid gland, which is especially important if the treating surgeon

was not the surgeon who operated on the initial presentation. In this situation, a residual superficial and deep lobe of the parotid are seen, with six separate masses identified. Pleomorphic adenoma on MRI is low intensity on T1-weighted images and high intensity on T2-weighted images with mild enhancement after contrast administration.

Diagnosis

Recurrent pleomorphic adenoma

Medical Management

The management of recurrent pleomorphic adenoma is a complicated problem. Each patient must be considered on a case-by-case basis. Considerations for treatment include location and number of masses, previous therapy, number of recurrences, status of facial nerve, patient preference, patient age, and patient co-morbidities. Malignant transformation is a serious complication of recurrent or persistent pleomorphic adenoma but occurs rarely, with reports being between 0 and 8%, with rates as high as 15% reported in cases of multiple recurrences. This has been suggested to have an increased prevalence in patients who had undergone previous radiotherapy. However, with malignant transformation occurring infrequently, it is usually not a consideration in the management of recurrent pleomorphic adenoma. The indications for treatment are cosmetic concerns of the patient, including status of the facial nerve and potential malignant transformation.

Nonsurgical management of recurrent pleomorphic adenoma includes observation or radiotherapy. Observation is a clear option in the management of recurrent pleomorphic adenoma. This is especially an option in medically infirm or older patients or in patients in whom the facial nerve has normal or near-normal function, but surgery would clearly place the nerve at considerable risk because of multiple recurrences or multiple foci of pleomorphic adenoma. Other patients who might qualify for observation include those with small burden of tumor; in these situations, the patient should be followed up closely with clinical examinations and ultrasound to determine any change in the status of the disease. If observation is the path of management, extensive discussion must occur between the surgeon and the patient concerning the risk for malignant transformation, cosmetic detriment, and the potential for facial nerve weakness.

An alternative nonsurgical management of recurrent pleomorphic adenoma is radiotherapy, especially neutron beam if available. This has been shown to be an effective alternative when surgery is contraindicated or the risks of surgery impose a barrier for the patient. One study demonstrated a 75% locoregional control rate at 15 years in patients with gross disease. It has been suggested, however, that radiotherapy may increase the risk of malignant transformation; therefore this should be considered with caution in younger patients.

Surgical Management

The surgical management of recurrent pleomorphic adenoma depends on the extent of tumor foci and the extent of normal parotid tissue remaining. The surgery should be performed with the goal of preserving facial nerve function, which may be facilitated by intraoperative facial nerve monitoring. Many hypotheses have been suggested to explain the occurrence of recurrent pleomorphic adenoma, such as cell biological and genetic factors, but these are controversial. It is most widely accepted that the reason for development of recurrent pleomorphic adenoma is tumor spillage, incomplete excision, or violation of the pseudocapsule. Therefore excising this tumor with a wide cuff of tissue has been advocated, and when this technique is used at initial removal of pleomorphic adenoma, recurrence rates are reported to be less than 4%. The recurrence rate of up to 4% has been attributed to the surgeon attempting to preserve facial nerve function and not maintaining an adequate cuff when the tumor opposes the facial nerve.

The choice of the specific surgical management is patient specific. It has been reported that the more widely the tissue is excised, the better the control rate and reduction of recurrent disease. However, this comes at the

expense of increased risk of facial nerve injury both in cases of attempts at nerve sparing and in case of intentional nerve sacrifice. The unintentional risk to the facial nerve in surgery for recurrent pleomorphic adenoma has been reported to be as high as 30%. In cases of planned facial nerve sacrifice, for situations of preoperative facial nerve weakness or because of tumor location and burden, a consideration for intraoperative nerve repair should be considered. A high rate of multiple recurrences does exist, and with each recurrence, the risk to the facial nerve is greater. Multiple foci of tumor also lead to more difficult surgical control of disease with increased risk to the facial nerve. In many studies the prevalence of multiple nodules is around 75%.

Some argue for the use of neutron radiotherapy in an adjuvant fashion for multinodular recurrences, obvious tumor spillage, large tumor size, deep location or location adherent to the facial nerve in facial nerve–sparing surgery, positive margins, and metachronous recurrences.

Rehabilitation and Follow-up

A secondary recurrence of recurrent pleomorphic adenoma is unfortunately commonly encountered. Various reports found in the literature described rates commonly seen at 43 to 45% at 10 years, with estimations calculated as high as 57% or higher at 20 years. With these high rates of recurrence, it is not only essential to counsel the patient preoperatively but also to follow up with the patient closely for years following the therapy. This follow-up should include focused clinical examinations, and some argue that ultrasound should be used regularly at these appointments. Ultrasound may be able to find subclinical recurrent disease.

In cases of facial nerve weakness with a known intact facial nerve, this should be monitored for improvement, with the understanding that the deficit may be permanent. In patients with known facial nerve sacrifice, appropriate therapy should be initiated, including referral for possible facial reanimation procedures.

Questions

1. A 45-year-old woman is referred by her primary care physician. She is concerned because she has noticed an abrupt onset of weakness of the muscles at the angle of her mouth. Her biggest concern is drooling. She mentions that she had weakness like this in the past that was temporary after a surgery for a benign parotid tumor. This surgery was 15 years ago. What is the most likely cause of her facial paralysis?
- **A.** Postoperative complication
- **B.** Recurrent pleomorphic adenoma
- **C.** Malignant transformation of persistent or recurrent pleomorphic adenoma
- **D.** Acoustic neuroma
- **E.** Complication from otitis media

2. A patient consults you for a suspected pleomorphic adenoma. What is the imaging modality of choice to evaluate the extent of disease and condition of the parotid gland?
- **A.** MRI
- **B.** CT
- **C.** Nuclear medicine study
- **D.** Ultrasound
- **E.** Positron emission tomography

3. Options for treatment of confirmed recurrent pleomorphic adenoma with a small burden of disease in an otherwise healthy but older patient with normal facial nerve function include all of the following except which one?
- **A.** Observation with close monitoring
- **B.** Surgical excision with a wide cuff of tissue
- **C.** Total parotidectomy with facial nerve preservation
- **D.** Total parotidectomy with facial nerve sacrifice
- **E.** Surgical excision with adjuvant neutron beam radiotherapy

Suggested Readings

Douglas JG, Einck J, Austin-Seymour M, Koh WJ, Laramore GE. Neutron radiotherapy for recurrent pleomorphic adenomas of major salivary glands. Head Neck 2001; 23(12):1037–1042

Koral K, Sayre J, Bhuta S, Abemayor E, Lufkin R. Recurrent pleomorphic adenoma of the parotid gland in pediatric and adult patients: value of multiple lesions as a diagnostic indicator. AJR Am J Roentgenol 2003;180(4): 1171–1174

Leonetti JP, Marzo SJ, Petruzzelli GJ, Herr B. Recurrent pleomorphic adenoma of the parotid gland. Otolaryngol Head Neck Surg 2005;133(3):319–322

Redaelli de Zinis LO, Piccioni M, Antonelli AR, Nicolai P. Management and prognostic factors of recurrent pleomorphic adenoma of the parotid gland: personal experience and review of the literature. Eur Arch Otorhinolaryngol 2008;265(4):447–452

38

Submandibular Gland Adenoid Cystic Carcinoma

Ojas Shah and David L. Steward

History

A 52-year-old white woman is referred by her primary care physician for a persistent lump under the right jaw. The mass first appeared 6 weeks ago, and the patient was placed on Augmentin for a presumed diagnosis of acute sialadenitis. However, after 2 weeks of antibiotic therapy, the size of the jaw mass was undiminished, nor has it changed size in the last month. She denies any fevers, chills, night sweats, or other lumps in the neck. She also denies weight loss, persistent cough, dysphagia, odynophagia, hoarseness, difficulty chewing or moving her tongue, or weakness of the face. Her medical history is significant only for two spontaneous vaginal deliveries and arthroscopic knee surgery. She currently takes metoprolol for high blood pressure and prophylactic aspirin. She denies ever using tobacco and consumes two or three glasses of wine socially every week. On physical examination, her eyes, ears, oropharynx, hypopharynx, and larynx are unremarkable. The face is symmetric in appearance. The tongue is mobile and is unremarkable; the base of tongue and floor of mouth are soft. There is no purulent drainage from the Wharton duct, nor is a stone palpated in the duct. There is a right-sided 2 × 2 cm submandibular mass that is firm, nonmobile, and nontender. No other palpable neck masses or lymph nodes are found.

Differential Diagnosis—Key Points

1. The differential diagnosis can be generally classified into inflammatory and noninflammatory causes. Inflammatory causes include acute and chronic infectious lesions from bacterial (e.g., *Staphylococcus*, *Streptococcus*, *Haemophilus*, *Escherichia coli*, *Bartonella*, *Mycobacterium*, syphilis, Lyme disease), viral (e.g., Paramyxovirus, cytomegalobirus, Epstein-Barr virus, human immunodeficiency virus), and fungal (e.g., histoplasmosis, coccoid mycosis) agents, autoimmune diseases (Sjögren syndrome), and granulomatous diseases (Wegener, sarcoidosis). Noninflammatory causes include sialothiasis, sialocele, sialadenosis, benign or malignant neoplasms, radiation injury, or trauma.
2. Acute sialadenitis typically presents with diffuse enlargement of the gland, tenderness, and induration of the gland. Pus can also be seen at the Wharton duct on massage

of the gland. With sialolithiasis, palpation of the gland may reveal the presence of a stone in the Wharton duct. Given the patient's age and presentation, a neoplasm should be suspected. A comprehensive evaluation of the head and neck should be performed to rule out the presence of a tumor that may have metastasized to the neck. The most appropriate method of evaluation at this point would include a fine-needle aspiration biopsy (FNA), which could distinguish an inflammatory versus noninflammatory lesion.

3. About 50% of submandibular gland neoplasms are benign. The most common benign neoplasm of the parotid and submandibular glands is pleomorphic adenoma. Other benign neoplasms of the salivary glands include oncocytoma, Warthin tumor, and monomorphic adenoma. Malignant neoplasms include mucoepidermoid carcinoma, adenoid cystic carcinoma, acinic cell carcinoma, adenocarcinoma, carcinoma ex-pleomorphic adenoma, squamous cell carcinoma, and lymphoma. A neoplastic mass that is tender or associated with paralysis of the facial nerve (or its branches) or the hypoglossal nerve is suspicious for a malignancy. In malignant neoplasms, pain is usually indicative of perineural invasion, which predicts a worse prognosis than a neoplasm that is not painful.

◆ Test Interpretation

1. Biopsy: FNA will be diagnostic in most cases and can be performed with the assistance of ultrasound or computed tomography (CT) guidance if the lesion is difficult to palpate. In cases of a nondiagnostic first FNA, most studies recommend a second FNA. An incisional biopsy of the submandibular gland is to be avoided because of the risk of potentially seeding tumor. For this patient, a biopsy revealed a cribriform pattern with nests and columns of epithelial and myoepithelial cells surrounding large glandlike spaces filled with hyaline periodic acid–Schiff (PAS)-positive material. At the time of surgical excision, the histologic analysis revealed a characteristic "Swiss cheese" pattern.

2. Basic imaging: Ultrasound is useful for detecting the presence of a mass in the salivary gland. Furthermore, it may be useful in distinguishing a cystic lesion from a solid lesion. CT scans provide valuable information about the extent of disease. A CT provides good detail about tumor volume, the relationship to bony and vascular structures, and the extent of lymphatic involvement. In addition, the CT scan may indicate involvement of perineural structures by demonstrating widening of specific foramina (e.g., mental foramen, foramen ovale). Magnetic resonance imaging (MRI) is useful in delineating soft tissue involvement. It is particularly useful in cases where there is concern for intracranial involvement or extension of tumor. In this case, a CT demonstrated involvement of the right submandibular gland with no evidence of invasion into any of the surrounding tissues or neurovascular structures. No lymphadenopathy was noted. An MRI scan was not performed.

3. Positron emission tomography (PET) scan: Several studies have been performed that demonstrate the usefulness of PET scans for the initial diagnosis and management of head and neck cancers. It has been particularly useful for detecting recurrent disease and distant metastases in the follow-up period. One series demonstrated significant changes in treatment strategy based on the results of the PET scan. For this patient, a PET scan demonstrated some increased uptake in the region of the right submandibular gland but no evidence of any distant metastatic disease.

◆ Diagnosis

Adenoid cystic carcinomas (ACCs) constitute 10% of all salivary gland neoplasms. ACC is the second most common malignant neoplasm of the parotids and the most common malignancy of the minor salivary glands and the submandibular glands. It occurs with equal frequency in males and females. It is derived from mucus-secreting glandular cells of the foregut. Histologically, ACCs are divided into the cribriform, tubular, and solid subtypes. The most common is the cribriform subtype,

which has a glandular pattern with vacuolated areas. The tubular pattern is represented by cords and groups of malignant cells. The solid subtype is characterized by solid sheets of malignant cells. The histologic subtype correlates with overall prognosis. The solid subtype has the worst prognosis in terms of recurrence and survival. The cribriform pattern has the best prognosis. It is important to note that the different histologic patterns can be found within the same tumor, thereby making the pathological evaluation difficult.

Medical Management

Chemotherapy is typically reserved for locoregional recurrences and distant metastases, which are not amenable to surgical resection or radiation therapy. Several trials that have demonstrated responses to cisplatin, vinorelbine, epirubicin, mitoxantrone, cyclophosphamide, doxorubicin, and anthracycline, either as single agents or as combination therapy. However, there is still no clearly superior regimen at this time. In addition, a few series have also investigated therapies that target the *c-kit* gene in ACC but have not demonstrated a clear survival benefit. A good initial response has been seen with radiation therapy alone, but most tumors will recur with time. The best responses are typically seen with a combination of surgery and radiation therapy. Radiation therapy, at doses greater than 60 Gy, may be best for treating unrecognized microscopic residual disease, in particular that resulting from ACC's propensity for perineural invasion.

Surgical Management

Surgery along with postoperative radiation therapy continues to remain the mainstay of treatment for ACC, which has a propensity for long-term recurrence and distant metastasis. This is due largely to the propensity of ACC toward perineural invasion. For malignant tumors, surgical excision usually involves the gland and the contents of the submandibular triangle. If the tumor involves the surrounding tissue, the surgical excision includes the involved tissue leaving a tumor-free margin. Nerves are not excised unless they are found to be infiltrated by tumor. Elective neck dissection in the absence of metastatic lymphadenopathy has no role because the incidence of lymphatic metastasis is very low.

Rehabilitation and Follow-up

Several reports over the years have documented the long-term recurrence of ACC. The recurrence has been found to be at both the local site and distant sites. For that purpose, several studies have advocated that patients be followed up for the rest of their life because of this risk of recurrence. Although there is no established surveillance protocol, routine follow-up with chest radiographs has been recommended. PET scans also serve as effective imaging studies for evaluating recurrence.

The patient in this case was treated with wide-excision surgery involving the right submandibular gland and the submandibular triangle, followed by local radiation therapy.

Questions

1. What is the most common type of malignancy of minor salivary glands and the submandibular glands?
- **A.** Adenoid cystic carcinoma
- **B.** Squamous cell carcinoma
- **C.** Acinic cell carcinoma
- **D.** Adenocarcinoma

2. What are the different imaging modalities that are useful in the evaluation of a salivary gland lesion?
- **A.** PET
- **B.** CT
- **C.** MRI
- **D.** All of the above

3. What characteristics of a salivary gland mass are suspicious for a malignancy?
- **A.** Pain
- **B.** Paralysis of the hypoglossal nerve
- **C.** Paralysis of the facial nerve
- **D.** All of the above

Suggested Readings

Chen AM, Bucci MK, Weinberg V, et al. Adenoid cystic carcinoma of the head and neck treated by surgery with or without postoperative radiation therapy: prognostic features of recurrence. Int J Radiat Oncol Biol Phys 2006;66(1):152–159

Chen AM, Garcia J, Granchi PJ, Johnson J, Eisele DW. Late recurrence from salivary gland cancer: when does "cure" mean cure? Cancer 2008;112(2):340–344

Chen AM, Garcia J, Lee NY, Bucci MK, Eisele DW. Patterns of nodal relapse after surgery and postoperative radiation therapy for carcinomas of the major and minor salivary glands: what is the role of elective neck irradiation? Int J Radiat Oncol Biol Phys 2007;67(4):988–994

Khafif A, Anavi Y, Haviv J, Fienmesser R, Calderon S, Marshak G. Adenoid cystic carcinoma of the salivary glands: a 20-year review with long-term follow-up. Ear Nose Throat J 2005;84(10):662–664, 664–667

Kokemueller H, Eckardt A, Brachvogel P, Hausamen JE. Adenoid cystic carcinoma of the head and neck: a 20 years experience. Int J Oral Maxillofac Surg 2004; 33(1):25–31

Laurie SA, Licitra L. Systemic therapy in the palliative management of advanced salivary gland cancers. J Clin Oncol 2006;24(17):2673–2678

Westra WH. The surgical pathology of salivary gland neoplasms. Otolaryngol Clin North Am 1999;32(5):919–943

IX

The Skin

39

Basal Cell Carcinoma

Tiffany Pickup and Hugh M. Gloster

History

A 42-year-old white woman had a 1-year history of an enlarging lesion involving her left nasal ala. During the past 3 months, the lesion bled intermittently. Medical history is significant for a 10 pack-year cigarette smoking history. Family history is notable for melanoma in her father.

Physical examination revealed a well-developed, well-nourished middle-aged white woman who appeared older than her stated age. Examination of her face revealed significant photoaging. Involving her entire left nasal ala and extending into the nasal vestibule was an ill-defined, approximately 3.0 × 2.5 cm brightly erythematous, telangiectatic, crusted, focally ulcerated plaque with pearly borders (**Fig. 39.1**). The remainder of the physical examination was within normal limits.

Differential Diagnosis—Key Points

Basal cell carcinoma (BCC) subtypes and the differential diagnosis:

1. Nodular BCC. The most common subtype, commonly found on sun-exposed areas (head, neck), often appears as a telangiectatic pearly papule with rolled borders and frequent ulceration. Differential diagnosis includes dermal nevus, seborrheic keratosis, amelanotic melanoma, and sebaceous carcinoma.
2. Pigmented BCC. This subtype appears as a hyperpigmented, sometimes pearly papule. Differential diagnosis includes compound nevus, blue nevus, appendageal tumor, seborrheic keratosis, and nodular melanoma.
3. Superficial BCC. This subtype appears most commonly as an erythematous patch or plaque on the trunk. Differential diagnosis includes a single plaque of dermatitis or psoriasis, Bowen disease, Paget disease, tinea corporis, and squamous cell carcinoma.
4. Morpheaform (sclerosing) BCC. This aggressive variant appears as a white or pink plaque. Differential diagnosis includes scar tissue and morphea.
5. Fibroepithelioma of Pinkus. This subtype often appears as a pink papule on the trunk. Differential diagnosis includes acrochordon and dermal nevus.

Test Interpretation

A biopsy is the diagnostic procedure of choice. Biopsy can be performed either by shave biopsy, incisional (punch) biopsy, or excisional biopsy with primary closure. The shave biopsy is usually preferred, with the exception of

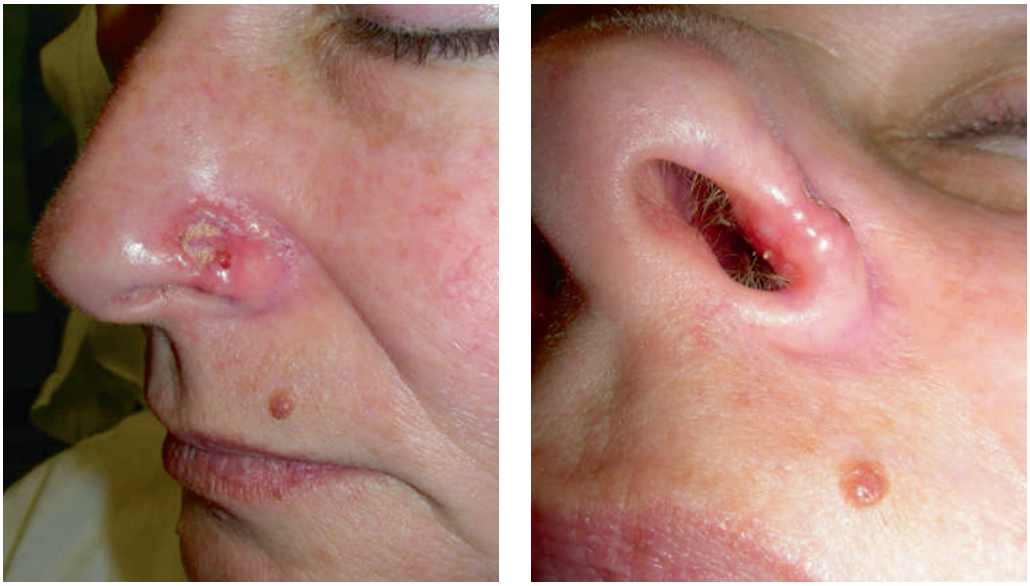

Fig. 39.1 Patient photos. **(A)** Entire left nasal. **(B)** Nasal vestibule.

morpheaform BCC or recurrent BCC in a scar, in which a punch biopsy is more effective. If an incisional biopsy is performed, it is important to include the thickest portion of the nodule and the periphery to show maturation and the deepest area of invasion. The result of the biopsy in this patient revealed nodular BCC invading deep into the subcutaneous tissues. Given the invasive nature of this tumor and the high risk of this location, the following additional evaluation should be considered:

1. A complete otolaryngology examination should be performed before removal of the primary tumor so that an estimate of the tumor size and possible spread can be evaluated.
2. If any signs of bone, orbital, or sinus invasion are evident, a preoperative high-resolution CT scan of the orbit and paranasal sinuses should be performed up through the skull base to rule out orbital, sinus, or intracranial invasion. If intracranial invasion is a concern, MRI may be helpful to evaluate dural invasion. If the tumor is extensive enough to warrant a craniofacial resection or orbital exenteration, a metastatic workup that includes a chest radiograph and bone and liver serology should be obtained at the minimum. More focal evaluations can be performed if any signs or symptoms of distant metastasis are evident. Metastatic BCC is exceedingly rare, occurring in less than 0.01% of cases.

◆ Diagnosis

BCC of the left nasal ala extending into the nasal vestibule

◆ Medical Management

This patient has a BCC in a significant functional and cosmetic area. The patient should have a complete cutaneous examination to rule out other cutaneous malignancies. A facial plastic surgery consultation should be obtained to ensure optimal treatment regarding closure and cosmesis.

Although not appropriate in this case, many studies have proposed the use of imiquimod 5% cream daily for 6 weeks to treat superficial BCC.

◆ Surgical Management

Treatment

Treatment of BCC in the nasal area is challenging. The various modalities of treatment include excision with frozen-section control,

Mohs micrographic surgery, cryosurgery, curettage, and radiation therapy. The cure rates with each of these techniques exceed 90% in most large series, depending on the subtype and location of the BCC. However, given the high-risk nature of this lesion in this critical anatomic location, it is vital to be initially aggressive to prevent recurrence. Excision with frozen section control is the historical standard. This technique has been criticized for not providing adequate control because of the difficulty in obtaining good frozen sections in this functionally crucial area. The concern is that to gain adequate control of frozen section, sacrifice of unnecessary normal tissue may be required. Mohs micrographic surgery has the advantage of using a microscopic sampling technique to enable better preservation of normal tissues. Most studies have suggested a less than 1% recurrence rate for primary BCCs treated by Mohs surgery. In addition, Mohs surgery is the treatment of choice for morpheaform and infiltrating BCCs, recurrent BCCs, and other high-risk BCCs (nasal, ocular, and postauricular regions) where tissue conservation is of the utmost importance. The disadvantages of Mohs surgery are that it is more time consuming and possibly may require two separate sittings for removal and reconstruction. Finally, radiation therapy offers advantages that include less patient discomfort and a nonsurgical option for patients in poor health who are not good surgical candidates. However, in large BCCs the cure rates are less than surgical removal. In addition, cosmesis is often rated inferior to surgical excision and actually may worsen over time. Curettage and cryosurgery are reserved primarily for the treatment of low-risk BCCs and are not appropriate treatment modalities for large, aggressive, deeply invasive tumors (**Fig. 39.2**).

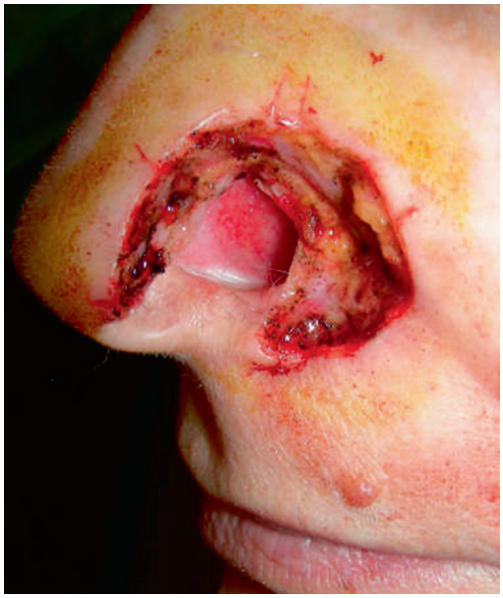

Fig. 39.2 After Mohs excision, the patient had an extensive full-thickness defect of the left lateral portion of the nose involving mucosa, cartilage, and skin.

Reconstruction

The key point to adequate reconstruction in the nasal area is to try to recreate function and cosmesis at the same time. This case required reconstructing the alar cheek junction, the alar tip and sidewall, and the floor of the nose and mucosal lining.

Because this was a full-thickness defect, the options for closure are limited. Surgical repair includes the following:

1. Reconstruction of the inner nasal lining using an inner nasal mucosal flap, reconstruction of the middle nasal layer using nasal septal or ear cartilage, and repairing the outer skin using a paramedian forehead flap (chosen in this case).
2. Another option is a nasal labial skin flap for outside lining.

Key Functional Points

1. For reconstructive planning of nasal skin defects, utilization of the nasal subunit principle is helpful. If greater than 50% of the subunit is involved, consideration can be made to excise the remaining skin within that subunit. In this way, the whole subunit can then be reconstructed to allow for the incisions and skin contour contraction to appear more natural. Nasal defects can also be classified by size. Defects are classified as *small* when they are smaller than 1.5 cm or as *large* when they are greater than 1.5 cm. If the skin defect is larger than 1.5 cm, then either a skin flap or skin graft are more likely used.
2. When there is a large defect with loss of nasal mucosa, vestibular lining must be replaced. The surgeon must choose a lining that has

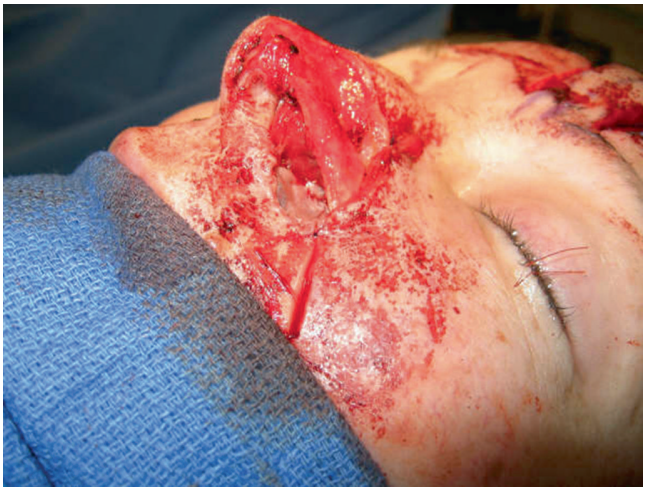

Fig. 39.3 Reconstruction of the nasal vestibule.

adequate vasculature to support cartilage grafts, pliable enough to conform to the proper shape, and thin enough that it will not obstruct the airway. In this case, a unilateral septal mucosa skin flap was used in combination with a bipedicle vestibular skin flap, based medially on the nasal septum and laterally on the nasal floor (**Fig. 39.3**).

3. Once the lining is restored, reconstruction of the lateral border of the nasal ala must ensue. In this case, the left ear conchal bowl cartilage was used to reconstruct the ideal contour of the ala sidewall and alar tip.
4. The surface of the nose is different depending on the nasal subunit. The dorsum and sidewall of the nose are nonsebaceous skin; however, the nasal tip and ala are covered by thick, sebaceous skin. Thus the forehead is acknowledged as the best tissue reservoir for the nasal tip and alar regions because of its equivalent color and texture (**Fig. 39.4**).

◆ Rehabilitation and Follow-up

Patients diagnosed with sun-related skin cancer have a 30 to 40% chance of developing another cutaneous malignancy. Thus, routine follow-up is important for tumor surveillance. Furthermore, patients must be closely followed to guard against local recurrence. Close follow-up is especially important in this functionally important area.

Because the nasofacial junction lies at embryologic fusion planes, it is theorized that this region can have a higher likelihood of deep penetration and mortality. As a result, close follow-up of this area is of utmost importance. Biopsy should be done at the earliest sign of recurrence and resection performed if the biopsy is positive. In addition, avoiding sun exposure and using sunscreens is recommended.

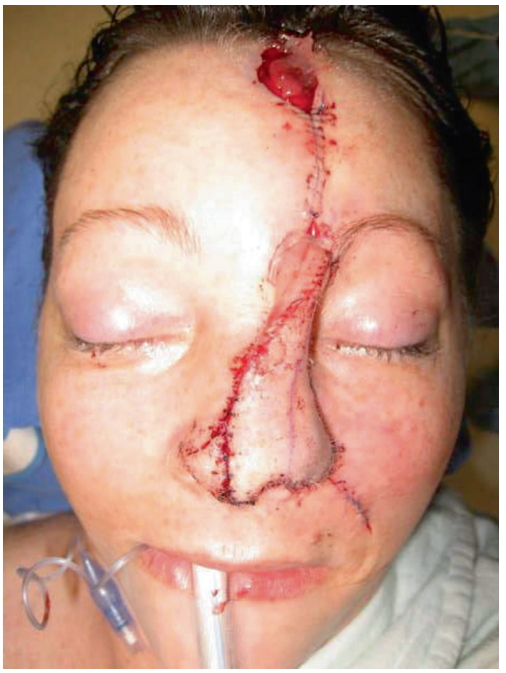

Fig. 39.4 Left paramedian forehead flap sutured into place.

Questions

1. What is the treatment of choice for recurrent basal cell carcinomas (BCCs)?
   - **A.** Cryosurgery
   - **B.** Curettage
   - **C.** Mohs micrographic surgery
   - **D.** Excision with frozen-section control
   - **E.** Radiation therapy
2. What is considered the best tissue source for reconstructing the nasal tip and alar regions?
   - **A.** Conchal bowl
   - **B.** Forehead
   - **C.** Preauricular region
   - **D.** Thigh
   - **E.** Supraclavicular region
3. What is considered to be the most aggressive subtype of BCC?
   - **A.** Superficial BCC
   - **B.** Nodular BCC
   - **C.** Pigmented BCC
   - **D.** Fibroepithelioma of Pinkus
   - **E.** Morpheaform BCC
4. Which anatomic location affected with a BCC is considered to have the highest risk for metastatic potential?
   - **A.** Forearm
   - **B.** Forehead
   - **C.** Nose
   - **D.** Cheek
   - **E.** Neck

Suggested Readings

Boyd CM, Baker SR, Fader DJ, Wang TS, Johnson TM. The forehead flap for nasal reconstruction. Arch Dermatol 2000;136(11):1365–1370

Burget GC, Walton RL. Optimal use of microvascular free flaps, cartilage grafts, and a paramedian forehead flap for aesthetic reconstruction of the nose and adjacent facial units. Plast Reconstr Surg 2007;120(5):1171–1207

Chiummariello S, Dessy LA, Buccheri EM, et al. An approach to managing non-melanoma skin cancer of the nose with mucosal invasion: our experience. Acta Otolaryngol 2008;128(8):915–919

Mureau MA, Moolenburgh SE, Levendag PC, Hofer SO. Aesthetic and functional outcome following nasal reconstruction. Plast Reconstr Surg 2007;120(5):1217–1227

Schulze HJ, Cribier B, Requena L, et al. Imiquimod 5% cream for the treatment of superficial basal cell carcinoma: results from a randomized vehicle-controlled phase III study in Europe. Br J Dermatol 2005; 152(5):939–947

40

Malignant Melanoma

Erica A. Mailler-Savage and Hugh M. Gloster

History

A 46-year-old white man presented with a rapidly enlarging black lesion on the right temple. He did not have a personal or family history of dysplastic nevi or melanoma. Physical examination revealed a 2-cm black irregular mobile nodule on the right temple **(Fig. 40.1**). No palpable preauricular, postauricular, anterior, or posterior cervical lymphadenopathy was noted. He denies weight loss.

Differential Diagnosis—Key Points

The following lesions should be considered in a patient with suspected melanoma (including lentigo maligna and amelanotic melanoma):

Benign lesions. Benign melanocytic nevus, dysplastic melanocytic nevus, seborrheic keratosis, lentigo simplex/solar lentigo

Malignant lesions. Pigmented basal cell carcinoma, squamous cell carcinoma, Merkel cell carcinoma, atypical fibroxanthoma, cutaneous metastasis

1. Diagnostic features. The ABCD rule is useful in differentiating acquired nevi from melanoma: A, asymmetry; B, irregular border; C, variegated color; D, diameter greater than 6 mm. Lesions that satisfy one or more ABCD criteria should be considered for biopsy. Note that congenital nevi tend to be larger, and size is not helpful unless there has been a rapid increase.
2. Full skin examination and regional lymph node palpation are essential in any patient with suspected melanoma.
3. Patients with dysplastic nevi have a 2 to 12% increased lifetime risk of melanoma. Prophylactic removal of dysplastic nevi does not eliminate risk of developing melanoma but serves as a marker of increased risk. Excision of dysplastic nevi with moderate to severe atypia is advised.

Test Interpretation

Given the location of this patient's lesion, an excisional biopsy with 2-mm margins was performed and revealed a nodular proliferation of markedly atypical melanocytes extending to the deep dermis (Breslow thickness 3.3 mm, Clark level IV, nonulcerated).

A properly performed biopsy is critical for accurate diagnosis of melanoma. In most cases this will entail an excisional biopsy; however, an incisional or punch biopsy can be performed on large lesions (>2 cm) or for those that would require elaborate reconstruction and planning. An incisional/punch biopsy

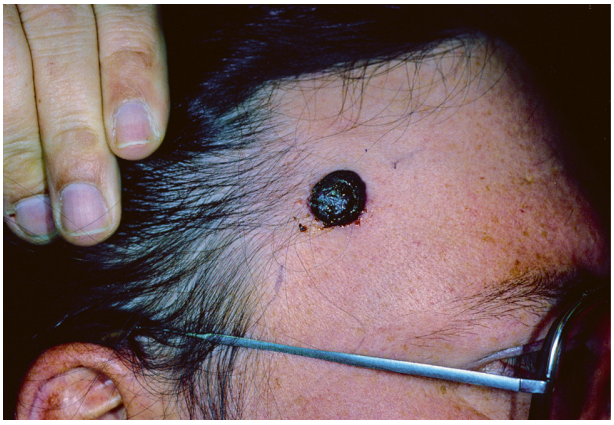

Fig. 40.1 Presenting lesion.

should be performed on the most raised and the darkest portion of the lesion and should include full-thickness skin and subcutaneous fat. Excisional biopsy should be down to the level of the underlying fascia. For lentigo maligna, a Wood's lamp may be helpful in delineating unclear borders of suspicious lesions.

As evidenced in the 2002 American Joint Committee on Cancer (AJCC) melanoma staging system (**Table 40.1**), Breslow thickness, the presence of ulceration, lymphatic invasion, and the presence of distal metastases are the most important prognostic indicators in primary cutaneous melanomas. Breslow thickness is measured from the granular layer of the epidermis to the deepest portion of the tumor. Clark level (level of dermal involvement) is often reported but is not of prognostic significance except in differentiating stage 1A and 1B melanomas.

◆ Diagnosis

Malignant melanoma, 3.3-mm Breslow thickness, Clark level IV, nonulcerated

◆ Medical Management

Although melanoma is a surgical disease, the role of radiation and other adjunctive therapy has been a popular source of debate. Recent studies have suggested that adjuvant radiation therapy results in good locoregional control rates (85–90%) in high-risk patients and comes with minimal complications. It has not, however, seemed to have an impact on overall survival. High-dose interferon-α has shown some survival benefit as adjunctive therapy for stage II and III disease, but it is of no benefit in metastatic disease. High-dose interleukin-2 (IL-2) has been beneficial in a small percentage of patients. Single-agent chemotherapy with dacarbazine is used for patients with metastatic disease because of favorable quality of life measures and modest cost impact. Numerous other immunotherapy and targeted therapies are currently being investigated.

◆ Surgical Management

The patient was referred for wide local excision and sentinel lymph node biopsy (SLNB). Lymphatic mapping and lymphoscintigraphy revealed the sentinel lymph node and an additional node within the deep portion of the parotid gland. Superficial parotidectomy was performed and the sentinel nodes were confirmed positive. The patient underwent wide local excision and modified radical neck dissection.

Definitive management of melanoma follows determination of local, regional, and distal extent of the disease. Lentigo maligna may be treated with surgery or imiquimod, but surgical management is the mainstay for other forms of melanoma in situ and stages I to III.

Table 40.1 American Joint Cancer Committee 2002 Revised Melanoma Staging

		Overall Survival		
Stage	**Histological Features/TNM Classification**	**1-yr**	**5-yr**	**10-yr**
0	Intraepithelial/in situ melanoma (TisN0M0)		100%	100%
IA	< 1 mm without ulceration and Clark Level II/III (T1aN0M0)		95%	88%
IB	< 1 mm with ulceration or Clark level IV/V (T1bN0M0)		91%	83%
	1.01–2 mm without ulceration (T2aN0M0)		89%	79%
IIA	1.01–2 mm with ulceration (T2bN0M0)		77%	64%
	2.01–4 mm without ulceration (T3aN0M0)		79%	64%
IIB	2.01–4 mm with ulceration (T3bN0M0)		63%	51%
	>4 mm without ulceration (T4aN0M0)		67%	54%
IIC	>4 mm with ulceration (T4bN0M0)		45%	32%
IIIA	Single regional nodal micrometastasis, nonulcerated primary (T1–4aN1aM0)		69%	63%
	2–3 microscopic regional nodes, nonulcerated primary (T1–4aN2aM0)		63%	57%
IIIB	Single regional nodal micrometastasis, ulcerated primary (T1–4bN1aM0)		53%	38%
	2–3 microscopic regional nodes, ulcerated primary (T1–4bN2aM0)		50%	36%
	Single regional nodal macrometastasis, nonulcerated primary (T1–4aN1bM0)		59%	48%
	2–3 macroscopic regional nodes, nonulcerated primary (T1–4aN2bM0)		46%	39%
	In-transit metastases/satellite lesion(s) without metastatic lymph nodes (T1–4a/bN2cM0)		30–50%	
IIIC	Single microscopic regional node, ulcerated primary (T1–4bN1bM0)		29%	24%
	2–3 macroscopic regional nodes, ulcerated primary (T1–4bN2bM0)		24%	15%
	4 or more metastatic nodes, matted nodes/gross extracapsular extension, or intransit metastases/satellite(s) and metastatic nodes (anyTN3M0)		27%	18%
IV	Distant skin, subcutaneous, or nodal metastases with normal LDH (any TanyNM1a)	59%	19%	16%
	Lung metastases with normal LDH (anyTanyNM1b)	57%	7%	3%
	All other visceral metastases with normal LDH or any distant metastases with increased LDH (anyTanyNM1c)	41%	9%	6%

LDH, lactate dehydrogenase.

Adequate surgical margins (**Table 40.2**) are critical for preventing recurrence but are often difficult to achieve in the head and neck because of cosmetic and functional considerations.

Sentinel lymph node status and the number of regional lymph nodes involved are important prognostic indicators for both recurrence and overall survival. SLNB has been a topic of

Table 40.2 Surgical Recommendations for Cutaneous Melanoma

Depth of Invasion	Presence of Ulceration	Recommended Margins (cm)	SLNB
Melanoma in situ	NA	0.5	No
<1 mm	No	1	No
<1 mm	Yes	1	Yes
<2 mm	Yes/no	1	Yes
2–4 mm	Yes/no	2	Yes
>4 mm	Yes/no	2	Offer to patient

SLNB, sentinel lymph node biopsy.

great debate in the past but has become the gold standard for melanoma head and neck staging for patients with Breslow thickness greater than 1 mm or less than 1 mm with ulceration (**Table 40.2**).

For known regional disease in the head and neck, a therapeutic radical or modified radical neck dissection is required to provide regional control, although there is a 21 to 24% risk of regional recurrence. The management of the N0 neck with elective lymph node dissection was previously debated but has since shown no survival benefit.

A few unique factors must be taken into consideration when surgically managing head and neck melanoma. First, there is unpredictable lymphatic drainage in the head and neck, especially in the scalp. It is not unusual for the sentinel node from scalp melanomas to be located in the parotid, postauricular, or suboccipital nodal basins. Second, the presence of intraparotid lymph nodes places the patient at high risk for facial nerve injury. Finally, SLNB can be technically challenging in the head and neck, as evidenced by lower sentinel lymph node recovery compared with other anatomic regions.

◆ Rehabilitation and Follow-up

The patient had a local recurrence over the right temple 3 months after wide local excision and modified radical neck dissection. He underwent re-excision with a split-thickness skin graft. His neck and temple were subsequently irradiated, and he was initiated on adjuvant vaccination therapy. Five months later, metastatic disease was found on the right side of his neck. The patient underwent radical resection and was initiated on temozolamide (Temodar) with IL-2 and interferon but subsequently died of metastatic disease.

Routine follow-up examinations for local, regional, and distant metastases are critical for melanoma patients. Patients should be followed up one to four times per year (depending on the thickness of the lesion and other risk factors) for the first 2 years and then once or twice per year thereafter. Routine blood work and imaging have limited usefulness in asymptomatic patients with tumors 4 mm or less thick for initial staging or follow-up and should be directed by thorough history and physical examination. Some studies have suggested that lactic dehydrogenase and chest radiograph may be helpful in detecting occult disease and alter management; however, there is no evidence to suggest improved survival with this type of monitoring.

The use of sunscreens and protective clothing should be advocated. Patients should also be instructed in self-examinations of the skin. Evidence suggests that most recurrences or metastases are discovered by the patient or the patient's family members. Finally, families of melanoma patients should be similarly counseled about self-examinations and sun protection and should be enrolled in a regular screening program.

Questions

1. A 49-year-old white man presents with a 3-cm black irregular plaque on the left temple. Melanoma is suspected. Choices for obtaining tissue for diagnosis include the following:
- **A.** Excisional biopsy
- **B.** Incisional biopsy
- **C.** Punch biopsy
- **D.** Shave excision
- **E.** B and C

2. What is the single most important prognostic indicator for overall melanoma survival?
- **A.** Breslow thickness
- **B.** Clark level
- **C.** Sentinel node status
- **D.** Presence of ulceration
- **E.** Presence of tumor-infiltrating lymphocytes on biopsy

3. A 59-year-old white man was diagnosed with malignant melanoma of the right cheek, 4-mm Breslow thickness, Clark level IV, sentinel lymph node positive. The patient underwent wide local excision with modified radical neck dissection. What is his risk of local recurrence?
- **A.** 10–15%
- **B.** 15–20%
- **C.** 20–25%
- **D.** 25–30%
- **E.** 35–40%

4. Adjuvant therapy for patients with advanced melanoma includes all of the following *except*:
- **A.** Imiquimod
- **B.** Radiation therapy
- **C.** Interleukin-2
- **D.** Interferon
- **E.** Temodar

Suggested Readings

Bene NI, Healy C, Coldiron BM. Mohs micrographic surgery is accurate 95.1% of the time for melanoma in situ: a prospective study of 167 cases. Dermatol Surg 2008;34(5):660–664

Hamm C, Verma S, Petrella T, Bak K, Charette M ; for the Melanoma Disease Site Group of Cancer Care Ontario's Program in Evidence-based Care. Biochemotherapy for the treatment of metastatic malignant melanoma: a systematic review. Cancer Treat Rev 2008;34(2): 145–156

Mendenhall WM, Amdur RJ, Grobmyer SR, et al. Adjuvant radiotherapy for cutaneous melanoma. Cancer 2008; 112(6):1189–1196

Rigual NR, Popat SR, Jayaprakash V, Jaggernauth W, Wong M. Cutaneous head and neck melanoma: the old and the new. Expert Rev Anticancer Ther 2008;8(3):403–412.

Sober AJ, Chuang TY, Duvic M, et al; for the Guidelines/ Outcomes Committee. Guidelines of care for primary cutaneous melanoma. J Am Acad Dermatol 2001;45(4): 579–586

X

Rhinology

41

Allergic Fungal Sinusitis

Aaron I. Brescia and Allen M. Seiden

History

A 47-year-old man's chief complaints are chronic nasal obstruction, repeated sinus infections, and a history of nasal polyps noted by his primary care physician. He complains of intermittent facial pain and pressure over both cheeks, thick postnasal drip, and sore throats, especially in the morning. He has received two 5-day courses of azithromycin for presumed infection over the past 12 months with little change in symptoms. In addition, he has been started on nasal steroid sprays and oral antihistamines for presumed allergic rhinitis. One week ago, he completed a 21-day course of oral fluoroquinolone therapy with perhaps a little improvement in his congestion. He has not previously been tested for allergies, has no history of asthma, and denies a history of aspirin sensitivity. He did undergo sinus surgery 5 years ago and seemed to feel better for 2 to 3 years, but his symptoms have gradually recurred. He does take medication for high blood pressure, but otherwise he is healthy and takes only a multivitamin.

On physical examination, the patient has normal visual acuity and extraocular movements remain intact, with no proptosis. However, he does have mild pseudohypertelorism. Examination of his nose reveals a midline septum, nasal polyps bilaterally, inferior turbinate hypertrophy, and very thick, pasty discharge from each middle meatus.

Differential Diagnosis—Key Points

1. Recurrent diffuse nasal polyposis represents a benign process and is closely associated with a variety of nasal pathologies, including allergic rhinitis, cystic fibrosis, allergic fungal rhinosinusitis, and bacterial rhinosinusitis, all of which need to be considered in the differential diagnosis. On histologic examination, benign respiratory polyp tissue exhibits inflammatory hallmarks of eosinophilia, lymphocytic infiltration, and hyperplastic stroma. Bilateral nasal polyps make the diagnosis of inverting papilloma less likely, though not impossible, and this diagnosis should be kept in mind as well.
2. Samter's triad of aspirin sensitivity (flushing, pruritic rashes, rhinorrhea, wheezing, or frank anaphylaxis), asthma, and nasal polyposis can develop later in life and so is not necessarily precluded by a relatively recent onset in a 47-year-old patient. However, this patient denies a history of asthma and has taken aspirin without untoward side effects.

3. Allergic fungal sinusitis (AFS) is a reaction, in part allergic, to aerosolized fungal spores, usually of the dematiaceous species. It is noninvasive and is present in immunocompetent hosts. This is in contrast to invasive forms of fungal sinus infections, which generally occur in patients who are immunocompromised. Acute invasive fungal sinusitis runs a rapidly fulminant course unless it is recognized early and treated aggressively. It is usually associated with saprophytic fungi of the order Mucorales, especially *Mucor*, and is characterized by evidence of necrotic tissue within the nasal cavity. Chronic invasive fungal sinusitis runs a slowly progressive course, is most often associated with *Aspergillus* spp., and is seen most often in diabetic patients. This patient has no history of immunodeficiency, making an invasive form of fungal sinusitis unlikely. However, thick mucus, evidence of expansion and medial erosion on computed tomography (CT) scan, and pseudohypertelorism are strongly suggestive for AFS.
4. AFS is generally characterized by Gell and Coombs type I immunoglobulin E (IgE)-mediated fungal hypersensitivity, eosinophilic or allergic mucin without evidence of mucosal invasion, positive fungal stain or culture, and nasal polyposis. Eosinophilic mucin is thick and has been described having a "peanut butter" consistency. It contains eosinophils, Charcot-Leyden crystals (eosinophilic debris or breakdown products), and fungal hyphae. There is no histologic evidence of invasion.
5. Another form of fungal sinusitis that needs to be considered is a mycetoma, aspergilloma, or fungus ball. However, this is an extramucosal infection that typically involves only a single sinus, most often the maxillary or sphenoid sinus. Once surgically removed, it tends not to recur.

Test Interpretation

Patients with AFS typically have nasal polyps and thick, inspissated mucus. The mucus may be tested for the presence of eosinophils, Charcot-Leyden crystals, and fungal hyphae, consistent with so-called allergic mucin. Culture for fungus may or may not be positive. Patients are usually atopic and have an elevated serum IgE and will demonstrate type 1 hypersensitivity to fungal antigens on allergy testing.

A noncontrast maxillofacial CT scan generally demonstrates characteristic findings. In this patient, pansinusitis is noted with evidence of prior surgery (**Fig. 41.1**). Extensive

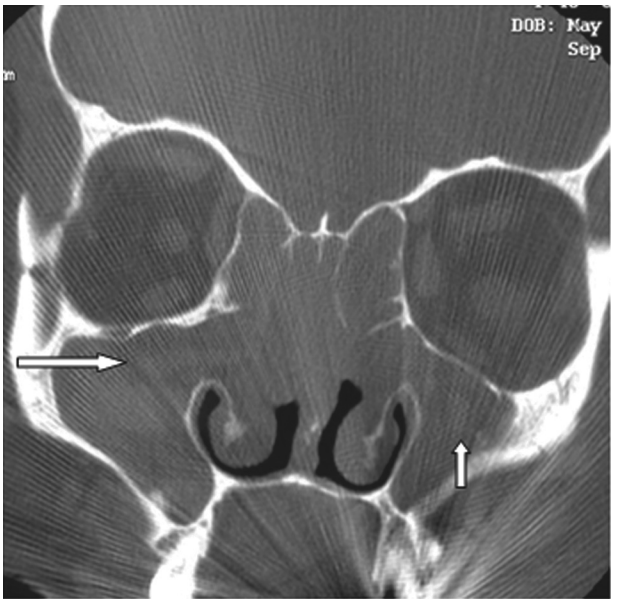

Fig. 41.1 Computed tomography scan demonstrating pansinusitis with an ethmoidectomy and maxillary antrostomy having been previously performed. Patchy areas of hyperdensity are noted within the maxillary sinuses (*arrows*).

opacification is typically present, often associated with expansion involving the lamina papyracea and skull base, resulting in thinning and possible dehiscence of surrounding bone. There is an inhomogeneous appearance with areas of hyperdensity, distinguishing AFS from other forms of chronic sinusitis.

◆ Diagnosis

Allergic fungal sinusitis

◆ Medical Management

Although the treatment of AFS is primarily surgical, ongoing medical management is important to try to prevent recurrence and alleviate symptoms. Systemic corticosteroids remain the most effective medical therapy for AFS, but the side effects make this approach less than ideal as a long-term treatment. Nevertheless, it continues to be the mainstay of medical management. Topical steroids are helpful and can be used long term, often at higher than recommended doses.

Antifungal medications, used both systemically and topically, have had mixed results and are not used routinely. Systemic antifungal therapy is hampered by the potentially harmful side effects and is reserved for cases of life-threatening invasive fungal sinusitis. Topical antifungal therapy remains controversial. Several small studies have been published with topical amphotericin or fluconazole sprays. At best, the patient's symptoms remain the same as with placebo, but some report worsening of their nasal symptoms.

Immunotherapy initially held promise, but longer-term studies have not found the benefit to be consistent.

◆ Surgical Management

The goals of surgical treatment are to debride the nasal cavity of polypoid tissue, irrigate and disimpact the involved sinuses of the mucinous secretions, and preserve the middle turbinate and other normal structures while optimizing natural sinus outflow tracts. In this particular case of revision sinus surgery with polyps, the surgeon may be well-advised to use computer navigation because inflammation is extensive and landmarks degraded. Again, the sinus secretions should be sent for microbiologic and pathologic analyses.

◆ Rehabilitation and Follow-up

Postoperative care should center on breaking the cycle of nasal and sinus inflammation while avoiding the retention of sinus secretions and superimposed bacterial infections. Skin allergy testing may guide the patient to avoidance measures or to immunotherapy. Nasal steroid use and leukotriene antagonists may help to delay the formation of new polyps.

◆ Questions

1. Allergic fungal sinusitis (AFS) is characterized by all of the following except:
- **A.** Nasal polyps
- **B.** Allergic mucin
- **C.** Type 1 hypersensitivity to fungi
- **D.** Asthma
- **E.** Positive fungal stain

2. Allergic mucin is characterized by all of the following except:
- **A.** Eosinophils
- **B.** Lymphocytes
- **C.** Charcot Leyden crystals
- **D.** Fungal hyphae

3. A maxillofacial computed tomography scan of a patient with AFS will characteristically demonstrate the following except:
- **A.** Bony remodeling
- **B.** Inhomogeneous opacification
- **C.** Areas of hyperdensity
- **D.** Areas of bone erosion

Suggested Readings

Jen A, Kacker A, Huang C, Anand V. Fluconazole nasal spray in the treatment of allergic fungal sinusitis: a pilot study. Ear Nose Throat J 2004;83(10):692–695, 694–695

Krouse JH. Allergic and non-allergic rhinitis. In: Bailey BJ, Johnson JT, Newlands SD, et al, eds. Bailey's Head and Neck Surgery—Otolaryngology, 4th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2004

Lanza DC, Dhong HJ, Tantilipikorn P, Tanabodee J, Nadel DM, Kennedy DW. Fungus and chronic rhinosinusitis: from bench to clinical understanding. Ann Otol Rhinol Laryngol Suppl 2006;196:27–34

Simon-Nobbe B, Denk U, Pöll V, Rid R, Breitenbach M. The spectrum of fungal allergy. Int Arch Allergy Immunol 2008;145(1):58–86

Sullivan DP, Bent JP III. Allergic fungal sinusitis: diagnosis and management. Oper Tech Otolaryngol–Head Neck Surg 2001; 12(1):2–6

42

Wegener Granulomatosis

Brian Matthew Harmych, Angela Donaldson, and Lee A. Zimmer

History

A 57-year-old woman is referred to the otolaryngology clinic for evaluation of chronic sinusitis. The primary physician treated the patient with multiple courses of antibiotics, nasal steroid sprays, saline irrigations, and topical decongestants over the past several years with no noticeable resolution of symptoms. On questioning in the otolaryngology clinic, the patient is complaining primarily of intermittent bilateral mucoid rhinorrhea, chronic intermittent epistaxis, aural fullness, decreased hearing, and nasal obstruction. This has been stable over the past several years. Review of systems revealed some malaise, generalized fatigue, diffuse arthralgia, and a 10-pound unintentional weight loss over the last year. She denies nasal trauma, nasal surgery, otolologic surgery or trauma, shortness of breath, stridor, hemoptysis, or skin changes. She does not abuse over-the-counter topical decongestants or cocaine. However, she is also concerned about the appearance of her nose which she feels has become less prominent recently. She attributes this to aging and is considering rhinoplasty.

Physical examination reveals a thin woman who appears her stated age. She is in no distress. She has an obvious deformity of the nasal dorsum with a significant depression overlying the cartilaginous septum. Anterior rhinoscopy reveals dry nasal mucosa with several excoriated areas of crusting along the anterior septum and inferior turbinates (**Fig. 42.1**). There is obstructing clot and crusts in the nasal cavities bilaterally. After removing the clot, a large septal perforation is revealed. Nasal endoscopy shows no evidence of mass or anatomic obstruction of the osteomeatal complex. The nasopharynx is clear. There is a unilateral effusion in the left middle ear space. Tuning fork examination is consistent with a conductive hearing loss in the left ear.

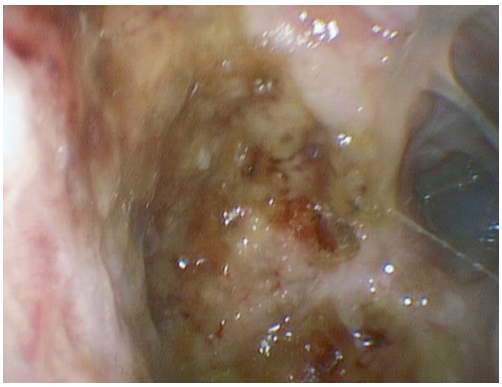

Fig. 42.1 Right nasal cavity of a patient with Wegener granulomatosis. Note the dried nasal mucus crusting to the lateral and posterior nasal wall.

Differential Diagnosis—Key Points

1. The differential diagnosis for this middle-aged woman with a large septal perforation includes nasal trauma, sinonasal surgery, overuse or abuse of topical decongestants or cocaine, neoplasm, systemic inflammatory and autoimmune diseases, and infectious processes.
2. The absence of a mass on thorough nasal endoscopic examination of the nasal cavities and nasopharynx makes neoplasm unlikely.
3. The history is inconsistent with nasal trauma or topical vasoconstrictive agent abuse because she denies use of these drugs. In addition, she also denies any history of nasal trauma or sinonasal surgery.
4. Infectious process is unlikely because this patient had been appropriately treated in the past with little to no response. Also, the examination is not consistent with infectious process and no purulence is noted.
5. The importance of obtaining a thorough review of systems should be highlighted. This patient is complaining of generalized malaise, fatigue, and arthralgias, raising concern for a systemic inflammatory disease process.

Test Interpretation

If the history and physical examination are suggestive of systemic inflammatory or autoimmune pathology, further diagnostic evaluation is indicated. Biopsy of diseased nasal mucosa is critical for the diagnosis. Initial laboratory studies include antineutrophil cytoplasmic antibody (cANCA) and perinuclear antineutrophil cytoplasmic antibody.

Chest radiography should be performed to evaluate for pulmonary involvement. Wegener granulomatosis (WG) can present with pulmonary lesions that have a cavitary appearance. An elevated erythrocyte sedimentation rate suggests the presence of a systemic inflammatory process.

Diagnosis

Sinonasal involvement of WG

Medical Management

Medical therapy with anti-inflammatory agents and immune modulators is the mainstay of treatment for patients with sinonasal WG. Corticosteroids and cyclophosphamide are commonly used. Disease progression should be monitored with serial cANCA titers.

The specific sinonasal symptoms of the disease can be treated locally. Nasal mucosal xerorhinia and crusting can be treated with topical ointment, saline sprays, and saline irrigations. Crusting and inflammation may cause functional obstruction, leading to acute bacterial rhinosinusitis. Antimicrobial therapy should be culture directed.

Surgical Management

There is little role for surgical management in sinonasal WG. In fact, the diagnosis of WG is often considered when patients develop complications following sinus surgery, such as extensive synechiae, scarring, excessive postoperative crusting, and need for frequent revision surgery.

The patient in the preceding scenario has a saddle-nose deformity secondary to loss of septal support of the nasal dorsum. In the absence of a systemic inflammatory condition, the surgical treatment for a saddle-nose deformity would be open rhinoplasty with bone graft to augment the nasal dorsum. However, surgical management should be delayed until the disease is adequately treated with medical therapy. Most authorities describe waiting until longstanding remission has been achieved.

Most authorities advise against repairing septal defects in patients with WG. Inflammation from the disease process itself and bacterial infection complicate postoperative wound healing in patients with WG. Functional endoscopic sinus surgery should be avoided in these patients for the same reasons. When aggressive nasal hydration and irrigation fail to remove crusting, the patient may undergo endoscopic debridement of crusts in the operating room.

Rehabilitation and Follow-up

Long-term remission can be achieved with medical therapy. Nasal irrigations and saline sprays should be used as lifelong treatments.

Acute exacerbations of bacterial rhinosinusitis should be treated adequately with microbial-specific, culture-directed antimicrobial therapy.

◆ Questions

1. Sinusitis associated with Wegener granulomatosis is often difficult to treat. What organism is typically cultured from these patients?
   - **A.** *Streptococcus pneumoniae*
   - **B.** *Staphylococcus aureus*
   - **C.** *Haemophilus influenzae*
   - **D.** *Streptococcus milleri*
2. Which type of vessels is affected by Wegener granulomatosis vasculitis?
   - **A.** Small vessel
   - **B.** Medium vessel
   - **C.** Large vessel
   - **D.** Small and medium-sized vessel
3. Which of the following statements about Wegener granulomatosis is false?
   - **A.** Nasal symptoms are typically seen early in the disease process.
   - **B.** If Wegener granulomatosis is left untreated, it is fatal within 2 years for over 90% of patients.
   - **C.** Wegener triad includes glomerulonephritis, vasculitis, and arthritis.
   - **D.** The most common oral cavity finding in Wegener granulomatosis is hyperplasia of the gingiva and gingivitis.
4. About what percentage of patients with Wegener granulomatosis will die of this disease or its therapy?
   - **A.** Less than 5%
   - **B.** 10%
   - **C.** 20%
   - **D.** 40%

Suggested Readings

Gubbels SP, Barkhuizen A, Hwang PH. Head and neck manifestations of Wegener's granulomatosis. Otolaryngol Clin North Am 2003;36(4):685–705

Oei A, Lemmerling M, Mortelé K, Vandekerckhove T, Praet M, Kunnen M. Skull base granuloma: an unusual location of idiopathic midline destructive disease. Eur Radiol 2001;11(6):1076–1078

Rasmussen N. Management of the ear, nose, and throat manifestations of Wegener granulomatosis: an otorhinolaryngologist's perspective. Curr Opin Rheumatol 2001;13(1):3–11

43

Orbital Complications of Sinusitis

Aaron I. Brescia and Allen M. Seiden

History

A 9-year-old boy goes to the emergency room after awakening that morning with extensive left eye swelling. His recent medical history is significant for 8 days of an upper respiratory illness, with symptoms of nasal congestion, rhinorrhea, headache, and fever. He is otherwise in good health and his immunizations are up to date.

Physical examination reveals a well-nourished though ill-appearing child with a normal ear examination, nasal congestion with purulent rhinorrhea bilaterally, 2+ tonsils that are not inflamed, and shotty lymphadenopathy. He is febrile to 101.9°F. He exhibits marked left periorbital edema that appears to be preseptal, with no apparent proptosis. Visual acuity is intact. His extraocular movements are intact. He has full flexion and extension of his neck. Based on his history and physical examination findings, a computed tomography (CT) scan of his sinuses and orbits is obtained.

Differential Diagnosis—Key Points

1. Swelling of the upper and lower eyelids should lead the investigator to consider both intrinsic periorbital abnormalities as well as orbital extensions of periorbital processes. Furthermore, it is important to distinguish periorbital cellulitis from orbital cellulitis.
2. Periorbital cellulitis can result from a traumatic insult to the area, as from an insect bite, bacteremia, or spread from a regional infection such as bacterial conjunctivitis, dacrocystitis, or sinusitis. Typically the upper and lower eyelids are erythematous and swollen shut. However, examination of the eye reveals that visual acuity and extraocular movements are intact.
3. Decreased visual acuity and restricted movement of the globe, or ophthalmoplegia, are more indicative of orbital cellulitis, as are chemosis and proptosis. These signs indicate that the infection and inflammation have spread to postseptal muscle and fat tissue. These findings are of concern because of the possibility of spread of the infection to the optic nerve and intracranially. Permanent eye damage can result. This patient had no evidence of ophthalmoplegia, making a diagnosis of orbital cellulitis unlikely.
4. The most common cause of orbital and periorbital cellulitis in children is sinus disease, most notably ethmoid sinus infection. Both are more common in children than in adults and may respond to intravenous antibiotic therapy without the need for surgical

intervention. Orbital cellulitis is more often associated with systemic signs, such as fever and malaise, and will usually require a more prolonged course of therapy.

5. Causative organisms in orbital cellulitis include mainly *Staphylococcus* and *Streptococcus* spp. Not surprisingly, *Haemophilus influenzae* spp. are also associated with these complications of sinus infection.

Test Interpretation

1. CT: A sinus CT scan with contrast is the study of choice to evaluate the paranasal sinuses and delineate the presence or absence of a subperiosteal phlegmon or abscess within the orbit (**Fig. 43.1**). Often it is difficult to determine definitively whether a fluid collection is phlegmonous inflammation or a true abscess, that is, whether surgery is warranted immediately or a trial of medical therapy may be used. Particular attention should be paid to the cavernous sinus for asymmetric enhancement or the absence of signal, which should alert the investigator to the possibility of spread of infection into this area with possible venous thrombosis (**Table 43.1**).
2. Magnetic resonance imaging (MRI): If spread of the infection into the optic nerve or anterior or middle cranial fossa is suspected as the result of ominous physical findings in the obtunded patient, or those with bilateral findings, MRI with contrast may help to determine the true extent of the infection.
3. Complete blood cell count (CBC) with differential: A baseline CBC should be obtained to help rule out immunodeficiencies such as neutropenia and also to provide a marker to follow during admission to monitor response to medical therapy, especially in patients in whom medical therapy alone is used as the result of ambiguous imaging findings.
4. Blood Gram stain and culture: One must determine the presence or absence of bacteremia, especially in pediatric patients.

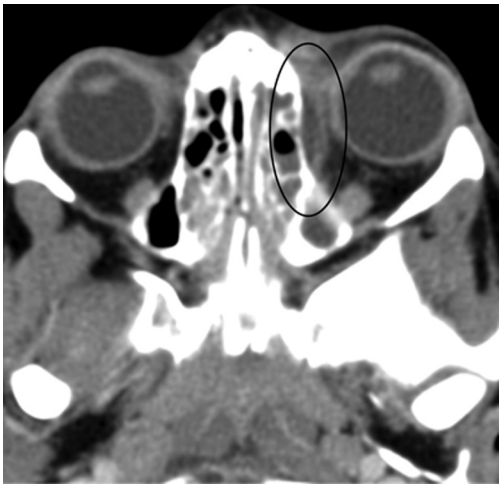

Fig. 43.1 Axial computed tomography scan of a patient with a subperiosteal abscess (circle).

Table 43.1 Modified Chandler staging system for orbital inflammation

Stage	Description
I	Preseptal cellulitis
II	Inflammatory orbital edema
III	Subperiosteal abscess
IV	Orbital abscess
V	Cavernous sinus thrombosis

Diagnosis

Periorbital cellulitis

Medical Management

As stated, CT findings may fail to indicate definitively a drainable fluid collection in the subperiosteal space of the orbit. In this instance, and if the patient's visual acuity remains intact per ophthalmologic evaluation, hospitalization with medical management is reasonable. Antibiotic therapy should be directed against the most common offending sinus organisms (i.e., ampicillin plus sulbactam or vancomycin plus a second- or third-generation cephalosporin in communities with a high incidence of, or in the patient with a history of, methicillin-resistant *Staphylococcus aureus* (MRSA). In addition, nasal hygiene with both

systemic and local decongestant therapy, as well as nasal saline irrigations, can help to promote sinus drainage. The use of steroids to both decrease periorbital edema and shrink the nasal lining to promote sinus drainage is controversial. Steroid therapy in the face of bacterial infection not only has inherent biochemical ramifications, but it also renders the CBC of little benefit as a clinical marker with which to judge response to medical therapy. Typically topical nasal decongestant therapy is given twice daily for 3 days only to avoid the chance of rhinitis medicamentosa. Daily evaluation of the orbit and visual acuity by the ophthalmology service during admission should be standard. The clinical examination should improve within 36 to 48 hours. If it does not, a repeat CT scan should be obtained to evaluate for progression of the phlegmon into an abscess.

◆ Surgical Management

Imaging that clearly indicates a rim-enhancing drainable fluid collection, a decrement in visual acuity, or failure to respond to medical therapy is an indication for surgery. The decision to proceed to the operating room is soon followed by the question of which approach to use, endoscopic versus open. The external approach through a Lynch incision provides wide access and potentially the best chance of completely draining the abscess. The endoscopic approach avoids an external facial scar for the child and allows access to the ethmoid sinuses and the lamina papyracea. It is widely used, especially in initial drainage procedures. Inadequate drainage, however, is a concern, as is the prospect of operating in an acutely inflamed and likely bloody endoscopic field.

◆ Rehabilitation and Follow-up

The postoperative course should include continued antibiotic therapy, monitoring of wound cultures to help tailor medical therapy, and regular ophthalmic examinations. Again, the examination should dramatically improve within 36 to 48 hours with adequate surgical drainage and appropriate medical adjunctive therapy. If it does not, a repeat CT scan should be obtained. If the initial attempt at surgical drainage was endoscopic, and the repeat scans indicate reformation of the abscess, one is probably well advised to proceed with open ethmoidectomy, especially if the visual examination has started to decline.

◆ Questions

1. Which of the following is a physical finding that would suggest orbital cellulitis versus periorbital cellulitis?
   - **A.** Preseptal edema
   - **B.** Tenderness
   - **C.** Ophthalmoplegia
   - **D.** Young age
2. Periorbital cellulitis may occur secondary to
   - **A.** Ethmoid sinusitis
   - **B.** Dacrocystitis
   - **C.** Trauma
   - **D.** Insect bite
   - **E.** All of the above
3. A patient has ethmoid sinusitis, periorbital cellulitis, and a subperiosteal abscess. Intravenous antibiotics are initiated, surgical drainage is performed, but after 2 days the patient still has substantial periorbital swelling. What is the next step?
   - **A.** Add cold compresses to the eye
   - **B.** Change antibiotic coverage
   - **C.** Order a computed tomography scan
   - **D.** Bring the patient immediately back to surgery for re-exploration and drainage.

Suggested Readings

Beech T, Robinson A, McDermott AL, Sinha A. Paediatric periorbital cellulitis and its management. Rhinology 2007;45(1):47–49

Giannoni CM, Weinberger DG. Complications of Rhinosinusitis. In Bailey BJ, Johnson JT, Newlands SD, et al, eds. Bailey's Head and Neck Surgery—Otolaryngology, 4th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2004.

Tanna N, Preciado DA, Clary MS, Choi SS. Surgical treatment of subperiosteal orbital abscess. Arch Otolaryngol Head Neck Surg 2008;134(7):764–767

44

Epistaxis

Gordon H. Sun and Allen M. Seiden

History

A 39-year-old man arrived in the emergency department complaining of intermittent epistaxis over the past 2 days. These episodes previously resolved on their own with a moderate amount of pressure applied to the anterior nose for about 5 to 10 minutes, with occasional use of oxymetazoline applied by nasal spray. However, this particular episode was much more severe; anterior nasal pressure reduced but did not completely staunch the flow of blood, and the patient could feel blood trickling down the back of his throat. He reported having had this episode of epistaxis for more than 2 hours before arrival in the emergency department.

His medical history was significant for recently diagnosed mild hypertension, not yet medically treated by his primary care provider. He also carried a diagnosis of idiopathic thrombocytopenic purpura (ITP), although the patient could not recall how he was discovered to have that condition initially. He denied a personal history of stroke, peripheral vascular disease, or previous episodes of easy bruising or bleeding, other than the recent epistaxis. There was no history of trauma to the nose, nor did he have a previous surgical history. He was a nonsmoker. The patient denied a family history of epistaxis or bleeding diatheses. He was not taking any medications.

On physical examination, the patient was awake, alert, and able to speak clearly, although he reported feeling increasingly light-headed during the examination. He was tachycardic, but his blood pressure was within normal limits. Oxygen saturation levels were normal. The patient was not stridulous. Rhinoscopy revealed active bilateral epistaxis, with a moderate amount of fresh blood and clots in the anterior nasal cavity, precluding visualization of the posterior nares despite repeated suctioning. The source of the epistaxis could not be identified anteriorly. Oral cavity and oropharyngeal examination demonstrated significant amounts of blood coursing down the posterior pharyngeal wall. No friable or hypervascular lesions were seen within the oral mucosa, lips, or face.

Two large-bore intravenous (IV) lines were quickly placed by the emergency department staff and rapid IV fluid resuscitation was initiated. Continuous cardiopulmonary monitoring and pulse oximetry were started. An attempt was made to pack both nares anteriorly and posteriorly with inflatable nasal packs, but the patient did not tolerate the procedure well and the epistaxis increased in intensity bilaterally. At this point, rapid sequence induction was initiated and the patient was expeditiously intubated for airway protection. Following orotracheal intubation, the nose and mouth were thoroughly suctioned, and bilateral Foley

catheters were placed and inflated to tamponade the nasopharynx while petrolatum-infused gauze was extensively packed into the anterior nasal vaults. Now in stable condition, the patient was transported to the intensive care unit for monitoring. Two units of packed red blood cells were transfused during his recovery.

◆ Differential Diagnosis—Key Points

1. The blood supply to the nose originates from both the external and internal carotid systems. The external carotid artery supplies the facial and internal maxillary arteries, the latter of which terminates in the sphenopalatine artery. The sphenopalatine artery enters the nasal cavity through a foramen on the posterior lateral nasal wall. This artery is well known as a major potential source of posterior epistaxis. The internal carotid artery feeds into the ophthalmic artery, which itself branches into anterior and posterior ethmoidal arteries that supply the upper lateral nasal wall and septum. The source of most nose bleeds is the Kiesselbach plexus or the Little area, found on the anterior nasal septum, and supplied by the sphenopalatine, greater palatine, superior labial and anterior ethmoid arteries.
2. Epistaxis has a lifetime incidence of up to 60% in the general population, with less than 10% of all cases requiring medical attention. The condition has a bimodal distribution, peaking in children younger than 10 years of age and adults over 50 years old. Roughly 80% of all cases of epistaxis are anterior. Minor bleeds, which are usually found in the anterior nares, are more frequently encountered in the pediatric population. Severe episodes of epistaxis requiring major intervention, more often originating posteriorly, are seen more often in adults. The distinction between anterior and posterior epistaxis has blurred recently with the introduction of various endoscopic methods of controlling severe epistaxis. Traditionally, anterior epistaxis can be seen on anterior rhinoscopy or controlled with anterior packing. Posterior epistaxis might not be seen easily even with rigid nasal endoscopy because of its tendency to be more severe; thus posterior nasal hemorrhage might be identified only by the necessity for posteriorly based packing to gain control.
3. The differential diagnosis of epistaxis is quite extensive. History-taking should be directed, especially if there is active epistaxis. The most common cause of epistaxis in any age group is digital trauma (nose picking). Mucosal dryness, maxillofacial trauma (including nasal bone fracture or septal deviation), active inflammation or infection (including nasal polyps), and previously undiagnosed tumors (including inverting papilloma or juvenile nasopharyngeal angiofibroma) are all potential local causes of epistaxis (**Fig. 44.1**). In adults, chronic alcoholism, use of aspirin and nonsteroidal anti-inflammatory drugs; use of warfarin, enoxaparin, and other anticoagulants; and even ingestion of "alternative" medications such as ginkgo biloba and ginseng have been implicated in epistaxis. Excessive use of intranasal corticosteroids, especially when directed against the nasal septum, may contribute to nosebleeds as well. Systemic conditions that predispose individuals to epistaxis include von Willebrand disease, hemophilia, and leukemia, all of which contribute to either poor platelet function or thrombocytopenia (low platelet counts). In this patient, a history of possible

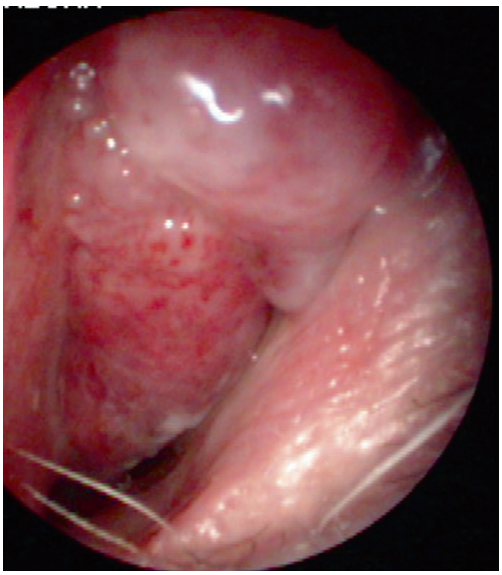

Fig. 44.1 Intranasal melanoma in the left nasal cavity of a patient presenting with epistaxis.

ITP or low platelets could have contributed to epistaxis. Osler-Weber-Rendu disease (hereditary hemorrhagic telangiectasia, or HHT) is an autosomal dominant disease that presents with arteriovenous malformations (AVMs) and telangiectasias in the nasal mucosa, lungs, and brain. Hypertension has not been firmly established as an independent risk factor for epistaxis, but it may make control of epistaxis more challenging.

◆ Test Interpretation

A CBC should be obtained in any patient presenting with recurrent or active epistaxis. Low hemoglobin and hematocrit may imply that a patient has either suffered massive acute blood loss or has had persistent blood loss from epistaxis. Although in many cases platelet counts will be normal, this patient with ITP was thrombocytopenic. A platelet count of less than 20,000/mm^3 significantly increases the difficulty of controlling an episode of epistaxis. Coagulation studies (prothrombin time/partial thromboplastin time) may be helpful if an underlying bleeding dyscrasia is present or suspected, which might warrant further workup. However, one study in 2008 concluded that a coagulation profile should be obtained only in patients receiving anticoagulant therapy or those who have a known history of chronic liver disease or coagulopathy because obtaining coagulation studies in otherwise healthy patients did not seem to change the plan of care.

HHT is a disease affecting multiple organ systems and would warrant workup, including CT of the chest and MRI of the head to identify pulmonary and intracranial AVMs. However, for most epistaxis patients, this additional workup is not necessary in the acute setting.

◆ Diagnosis

Posterior epistaxis

◆ Medical Management

For minor epistaxis, medical management is conservative. This may range from simple observation to topical application of 0.05% oxymetazoline or 1% phenylephrine solution for vasoconstriction, combined with direct pressure applied to both nares for 15 to 20 minutes. To help prevent recurrence, it is important to reduce mucosal dryness, which may be achieved with saline nasal sprays, application of petrolatum or mupirocin ointment, or use of humidifiers at home. Patients who require continuous use of nasal cannula or face mask–delivered oxygen for other medical issues may benefit from simple humidification added to their oxygen.

Patients demonstrating severe nasal hemorrhage may be at risk for both airway compromise from aspiration of blood and hemodynamic instability, leading to substantial morbidity and possibly death. In these instances, following the "ABCDE" protocol of trauma is critical. An artificial airway should be placed first, under controlled circumstances if at all possible. Typically this can be achieved by rapid sequence induction, followed by orotracheal intubation. Nasotracheal intubation should be avoided because the trauma from endotracheal tube placement may exacerbate the nasal hemorrhage. Cricothyrotomy and tracheotomy should only be performed if intubation cannot be performed owing to an inability to visualize the airway as a result of the hemorrhage. Two large-bore IVs should be placed and aggressive fluid resuscitation should be performed, with blood transfusions as needed. If the patient is being seen in an emergency department setting, recruiting nearby medical assistance to manage the patient's resuscitation may be immensely helpful while the otolaryngologist attempts to control the source of epistaxis.

◆ Surgical Management

Surgical management of epistaxis varies widely, depending on the severity of the hemorrhage and familiarity of the treating physician with the numerous tools available to control nosebleeds. After local anesthetic and vasoconstrictive medications have been applied to the nasal cavity, small anterior mucosal bleeds may respond well to topical application of silver nitrate cautery, which causes local chemical damage to bleeding tissues. There is a risk of septal perforation if both sides of the nasal septum are aggressively cauterized in this manner. Other methods of controlling anterior

epistaxis include nasal tampons such as Merocel (Medtronic, Minneapolis, MN) or Rhino Rocket (Shippent Medical Technology, Centennial, GA) sponges, Rapid Rhino (ArthroCare, Austin TX) inflatable packs, or in more severe cases, formal packing using layered ribbon gauze impregnated with Vaseline or bismuth-iodoform pastes. A nasal speculum and strong light source (headlight) should always be used when performing cautery or packing. If further bleeding is seen, consider packing the opposite nasal passage before removing the already placed pack.

Posterior epistaxis can be difficult to treat owing to its location and often its severity. Several specially designed balloon systems and packs are available for posterior nosebleeds, including the Epistat and anterior-posterior version of the Rapid Rhino. An alternative, and possibly more available method involves placement of a 12 or 14 French Foley catheter into the nasal cavity, inflation of the balloon with 10 mL of saline, and pulling of the catheter anteriorly until the balloon lodges in the posterior choanae. The anterior nasal passage is then packed tightly with layered petrolatum gauze. Up to 70% of posterior packing procedures are successful in stopping epistaxis; repositioning the packs may help if a small amount of bleeding is still observed. Note that many patients do not tolerate posterior nasal packing, and thus elective endotracheal intubation and sedation may need to be performed beforehand.

In patients refractory to formal packing, operative intervention is required. Common methods of surgical intervention include electrocautery of the affected area under nasal endoscopic guidance, septoplasty, and ligation of the anterior ethmoidal or sphenopalatine arteries. The former is generally done via an external Lynch incision, and the latter is now generally done through an endoscopic approach (**Fig. 44.2**).

Angiographic embolization of selected arteries was first described in 1972 and is performed in some institutions for treatment of severe, intractable epistaxis, particularly in patients considered poor candidates for general

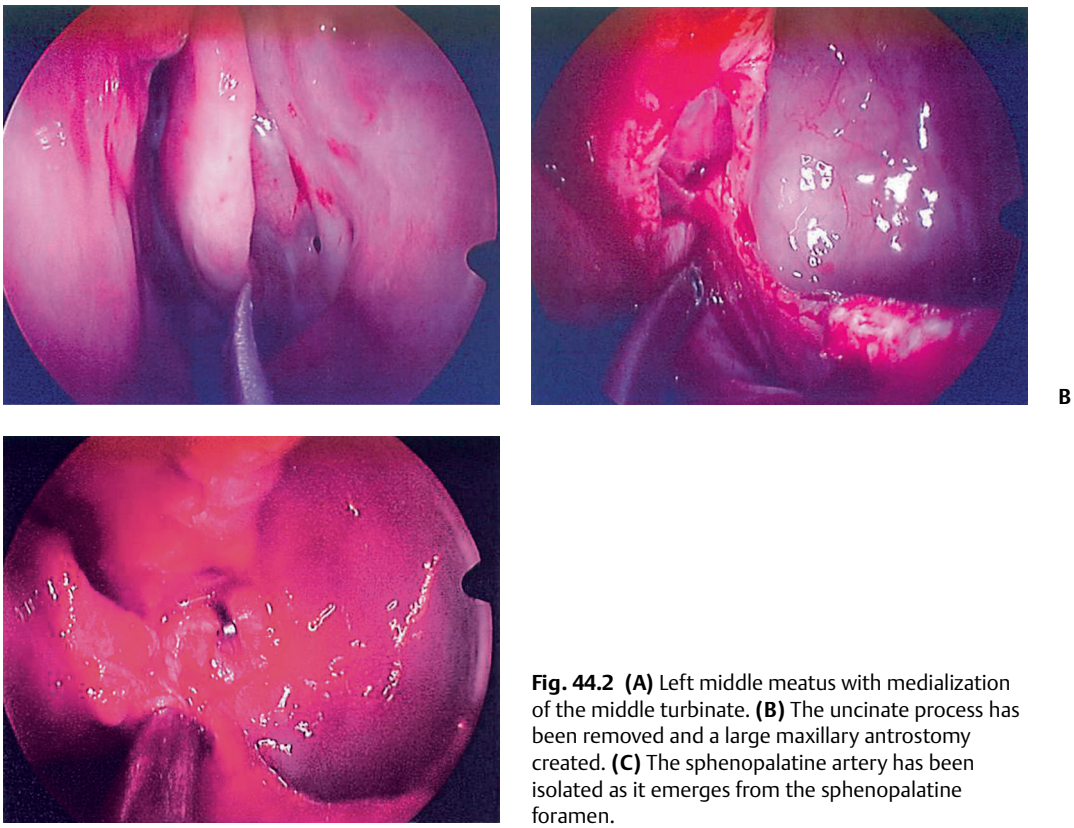

Fig. 44.2 **(A)** Left middle meatus with medialization of the middle turbinate. **(B)** The uncinate process has been removed and a large maxillary antrostomy created. **(C)** The sphenopalatine artery has been isolated as it emerges from the sphenopalatine foramen.

anesthesia. The success rate is as high as 87%, although the technology needed to perform this procedure is not widely available.

Every patient undergoing posterior packing or other intervention for a posterior nosebleed should be admitted and their oxygen saturation monitored due to the potential for hypoxia and cardiac arrhythmias. Other complications that can arise from prolonged packing include ulceration and necrosis of the nasal skin, septum, nasopharyngeal soft tissue, septal perforation, sinusitis, and synechiae. Antibiotic therapy should be initiated in patients with nasal packing in place to prevent toxic shock syndrome or the development of secondary bacterial sinusitis. Nasal packs may be removed after 48 to 72 hours if no further bleeding is seen.

Rehabilitation and Follow-up

Preventive measures are strongly recommended for all patients. These include refraining from nose-blowing or picking; abstinence from alcohol or hot drinks that can vasodilate nasal vessels; using saline nasal sprays; petrolatum ointments, or humidification to moisturize the nasal passages; and appropriate management of co-morbidities that can contribute to epistaxis, including hypertension. Medication use (e.g., aspirin or warfarin) should be closely monitored. All patients with severe or recurrent epistaxis should have a formal evaluation of the nasal cavity performed, either while hospitalized or in an outpatient setting to rule out neoplastic lesions or anatomic anomalies.

Patients with HHT should always be closely monitored because they are susceptible to recurrent epistaxis (even with repeated interventions) and may need numerous blood transfusions over their lifetime. Family members of patients affected by HHT should be screened for the disorder. Patients who have a personal or family history of a bleeding disorder or are suspected to have a bleeding dyscrasia based on hematologic or coagulation testing may benefit from a hematology consultation.

Questions

1. Most cases of epistaxis arise from where?
   - **A.** The sphenopalatine artery
   - **B.** The anterior ethmoid artery
   - **C.** The Kiesselbach plexus
   - **D.** The Woodruff plexus
2. If a patient presents with elevated blood pressure and severe epistaxis, the blood pressure must be controlled *before* the epistaxis can be controlled.
   - **A.** True
   - **B.** False
3. The Keisselbach plexus is supplied by each of the following arteries except which one?
   - **A.** Sphenopalatine
   - **B.** Greater palatine
   - **C.** Superior labial
   - **D.** Inferior labial
   - **E.** Anterior ethmoid
4. Potential complications from nasal packing include which of the following?
   - **A.** Hypoxia
   - **B.** Toxic shock syndrome
   - **C.** Nasal adhesions
   - **D.** Septal perforation
   - **E.** All of the above

Suggested Readings

Awan MS, Iqbal M, Imam SZ. Epistaxis: when are coagulation studies justified? Emerg Med J 2008;25(3):156–157

Damrose JF, Maddalozzo J. Pediatric epistaxis. Laryngoscope 2006;116(3):387–393

Gallitelli M, Pasculli G, Fiore T, Carella A, Sabbà C. Emergencies in hereditary haemorrhagic telangiectasia. QJM 2006;99(1):15–22

Gifford TO, Orlandi RR. Epistaxis. Otolaryngol Clin North Am 2008;41(3):525–536, viii

Kucik CJ, Clenney T. Management of epistaxis. Am Fam Physician 2005;71(2):305–311

Pope LE, Hobbs CG. Epistaxis: an update on current management. Postgrad Med J 2005;81(955):309–314

45

Hereditary Hemorrhagic Telangiectasia

Brandon Scott Hopkins and Allen Seiden

History

A 19-year-old man with a history of nosebleeds that are increasing in frequency and becoming harder to control recently required an anterior nasal pack to control bleeding. This patient reports no nosebleeds before age 15, no history of nasal trauma, no history of sinusitis or recurrent upper respiratory tract infections, and no history of prolonged bleeding associated with cuts or bruising. The nosebleeds seem to begin spontaneously. Ordinarily he has no complaints of nasal obstruction except that the bleeding has caused some persistent nasal crusting.

The patient notes that his father had recurrent nosebleeds over the years, requiring frequent nasal packing and even blood transfusions on occasion, but had died unexpectedly from a stroke several years earlier. His mother reports no history of nosebleeds. The patient has an older brother and younger sister, both without a history of nosebleeds.

On physical examination the patient is a well-appearing man with no signs of bruising. On head and neck examination, a small hyperemic blemish is noted on his lower lip (**Fig. 45.1**) and conjunctivae. Nasal endoscopy is performed and demonstrates these same blemishes along his nasal septum with residual crusting from his previous bleed and a small telangiectasia on the right inferior turbinate (**Figs. 45.2 and 45.3**).

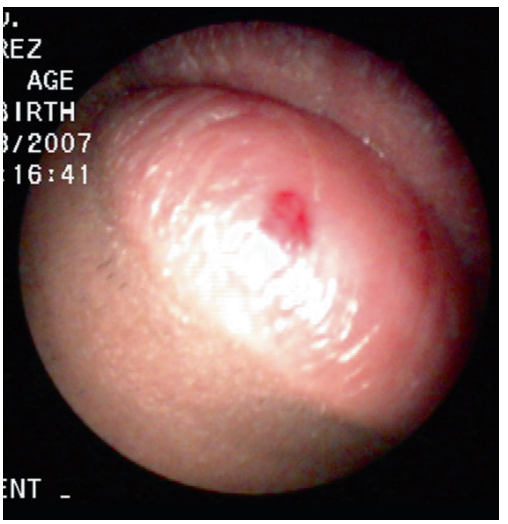

Fig. 45.1 Telangiectasia noted on lower lip suggestive for hereditary hemorrhagic telangiectasia.

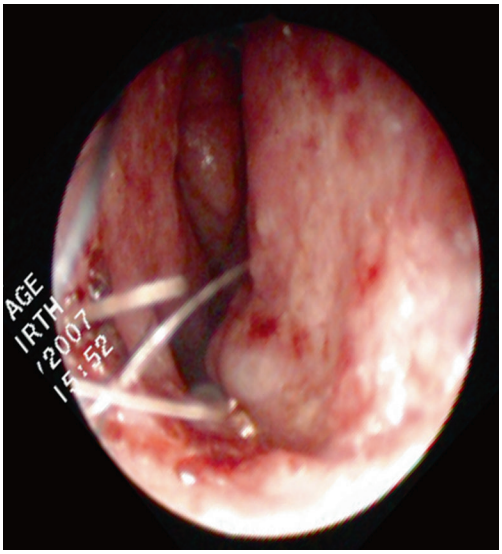

Fig. 45.2 Multiple telangiectasias noted on the right anterior nasal septum.

Differential Diagnosis—Key Points

1. The differential diagnosis for nasal bleeding is large.
   - **A.** In young children nasal trauma predominates and usually occurs on the side of their dominant hand. Other sources of mucosal irritation can also predispose to bleeding, including inflammation from infection, drying of the nasal mucosa, foreign bodies, and allergic rhinitis. The Kiesselbach plexus on the anterior septum is the region most likely to bleed with nasal trauma.
   - **B.** Granulomatous diseases, including Wegener and syphilis, can cause bleeding.
   - **C.** Hypertension often associated with renal disease and aortic coarctation can be a cause.
   - **D.** A large array of coagulapathies, including von Willebrand disease and the hemophilias, are relatively common and cause variable bleeding presentations that may include the nose.
   - **E.** Although rare, vitamin C deficiency with scurvy causes nasal bleeding as well as bleeding from other sites.
   - **F.** Any nasal mass can cause epistaxis, whether it is inflammatory or neoplastic. Be particularly wary with juvenile nasopharyngeal angiofibroma in the setting of unilateral nasal obstruction and epistaxis in an adolescent male.
   - **G.** An array of vascular malformations, including hereditary hemorrhagic telangiectasia (HHT), can be the cause of recurrent bleeding.
2. HHT, also known as *Osler-Weber-Rendu syndrome*, has dilated subepithelial vessels as the hallmark of the disease. Multiple arteriovenous malformations (AVMs) are present that lack intervening capillaries, resulting in direct connections between arteries and veins. These AVMs lack the usual contractile elements and therefore tend to bleed easily and profusely.
3. Small AVMs are referred to as *telangiectasias*, and are the visible signs of this disease. They are hyperemic, discrete, spiderlike, bright red maculopapules that usually measure 1 to 4 mm in diameter. They tend to occur on the skin and mucosal surfaces. They are frequently seen on the face, fingertips, conjunctivae, lips, oral cavity, pharynx, and gastrointestinal tract.
4. Large telangiectasias or AVMs tend to occur in the brain, lung, and liver. Rather than hemorrhage, these tend to be associated more often with significant shunting of blood, thrombosis, and embolus.

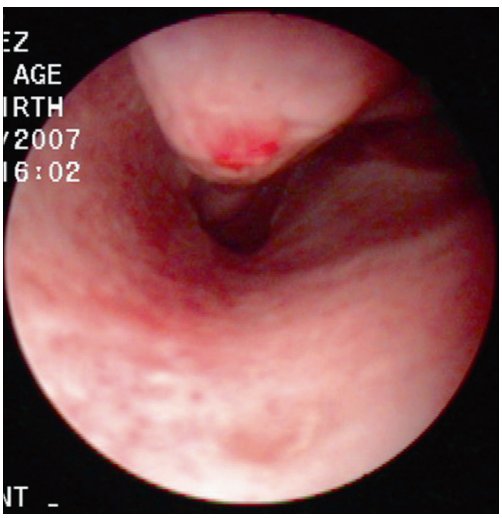

Fig. 45.3 Small telangiectasia on the inferior aspect of right inferior turbinate.

A. Intracranial (15%): Can cause hemorrhage, stroke, and abscess formation
B. Pulmonary (20–35%): Probably are present at birth, but symptoms manifest in patients in their 20s and 30s. These malformations cause shunting with resulting symptoms that include dyspnea and exercise intolerance but may lead to stroke, brain abscess, systemic infection, and high-output cardiac failure.
C. Gastrointestinal (11–40%): Painless bleeding, usually at age 50 to 60
D. Hepatic (8–16%): Causing liver disease and pulmonary hypertension. These patients may need a liver transplant.

5. Diagnosis of HHT is made clinically by the Curañao criteria, established in June 1999 by the Scientific Advisory Board of the HHT Foundation International, Inc. The disease is considered present if three of four criteria are met:
A. Recurrent spontaneous epistaxis
B. Mucocutaneous telangiectasias
C. Visceral involvement by AVMs
D. A family history of HHT

6 The incidence of HHT is 1 in 5000 to 1 in 10,000, affecting men and women equally. Symptoms generally manifest after puberty, with epistaxis usually the first manifestation. It affects 78 to 96% of HHT patients and usually manifests before age 20, although it may not be seen until later in life. It progresses in frequency and difficulty to control, such that initially the symptoms may be quite subtle.

7. HHT is caused by dysregulation of serine threonine kinase signaling in endothelial cells. It consists of two major disease types: HHT-1, caused by mutation in an endoglin gene on chromosome 9, and HHT-2, caused by a mutation in *ACVRL1* on chromosome 13. Other mutations have also been found. These different mutations make up for the variable presentation and symptom expression of these patients. Vascular stress may also affect disease presentation.

◆ Test Interpretation

Initial evaluation should include a complete blood count. The patient should then be screened for visceral AVM involvement. This screening is also appropriate for family members at high risk. Pulmonary AVM screening can be done with contrast echocardiography. If this demonstrates shunting, or if this test is not available, then thin-cut CT scanning is appropriate. Any pulmonary AVM with a feeding vessel larger than 3 mm deserves intervention. A head MRI with and without gadolinium is ordered to look for cerebral AVMs. Hepatic involvement can be determined with either ultrasound or CT but is not routine, as hepatic AVMs generally do not result in sudden and catastrophic symptoms. Further testing can be pursued as needed with genetic testing available in select centers.

◆ Diagnosis

Hereditary hemorrhagic telangiectasia

◆ Medical Management

The goal of therapy in these patients is to rule out visceral involvement and then to try to minimize the frequency of epistaxis. The tendency toward spontaneous and often severe nose bleeds greatly curtails social and professional activity. Conservative management includes lipid-based nasal moisturizers and unfortunately frequent nasal packing. Patients should avoid anticoagulants, including aspirin and nonsteroidal anti-inflammatory agents, unless necessary for other conditions. Systemic hormone therapy (both estrogen and estrogen-progesterone combinations) has shown some success by encouraging squamous metaplasia, but side effects must be monitored closely. Aminocaproic acid, an antifibrinolytic, has been administered both systemically and more often as a topical spray, also with limited benefit. Its mechanism of action is believed to be the inhibition of fibrinolysis in the telangiectatic vessel wall. Brachytherapy with iridium^{192} has been attempted but only in recalcitrant cases. Persisting anemia may benefit from supplemental iron therapy. Patients also frequently need blood transfusions throughout their life, and they run the risk of infectious disease transmission via blood products.

Surgical Management

If medical management fails, infrared lasers, including the Nd:YAG (neodymium:yttrium-aluminum-garnet), argon, and KTP (potassium-titanyl-phosphate), have been used with some success. These treatments generally need to be repeated because lesions typically return in 3 to 12 months. Septal dermoplasty is another approach. This involves stripping the mucous membrane over the septum and lateral nasal wall with placement of a split-thickness skin graft. This can be effective, but over time AVMs will recur. For persistent bleeding despite these conservative surgical therapies, other approaches exist. These include internal maxillary artery ligation or embolization and the Young procedure, which is anterior closure of the nasal cavity.

Surgical management may also be needed for systemic AVMs. Pulmonary AVMs with feeding vessels larger than 3 mm require occlusion, usually via transcatheter embolotherapy. A chest CT should be repeated 1 year later to ensure that reperfusion has not occurred. Cerebral AVMs larger than 1 mm are treated by neurovascular surgery, embolotherapy, or stereotactic radiosurgery. Liver AVMs rarely require therapy, but if a patient does become symptomatic, liver transplantation has been the only effective intervention.

Rehabilitation and Follow-up

Despite the array of treatments available, all therapies are temporary because of recurrent AVMs. Ongoing therapy will likely be required for recurring epistaxis. A complete blood count should be checked periodically. An evaluation for pulmonary AVMs should be repeated every 5 years, either with contrast echocardiography or CT, unless symptoms develop. If an MRI has ruled out cerebral AVMs, it generally does not need to be repeated. It is recommended that a multidisciplinary team be involved to manage the wide range of organ systems affected by this disease.

Questions

1. To diagnose hereditary hemorrhagic telangiectasia (HHT), the following should be present:
- **A.** Recurrent epistaxis
- **B.** Family history of HHT
- **C.** Anemia
- **D.** Mucocutaneous telangiectasias
- **E.** A, B, and C
- **F.** A, B, and D

2. A patient with HHT begins to complain of fatigue, with subsequently limited activity. This should raise concern for:
- **A.** Anemia
- **B.** Cerebral AVM
- **C.** Pulmonary AVM
- **D.** Hepatitis

3. The nose is the most common source of bleeding in HHT, but where are other common sites located?
- **A.** Fingers
- **B.** Liver
- **C.** Lip
- **D.** Gastrointestinal tract
- **E.** Lung

Suggested Readings

Abdalla SA, Letarte M. Hereditary haemorrhagic telangiectasia: current views on genetics and mechanisms of disease. J Med Genet 2006;43(2):97–110

Cottin V, Dupuis-Girod S, Lesca G, Cordier JF. Pulmonary vascular manifestations of hereditary hemorrhagic telangiectasia (Rendu-Osler disease). Respiration 2007;74(4):361–378.

Elluru RG, Azizkhan RG. Cervicofacial vascular anomalies. II. Vascular malformations. Semin Pediatr Surg 2006; 15(2):133–139

Espinosa PS, Pettigrew LC, Berger JR. Hereditary hemorrhagic telangectasia and spinal cord infarct: case report with a review of the neurological complications of HHT. Clin Neurol Neurosurg 2008;110(5):484–491

Geisthoff UW, Fiorella ML, Fiorella R. Treatment of recurrent epistaxis in HHT. Curr Pharm Des 2006;12(10): 1237–1242

Giordano P, Nigro A, Del Vecchio GC, Sabbà C, De Mattia D. HHT in childhood: screening for special patients. Curr Pharm Des 2006;12(10):1221–1225

Jameson JJ, Cave DR. Hormonal and antihormonal therapy for epistaxis in hereditary hemorrhagic telangiectasia. Laryngoscope 2004;114(4):705–709

Shovlin CL, Guttmacher AE, Buscarini E, et al. Diagnostic criteria for hereditary hemorrhagic telangiectasia (Rendu-Osler-Weber syndrome). Am J Med Genet 2000;91(1):66–67

46

Anosmia

Charles M. Myer IV and Allen M. Seiden

History

A 54-year-old white man complains of olfactory loss that seemed to persist following a severe upper respiratory infection that he suffered 15 months ago. The infection has since cleared, and he reported no residual nasal obstruction, postnasal drainage, or recurrent infections. The patient denied any history of allergies or asthma. He reported a history of a broken nose and resultant deviated septum from an injury 5 years ago but no other facial trauma. He denied any history of toxin, chemical, or solvent exposure.

The patient noted that on occasion he could detect an unpleasant odor that was difficult to describe, but he characterized it as rotten. It seemed to occur spontaneously, without relation to any obvious environmental stimulus, although it had diminished somewhat in frequency over the past few months. He did report that his taste perception had decreased the same time as his loss of smell, although he noted that he could clearly distinguish salty, sour, sweet, and bitter taste sensations. The patient further reported an unremarkable medical history, was on no medications, and did not smoke or drink alcohol.

On physical examination, anterior rhinoscopy demonstrated a patent nasal airway with mild deviation of the septum. No erythema, rhinorrhea, crusting, discharge, or nasal polyps were visible. Oral examination also showed no erythema, lesions, or excessive dryness. The tongue showed normal-appearing mucosa and papillae on the dorsal surface. The remainder of the head and neck examination was unremarkable. Olfactory testing was performed (see below) and was consistent with severe hyposmia.

After application of topical anesthesia and decongestion, nasal endoscopy was performed. In both the right and left nasal passages, the middle meatus and superior meatus were clear of polyps, discharge, or signs of inflammation. No tumors or scarring were present, and the airspace surrounding the olfactory cleft was clear.

Differential Diagnosis—Key Points

1. Dysfunction of the olfactory system may be due to a conductive or sensorineural loss. A conductive loss is due to obstruction of the airflow carrying odorant molecules to the neuroepithelium located in the olfactory cleft, which consists of the superior nasal septum, the upper aspect of the superior turbinate, and the superior-lateral nasal wall. This cleft is approximately 1 mm wide. Sensorineural losses result from dysfunction

along the neural pathway from the neuroepithelium to central processing locations via the olfactory bulb.

2. Nasal or sinus pathology is the only cause of a conductive olfactory deficit and may include diseases such as sinonasal neoplasms, nasal inflammation and polypoid disease consistent with chronic sinusitis and even adenoidal hypertrophy where transport of odorant molecules to the olfactory cleft is decreased or absent altogether as the result of absence of nasal airflow. Nasal airflow obstruction is suggestive of a transport defect, but only around a third of patients with an obstructive loss report such symptoms, and its absence should not rule out an obstructive cause. A fluctuation in the loss of smell suggests variation with changes in nasal congestion and occurs only with an obstructive loss, commonly in patients with chronic sinus disease and nasal polyposis. Unilateral symptoms and epistaxis should raise suspicion of neoplastic disease, which only rarely manifests with isolated anosmia. Nasal endoscopy should be performed in all patients with decreased olfaction and can detect more subtle pathology, such as within the nasal vault, ostiomeatal complex, or sphenoethmoidal recess, which might not be evident on anterior rhinoscopy (**Fig. 46.1**).

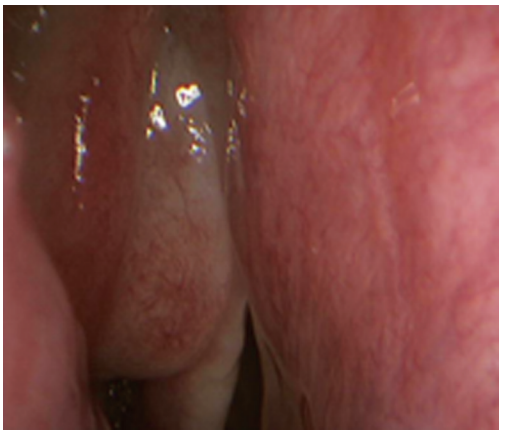

Fig. 46.1 Nasal endoscopic view of a small polyp within the right middle meatus in a 50-year-old woman complaining of olfactory loss but without a history of sinusitis, demonstrating the importance of a thorough nasal endoscopic examination.

3. Common causes of a sensorineural olfactory loss include an upper respiratory infection (URI), head trauma, and chemical or toxic exposure. Less common causes include dementia (olfactory loss has been identified as a possible early symptom in several neurodegenerative diseases, including Alzheimer and Parkinson diseases), medication effects, and insults to the sensory neuroepithelium, such as radiation therapy. There is evidence to suggest that chronic inflammatory sinus conditions may be associated with inflammatory changes in the olfactory receptor cells, thereby possibly causing a sensorineural component along with obstruction.
4. This patient describes a severe upper respiratory infection, after which his sense of smell never seemed to return. One needs to rule out the possibility of underlying sinonasal inflammation, but such a history suggests this loss to be postviral. The pathophysiology behind postviral olfactory loss is unclear, but studies support both destruction of the peripheral receptors and degeneration of the central olfactory pathways as possible insults. Most frequent in women and older age groups, the postviral loss can present with either hyposmia, a reduced ability to smell, or complete anosmia. The diagnosis is based on a consistent history of olfactory deficit following URI and the absence of any other pathology. Often this infection is described as having been particularly severe. This patient's history suggests a viral cause, and the lack of physical examination findings suggestive of any other cause makes this the most likely diagnosis.
5. In this patient, the history of nasal trauma might suggest a traumatic cause, but this event occurred many years before the patient noted a loss of smell. He does have a deviated septum, but this is generally not associated with a conductive olfactory loss.
6. This patient complained of a taste loss associated with his loss of smell, common in many patients with a decrease in olfaction. Given the more extensive and redundant neural inputs mediating taste (including the chorda tympani, glossopharyngeal and vagus nerves bilaterally), a true measurable taste loss is uncommon. However, the flavor of foods is determined by both gustatory

and olfactory inputs in addition to temperature and texture information. In the absence of olfactory input, patients perceive the lack of flavor as a loss of taste input. In the history, assessing the ability of the patient to detect the four basic taste qualities can determine whether a taste loss is present.

7. Dysosmia, an abnormal or distorted sense of smell, may be precipitated by an environmental stimulus (parosmia or troposmia), or it may occur spontaneously (phantosmia). It has been described following postviral and traumatic damage to the sensory neuroepithelium as well as in sinonasal disease. Dysosmia may also be seen in psychiatric disorders and as an aura symptom in temporal lobe seizures. It is most common in postviral hyposmia or anosmia, occurring in two thirds of patients.

◆ Test Interpretation

Testing a patient's olfactory ability is mandatory to verify and quantify the patient's complaint as well as to establish a baseline against which interventions and recovery may be measured. Several commercial testing systems are available for clinical settings using odor identification or odor threshold testing. Of the most widely used tests, the University of Pennsylvania Smell Identification Test (Sensonics, Haddon Heights, NJ), is a well-validated forced-choice test that provides a method to determine olfactory sensitivity and detect malingering using 40 microencapsulated odorants in a "scratch-and-sniff" format with four answer choices for each.

If sinonasal pathology is suspected based on the history and physical examination, imaging with computed tomography is the procedure of choice. Magnetic resonance imaging (MRI) is preferred when evaluating for intracranial causes of olfactory dysfunction. As previously discussed, the likelihood of an intracranial process causing an isolated anosmia without other findings is quite rare and, therefore, routine MRI is not indicated in most cases. In cases where the clinical presentation does not match the common causes presented previously, or symptoms or signs such as cranial nerve palsies or papilledema exist, MRI should be performed. Because this patient fits the clinical picture of postviral loss, with no evident sinonasal pathology, no imaging is necessary.

In the patient with sinus pathology and an absence of a clear history of fluctuation, establishing reversibility can support an obstructive cause and exclude sensorineural causes. Systemic corticosteroids may be prescribed as a diagnostic tool to facilitate at least a temporary improvement in olfactory sensitivity, presumably by decreasing the mucosal edema in sinonasal disease. Topical steroids alone do not appear to be as effective in the short-term reversal of such a loss.

◆ Diagnosis

Postviral olfactory loss

◆ Medical Management

No known medical therapy has been shown to be effective in the treatment of postviral olfactory loss. Zinc, believed for many years to be an effective remedy for all chemosensory disorders, has been shown in recent studies to have no benefit over placebo. Many new therapies are being tested and show promise, but controlled and randomized data are lacking.

In cases of obstructive olfactory loss, as noted earlier, systemic steroids will often reverse the decreased sense of smell. However, given the adverse effects of long-term steroid use, only a short tapering course should be used. A topical steroid spray may then be started, which may or may not be able to maintain any improvement.

Elimination of any inciting factors in nasal inflammation is important, and as such any associated allergies should be evaluated and adequately treated. If active infection is found, appropriate antibiotic therapy is indicated.

◆ Surgical Management

In cases of obstructive olfactory loss in which normal function cannot be maintained medically, surgical intervention may be considered. The surgical approach will depend on the extent and nature of the underlying pathology but can be effective in restoring an

obstructive loss. Special attention in the area of the olfactory cleft to prevent scarring or injury is vital to the success of the procedure.

Rehabilitation and Follow-up

The olfactory receptors undergo constant turnover, and this regenerative capacity creates the potential for recovery following a sensorineural loss. However, such recovery is variable and seems to occur more often after a viral rather than traumatic insult, although a recent study suggested that recovery is inversely related to the severity of loss regardless of the cause. Approximately two thirds of patients suffering a viral olfactory loss will get some, albeit incomplete, recovery.

Questions

1. Olfactory loss is commonly associated with the following except:
- **A.** Head injury
- **B.** Deviated nasal septum
- **C.** Viral upper respiratory infection
- **D.** Parkinson disease

2. When a patient reports a loss of smell that seems to fluctuate with exercise or showering, this suggests that the loss is due to which of the following?
- **A.** A viral infection
- **B.** Nasal or sinus disease
- **C.** Hypothyroidism
- **D.** Toxic exposure

3. The flavor of food is determined by what?
- **A.** Gestation
- **B.** Olfaction
- **C.** Texture
- **D.** Temperature
- **E.** All of the above

4. Dysosmia occurs most commonly in association with olfactory loss due to:
- **A.** Head injury
- **B.** Nasal and sinus disease
- **C.** Viral upper respiratory infection
- **D.** Toxic exposure

Suggested Readings

Doty RL. Office procedures for quantitative assessment of olfactory function. Am J Rhinol 2007;21(4):460–473

Duncan HJ, Seiden AM. Long-term follow-up of olfactory loss secondary to head trauma and upper respiratory tract infection. Arch Otolaryngol Head Neck Surg 1995;121(10):1183–1187

London B, Nabet B, Fisher AR, White B, Sammel MD, Doty RL. Predictors of prognosis in patients with olfactory disturbance. Ann Neurol 2008;63(2):159–166

Seiden AM. Postviral olfactory loss. Otolaryngol Clin North Am 2004;37(6):1159–1166

Seiden AM, Duncan HJ. The diagnosis of a conductive olfactory loss. Laryngoscope 2001;111(1):9–14

Spielman AI. Chemosensory function and dysfunction. Crit Rev Oral Biol Med 1998;9(3):267–291

Wrobel BB, Leopold DA. Clinical assessment of patients with smell and taste disorders. Otolaryngol Clin North Am 2004;37(6):1127–1142

47

Cerebrospinal Fluid Rhinorrhea

Gordon H. Sun and Allen M. Seiden

History

A 27-year-old man was involved in a low-speed motor vehicle collision as a front passenger and was brought urgently to the emergency department for evaluation. After his respiratory status was deemed stable and his cervical spine was cleared, he underwent a maxillofacial computed tomography (CT) scan that demonstrated a comminuted nasal fracture with septal deviation to the patient's right side. Evidence was seen of a bony dehiscence of the skull base at the right posterior ethmoid sphenoid junction (**Fig. 47.1**). The otolaryngology service was consulted for both the nasal fracture and mild bloody rhinorrhea that had failed to resolve spontaneously despite direct pressure and topical nasal decongestants. A directed history elicited no medical history, no previous maxillofacial trauma (until the motor vehicle accident), and no prior sinonasal surgeries. He was not on medications at the time of the collision, and he denied drug or environmental allergies. He denied visual or auditory deficits, symptoms of nasal obstruction, or anosmia.

On physical examination, the patient was afebrile with stable vital signs. He was awake, alert, and fully oriented. He demonstrated

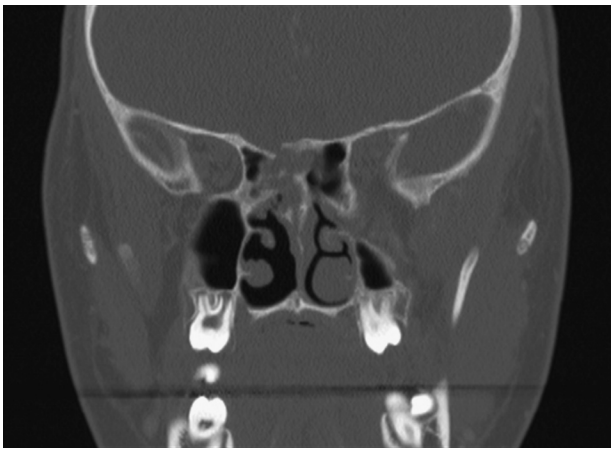

Fig. 47.1 Coronal computed tomography image demonstrates a traumatic encephalocele extending into the right ethmoid and sphenoid sinuses.

several superficial skin lacerations and abrasions as well as a C-shaped deformity of the nasal septum. He also had a small amount of blood-tinged clear drainage emanating from both nares, which was more obvious when the patient leaned forward. Drainage that had seeped onto his pillowcase showed a "halo" sign. Rigid nasal endoscopy at the bedside did not demonstrate an obvious intranasal source. The remainder of his head and neck examination was unremarkable.

Some of the fluid was captured in a test tube and sent for laboratory studies. He was observed in the hospital for an additional 2 days with bed rest precautions. The rhinorrhea resolved spontaneously within 48 hours of the initial consultation. The patient was subsequently discharged in stable condition, with closed reduction of the nasal fracture scheduled as an outpatient procedure.

◆ Differential Diagnosis—Key Points

1. In this case, the patient clearly had a nasal fracture, and mild epistaxis would be expected. However, as it did not stop with the usual measures, seemed more clear than frankly bloody, and was exacerbated by leaning forward, the possibility of a post traumatic cerebrospinal fluid (CSF) leak needs to be considered.
2. CSF is formed in the choroid plexus within the lateral, third, and fourth ventricles of the brain at a rate of 20 mL per hour. The total volume of CSF in an adult is 90 to 150 mL. Normal CSF pressure is 140 mm H_2O (10–15 mm Hg) in adults and 40 mm H_2O in children, although pressure varies with the cardiac and respiratory cycles, activity level, and changes in head position. Maintaining CSF pressure requires a delicate balance between CSF secretion and resorption.
3. Between 1 and 3% of all closed-head injuries result in CSF leaks. Roughly 80% of all CSF leaks manifest as rhinorrhea, whereas the remaining patients demonstrate otorrhea. The most common cause of CSF rhinorrhea is accidental head trauma (44%), followed by iatrogenic injury from endoscopic sinus surgery, skull-base surgery, septoplasty, neurosurgery (29%), and tumors (22%). Congenital and "spontaneous" CSF leaks account for the remaining 15% of cases. Spontaneous CSF leaks, which have no obvious inciting factor or cause, are most commonly encountered in obese, middle-aged women, who usually complain of headaches, pulsatile tinnitus, and disequilibrium. This may be a manifestation of benign intracranial hypertension or another cause of increased intracranial pressure (ICP). This elevation exerts pressure on the anterior skull base, with thinning and remodeling of bone, particularly in areas such as the lateral sphenoid recess and cribriform plate. Once a defect occurs, the dura begins to herniate (meningocele), which can then incorporate brain parenchyma as it enlarges (encephalocele) (**Fig. 47.2**).
4. The most common location for CSF rhinorrhea is the cribriform plate (35%), followed by the sphenoid sinus (26%), anterior ethmoid (18%), frontal sinus (10%), posterior ethmoid (9%), and inferior clivus (2%). The site of the leak is important in determining the best surgical approach for repair when surgery is indicated.
5. If left untreated, CSF rhinorrhea carries at least a 10% risk of developing meningitis

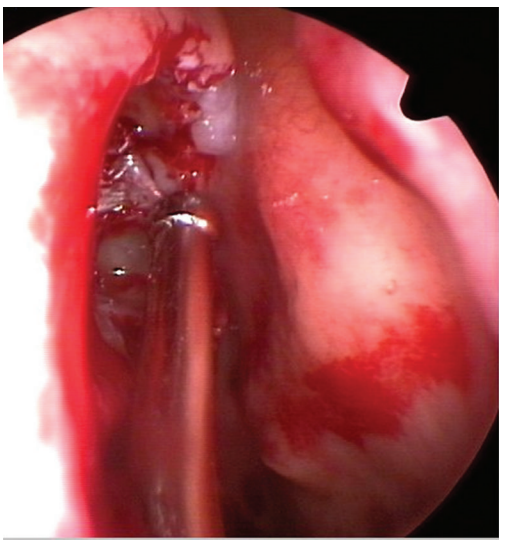

Fig. 47.2 Curved olive tip suction points toward an encephalocele protruding through the right cribriform plate. The middle turbinate has largely been removed.

annually; the risk becomes higher as the CSF leak persists for longer durations. Some studies have demonstrated a clear reduction in acquiring meningitis when the leak is closed endoscopically. Nonetheless, traumatic CSF rhinorrhea is also associated with a high rate of spontaneous closure (>70%).

Test Interpretation

Traditionally a rapid bedside test for CSF rhinorrhea is the "halo" sign, which is positive when a clear ring surrounding a central blood spot appears after bloody nasal discharge is placed on a paper towel. However, tears or saliva are also well known to give a false-positive appearance. Sometimes a thorough nasal endoscopic examination can provide evidence of clear watery discharge and can provide clues as to the source or location (**Fig. 47.3**).

The most commonly used laboratory study for detecting CSF leaks is the assay for β-2 transferrin, a compound found nearly exclusively in CSF. The test is extremely sensitive (97%) and specific (93%) for the presence of CSF; however, this assay is not widely available, and collecting a sufficient sample for analysis may be challenging in patients who have minimal or intermittent CSF rhinorrhea.

The gold standard for localizing the source of CSF rhinorrhea is CT cisternography, with a sensitivity of 92% in localizing active leaks and 40% in intermittent or inactive leaks. Coronal and axial CT imaging may delineate areas of bony dehiscence; a contrast agent delivered by lumbar puncture (followed by maneuvers to force an active CSF leak) may identify the specific location of the rhinorrhea. Its primary shortcomings are a limited ability to assess intermittent or very slow leaks and the small risk of infection or lumbar CSF leak. In addition, its resolution is such that it often does not delineate the precise site of leak but may simply confirm that there is a leak. Because of these flaws, many otolaryngologists regard plain thin-section CT imaging as sufficient for identification of likely sites of CSF rhinorrhea. With fine-cut CT techniques, sensitivity in detecting osseous defects may be as high as 92% and specificity approaching 100%. Magnetic resonance imaging may be useful if the suspected cause of the leak is an encephalocele or meningoencephalocele but has the significant disadvantage of poor bone detail. Magnetic resonance cisternography is a relatively new modality for detecting CSF leak sites, with early studies demonstrating sensitivities of 87% and accuracy of 78 to 100%. However, at this time the gadolinium-enhanced contrast material used for this study is considered off label. Another frequently used test is radionuclide cisternography, which is usually done by placing nasal pledgets endoscopically, then administering intrathecal radiotracers and measuring the radiotracer activity within the pledgets after they are removed 24 hours later. While moderately sensitive, radionuclide cisternography requires substantial cooperation from the patient, who may have to tolerate up to six pledgets at a time.

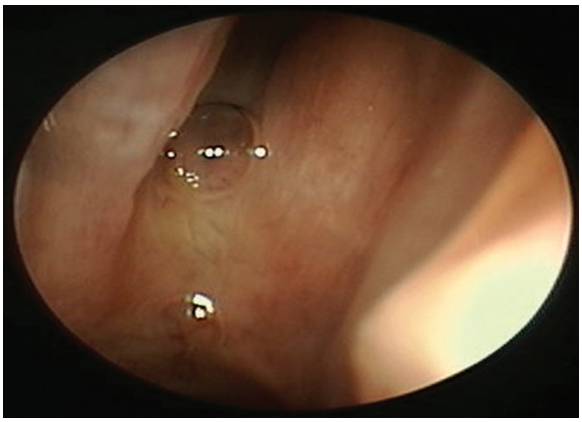

Fig. 47.3 Clear fluid noted from the right sphenoid ostium on nasal endoscopy, suggesting this to be the location of the patient's cerebrospinal fluid leak.

Diagnosis

Traumatic CSF rhinorrhea

Medical Management

Most CSF leaks, especially those caused by head trauma, will resolve with conservative measures. These include bed rest, head elevation, use of stool softeners, and avoidance of straining, nose blowing, or Valsalva maneuvers. Positive pressure ventilation, in particular, should be avoided to prevent the development of pneumocephalus. Another method of treatment is to place a lumbar drain for passive drainage of excess CSF. While the drain is in place, daily CSF laboratory studies, including CSF cell counts, glucose, protein, and cultures, should be obtained, and a goal drainage rate of 5 to 10 mL should be achieved. Persistent CSF leaks have a higher rate of infection, and surgical management becomes mandatory. Nonetheless, prophylactic antibiotics in patients with newly diagnosed CSF leaks are generally not indicated, although antibiotics are usually given in the perioperative setting. Ceftriaxone, trimethoprim-sulfamethoxazole, and levofloxacin are frequently used antibiotics because of their ability to penetrate the blood–brain barrier.

Surgical Management

In many cases, placing a lumbar drain before operative CSF leak repair is useful to establish better control of ICP during surgery. Fluorescein can be injected into the intrathecal space after the drain is placed to identify more precisely the location of the leak. Rapid-sequence intubation should be performed to minimize the need for bag-mask ventilation and the possibility of pneumocephalus.

The location of the CSF leak is critical in selecting the most effective surgical approach for repair. Bony defects in the ethmoid roof, cribriform plate, central sphenoid, and perisellar areas of the skull base can be repaired via transethmoid or transseptal endoscopic approaches. Lateral sphenoid defects are more difficult to access in this fashion and may require a transpterygoid endoscopic approach after performing a total ethmoidectomy and maxillary antrostomy. Frontal sinus CSF leaks can be further subdivided into leaks originating from the frontal recess, adjacent to the frontal recess, and within the frontal sinus itself. If the defect is located in the inferior or anterior position, a combined endoscopic and open procedure (e.g., frontal trephination) may be appropriate to access the source of the CSF rhinorrhea. Laterally and superiorly based CSF leaks may not be accessible with nasal endoscopy and thus usually require an extracranial approach, specifically an osteoplastic flap with or without frontal sinus obliteration. Defects at multiple sites, excessively large or comminuted bony injuries, obvious skull-base deformities, high-pressure CSF leaks, or tumors with intracranial extension limit the usefulness of purely endoscopic approaches and will generally require an extracranial approach. Bone and mucosal grafts from the nasal septum, turbinates, mastoid, or maxilla, temporalis fascia, and even vascularized tissue transfer are all potential sources of tissue for closing a CSF leak site. The tissue may be placed as either an underlay or overlay graft, although mucosal grafts placed in an underlay fashion may theoretically result in mucoceles, meningitis, or other neurologic problems. Accidental obstruction of other sinuses after placing the graft(s) can lead to mucocele formation as well. Absorbable packing material (e.g., abdominal fat, Gelfoam, Surgicel) should be placed over all of the grafts to prevent inadvertent removal or displacement. Successful surgical closure of CSF leaks without the need for a craniotomy now exceeds 90% with current endoscopic and extracranial techniques.

Rehabilitation and Follow-up

As long as a lumbar drain is present, all patients with CSF rhinorrhea should remain on strict bed rest with the head of bed elevated to at least 15 degrees. Once the drain is removed, ambulation is gradual and light activity is encouraged for up to 6 weeks after surgery. Straining should be avoided, including Valsalva maneuvers or other activities that increase CSF pressure. Continuous positive airway pressure should be avoided in the immediate postoperative period to prevent pneumocephalus, but it may be restarted about 4 to 6 weeks after

surgery. In some cases, acetazolamide may be prescribed to decrease CSF production. Routine follow-up every 1 to 2 weeks is scheduled with conservative endoscopic debridement of the nasal passages to prevent crusting, sinusitis, and other complications. The graft and packing sites are avoided to allow appropriate healing to take place. After about 6 weeks, the repair site should be healed and normal activity may be resumed.

◆ Questions

1. What is the most accurate method for determining whether nasal discharge is spinal fluid?
   A. Halo sign
   B. Measure for glucose
   C. Measure for β-2 transferrin
   D. CT cysternography

2. What is the most common location for CSF rhinorrhea to occur?
   A. Ethmoid roof or fovea ethmoidales
   B. Cribriform plate
   C. Sphenoid sinus
   D. Frontal sinus

3. What is the first step in properly managing traumatic CSF rhinorrhea?
   A. Bed rest
   B. Lumbar drain
   C. Placement of nasal packing
   D. Operative intervention to repair the leak

Suggested Readings

Daudia A, Biswas D, Jones NS. Risk of meningitis with cerebrospinal fluid rhinorrhea. *Ann* Otol Rhinol Laryngol 2007;116(12):902–905

Lloyd KM, DelGaudio JM, Hudgins PA. Imaging of skull base cerebrospinal fluid leaks in adults. Radiology 2008;248(3):725–736

Martin TJ, Loehrl TA. Endoscopic CSF leak repair. Curr Opin Otolaryngol Head Neck Surg 2007;15(1):35–39

Reh DD, Metson R, Sindwani R. Sinus surgery in patients with previously repaired cerebrospinal fluid leaks. Arch Otolaryngol Head Neck Surg 2008;134(11):1187–1190

Schlosser RJ, Bolger WE. Endoscopic management of cerebrospinal fluid rhinorrhea. Otolaryngol Clin North Am 2006;39(3):523–538, ix

Schlosser RJ, Bolger WE. Nasal cerebrospinal fluid leaks: critical review and surgical considerations. Laryngoscope 2004;114(2):255–265

48

Inverting Papilloma

Brandon Scott Hopkins and Allen M. Seiden

History

A 45-year-old man seen by his primary care physician over several months complains of difficulty breathing through his right nasal cavity for longer than a year, with frequent nasal drainage and occasional bleeding. He is referred after being thoroughly treated for chronic sinusitis with antibiotics and nasal steroid sprays and has had no resolution of his symptoms. He reports having had surgery for a deviated septum and right nasal polyp 2 years ago but has not since been seen by an otolaryngologist. The patient is a social drinker with no smoking history.

On examination he appears to have decreased movement of air through his right nasal cavity. Anterior rhinoscopy is essentially clear, but on nasal endoscopy a fairly large polypoid mass can be seen projecting from beneath the right middle turbinate (**Fig. 48.1**). It appears polypoid and not terribly erythematous, friable, or firm. The mass does not appear to be vascular. Endoscopic examination of the left nasal cavity is clear, demonstrating no evidence of polyps or ostiomeatal disease. A subsequent computed tomography (CT) scan of the sinuses was obtained (**Fig. 48.2**).

Differential Diagnosis—Key Points

1. The most common nasal mass is a benign inflammatory polyp. However, these tend to be bilateral and have a gray, translucent appearance. The finding of a unilateral nasal mass opens a broad differential that includes

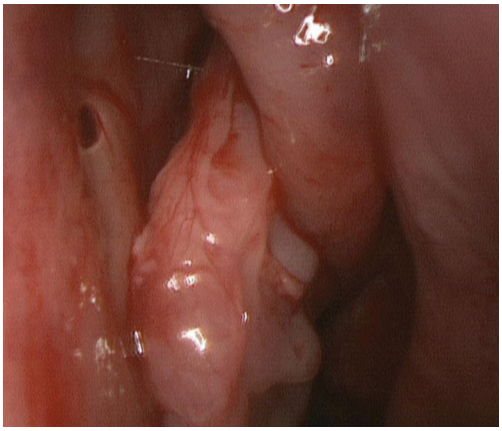

Fig. 48.1 Right nasal polypoid mass protruding from beneath the middle turbinate.

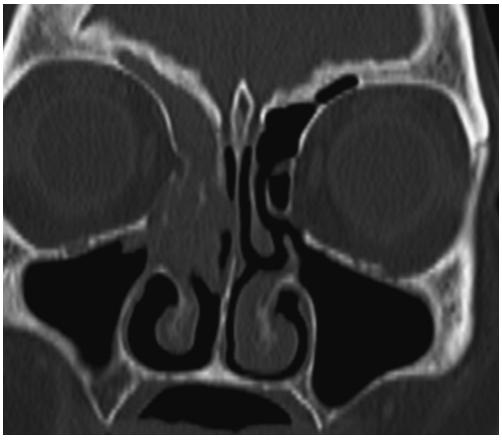

Fig. 48.2 CT demonstrating a polypoid mass within the right anterior ethmoid sinus and middle meatus, with possibly bony disruption of the uncinate process and middle turbinate.

infectious processes as well as benign and malignant tumors. Antrochoanal polyps can present as unilateral polypoid masses filling the middle meatus. Benign tumors include osteomas, chondromas, ossifying fibromas, schwannomas, neurofibromas, meningiomas, vascular tumors, hamartomas including ameloblastomas, and, most commonly, papillomas. Malignant tumors of the nasal cavity are most commonly squamous cell cancer but can include adenocarcinoma, esthesioneuroblastoma, lymphoma, melanoma, and others.

2. Inverting papilloma is a lesion arising from the Schneiderian membrane. This is the transition between the endoderm-derived respiratory epithelium and the ectodermally derived squamous epithelium. Other lesions can arise from this membrane, including fungiform papillomas, which are often found on the nasal septum and are human papillomavirus (HPV) related, and cylindrical papillomas, which appear different but act similar to inverting papillomas. Debate exists as to whether these are different entities or expressions of the same process in different locations.
3. Inverting papillomas histologically have an epithelium that inverts into the underlying connective tissue stroma. They often appear exophytic, polypoid, more vascular than an inflammatory polyp, and with a gray to pink appearance. In contrast to the lesion in this case, they may sometimes appear more fibrous and friable. No mucus cells, koilocytosis, or eosinophils are present. This is in contradistinction to inflammatory nasal polyps. The basement membrane remains intact. Although invasion into surrounding structures such as occurs with malignancy is not present, pressure and mass effect can cause bone remodeling and destruction.
4. Inverting papilloma is the most common benign epithelial tumor of the nose and paranasal sinuses. Incidence is in the range of 0.2 to 0.6% per 100,000 people per year, making up 0.5 to 4% of primary nasal tumors. The male-to-female ratio is 3:1, and predominance in white patients is suggested in the literature. The most common sites of origin include the lateral nasal wall at the root of the middle turbinate and the maxillary sinus.
5. There is no widely accepted cause, but there may be an association with HPV. Various types, including 6, 11, 16, and 18, have been found in these lesions, but no clear causative factor has been found. Further study is needed in this area and in establishing a possible link with smoking. Allergy does not appear to have any association with inverting papillomas.
6. A universally accepted staging system does not exist, but the one by Krouse is often used (**Table 48.1**).
7. Malignancy, usually squamous cell cancer, may develop as a focus in an inverting papilloma, as a separate lesion, or as a metachronous lesion presenting after an inverting papilloma resection. Metachronous lesions may present years later. Malignancy risk has been quoted as anywhere between 1 and 53%, but 10% is a more likely rate. This risk, along with its tendency to recur and cause local destruction, is what creates the indication for aggressive surgical removal.

Table 48.1 Staging system for inverting papilloma

Stage	Description
T1	Tumor confined to the nasal cavity
T2	Tumor involving the ostiomeatal complex, ethmoid sinus, and/or medial wall maxillary sinus
T3	Tumor involving lateral, inferior, superior, anterior, or posterior walls of maxillary sinus, sphenoid sinus, and/or frontal sinus
T4	Tumors with extranasal and extrasinus extension and all malignant tumors

Source: Adapted from Krouse JH. Development of a staging system for inverted papilloma. Laryngoscope 2000;110:965–968.

◆ Test Interpretation

Confirmative diagnosis lies in biopsy. This is best done before plans are made for definitive surgical management. This patient undewent biopsy in the clinical setting, with minor bleeding controlled with silver nitrate. On hematoxylin and eosin staining, fronds of epithelium inverting into the surrounding stroma were demonstrated (**Fig. 48.3**). In such cases, the epithelium can be squamous, transitional, or respiratory.

If a vascular tumor such as a hemangioma, hemangiopericytoma, juvenile nasopharyngeal angiofibroma, or pyogenic granulama is suspected an office biopsy may be contraindicated because it may cause extensive bleeding. With this in mind, any biopsy of a nasal mass should be approached cautiously in a controlled environment with resources available to manage epistaxis.

Imaging studies are important to define the extent of the lesion. A friable, vascular-looking lesion whose upper extent cannot be visualized raises the concern for a possible encephalocele or other intracranial pathology. In such situations, it is helpful to obtain imaging studies prior to biopsy.

CT is the imaging modality of choice, depicting both soft tissue and bony changes. A CT scan will often support a diagnosis of inverting papilloma versus benign respiratory polyps by demonstrating an expansile appearance with surrounding bone destruction. However, it is important to note that benign polypoid disease associated with allergic fungal sinusitis, antrochoanal polyp, and other conditions can cause bony destruction of the uncinate process and medial wall of the maxillary sinus.

Magnetic resonance imaging is valuable in selected cases to distinguish tumor from retained secretions and to differentiate from other soft tissue structures in the event of extrasinus extension.

◆ Diagnosis

Inverting papilloma

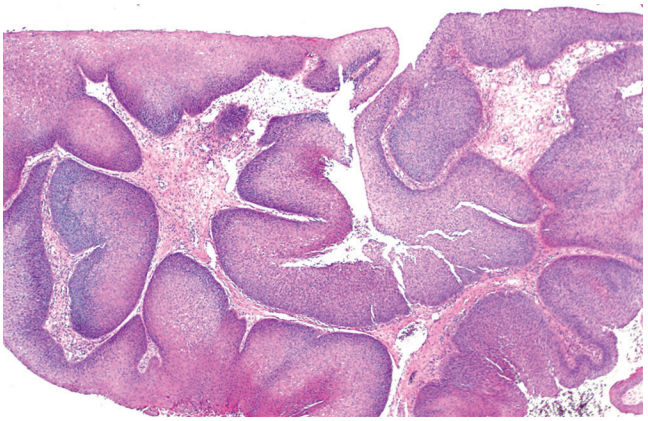

Fig. 48.3 Histological examination reveals hyperplastic epithelium invaginating into the fibrovascular connective tissue stroma.

Medical Management

Both radiation and chemotherapy have been used in the treatment of inverting papillomas. There are no clear guidelines for when these are indicated, but these modalities are usually considered when there is either an associated malignancy or an incomplete resection. Steroids and antibiotics have also been used to alleviate any inflammatory or infectious component before surgery, which can allow the extent of the papilloma to be more clearly visualized.

Surgical Management

Surgery is the modality of choice for treating inverting papilloma. Before the pathology of these lesions was fully understood, an intranasal polypectomy was associated with recurrence rates greater than 70%, leading to the development of more aggressive, en bloc resections. In the 1980s, a lateral rhinotomy with medial maxillectomy became the standard of care to remove completely the lateral nasal wall, with recurrence rates less than 15%. Complications include epiphora, a misplaced medial canthus, and a large facial scar. An alternative approach that spares an external incision is the midface degloving technique.

With the advent of endoscopic surgical techniques, many inverting papillomas are now being addressed in this manner. Recurrence rates have been comparable to traditional approaches. Initially only patients with small tumors were selected, but this has now been expanded to include most cases. Limited tumors can be addressed by limited resections, but if necessary a medial maxillectomy can also be performed endoscopically. A Caldwell-Luc approach in which a sublabial incision with maxillary puncture is performed may aid in access to the lateral, anterior, and inferior aspects of the maxillary sinus. En bloc resection via an endoscopic approach is possible and may be beneficial in cases associated with malignancy. However, a diagnosis of cancer often necessitates an external approach as well as adjunctive therapy. The most important aspect of inverting papilloma surgery is that the site of tumor attachment must be adequately addressed, often with removal or drilling of the underlying bone. Even though the inverting papilloma may be large at presentation, the site of attachment is often quite localized, but it may be beyond the confines of the lateral nasal wall. If the attachment site is not addressed, recurrence is likely. After surgery, endoscopic surveillance can usually be performed.

Rehabilitation and Follow-up

The risk of recurrence of inverting papilloma can approach 13% even with aggressive surgery. The risk of malignancy also makes monitoring an important aspect of care. It has been shown that lesions can occur 1 to 24 years later, but most present within 24 months. Therefore, regular follow-up with a thorough endoscopic examination and imaging when appropriate is important.

Questions

1. Why does inverting papilloma need to be treated aggressively?
- **A.** It can cause severe bleeding
- **B.** It is malignant.
- **C.** It may cause local destruction.
- **D.** It can metastasize.
- **E.** All of the above are correct.

2. Along with the middle turbinate and middle meatus, what is the most common site of origin of inverting papilloma?
- **A.** Frontal sinus
- **B.** Ethmoid sinus
- **C.** Maxillary sinus
- **D.** Sphenoid sinus

3. What is the rate of associated malignancy with inverting papilloma?
- **A.** 10%
- **B.** 15%
- **C.** 20%
- **D.** 25%

Suggested Readings

Cannady SB, Batra PS, Sautter NB, Roh HJ, Citardi MJ. New staging system for sinonasal inverted papilloma in the endoscopic era. Laryngoscope 2007;117(7):1283–1287

Eggers G, Mühling J, Hassfeld S. Inverted papilloma of paranasal sinuses. J Craniomaxillofac Surg 2007; 35(1):21–29

Karkos PD, Fyrmpas G, Carrie SC, Swift AC. Endoscopic versus open surgical interventions for inverted nasal papilloma: a systematic review. Clin Otolaryngol 2006;31(6):499–503

Krouse JH. Development of a staging system for inverted papilloma. Laryngoscope 2000;110(6):965–968

Landsberg R, Cavel O, Segev Y, Khafif A, Fliss DM. Attachment-oriented endoscopic surgical strategy for sinonasal inverted papilloma. Am J Rhinol 2008;22(6):629–634

Melroy CT, Senior BA. Benign sinonasal neoplasms: a focus on inverting papilloma. Otolaryngol Clin North Am 2006;39(3):601–617

Myers EN. Operative Otolaryngology—Head and Neck Surgery, 2nd ed. Philadelphia, PA: WB Saunders; 2008

Sauter A, Matharu R, Hörmann K, Naim R. Current advances in the basic research and clinical management of sinonasal inverted papilloma. Oncol Rep 2007;17(3): 495–504

49

Esthesioneuroblastoma

Aaron I. Brescia and Allen M. Seiden

History

A 37-year-old man, a nonsmoker, presents with a 3-month history of left nasal congestion and intermittent epistaxis. The bleeding has been fairly slight and has not required trips to the emergency department or placement of nasal packing. He has a history of mild allergic rhinitis but denies chronic sinusitis. He does not ordinarily take any medication for his allergies, and he is not on any medicine that might affect his bleeding time or coagulation. When questioned, he does admit to mild hyposmia. He denies weight loss, fevers, or night sweats.

Examination reveals anterior deviation of the nasal septum to the left with slight crusting, making the left posterior and superior nasal cavity somewhat difficult to visualize. However, no other lesions are noted on anterior rhinoscopy. His basic head and neck examination is otherwise unremarkable. Visual acuity and extraocular movements are intact. Pupils are equal, round, and reactive.

Nasopharyngoscopy reveals no polyps or evidence of ostiomeatal disease within the right nasal cavity, but does suggest some mild allergic inflammation of the inferior and middle turbinates. Examination of the left side does reveal a polypoid, slightly fibrous irregular mass in the superior nasal vault, medial to the middle turbinate (**Fig. 49.1**). It appears slightly friable. Maxillofacial computed tomography (CT) confirms a soft tissue mass filling the left nasal vault, with no apparent erosion of the cribriform plate, but involvement of the left ethmoid sinus and possible erosion of the left lamina papyracea. A subsequent magnetic resonance imaging (MRI) scan of the head and neck with contrast also reveals a large homogeneously enhancing mass filling the left nasal vault, with no intracranial

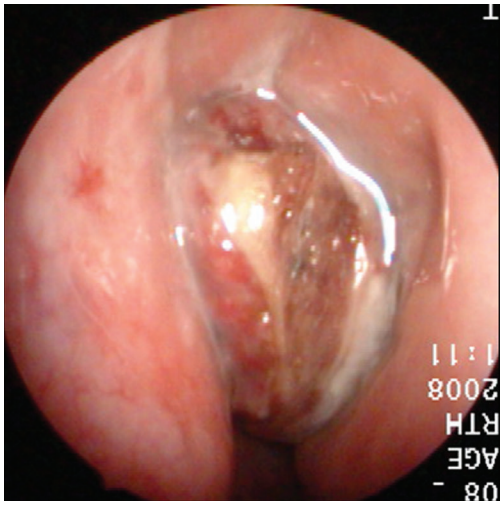

Fig. 49.1 Fibrous, irregular mass left upper nasal cavity.

extension, and sparing of the soft tissues of the orbit. Because of the possible vascular nature of the mass, he is scheduled for nasal endoscopy and nasopharyngoscopy with biopsy in the operating room.

Differential Diagnosis—Key Points

1. The etiology of this patient's symptoms could easily have been attributed to his deviated septum. However, a 3-month history of recurrent epistaxis warrants a more thorough examination of the nose. A unilateral nasal mass should immediately raise concerns that this might be something other than a benign respiratory polyp. Benign neoplasms to consider include inverting papilloma, hemangioma, schwannoma, antrochoanal polyp, and meningocele or meningoencephalocele.
2. Malignant nasal masses that may present in the nose include squamous cell carcinoma, both within the setting of a preexisting inverting papilloma and as a new, isolated neoplasm. Other malignancies include adenocarcinoma, adenoid cystic carcinoma, mucoepidermoid carcinoma, hemangiopericytoma, blue cell tumors such as esthesioneuroblastoma, rhabdomyosarcoma, plasmacytoma, malignant mucosal melanoma, lymphoma, sinonasal undifferentiated carcinoma, and nasopharyngeal carcinoma.
3. Clearly, in this case a biopsy is necessary to determine the specific histology. If there is concern for bleeding, biopsy is best undertaken in the operating room under controlled conditions. This should be done after appropriate imaging, which usually will include both CT and MRI with contrast.
4. Granulomatous diseases such as sarcoidosis, Wegener granulomatosis, and midline lethal granuloma may present in a similar fashion as a sinonasal malignancy (i.e., as a unilateral friable nasal mass). The lack of any other systemic findings or history in this 37-year-old patient makes these diagnoses less likely. Chronic rhinorrhea with extensive crusting, septal perforation, and a history of pulmonary or renal troubles or rheumatoid arthritis would guide the investigator toward these possibilities.

Test Interpretation

1. A direct coronal CT scan with cuts 3 mm or less is the preferred initial radiologic test. Most nasal tumors will appear as a homogeneous soft tissue mass with uniform moderate contrast enhancement. It is important to evaluate for possible erosion of the cribriform plate, fovea ethmoidalis, or lamina papyracea, and to stage the tumor (**Fig. 49.2**). If the tumor obstructs sinus drainage, accumulated secretions may be difficult to differentiate from tumor on a CT scan.
2. An MRI scan is useful for differentiating tumor from secretions, as well as from intracranial and orbital contents. Therefore, it is usually complementary to the CT (**Fig. 49.3**).
3. Biopsy. Histological analysis reveals discrete nests of tumor cells with dense stromal blood vessels. The tumor cells are of the small blue cell type, showing small dark round or oval nuclei, absence of nucleoli, and little cytoplasm. In addition there is fine eosinophilic fibrillary material that stains with S100 stain. The tumor cells exhibit pseudorosette configurations around this fibrillary eosinophilic material.

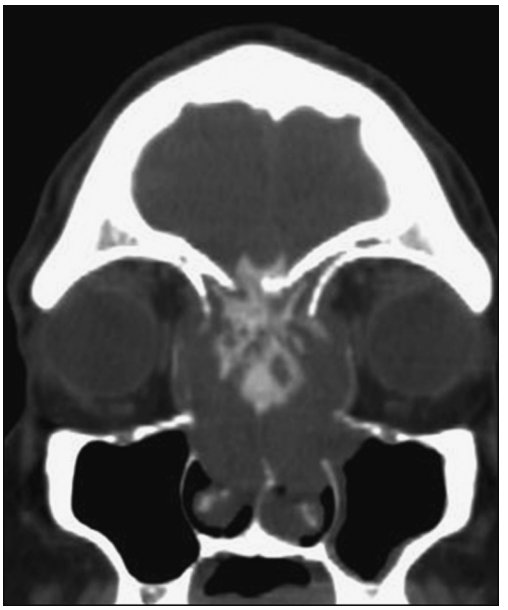

Fig. 49.2 Computed tomography scan of an advanced esthesioneuroblastoma, with erosion of the skull base and bilateral lamina papyracea.

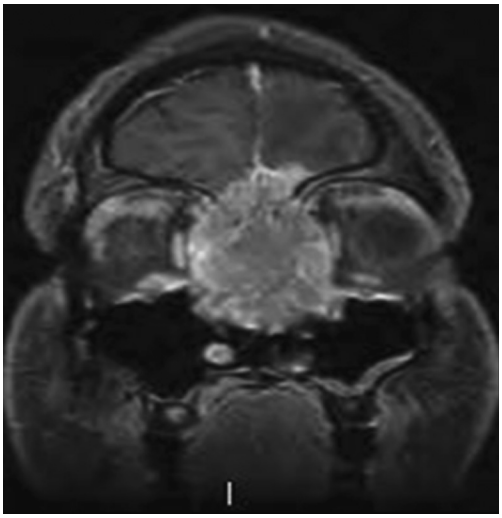

Fig. 49.3 Magnetic resonance imaging scan of the same patient shown in **Fig 49.2**, with an advanced esthesioneuroblastoma. Intracranial extension is demonstrated, with clear delineation between tumor and brain.

Diagnosis

Esthesioneuroblastoma

Medical Management

Esthesioneuroblastoma is a malignant neoplasm of olfactory and neuroectodermal origin. This tumor arises in the nasal cavity and is one of the families of small blue cell tumors that can be found in the sinonasal cavity. This tumor typically presents at an advanced stage as it arises high in the nasal vault, and nonspecific symptoms such as nasal obstruction, epistaxis, anosmia, or headache may delay further investigation until the symptoms become severe. The most widely used staging system is the Kadish classification: (A) confined to the nasal cavity, (B) extension to the paranasal sinuses, (C) extending beyond these limits.

Treatment is generally surgical, but when necessary because of poor general health, distant metastases, or extensive local disease, chemotherapy and radiation may be provided in a palliative fashion. Chemotherapeutic agents such as cisplatin, vincristine, and etoposide have shown efficacy when combined with radiation. Generally radiation regimens consist of 40 to 60 Gy over 4 to 6 weeks.

Surgical Management

Wide resection of the tumor is usually achieved through anterior craniofacial resection, with anterior skull base resection and reconstruction as necessary. Under selective circumstances, contemporary anterior endoscopic cranial base techniques may be employed for tumor extirpation. Postoperative radiation therapy is usually recommended unless the lesion is very small and isolated below the cribriform plate. Orbital exenteration should be undertaken when periorbital soft tissues are involved, with postoperative radiation and chemotherapy as adjuvant treatment. Endoscopic resection has been described in the literature, even in cases of stage C disease. Neck dissection for clinically positive neck disease is undertaken at the time of primary resection, but the role of elective neck dissection in N0 disease remains controversial.

Rehabilitation and Follow-up

Postoperative complications include those associated with sinus and skull base surgeries, such as infection, cerebrospinal leak, meningitis, pneumocephalus, and seizures. Nasal crusting may be a significant problem that requires daily maintenance. Long-term follow-up surveillance for recurrence requires regular office evaluation and radiologic study. Average reported survival rates based upon Kadish stages A, B, and C are 72%, 59%, and 47%, respectively. Recurrence is most often local and may not occur until many years after initial therapy.

Questions

1. Symptoms that might be consistent with a neoplastic process in the nose include:
A. Unilateral nasal obstruction
B. Epistaxis
C. Purulent nasal discharge
D. Unilateral facial pain
E. All of the above

2. Physical findings that raise concern for a neoplastic process in the nose include:
- **A.** Unilateral nasal mass
- **B.** Generalized turbinate congestion
- **C.** Purulent nasal discharge
- **D.** Generalized nasal crusting
- **E.** All of the above

3. The lesion described in this case would be Kadish classification stage
- **A.** A
- **B.** B
- **C.** C

Suggested Readings

Castelnuovo PG, Delù G, Sberze F, et al. Esthesioneuroblastoma: endonasal endoscopic treatment. Skull Base 2006;16(1):25–30

Devaiah AK, Larsen C, Tawfik O, O'Boynick P, Hoover LA. Esthesioneuroblastoma: endoscopic nasal and anterior craniotomy resection. Laryngoscope 2003;113(12): 2086–2090

Turakhia S, Patel K. Esthesioneuroblastoma. Indian J Radiol Imag 2006;16(4):669–672

50

Sphenoid Sinus Mucocele

Angela Donaldson and Lee A. Zimmer

History

A 46-year-old black woman arrived at the clinic with a 3-month history of headaches and visual changes. The patient described the headaches as localized to the left frontal region, worsened by lying supine, and constant in nature. The patient denied photophobia, nausea, and vomiting and had no history of migraines. She also complained of diplopia, which had been present for the past month. The diplopia improved when she turned her head to the left. An eye examination 3 months earlier had shown 20/20 vision bilaterally, and she had no current complaints. For the past 2 weeks she had been experiencing pain behind her left eye. The patient denied recent trauma, vertigo, nausea, vomiting, or weight loss.

The patient's medical history includes hypertension, allergic rhinitis, and chronic sinusitis. She works for an interior design firm. Patient denied the use of tobacco, alcohol, and illicit drugs. Family history is noncontributory.

On physical examination, the patient appears well. Pupils are equal, round, and reactive to light. The left eye has mobility superiorly, medially, and inferiorly, but it does not move laterally. Extraocular movement is intact on the right. There is mild swelling of the inferior tuberinates bilaterally and no septal deviation or polyps. Nasal endoscopy showed a smooth cystic swelling in the area of the sphenoid with no focal ulcerations.

Differential Diagnosis—Key Points

1. Taking a thorough history regarding the patient's headache is important to the diagnosis. The differential diagnosis of headaches includes, but is not limited to, tension headache, cluster headache, migraine, meningitis, intracranial mass, sinusitis, fungus ball, and temporomandibular joint (TMJ) syndrome. This patient describes her headaches as retro-orbital and worse when lying in the supine position. The patient also denied aura symptoms and has no family history of migraine, therefore making this diagnosis of migraine less likely. Because there is no history of trauma and the headaches have been present for an extended period, the concern for a space-occupying lesion should be high on the differential diagnosis.
2. The symptoms of diplopia and decreased lateral eye movement are consistent with cranial nerve (CN) VI palsy. Patients with this type of palsy tend to turn their head

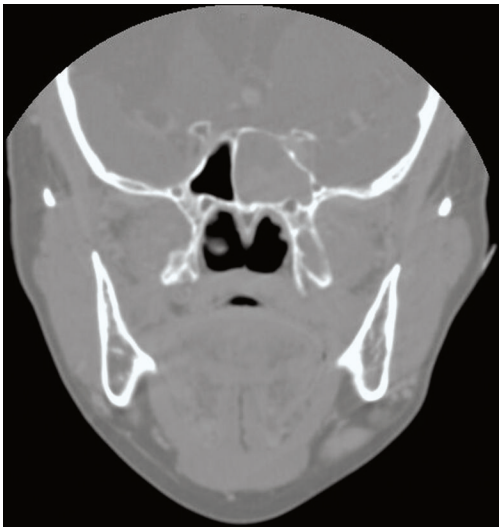

Fig. 50.1 Coronal computed tomography image showing complete opacification of the left sphenoid sinus with a mucocele.

toward the side of the lesion to reduce diplopia. Given the location of CN VI just lateral to the sphenoid sinus, clinicians should evaluate for injury to surrounding structures such as CNs III, IV, and V, which are all located in this area. With the concern for a space-occupying lesion and evidence of CN VI involvement, tension headache, cluster headache, sinusitis, and TMJ syndrome are unlikely causes. Meningitis can cause CN VI palsy, but this patient does not exhibit other signs and symptoms of meningitis, such as fever, neck rigidity, and altered mental status.

3. Imaging studies are crucial to this diagnosis. The computed tomography (CT) maxillofacial scan showed a homogeneous, nonenhancing mass that completely filled the sphenoid sinus cavity (**Fig. 50.1**).

◆ Test Interpretation

Nasal endoscopy typically shows smooth cystic swelling in the sphenoid area with no ulcerations. A CT maxillofacial scan is ordered to look for bone erosion. If a space-occupying lesion is noted on CT, magnetic resonance imaging (MRI) is performed. The CT characteristics of mucocele include a homogeneous, nonenhancing, expansile sinus mass that completely fills the potential sinus cavity expanding or remodeling surrounding bone margins. MRI is superior for demonstrating the connection of the mucocele with surrounding cranial structures. MRI intensity varies depending on the contents of the mucocele. Mucoceles usually have low-signal intensity on T1-weighted images and high signal intensity on T2-weighted images.

◆ Diagnosis

Sphenoid sinus mucocele

◆ Medical Management

Sphenoid sinus mucoceles are managed through a surgical approach. Evaluation by medicine or anesthesia may be warranted if the patient has significant co-morbidities. Medical management for this patient would include intraoperative intravenous fluids and prophylactic antibiotics.

◆ Surgical Management

The gold standard for surgical treatment is transnasal endoscopic sphenoidotomy with mucocele marsupialization. The procedure allows the best visualization of the sphenoid sinus and grants better restoration of normal sinus function.

◆ Rehabilitation and Follow-up

Many case studies report that headache and visual changes resolve immediately after sphenoidotomy. Physicians may choose to get repeat MRI after surgery and others follow the patient clinically. Currently no published studies address the need for postoperative imaging.

Questions

1. Which of the following describes the histologic pathology of sphenoid sinus mucocele?
   - **A.** Transformation of ciliated columnar cells to cuboidal
   - **B.** Transformation of columnar cells to cuboidal
   - **C.** Transformation of cuboidal cells to columnar cells
   - **D.** Transformation of cuboidal cells to squamous
2. All the signs and symptoms listed below are associated with sphenoid sinus mucocele except:
   - **A.** Blindness
   - **B.** Ptosis
   - **C.** Sudden onset of anosmia
   - **D.** Headaches that are worse when the patient is in the supine position
3. Which of the following statements is most true regarding sphenoid sinus mucocele?
   - **A.** Lateral and upward is the common extension of the mucocele.
   - **B.** Sphenoid sinus mucocele is most likely to extend anterior because of lower resistance.
   - **C.** Sphenoid sinus mucocele is the most common mucocele of the paranasal sinuses.
   - **D.** Medical management is first-line therapy for sphenoid sinus mucocele.
4. Sphenoid sinus mucocele routes of extension include which of the following? (Check all that apply.)
   - **A.** Upward extension toward the floor of the sella
   - **B.** Anterior extension toward the ethmoid cells
   - **C.** Lateral and upward extension toward the cavernous portion of the carotid artery
   - **D.** All of the above

Suggested Readings

Friedman A, Batra PS, Fakhri S, Citardi MJ, Lanza DC. Isolated sphenoid sinus disease: etiology and management. Otolaryngol Head Neck Surg 2005;133(4):544–550

Giovannetti F, Filiaci F, Ramieri V, Ungari C. Isolated sphenoid sinus mucocele: etiology and management. J Craniofac Surg 2008;19(5):1381–1384

Lawson W, Reino AJ. Isolated sphenoid sinus disease: an analysis of 132 cases. Laryngoscope 1997;107(12 Pt 1): 1590–1595.

Metson R, Gliklich RE. Endoscopic treatment of sphenoid sinus disease: report of 39 cases. Otolaryngol Head Neck Surg 1996;114:736–744.

Sethi DS. Isolated sphenoid lesions: diagnosis and management. Otolaryngol Head Neck Surg 1999;120(5): 730–736.

51

Frontoethmoidal Mucocele

Maria V. Suurna and Lee A. Zimmer

History

A 78-year-old man complains of diplopia, frequent headaches, and swelling of the right eye for 3 months. The patient underwent functional endoscopic sinus surgery for chronic sinusitis several years ago.

The head and neck examination notes swelling over the right side of the forehead and right upper eyelid. The right eye has mild proptosis, inferior-lateral displacement, and decreased extraocular mobility. Nasal endoscopy reveals bilateral scarring of the middle meatus.

Differential Diagnosis—Key Points

1. Mucocele is a chronic cystic mucus-filled lesion of the paranasal sinuses secondary to obstruction of sinus outflow tracts. The outflow obstruction leads to the development of mucoceles. This may be a result of trauma, chronic sinusitis, sinonasal polyposis, neoplasms, or sinus surgery. Mucoceles may expand over a period of many years before becoming symptomatic. The mucocele may become infected, forming a mucopyocele. Frontoethmoidal mucocele is the most common paranasal sinus mucocele. Patients often present with frontal headache, proptosis, and diplopia secondary to downward and outward displacement of the globe.
2. Osteoma is the most common tumor of the paranasal sinuses and is commonly found in the frontoethmoidal region. Presenting symptoms are related to slow expansion of the lesion leading to impingement and displacement of normal structures. Other fibro-osseous and bony lesions such as fibrous dysplasia and ossifying fibroma should also be considered in the differential diagnosis.
3. Inverted papilloma is a benign tumor of the paranasal sinuses characterized by inverted growth pattern. There is about a 10% risk of associated squamous cell carcinoma.
4. Other benign neoplasms include meningiomas, schwannomas, neurofibromas, hamartomas, hemangiomas, and chordomas.
5. The Pott puffy tumor is a subperiosteal abscess of the frontal bone associated with frontal osteomyelitis. It can result from acute and chronic frontal sinusitis and often manifests with localized swelling overlying the forehead.
6. Invasive fungal sinusitis results from the rapid spread of fungal infection from the paranasal sinuses invading the surrounding structures and is most commonly seen in immunocompromised individuals. Diagnosis

requires histopathologic evidence of fungal invasion of mucosa, submucosa, blood vessels, or bone.

7. Malignant neoplasms such as squamous cell carcinoma, adenocarcinoma, adenoid cystic carcinoma, melanoma, and sarcoma should be considered as part of the differential diagnosis. The malignant tumors of frontal and ethmoid sinuses often present with evidence of an advanced disease.

◆ Test Interpretation

A computed tomography (CT) scan in the axial and coronal planes is the preferred imaging modality for evaluating mucoceles. They appear as hypodense, nonenhancing masses filling the sinus cavity. Imaging often reveals bone expansion and remodeling that result from pressure within the sinus cavity as a result of mucocele formation.

Magnetic resonance imaging is used to confirm the diagnosis. Mucoceles are usually hypointense on T1-weighted images and hyperintense on T2-weighted images. However, signal intensity can vary depending on water or blood content within the mucocele. Gadolinium-contrasted T1-weighted images show no enhancement of the mucocele.

In the present case, a CT scan of the sinuses was obtained. The study revealed a large expansile right frontal mass with erosion into the orbital plate and the anterior and posterior frontal sinus table (**Fig. 51.1**).

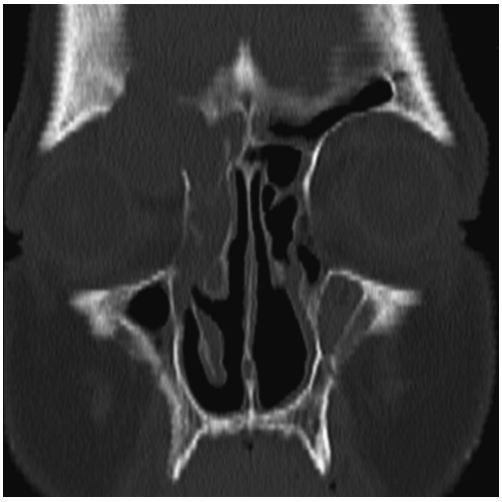

Fig. 51.1 Frontoethmoidal mucocele. Computed tomography scan, in coronal view, demonstrating opacification of right frontal and ethmoidal sinuses.

◆ Diagnosis

Frontoethmoidal mucocele

◆ Medical Management

None

◆ Surgical Management

The recommended treatment of frontoethmoidal mucocele is surgery. The osteoplastic flap with fat obliteration has been the gold standard for management of frontal sinus mucoceles in the past. With technological advancements, endoscopic drainage and marsupialization procedures have mostly replaced the osteoplastic flap technique, with reports of low morbidity and low recurrence rates.

In the present case, the patient underwent endoscopic image–guided right frontal sinusotomy with marsupialization of the frontoethmoidal mucocele. Following the procedure the patient reported resolution of ocular symptoms and had no evidence of recurrence 2 years later.

◆ Rehabilitation and Follow-up

Even though mucoceles of the paranasal sinuses are treated by transnasal endoscopic surgery, external approaches with or without obliteration of the sinuses are still used. Published data suggest a low recurrence rate of mucocele following either surgical modality. However, because of the nature and pathophysiology of the disease, patients should have long-term follow-up to monitor for recurrence.

Questions

1. What is the most likely cause for frontoethmoidal mucocele formation?
   - **A.** Fracture of the anterior frontal sinus table
   - **B.** Obstructed frontal sinus outflow tract
   - **C.** Acute sinusitis
   - **D.** Sinonasal polyposis
2. What is a goal of endoscopic surgical treatment of frontoethmoidal mucocele?
   - **A.** Reestablishment of sinus drainage
   - **B.** Complete removal of the sinus mucocele
   - **C.** Frontal sinus obliteration
   - **D.** Easier access to the frontal sinus
3. The patient underwent endoscopic marsupialization of frontoethmoidal mucocele. On postoperative day 1, he complains of a headache and clear nasal drainage. What is the likely cause?
   - **A.** Acute sinusitis
   - **B.** Recurrent mucocele
   - **C.** Cerebrospinal fluid leak
   - **D.** Normal postoperative course
4. The predisposing factors for mucocele formation are all except which of the following?
   - **A.** Prior sinus surgery
   - **B.** Osteoma
   - **C.** Family history
   - **D.** Facial trauma

Suggested Readings

Har-El G. Endoscopic management of 108 sinus mucoceles. Laryngoscope 2001;111(12):2131–2134

Herndon M, McMains KC, Kountakis SE. Presentation and management of extensive fronto-orbital-ethmoid mucoceles. Am J Otolaryngol 2007;28(3):145–147

Khong JJ, Malhotra R, Selva D, Wormald PJ. Efficacy of endoscopic sinus surgery for paranasal sinus mucocele including modified endoscopic Lothrop procedure for frontal sinus mucocele. J Laryngol Otol 2004;118(5):352–356

Lloyd G, Lund VJ, Savy L, Howard D. Optimum imaging for mucoceles. J Laryngol Otol 2000;114(3):233–236

XI

Laryngology

52

Unilateral Vocal Fold Paralysis

Sid M. Khosla

History

A 70-year-old man has shown varying degrees of dysphonia since his aortic aneurysm repair 9 years ago. He reports having a thyroplasty type I 8 years ago and again 3 years ago. He reports that the medialization operations helped his voice "a little," but neither operation improved the loudness of his voice. He also reports inspiratory stridor on exertion that developed a few months after his aneurysm repair. Both medialization operations had no effect on his stridor. His voice is presently soft and breathy. He has no aspiration.

On physical exam, he is found to have an immobile left vocal cord. The left vocal process is significantly anterior and inferior, relative to the normal right side. On inspiration, it is seen that both the arytenoid mucosa and the right vocal fold move toward the intraglottal space, producing significant collapse of the airway (**Fig. 52.1**). This is not seen during expiration. There is a moderate vocal fold gap, and his voice is breathy.

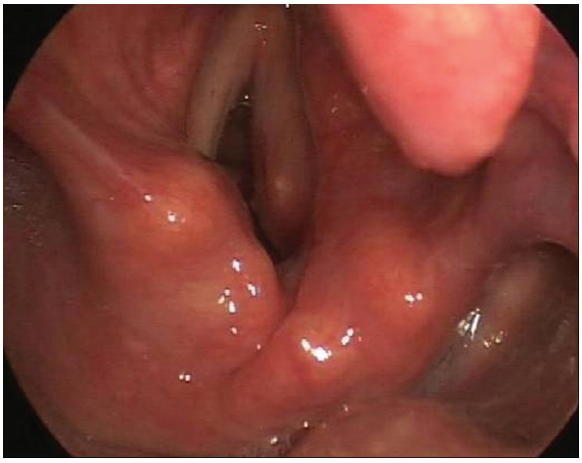

Fig. 52.1 Left vocal fold paralysis with the left arytenoid prolapsed anteriorly.

Differential Diagnosis—Key Points

Left vocal fold paralysis secondary to aortic aneurysm repair: In addition to a vocal fold gap, there is an asymmetry in vocal fold length, height, and tension. There is also a dynamic airway obstruction.

1. Unilateral vocal fold paralysis (UVFP) usually presents with a glottal gap; this gap may increase in the first 6 months if the thyroarytenoid muscle atrophies, or it may improve if there is some reinnervation. If there is reinnervation, this may increase tone and bulk of the vocal fold; but, because of synkenesis, there may not be any movement. Many symptoms are due to a membranous vocal fold gap, such as a breathy, soft voice; decreased phonation time, decreased ability to cough. If the reinnervation is minimal, the position of the arytenoid will also change. For example, if there is minimal tone in the posterior cricoarytenoid, the arytenoid can tilt anteriorly, producing a vocal process that is anterior and inferior to the normal vocal process. In addition to atrophy of the vocalis and thyroarytenoid, this abnormal positioning results in less tension in the left fold relative to the right. This asymmetric tension may produce an abnormal voice.
2. In general, stridor is not seen in UVFP, but it can be seen in bilateral paralysis. In this case however, the anterior-inferior position of the arytenoid allows it to be close to the mucosa of the right fold, and the surfaces are close enough that negative pressures, owing to Bernoulli's law, cause a dynamic collapse during inspiration. In Bernoulli's law, the negative pressures increase as the air velocity increases. Thus, if a person is breathing harder, the collapse will worsen.
3. Cricoarytenoid fixation may need to be ruled out. Arytenoid dislocations can sometimes be recognized by the position of the arytenoid. Trauma, previous surgery, or diseases such as rheumatoid arthritis can also cause fixation. A good history and examination will help determine when this diagnosis needs to be worked up. To check definitively for fixation, the joint must be palpated.

Test Interpretation

1. Computed tomography (CT): In this case, it is assumed that the repair of the aortic aneurysm injured the recurrent laryngeal nerve as it loops around the aortic arch. If the cause is not known, imaging needs to be done along the course of the nerve. A CT scan of the neck (skull base to superior mediastinum) is indicated. A full chest CT scan is not needed so long as the neck CT extends to the aortic arch. Some physicians still use a chest radiograph to image the lungs; a head, neck, and chest CT is done down to the aortic arch on the left but is not needed on the right.
2. Video stroboscopy or flexible nasolaryngoscopy: A recorded examination helps with diagnosis, follow-up, and surgical planning, if needed. If the folds are vibrating, video stroboscopy is necessary to evaluate vibrations.
3. Maximum phonation time (MPT): This measurement can easily be taken in the clinic. Although MPT can vary depending on the patient's effort, it is a good general measure of air leakage and can be used to monitor progress. This patient had an MPT of 5 seconds, which is very low.
4. Patient assessment of the effect of his or her voice problems: Questionnaires such as the Voice Handicap Index (Hogikyan) can be used for this purpose.

The following tests can be done but are not necessary.

5. Laryngeal electromyelography (EMG): EMG shows spontaneous activity without any voluntary motor units. Return of function is unlikely. In general, EMG can help predict whether recover will occur. In addition it will help differentiate between recurrent laryngeal nerve injury and cricoarytenoid fixation.
6. Acoustic tests: Acoustic tests will reveal a high amount of broadband noise, which will be reflected by a low signal-to-noise ratio, a

low harmonic-to-noise ratio, or a low cepstral peak prominence.

7. Aerodynamic tests. These tests will demonstrate leakage of air during phonation.

Diagnosis

Left vocal fold paralysis

Medical Management

The management of vocal cord paralysis is voice therapy, which can be used to add bulk to the paralyzed fold and help to avoid compensatory habits, such as increased muscle tension or false fold phonation. Many patients with unilateral vocal cord paralysis do well with voice therapy alone, provided the paralyzed vocal fold assumes a median or paramedian position.

Surgical Management

Management of vocal fold paralysis is based on the length of the paralysis and whether the patient has significant voice or aspiration issues. Treatment is divided into short-term or long-term solutions. Long-term solutions should not be attempted before 6 to 12 months after the onset of paralysis.

Short-term solutions involve injecting an absorbable substance into the paraglottic space, known as an *injection laryngoplasty*. This is often done in the clinic and can be performed through four approaches: transoral, just superior to the thyroid notch, through the cricoid cartilage, or through the cricothyroid membrane. A variety of substances are currently being injected, such as fat, acellular cadaveric dermis (Cymetra, LifeCell Corp., Branchburg NJ), and calcium hydroxapatite (Radiesse, BioForm Medical, San Mateo, CA).

Long-term solutions include a thyroplasty type 1, arytenoid adduction, adduction arytenopexy, and ansa cervicalis-to-recurrent laryngeal nerve reinnervation. Thyroplasty is the most commonly used procedure and will reduce or close the vocal fold gap. This procedure can be performed under local anesthesia usually with intravenous sedation. In addition, it can be reversible. Arytenoid adduction mimics the lateral cricoarytenoid and can be used to close a posterior gap and partially restore the prephonatory position of the vocal fold. Adduction artenopexy mimics the effect of many of the intrinsic muscles, including the intra-artytenoid; it is also used to stabilize the arytenoid in the anterior-posterior direction, much in the same way that the posterior cricoarytenoid does. Both these arytenoid procedures are done in addition to a thyroplasty type 1.

A reinnervation procedure involves an anastomosis between the recurrent laryngeal nerve and a branch of the ansa cervicalis. It does not produce movement but can result in good muscle tone and bulk and a midline position. This usually takes 3 to 6 months to achieve, so an injection laryngoplasty is often done.

In this case, a thyroplasty type 1 and an arytenoid adduction were done. The patient's stridor completely resolved, and he was very happy with his voice results.

Rehabilitation and Follow-up

Following surgery, some voice therapy is often very helpful for two reasons. The first is that many patients develop compensatory techniques (such as false fold phonation) when the fold is lateralized, and voice therapy is needed for retraining. The second is to teach patients how to use their voice most efficiently (such as teaching proper breath support).

Questions

1. Which of the following will thyroplasty will most likely do?
   - **A.** Increase the maximum phonation time
   - **B.** Close the posterior gap
   - **C.** Reduce the amount of asymmetry in stiffness
   - **D.** Result in an improved airway

2. A patient has a right vocal fold paralysis. What is the most necessary testing?
- **A.** Neck CT
- **B.** Head and neck CT down to just below the clavicle
- **C.** Head and neck CT; CT of mediastinum to the level of aortic arch
- **D.** Chest radiograph

3. Treatment for unilateral paralysis includes all of the following except which one?
- **A.** Injection of the vocal fold
- **B.** Reinnervation of recurrent laryngeal nerve
- **C.** Arytenoid adduction
- **D.** Reinnervation of superior laryngeal nerve

Suggested Readings

Bastian RW, Richardson BE. Vocal fold paralysis: clinical evaluation of vocal fold paralysis. Otolaryngol Clin North Am 2004;37(1):45–59

Paniello RC. Vocal fold paralysis: laryngeal reinnervation. Otolaryngol Clin North Am 2004;37(1):161–181

Zeitels SM. Vocal fold medialization: injection and laryngeal framework surgery. In: Rubin JS, Sataloff, RT, Korovin, GS, eds. Diagnosis and Treatment of Voice Disorders. San Diego, CA: Plural Publishing; 2006: 713–726

53

Spasmodic Dysphonia

Lisa Lee and Keith M. Wilson

History

A 46-year-old white woman has had chronic hoarseness of several years' duration. She describes her voice as breaking often and "going in and out." The patient first noticed the hoarseness after a sore throat during a particularly stressful time in her life. Her voice is worse when talking on the phone and during stressful situations. Intermittently her voice cannot be understood by others. She notes that her voice gets better after an alcoholic beverage. Her voice causes her great personal distress. Her medical history is remarkable only for mild anxiety. She denies smoking, heartburn, hemoptysis, throat clearing, and allergy problems. She has seen several health professionals about her chronic hoarseness, including her primary care physician, two otolaryngologists, a psychiatrist, and a speech therapist. Prior treatments included speech therapy, antibiotics, and reflux medication with minimal improvement.

During the conversation, the patient's voice is noted to have a strained and strangled quality with abrupt cessation and return of phonation. At times there is a notable tremulousness to her voice. Although she cannot count to 10 without intermittent stoppages, she is able to sing her favorite song smoothly. Her head and neck examination is otherwise unremarkable; there is no evidence of abnormal tongue or palate mobility. Her neurologic examination is notable only for a fine hand tremor. Flexible laryngoscopy shows normal-appearing, fully mobile vocal cords without any masses or sulci. No evidence of supraglottic erythema or edema is seen. During speech, the patient's vocal cords abruptly adduct forcefully corresponding to phonatory stoppages.

Differential Diagnosis—Key Points

1. Spasmodic dysphonia (SD). This neurologic disorder most often manifests in the larynx as a strangled vocal quality with abrupt phonatory stoppages associated with a "choppy" voice. The three forms of laryngeal dystonia consist of adductor SD, abductor SD, and mixed SD. *Adductor SD* is the most common form and is characterized by a strained voice quality with breaks in vocalization. Abductor SD is characterized by a breathy voice quality with intermittent breaks in vocalization. Mixed SD has characteristics of both adductor and abductor laryngeal dystonias. Although many SD patients have a family history of dystonias, which may be associated with chromosome 9, the cause of the underlying SD is largely unknown. SD affects more women than men, with age at presentation usually during the 40s or 50s. Occasionally, there is an associated hand tremor. However, it is

important to distinguish between essential tremor and SD. Unfortunately, patients with SD are often undiagnosed or misdiagnosed for several months to years.

2. Essential laryngeal tremor. Essential tremor causes shaking of the head, hands, and voice. The tremor is typically not present at rest and is worse with emotional stress or fatigue. Essential laryngeal tremor manifests as a regular, 4- to 12-Hz tremor seen in the vocal cords during speech and respiration. In contrast, the tremor found in SD is irregular. Vocal tremor can be the only symptom in some patients. The vocal tremor represents rhythmic laryngeal movement, presenting as rhythmic pitch and loudness alterations during speech. Essential tremor can be seen concomitantly with SD.
3. Muscle tension dysphonia. Affected individuals are usually unable to sing or whisper without voice breaks. The dysphonia is worse under stressful situations. Unlike SD, intraword phonatory breaks are infrequent. Patients are more likely to exhibit excessive contraction of the intrinsic and extrinsic laryngeal muscles, with an overadduction of the true or false vocal cords or even the supraglottis. Muscle tension dysphonia is most commonly treated with voice therapy. Occasionally botulinum toxin type A (Botox) is used in conjunction with voice therapy to release the abnormal muscle activation patterns.
4. Neurologic disorders. Although not likely to be the initial or sole manifestation, systematic neurologic diseases must be considered during the initial evaluation. Movement disorders such as Parkinson disease, Huntington disease, tardive dyskinesia, and cerebellar disorders need to be excluded with a complete neurologic examination. Other neurologic diseases to consider include myoclonus and Meige syndrome. Oculopalatal myoclonus presents as an involuntary movement of the head and neck region, including the soft palate, pharynx, larynx, and eyes. Laryngeal involvement presents as broken speech. Indirect laryngoscopy would show that the vocal cords have a slow, rhythmic adduction and abduction at the same frequency as the other myoclonic tics. Meige syndrome consists of myoclonic spasms of the eyelids, pharynx, tongue, floor of the mouth, and larynx.

◆ Test Interpretation

When no overt laryngeal pathology is found, physiologic tests are indicated to further investigate the cause of the hoarseness.

Speech Evaluation

The patient's voice is notable for intermittent glottal stops and pitch breaks. These episodes are associated with excessive and untimely contraction of the adductor laryngeal muscles, which account for the strained vocal quality and abrupt stoppages as the glottis squeezes shut. The muscle largely responsible for adductor SD is the thyroarytenoid muscle. The patient is also unable to carry a constant pitch and exhibits reduced voice loudness. The voice quality may be monotonal with glottal fry and vocal tremor. The patient would be able to read sentences with words that alternate with little difficulty between voiceless consonants and vowels. She would have severe difficulty reading sentences that have voiced consonants followed by vowels. The glottal breaks are intraword and may be elicited with sentences such as, "I eat apples and eggs" or "The dog has a new bone." Interestingly, the patient is able to whisper or sing without the usual strangled vocal quality or glottal breaks. She is also able to laugh or say phrases just fine when using a falsetto voice.

Abductor SD is characterized by incomplete glottic closure and abrupt glottal opening, which account for the breathy vocal quality and abrupt stoppages. The vocal cords at these times abruptly hyerabduct. Laryngeal tremor may be noted. The posterior cricoarytenoid (PCA) is largely responsible for abductor spasmodic dysphonia. The quality of the abductor SD may be best appreciated and elicited with sentences that alternate between voiceless consonants (h, f, l, p, s, t, th) and vowels. Such spasm trigger sentences include "How high is Harry's hat?" and "Did he go to the right or to the left?"

Acoustic Analysis

Aperiodic segments primarily characterized the phonation of adductor SD, followed by frequency shifts and phonatory breaks. The location of each of these acoustic events was

within the midportion of the vowel production. SD patients also exhibit high jitter and shimmer. Increased perturbations and a reduced signal-to-noise ratio are seen in both spasmodic dysphonia and essential tremor. Unlike essential tremor patients, tremor analysis does not reveal a laryngeal tremor in SD patients. Also, the signal-to-noise ratio is even lower with abductor spasmodic dysphonia because of periods of voicelessness. SD characterized as a "task-specific" laryngeal dystonia; that is, the severity of dysphonia varies depending on the demands of the vocal task. Voice produced in connected speech as compared with sustained vowels is said to provoke more frequent and severe laryngeal spasms. Reduced dysphonia severity during sustained vowels supports task specificity in adductor SD but not muscle tension dysphonia.

Electromyography

Although electromyography (EMG) is not necessary for the diagnosis of spasmodic dysphonia, it does provide important information to guide therapy. During phonation, EMG would show bursts of electrical activity at rest and enlarged motor unit action potentials in the affected muscle(s). Botulinum toxin injections can be directed to specific muscles using EMG guidance. EMG shows abnormally high resting potential in the thyroarytenoid muscle. There is imbalance between the thyroarytenoid and cricothyroid musculature that contributes to the increased tension in the laryngeal anteroposterior dimension.

◆ Diagnosis

Adductor spasmodic dysphonia

◆ Medical Management

Botulinum toxin type A (Botox) is used to denervate laryngeal muscles chemically by blocking the release of presynaptic acetylcholine at the neuromuscular junction. EMG-guided injection of Botox is used widely for the treatment for SD. A monopolar, hollow-bored Teflon-coated EMG needle connected to an EMG recorder is used to inject botulinum toxin. For adductor SD, botulinum toxin is injected transorally or percutaneously into the thyroarytenoid muscle. During percutaneous injection, the needle is passed through the cricothyroid membrane just off midline and directed just lateral to each thyroarytenoid muscle. For abductor SD, botulinum toxin is injected into the PCA muscle. Abductor injection requires grasping the larynx and rotating it to expose the PCA. The patient is asked to sniff, which causes abduction as the EMG indicates action potentials. The effective dose of botulinum toxin to achieve the desired vocal results varies from one individual to another and thus requires titration. The dose can vary from 1 mouse unit to up to 10 mouse units to one or both muscles. Some practitioners stage PCA botulinum toxin injections by 2 weeks after confirming that there is adequate abduction of the previously injected vocal cord. After injection, botulinum toxin can take up to 72 hours to take effect. Patients will note an improvement in their voice within this period. Although the voice is not totally normal, patients are generally satisfied with the improvement in speech following treatment. Side effects include temporary breathiness, possibly mild aspiration, and possibly mild dysphagia for up to 7 days after initial injection. The effects of botulinum toxin may last up to 6 months. Individual titration of botulinum toxin and repeated injections are necessary.

Voice therapy has been used with little success to treat SD primarily. Most techniques attempt to reduce the degree of vocal tightness and the incidence of voice breaks. Biofeedback, inverse phonation (speaking during inspiration), and identification and correction of dysfunctional vocal habits have all been used with low levels of success.

◆ Surgical Management

A variety of surgical techniques have been used to treat SD. Interventions include recurrent laryngeal nerve denervation alone, selective recurrent laryngeal nerve denervation and reinnervation, thyroplasty type II, and intrinsic laryngeal muscle myectomies. Alhough recurrent laryngeal nerve section had initially favorable results, long-term results showed recurrence of SD symptoms. More recently,

recurrent laryngeal nerve denervation and reinnervation technique have been used for SD. One method involves the denervation of the adductor branch of the recurrent laryngeal nerve, followed by reanastomosis of distal branches to the ansa cervicalis. Complications included moderate to severe breathiness. Variations in thyroarytenoid myectomies have been applied to adductor SD using microlaryngeal techniques and radiofrequency myothermy. Although current surgical endeavors may be promising for the amelioration of SD symptoms, the current standard of care consists of botulinum toxin injection to the affected vocal muscles with good results and improved quality of life.

Rehabilitation and Follow-up

Patients with SD would require repeat botulinum toxin injections every 3 to 6 months. They may participate in voice therapy to address any negative compensatory vocal behavior, but it generally plays a minor role in SD treatment.

Questions

1. A 56-year-old woman has chronic hoarseness of several months' duration. Although she has a slight hand tremor, the remainder of her neurologic examination is normal. What finding would most likely suggest muscle tension dysphonia?
- **A.** She whispers without voice breaks.
- **B.** Occasional vocal tremor is detected.
- **C.** Spectral analysis shows frequent intraword phonatory breaks.
- **D.** Laryngoscopy shows adduction of false vocal cords and supraglottic squeeze.
- **E.** Severity of phonation is not task dependent.

2. A 59-year-old woman has chronic hoarseness of several months' duration. Her voice is not raspy or breathy but does have a tremulous quality. On indirect laryngoscopy, her vocal cords are mobile, although mild bowing is present. What finding would most likely exclude a diagnosis of essential tremor?
- **A.** Absence of a hand tremor
- **B.** 12-Hz regular laryngeal tremor
- **C.** Acoustic analysis shows reduced signal-to-noise ratio.
- **D.** Dysphonia worsens during stressful situations.
- **E.** Tremor analysis does not show a laryngeal tremor.

3. A patient with abductor spasmodic dysphonia would most likely have a pronounced problem phonating which of the following sentences without breaks?
- **A.** "I eat apples and eggs."
- **B.** "The dog has a new bone."
- **C.** "Did he go to the right or to the left?"
- **D.** "I went to the store."
- **E.** "Mary has been there, too."

4. The injection of botulinum toxin type A works to improve phonation in patients with spasmodic dysphonia by doing what?
- **A.** Paralyzing the recurrent laryngeal nerve
- **B.** Competitively blocking acetylcholine at end motor plates
- **C.** Inhibiting the release of presynaptic acetylcholine
- **D.** Producing an immunologic reaction causing laryngeal muscle scarring
- **E.** Inhibiting proximal neuronal pathways to the basal ganglion

5. Which intervention has not been used to treat spasmodic dysphonia?
- **A.** Recurrent laryngeal nerve denervation
- **B.** Thyroplasty type II
- **C.** Injection laryngoplasty
- **D.** Thyroarytenoid myotomy
- **E.** Botulinum toxin type A injection

Suggested Readings

Casserly P, Timon C. Botulinum toxin A injection under electromyographic guidance for treatment of spasmodic dysphonia. J Laryngol Otol 2008;122(1):52–56

Chan SW, Baxter M, Oates J, Yorston A. Long-term results of type II thyroplasty for adductor spasmodic dysphonia. Laryngoscope 2004;114(9):1604–1608

Chang CY, Chabot P, Thomas JP. Relationship of botulinum dosage to duration of side effects and normal voice in adductor spasmodic dysphonia. Otolaryngol Head Neck Surg 2007;136(6):894–899

Chhetri DK, Berke GS. Treatment of adductor spasmodic dysphonia with selective laryngeal adductor denervation and reinnervation surgery. Otolaryngol Clin North Am 2006;39(1):101–109

Grillone GA, Chan T. Laryngeal dystonia. Otolaryngol Clin North Am 2006;39(1):87–100

Merati AL, Heman-Ackah YD, Abaza M, Altman KW, Sulica L, Belamowicz S. Common movement disorders affecting the larynx: a report from the neurolaryngology committee of the AAO-HNS. Otolaryngol Head Neck Surg 2005;133(5):654–665

Nakamura K, Muta H, Watanabe Y, Mochizuki R, Yoshida T, Suzuki M. Surgical treatment for adductor spasmodic dysphonia: efficacy of bilateral thyroarytenoid myectomy under microlaryngoscopy. Acta Otolaryngol 2008;128(12):1348–1353

Rees CJ, Blalock PD, Kemp SE, Halum SL, Koufman JA. Differentiation of adductor-type spasmodic dysphonia from muscle tension dysphonia by spectral analysis. Otolaryngol Head Neck Surg 2007;137(4):576–581

Roy N, Gouse M, Mauszycki SC, Merrill RM, Smith ME. Task specificity in adductor spasmodic dysphonia versus muscle tension dysphonia. Laryngoscope 2005;115(2):311–316

Roy N, Whitchurch M, Merrill RM, Houtz D, Smith ME. Differential diagnosis of adductor spasmodic dysphonia and muscle tension dysphonia using phonatory break analysis. Laryngoscope 2008;118(12):2245–2253

Sapienza CM, Walton S, Murry T. Acoustic variations in adductor spasmodic dysphonia as a function of speech task. J Speech Lang Hear Res 1999;42(1):127–140

54

Laryngopharyngeal Reflux

Keith M. Wilson

History

A 55-year-old man complains of "chronic sinus drainage" for many years. He also complains of a need to clear his throat often to clear thick phlegm. On further questioning, he reports periodic soreness of his throat, intermittent hoarseness, and the sensation of a lump in the throat. He denies hemoptysis, rhinorrhea, and weight loss. He currently smokes one pack of cigarettes per day and has done so for 35 years.

On physical examination, he is an overweight male with a mildly raspy voice. His tonsils are atrophic. Flexible nasolaryngoscopy reveals moderately edematous vocal folds, edema and erythema of the arytenoid mucosa, and thickening at the posterior commissure (**Fig. 54.1**). The rest of the head and neck examination is unremarkable.

Differential Diagnosis—Key Points

1. The chronic nature of this patient's complaints points to an insidious process. Hoarseness, mucus accumulation in the throat (postnasal drip), sore throat, and lump in the throat sensation (globus pharyngeus) are vague symptoms frequently associated with laryngopharyngeal reflux (LPR).

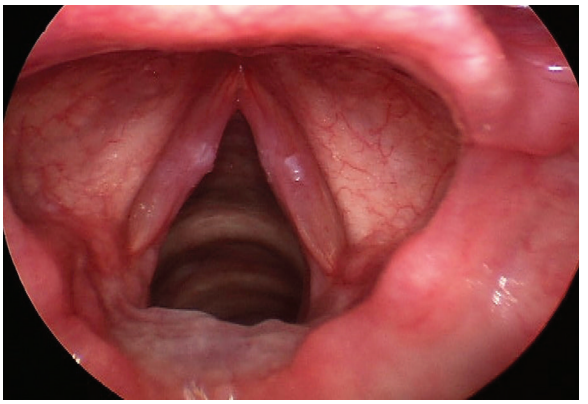

Fig. 54.1 Video stroboscopic view of the endolarynx reveals mid-to-moderate edema of the true vocal folds with erythema and edema of the arytenoid mucosa. Mild thickening (pachyderma) of the posterior commissure is also seen.

Additional symptoms of LPR include regurgitation of acid contents into the throat causing choking, burning and laryngospasm, asthma, cervical dysphagia, excess salivation, and repeated swallowing. Chronic nonproductive cough and halitosis are frequent complaints. Otalgia may be described and is thought to be mediated by the vagus nerve.

2. On laryngoscopy, the physical presentations of LPR are protean and consist of the following:
   - **A.** Erythema and edema of the arytenoid
   - **B.** Pachydermia of the posterior commissure
   - **C.** Reinke edema with or without significant erythema
   - **D.** Diffuse erythema with friable mucosa of the supraglottis
   - **E.** Vocal process granulomata
   - **F.** "Cobblestoning" of the pharyngeal mucosa
   - **G.** Vocal cord polyps with any of the above findings
   - **H.** Formation of contact ulcers
   - **I.** Pseudosulcus of the vocal folds
   - **J.** Glottis or subglottic stenosis
   - **K.** Paradoxical vocal fold movement
   - **L.** Laryngeal dysplasia or cancer
3. Classic symptoms of gastroesophageal reflux disease (GERD) include heartburn, a feeling of regurgitation, excessive belching, or intermittent substernal and epigastric chest pain. Only 20% of patients with LPR will complain of heartburn.
4. The same conditions that predispose to GERD also cause LPR. These conditions include obesity, straining, lying in a supine position, prolonged intubation, nasogastric tube placement, and foods that increase acid production such as fats, chocolate, carminatives (peppermint, spearmint, cinnamon, garlic, and onions), milk, alcoholic beverages, orange juice, and tomato juice.
5. Medications that have been associated with GERD include antidepressants, progesterone, β-blockers, calcium channel blockers, α-blockers, valium, and morphine. Nicotine and caffeine have a similar effect. These medications, and many of the abovementioned foods, induce reflux by reducing lower esophageal sphincter tone.
6. The complaint of globus raises concerns about pathology affecting the upper esophageal sphincter (cricopharyngeus). Globus pharyngeus is described as a choking sensation or lump in the throat. Edema of the mucosa in this area can present in this manner. Zenker diverticulum can also present with this complaint. In an immunocompromised patient, esophageal candidiasis should be considered.
7. In a patient with vague laryngopharyngeal complaints, other disease processes must be considered. The differential diagnostic list includes multiple sclerosis, amyotrophic lateral sclerosis, Gullain-Barré, diabetes, myasthenia gravis, scleroderma, esophageal or gastric carcinoma, caustic burn, cervical osteophytes, myoma or other benign tumor of the esophagus, esophageal web, or Plummer-Vinson syndrome and previous radiation.
8. Severe LPR can play a role in the development or exacerbation of subglottic stenosis, cricoarytenoid fixation, and cancer of the aerodigestive tract.

◆ Test Interpretation

The diagnosis of LPR is often based on the history and physical examination. Treatment with conservative or medical therapy is usually instituted. However, if the history or physical examination is not classic for the condition or if medical therapy fails and surgical therapy is considered, several tests may be useful in diagnosing LPR or providing information useful in targeting therapy.

1. Endoscopy. Endoscopic evaluation should include flexible or rigid laryngoscopy in all suspected cases. Endoscopic laryngeal biopsy in the office or formal direct laryngoscopy with biopsy in the operating room should be performed if there is suspicion of malignancy or if medical management fails to resolve a persistent suspicious abnormality of the larynx. Transnasal esophagoscopy (TNE) and esophagogastroduodenoscopy (EGD) are useful in detecting characteristic associated mucosal injury, esophagitis, and Barrett esophagus. It is important to inspect the esophagus because LPR symptoms were found to be more prevalent in patients with esophageal adenocarcinoma than were typical gastroesophageal symptoms and often represented the only sign of disease.

2. pH monitoring. Although it is considered the gold standard for GERD, pH monitoring is less reliable in confirming LPR. Variability in testing methods and lack of agreement on normative values have raised questions about the sensitivity of pH monitoring for detecting LPR. Placement of the upper probe 2 cm above the upper esophageal sphincter is considered critical for accuracy.
3. Ambulatory multichannel intraluminal impedance (MCII) and pH monitoring allow for a more complete description of reflux events. This approach allows for acid and nonacid reflux events to be identified as well as liquid (decreased impedance) and gaseous (increased impedance). This information is useful for targeting therapy. MCII can be combined with manometry to evaluate esophageal motility, sphincter competence, and bolus transport, possibly obviating barium esophagram.
4. Barium esophagram. This test identifies problems with esophageal motility, including nonprogressive (tertiary) contractions, increased amplitude and duration of contractions, and increased tone. Barium esophagram can also identify hiatal hernia, lumen integrity, aspiration, intrinsic and extrinsic masses. The patient's position can be modified during the test to look for reflux with change in position.

◆ Diagnosis

Laryngopharyngeal reflux

◆ Medical Management

Initial management of LPR should begin with lifestyle and dietary changes. Some lifestyle changes, which include weight reduction if applicable, smoking cessation, elevation of the head of bed (~6 inches) while sleeping, and avoidance of tight-fitting clothing. Dietary changes should restrict chocolate, caffeine, fats, citrus fruits, carbonated beverages, spicy tomato-based products, red wines, mints, and late-night meals.

The four categories of medications used to treat LPR are proton pump inhibitors (PPIs), histamine 2 (H_2) receptor antagonists, prokinetic agents, and mucosal cytoprotectants. A 4- to 6-week trial of an H_2 receptor inhibitor like ranitidine (Zantac), cimetine (Tagamet), or famotidine (Pepcid) can be tried as a first line of medical management. Milder forms of LPR may respond favorably. Prokinetic agents have never been shown to be effective in LPR but still may be beneficial in patients with gastroparesis or as part of a multidrug treatment strategy against LPR. Similarly, mucosal cytoprotectants, like sucralfate, may be helpful in an adjunctive role, protecting injured mucosa from harmful effects of pepsin and acid.

The mainstay of medial management for LPR is a 3-month empirical trial of a PPI. Twice-daily dosing is recommended, but many patients respond to once-a-day therapy. Best results are achieved when the medication is taken 30 to 60 minutes before a meal. It is important to inform the patient that the symptoms of LPR will resolve slowly to prevent premature discontinuation of therapy by the patient. Complete resolution of symptoms and physical findings often takes at least 6 months.

If no signs of improvement have been seen by 2 to 3 months, studies to confirm the diagnosis of LPR should be instituted. Currently, ambulatory MCII with pH monitoring is the most effective way to identify LPR. If MCII is not available, multichannel pH monitoring is the best choice. TNE, EGD, and barium-swallow studies can be used to evaluate the aerodigestive tract further.

In our patient, lifestyle and dietary recommendations were made initially. The patient was encouraged to decrease his smoking habit, start a weight loss program, and elevate the head of his bed. He was also given some specific dietary recommendations that included decreasing his consumption of caffeine, citrus fruits, and carbonated beverages. He was also started on ranitidine twice daily. After 6 weeks, no improvement was noticed and he was started on a PPI for 6 weeks. When he was reevaluated, he noticed significant improvement in his symptoms but with occasional throat clearing still present. After 6 additional weeks of therapy, his symptoms completely resolved, and the PPI was discontinued. He was informed that if his symptoms return, he should restart the medication.

Surgical Management

The goal of surgical management is restoration of competence of the lower esophageal sphincter. Fundoplication is the most common procedure performed. Currently the endoscopic approach is most commonly used. Endoscopic techniques using endoluminal plication, radiofrequency-induced thermal injury, and liquid polymer injections to improve function of lower esophageal sphincter are currently being investigated.

Rehabilitation and Follow-up

Around 25% of LPR patients experience spontaneous resolution of symptoms, and 50% have a chronic course of disease, with intermittent exacerbations and remissions. Patients whose LPR fails to resolve after definitive medical or surgical treatment must be followed up indefinitely with careful examination of the upper aerodigestive tract for signs of complications and malignancy.

Questions

1. Laryngopharyngeal reflux can present with all of the following symptoms except:
   - **A.** Intermittent hoarseness
   - **B.** Globus pharyngeus
   - **C.** Nausea and vomiting
   - **D.** Cough
   - **E.** Sore throat
2. Which of the following is a common physical finding in patients with laryngopharyngeal reflux?
   - **A.** Laryngomalacia of the epiglottis
   - **B.** Erythema of the posterior larynx
   - **C.** Frequent throat clearing
   - **D.** Laryngeal papillomata
   - **E.** Vallecular cyst
3. What is the current best diagnostic test for laryngopharyngeal reflux?
   - **A.** Barium esophagram
   - **B.** Single-probe pH study
   - **C.** CT scan
   - **D.** Dual-probe pH study
   - **E.** Ambulatory multichannel intraluminal impedance combined with pH monitoring

Suggested Readings

Ford CN. Evaluation and management of laryngopharyngeal reflux. JAMA 2005;294(12):1534–1540

Koufman JA. The otolaryngologic manifestations of gastroesophageal reflux disease (GERD): a clinical investigation of 225 patients using ambulatory 24-hour pH monitoring and an experimental investigation of the role of acid and pepsin in the development of laryngeal injury. Laryngoscope 1991;101(4 Pt 2, Suppl 53):1–78

Park W, Hicks DM, Khandwala F, et al. Laryngopharyngeal reflux: prospective cohort study evaluating optimal dose of proton-pump inhibitor therapy and pretherapy predictors of response. Laryngoscope 2005;115(7):1230–1238

Reavis KM, Morris CD, Gopal DV, Hunter JG, Jobe BA. Laryngopharyngeal reflux symptoms better predict the presence of esophageal adenocarcinoma than typical gastroesophageal reflux symptoms. Ann Surg 2004;239(6):849–856

Srinivasan R, Vela MF, Katz PO, Tutuian R, Castell JA, Castell DO. Esophageal function testing using multichannel intraluminal impedance. Am J Physiol Gastrointest Liver Physiol 2001;280(3):G457–G462

55

Subglottic Stenosis

Sid M. Khosla

History

A 40-year-old white man has a 3-month history of progressive dyspnea, now occurring while at rest. He reports facial pain overlying the left maxillary sinus, nasal obstruction, chronic cough, and recent mild hoarseness. He denies any dysphagia or aspiration, but he admits that he has had severe heartburn for several years. He reports that after a severe motor vehicle accident, he was intubated for 10 days and subsequently had a tracheotomy for 3 months.

The physical examination reveals biphasic stridor with moderate retractions while breathing. There is also significant tenderness over the left maxillary sinus and erythematous, boggy nasal mucosa in the left nares. Laryngeal examination reveals slightly erythematous folds. The subglottis cannot be evaluated.

Differential Diagnosis—Key Points

1. Prolonged intubation can cause subglottic or tracheal stenosis. A tracheotomy can also cause this, especially if it is placed too high. This could be the cause in this case, but given his other symptoms, other causes should also be explored.
2. Laryngopharyngeal reflux can make subglottic stenosis worse and can prevent it from healing. A dry cough is also seen with laryngopharyngeal reflux. Although postnasal drip has been associated with reflux, sinus pain and nasal obstruction usually are not.
3. With nasal, laryngeal, and possibly pulmonary symptoms, computed tomography needs to be done of the sinuses, neck, and lungs.
4. Inflammatory or infectious disorders may cause laryngotracheal stenosis. Wegener granulomatosis (subglottic), sarcoid (supraglottic), relapsing polychondritis, and tuberculosis are potential causes. Amyloidosis can also be a cause.

Test Interpretation

CT scans showed nonspecific left maxillary sinus disease and three nodules in the left lung.

A purified protein derivative skin test was placed and was negative. Acetylcholinesterace and calcium levels were normal (for sarcoid). Cytoplasmic-staining anti-neutrophil cytoplasmic antibody was strongly positive, as was anti-proteinase 3. Perinuclear-staining anti-neutrophil cytoplasmic antibody was negative.

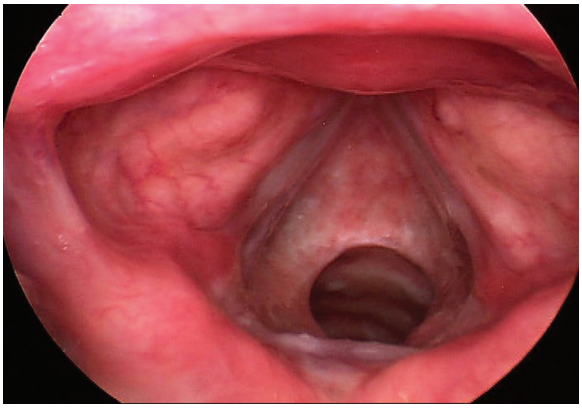

Fig. 55.1 Anterior subglottic stenosis.

The patient was scheduled for a microlaryngoscopy and bronchoscopy with possible tracheostomy. A rigid 4-mm Hopkins rod telescopic unit was used for evaluation. Again the larynx appeared normal. However, in the immediate subglottis, an anterior stenosis was seen that was around 5 mm thick (**Fig. 55.1**). Using the Hopkins rod as a reference, the airway was about 3 mm in diameter. Distally the tracheobronchial tree was normal. A CO_2 laser and a balloon dilator were used to open the stenosis (see Surgical Management), and a tracheotomy was not performed.

◆ Diagnosis

Subglottic stenosis secondary to Wegener granulomatosis

◆ Medical Management

In the past, a combination of cyclophosphamide and steroids was used for long-term treatment. More physicians are now using a combination of methotrexate with or without steroids. Trimethoprim-sulfamethoxazole can also be used as an adjunct or alone if the symptoms are mild to moderate.

◆ Surgical Management

For webs that are around 1 cm or smaller, endoscopic radial cuts with a laser can be used, followed by balloon dilation. Mitomycin C is given at the time of the operation.

In terms of airway reconstruction, a laryngotracheoplasty with a rib graft could be done; in this case, an anterior graft alone or an anterior and posterior graft could be used. A cricotracheal reconstruction is also an option.

◆ Rehabilitation and Follow-up

The patient should be followed up every 1 to 2 weeks for the first 2 to 3 months. After the physician thinks the airway is stable, the patient can be followed up every 3 months. Early on endoscopic evaluation in the operating room should be done if removal of granulation tissue or dilation is expected. When the airway is stable, bronchoscopy can be done in the clinic.

◆ Questions

1. Possible treatments for subglottic stenosis include all of the following except:
- **A.** Balloon dilation
- **B.** Laryngotracheal reconstruction
- **C.** Tracheal resection
- **D.** Cricotracheal resection

2. What laboratory value is usually seen with Wegener granulomatosis?
- **A.** Increased cytoplasmic-staining antineutrophil cytoplasmic antibody
- **B.** Increased angiotensin-converting enzyme level

C. Positive purified protein derivative
D. Increased calcium

3. Treatment for Wegener granulomatosis includes all of the following except:
A. Cyclophosphamide
B. Steroids
C. Methotrexate
D. Trimethoprim-sulfamethoxazole
E. Angiotensin-converting enzyme inhibitors

Suggested Readings

Colice GL, Stukel TA, Dain B. Laryngeal complications of prolonged intubation. Chest 1989;96(4):877–884

Lebovics RS, Hoffman GS, Leavitt RY, et al. The management of subglottic stenosis in patients with Wegener's granulomatosis. Laryngoscope 1992;102(12 Pt 1):1341–1345

Sandhu S, Howardd DJ. Adult acquired laryngotracheal stenosis. In: Rubin JS, Sataloff R, Korovin GS, eds. Diagnosis and Treatment of Voice Disorders. San Diego, CA: Plural Publishing; 2006:693–712

56

Laryngeal Trauma

Sid M. Khosla

History

A 50-year-old man is brought into the emergency department after a motor vehicle accident. A cricothyrotomy had been done emergently in the field. Examination shows significant anterior neck swelling with crepitus. Fiberoptic examination reveals a right fold only. The left true and false folds cannot be identified. The right fold is intact with minimal swelling. Individual thyroid cartilage fragments can be palpated on the left. He is judged to be stable, and imaging is obtained.

Differential Diagnosis—Key Points

1. Resuscitative measures following the Advanced Trauma Life Support guidelines are implemented for all serious injuries.
2. Presenting symptoms of blunt laryngeal trauma include dyspnea, hoarseness, tenderness, cough, hemoptysis, and dysphagia. Signs of laryngeal trauma are edema, crepitance, subcutaneous emphysema, and flattening of the laryngeal prominence. Laryngotracheal separation can also manifest in this manner.
3. A grading system for laryngeal trauma and management of the acute airway is reviewed in **Table 56.1**.
4. A complete history of the mechanism of injury should be obtained, as well as any preexisting medical conditions. A thorough physical examination includes both palpation of the neck and a flexible fiberoptic laryngoscopy to evaluate endolaryngeal trauma.
5. Concomitant injuries. Cervical spine injuries have been reported in up to 50% of patients with blunt laryngeal trauma. Recurrent laryngeal nerve palsies suggest a possible cricoid crush fracture. Pharyngoesophageal tears must also be evaluated.

Test Interpretation

Cervical spine and chest radiograph are clear. Neck computed tomography reveals a vertical thyroid cartilage fracture just lateral to midline, extending from the thyroid notch to the inferior edge of the thyroid cartilage. The right side of the cartilage appears to be in the proper place, but multiple pieces of the left cartilage appear to be posteriorly displaced. Significant subcutaneous emphysema is also noted. There are no other injuries of the airway. Facial CT reveals no fractures.

Diagnosis

Grade 4 blunt laryngeal fracture

Table 56.1 Grading laryngeal trauma and managing the acute airway

Group	Features	Management
1	Minimal or no airway compromise; minor endolaryngeal hematomas or lacerations	Conservative: humidified oxygen, 24-hour observation
2	Moderate edema, hematomas, and airway compromise; minor mucosal lacerations without cartilage exposure and nondisplaced laryngeal cartilage fracture	Tracheotomy under local anesthesia to prevent further damage to the airway by either endotracheal intubation or cricothyrotomy
3	Massive edema, exposed cartilage with large mucosal lacerations, and vocal fold immobility with displaced fractures	Same as group 2
4	Similar findings to group 3, plus more than two fracture lines and a disrupted anterior larynx or an unstable laryngeal skeleton	Same as group 2
5	Full cricotracheal separation	Endotracheal intubation; the distal airway usually retracts into the chest, so tracheotomy may compound the problem

◆ Medical Management

Conservative management is appropriate only for patients in group 1. These patients should be hospitalized for at least 24 hours in a monitored setting because the edema may progress, especially in the 6- to12-hour period. Humidified air or oxygen is supplied, and voice rest is instituted. The patient can expect a good result in both vocal quality and airway.

◆ Surgical Management

When he was stabilized, the patient was taken to the operating room. The cricothyrotomy was converted to a formal, low tracheostomy. Midline exploration revealed a severely comminuted thyroid cartilage. Multiple mucosal tears were seen. The anterior on the right was intact, but the left fold had no anterior attachment to cartilage. Surgical repair consisted of irrigating the endolarynx and repairing the mucosal lacerations. The anterior aspect of the left fold was sutured to the inner perichondrium of the appropriated piece of thyroid cartilage. The multiple pieces of thyroid cartilage were then put together and fixed with miniplates. Subglottic and glottic stents were then placed.

Group 3 or 4 patients should undergo tracheotomy and surgical exploration; group 5 should have immediate exploration and primary repair of the trachea. Indications for open exploration in group 2 patients are as follows:

1. Severe upper airway obstructions not from edema or hematoma
2. Displaced laryngeal skeleton fracture
3. Internal (endolaryngeal) derangement or exposed cartilage
4. Active hemorrhage
5. Increasing subcutaneous emphysema

- Tracheotomy. Tracheotomy is preferred to reduce the exacerbation of intubating an injured larynx and to provide a more stable airway. For patients who need general anesthesia and have mild to moderate trauma to the larynx (group 2), but no significant airway distress, tracheotomy is still the preferred method to prevent long-term edema and subsequent airway compromise.
- Laryngofissure. When the thyroid cartilage or endolaryngeal mucosa has been severely disrupted or cartilage is exposed, a laryngofissure is needed to gain access to repair the injury. Anterior commissure lacerations and disruption of the

free edge of the vocal fold should be approached similarly and repaired primarily. Often a keel is used to prevent blunting of the anterior commissure. Minor mucosal lacerations and noncomminuted thyroid cartilage fractures do not need a laryngofissure for repair.

- Stenting. Stenting is necessary for comminuted thyroid cartilage fractures that are unstable even after open reduction and internal fixation. It is also needed to prevent adhesions in massive endolaryngeal lacerations and to act as a keel in trauma resulting in a disrupted anterior commissure. Stents are maintained for a period of 2 to 3 weeks.

Rehabilitation and Follow-up

In mild (group 1) injuries, normal voice and airway are the standard outcome. With increasing severity of trauma and a delay greater than 24 to 48 hours for operative intervention, the vocal results suffer more than the airway: Group 4 patients have a 33% chance of a "fair" voice and a small, but real, possibility of a poor airway.

Long-term follow-up is needed to assist patients who have not returned to their pretrauma status. Speech therapy, thyroplasty, and laryngotracheoplasty are all possibilities for improving a less than adequate outcome.

Questions

1. What is the appropriate treatment for group 1 laryngeal trauma?
   - **A.** Tracheotomy
   - **B.** Open neck exploration, including tracheotomy
   - **C.** Close observation
   - **D.** Tracheotomy and repair of thyroid cartilage fractures
2. Indications for an open-neck exploration in group 2 patients include all the following except:
   - **A.** Severe upper airway obstructions not from edema or hematoma
   - **B.** Displaced thyroid cartilage fracture
   - **C.** Exposed cartilage
   - **D.** Aphonia
   - **E.** Active hemorrhage
   - **F.** Increasing subcutaneous emphysema
3. Which of the following should all patients with laryngeal fractures have?
   - **A.** Cricothyrotomy
   - **B.** Open reduction and internal fixation of the fractures
   - **C.** Cervical spine radiographs
   - **D.** Open neck exploration

Suggested Readings

Bent JP III, Porubsky ES. The management of blunt fractures of the thyroid cartilage. Otolaryngol Head Neck Surg 1994;110(2):195–202

Fried MP, ed. Laryngeal trauma. In: The Larynx. St Louis, MO: Mosby–Year Book; 1996:378–396

Schaefer SD. The acute management of external laryngeal trauma: a 27-year experience. Arch Otolaryngol Head Neck Surg 1992;118(6):598–604

Snow JB Jr. Diagnosis and therapy for acute laryngeal and tracheal trauma. Otolaryngol Clin North Am 1984;17(1):101–106

57

Reinke Edema

Sid M. Khosla

History

A 72-year-old woman presents with a history of hoarseness for 20 years that has been progressively getting worse in the last 6 months. She notes that the main changes in her voice include vocal fatigue and a decrease in pitch. She has no odynophonia, no dysphagia, and no odynophagia. She does report having increased shortness of breath for the past 6 months; initially she noted this on exertion only but now reports that she is constantly short of breath. On further questioning, she admits that the shortness of breath is the main reason she came into the clinic. She admits to smoking two packs a day for 50 years. She is a social drinker. She is not taking any medicines and has no cardiac, pulmonary, or neurologic history.

On physical examination, she has audible inspiratory stridor. Fiberoptic examination shows her vocal folds when she inspires (**Fig. 57.1**) and when she expires (**Fig. 57.2**). On video stroboscopy, the vocal folds are seen to have generalized pale edema (best seen in **Fig. 57.2**) with some submucosal hypervascularity (best seen in **Fig. 57.1**). No discrete masses or lesions are appreciated. The mucosal waves are irregular and the mucosa appears to be very floppy.

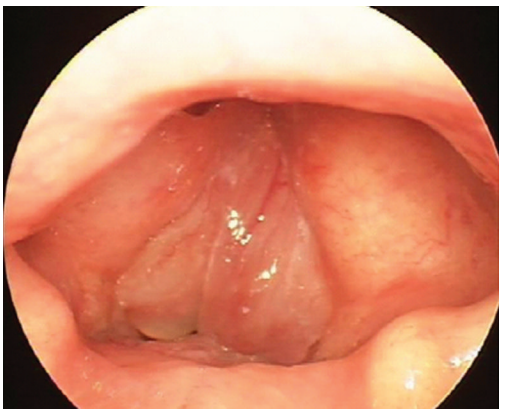

Fig. 57.1 Vocal folds during inspiration.

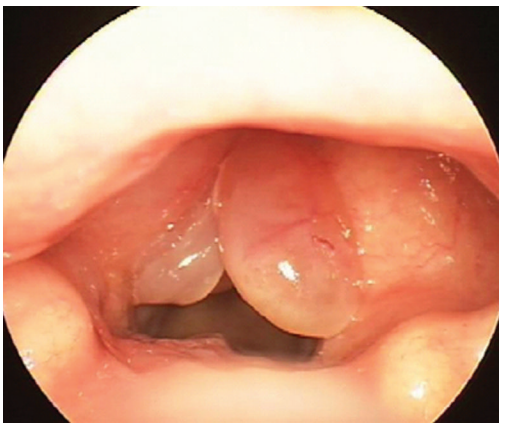

Fig. 57.2 Vocal folds during expiration.

Differential Diagnosis—Key Points

1. Reinke edema is an accumulation of fluid in the superficial layer of the lamina propria (SLLP), also known as Reinke's space. The fluid is usually along the entire length of the fold, greater in the superior surface of the fold, but it also accumulates along the medial surface.
2. Early on, the fluid is watery and relatively clear. Later in the disease, the fluid is very thick. Histologically, in both cases, the lamina propria is edematous with small pools of mucoid fluid between areas of elastic fibers.
3. The edema is usually bilateral but is often asymmetric, as in this case.
4. Compared with the normal fold, the SLLP in Reinke edema has increased mass and decreased stiffness. In general, the fundamental frequency is proportional to stiffness and inversely proportional to mass. Thus the fundamental frequency will be lower, which produces a lower pitch in the patient with Reinke edema.
5. If the folds are edematous enough, dynamic collapse during inspiration will occur, as it has in this case.
6. The major etiologic factor is thought to be tobacco smoking, although laryngopharyngeal reflux and voice abuse have also been suggested.

Test Interpretation

Fiberoptic laryngoscopy reveals watery folds consistent with Reinke edema. Video stroboscopy is necessary to evaluate the mucosal waves; however, in moderate to severe cases, mucosal waves might not be present. In addition, vibration is often irregular, and the strobe cannot be used.

Diagnosis

Reinke edema with hoarseness and significant airway obstruction

Medical Management

Voice therapy is helpful if the edema is mild; surgery is also necessary for more significant edema. In either case, the edema will recur if the patient continues to smoke.

Surgical Management

Indications for surgery, even if the patient continues to smoke, include the following:

1. If there is any concern of a neoplastic lesion
2. If there is airway obstruction, as is the case with this patient

Surgery usually involves making an incision in the superior lateral aspect of the fold. Mucosal flaps are raised, and the thick fluid is suctioned. Care is taken to leave some of the SLLP over the vocal ligament. After suctioning, any excess mucosa is trimmed.

Rehabilitation and Follow-up

If the patient does quit smoking, voice therapy can be useful. Smoking cessation should be heavily encouraged.

Questions

1. What is thought to be the main cause of Reinke edema?
   A. Alcohol
   B. Smoking tobacco
   C. Laryngopharyngeal reflux
   D. Voice abuse
2. In a patient with Reinke edema, the voice usually has:
   A. Increased pitch
   B. Decreased pitch
   C. Increased fundamental frequency
   D. Increased loudness

3. Surgery for Reinke edema involves:
- **A.** Resection of the vocal ligament
- **B.** Resection of all the vocal fold mucosa
- **C.** Resection of part of the superficial layer of the lamina propria
- **D.** Resection of all the superficial layer of the lamina propria

Suggested Readings

Vecerina Volić S, Kirincić N, Markov D. Some morphological, histological, cytological and histochemical aspects of Reinke's oedema. Acta Otolaryngol 1996;116(2): 322–324

Zeitels SM, Hillman RE, Bunting GW, Vaughn T. Reinke's edema: phonatory mechanisms and management strategies. Ann Otol Rhinol Laryngol 1997;106(7 Pt 1): 533–543

XII

General Otolaryngology

58

Epiglottitis

Gordon H. Sun and Allen M. Seiden

History

A 43-year-old woman was admitted to the hospital with chief complaints of hoarseness and sore throat after smoking crack cocaine roughly 4 hours before arrival. She describes her voice as being muffled. These symptoms had progressed rapidly, and her throat pain was increased with swallowing to the point that she was now unable to tolerate solid food or liquids by mouth. Additionally, she reported mild difficulty breathing without wheezing.

She was otherwise healthy, without any significant medical history, including reflux, asthma, or heart disease, other than an adenotonsillectomy done about 30 years ago for chronic tonsillitis. The patient denied recent neck trauma, foreign body ingestion, or other caustic exposure. She denied recent fevers or illnesses preceding onset of her current symptoms. The patient's immunization status was up to date. She was a 20-pack per year cigarette smoker and had a history of weekly crack cocaine and marijuana abuse. Other than acetaminophen for pain control, she was not taking any medications regularly. She had no known drug or environmental allergies.

On physical examination, the patient was anxious and appeared to be in moderate distress. She was sitting erect but leaning slightly forward and was seen to be drooling. She was mildly stridulous, with a muffled voice, but was not tachypneic or retracting. She had a fever of around 102°F and was tachycardic but not hypotensive. Oral examination demonstrated no erythema or exudate and the absence of tonsils consistent with her prior surgery. The uvula was not edematous and was midline. Indirect mirror examination of the larynx revealed diffuse edema and erythema of the supraglottic structures, including the epiglottis, aryepiglottic folds, arytenoids, and false vocal cords. The true vocal cords were poorly visualized, and assessment of their mobility was subsequently limited.

The neck was palpated and found to be tender anteriorly at the level of the larynx. Shotty bilateral cervical lymphadenopathy without overlying erythema or other skin changes was also appreciated. There was no thyromegaly.

Differential Diagnosis—Key Points

1. Although tonsillitis in this patient is ruled out by her absence of tonsils, this patient's sore throat could still be attributable to pharyngitis, or even possibly a deep neck infection. However, the onset of symptoms was quite rapid, decreasing the likelihood of a deep neck space infection, as well as other causes, such as malignancy. The history excludes other causes such as foreign

body impaction or other trauma. The combination of fever, progressive odynophagia, and sore throat should therefore raise the suspicion of epiglottitis.

2. Until the early 1990s, epiglottitis was typically considered a disease of childhood, most commonly seen in children aged 2 to 7 years. However, after the introduction of the *Haemophilus influenzae* type B (HiB) vaccine in the 1980s, the incidence in children declined significantly. This disease is now more commonly observed in adults, with an incidence of 1 to 3.1 per 100,000 people. Adult mortality rates approach 7%, in many cases owing to a delay in diagnosis or adequate care. Therefore, a high index of suspicion should be maintained.
3. Physical examination is diagnostic and requires the use of either indirect mirror or fiberoptic laryngoscopy. Classically there is concern that performing such an examination in children may precipitate an acute airway obstructive event, so it is still recommended that a child with suspected epiglottitis be immediately transported to the operative suite to secure the airway. In adults several studies have demonstrated that indirect laryngoscopy is a safe maneuver in patients who are not in significant respiratory distress. Alternatively, one could obtain a lateral airway radiograph, which may demonstrate the "thumbprint" sign, indicative of severe epiglottic edema (**Fig. 58.1**). However, keep in mind that the sensitivity and specificity of this imaging study are relatively low (38% and 78%, respectively). Also note that in adults, the inflammation may involve not only the epiglottis, but other supraglottic structures as well, and may even extend toward the pharynx and uvula; in children, the infection tends to manifest as a swollen, cherry-red epiglottis.
4. This patient was sitting in the "tripod" position, erect and leaning slightly forward, which helps to reduce obstructive symptoms caused by supraglottic edema. A muffled or so-called hot potato voice and tenderness to direct palpation over the larynx are also characteristic of epiglottitis. However, the three most common signs of epiglottitis are fever, respiratory difficulty, and irritability, regardless of age.

◆ Test Interpretation

A lateral neck (airway) radiograph may be useful in narrowing the differential diagnosis, especially in children, who are at significant risk of laryngospasm or other airway obstruction from indirect laryngoscopy. However, in adults this imaging modality is generally not favored if a full physical examination is possible because of its low sensitivity and specificity and a false-negative rate of 16%. Computed tomography (CT) imaging of the neck can rule out the presence of epiglottic or tongue-base abscesses, a complication found nearly exclusively in adults, or other deep neck space infection. However, because of the potential for complete airway compromise in epiglottitis, this imaging study should be ordered with extreme caution and only once the patient has been stabilized.

A complete blood count will usually reveal leukocytosis. Blood cultures may demonstrate

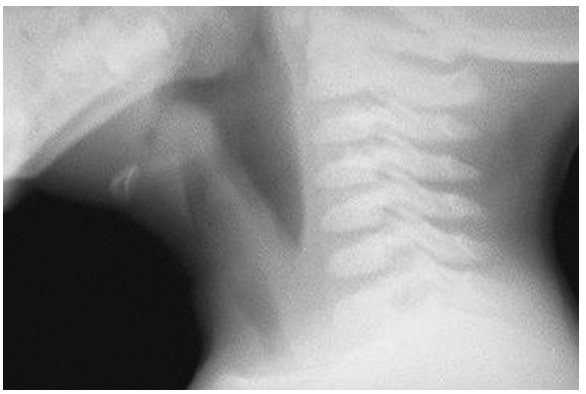

Fig. 58.1 Lateral airway radiograph demonstrates epiglottic edema seen in epiglottitis. (From eMedicine.com, 2009. Available at: http://www.emedicine.com/emerg/TOPIC169.HTM. Image used with permission.)

positive results for *H. influenzae*, *Streptococcus* spp., *Staphylococcus aureus*, or other organisms in 8 to 31% of cases, though prior to introduction of the HiB vaccine, *H. influenzae* bacteremia was seen in 90 to 95% of patients. *H. influenzae* and streptococci still constitute most of the bacteriology seen in adult patients with epiglottitis, while anaerobes, fungi, and viral agents are only rarely cited as pathogens. If epiglottitis is suspected, blood cultures should be obtained before initiating antibiotic therapy. Throat swab cultures are of little benefit because the infection is typically limited to submucosal cellulitis in adults. Note that there are numerous noninfectious causes of epiglottitis in adults, such as foreign body trauma, caustic injury, or thermal injury, which may explain the variance in positive blood cultures seen in several studies.

◆ Diagnosis

Epiglottitis

◆ Medical Management

Initial medical therapy involves aggressive intravenous fluid resuscitation and broad-spectrum antibiotic coverage for *H. influenzae*, staphylococci, and streptococci, such as ampicillin/sulbactam or a third-generation cephalosporin such as ceftriaxone. Antibiotic therapy can then be adjusted as culture results are obtained. Duration of antibiotic therapy (including oral medications) ranges from 1 to 2 weeks in total, depending on the clinical condition of the patient. Corticosteroid use to reduce upper airway edema is controversial but is recommended in most cases.

Morbidity and mortality rates from epiglottitis in the modern era are typically associated with airway obstruction. In children, automatically placing an artificial airway may reduce the incidence of morbidity, but this approach is somewhat more controversial in adults. One study from 1995 recommends automatic airway intervention not only for patients with obvious signs of airway obstruction but for all patients 5 years of age or younger; those with respiratory distress when sitting upright; those with stridor, drooling, or symptom progression over less than 24 hours; or significant enlargement of the epiglottis on laryngoscopy or radiography.

Several studies indicate that in adults without respiratory distress, close observation with humidified oxygen in an intensive care unit may be all that is required for airway management, with endotracheal intubation and tracheotomy equipment available at the bedside in case the patient develops more significant signs of respiratory distress. It should be noted that delayed respiratory compromise is known to occur in epiglottitis, making emergent airway intervention mandatory. Emergent intubation and tracheotomy are performed in adults with epiglottitis at a rate of 9 to 16%. Extubation trials may be attempted after a 48-hour trial of empiric antibiotics, an air leak is noted around the endotracheal tube, or significantly decreased supraglottic edema and erythema are noted on flexible laryngoscopy.

◆ Surgical Management

Surgical management for this disease may be indicated for establishing a secure airway. Intubation can be quite difficult because of significant edema and obscuration of landmarks. If intubation with or without fiberoptic guidance fails, a cricothyrotomy or tracheotomy is required. If an epiglottic abscess is suspected (or noted on CT imaging), incision and drainage of the abscess should be performed in the operative suite after the airway has been secured.

◆ Rehabilitation and Follow-up

Recurrent epiglottitis is rare and should raise the suspicion of an underlying medical problem such as sarcoidosis, collagen vascular disease, or even malignancy. One complication of epiglottitis that might not be immediately apparent while a patient is sedated and intubated is meningitis. Note that antibiotic regimens suitable for treatment of epiglottitis may not meet the recommended therapeutic dosages for meningitis.

As a result of the HiB vaccine, the incidence of pediatric epiglottitis is on the decline. Even though many adults have never received the HiB vaccine, it has been documented that

protection from *H. influenzae* can be transmitted through populations by the herd immunity phenomenon. Nonetheless, a decrease in adult epiglottitis patients has not yet been observed. Medical literature also documents cases in which HiB-vaccinated children were nonetheless diagnosed with *H. influenza*–mediated epiglottitis, with or without bacteremia, because of an inability to produce HiB antibodies or the production of anticapsular antibodies, which provides less protection from *H. influenzae*.

Questions

1. Epiglottitis is rarely seen in adults.
- **A.** True
- **B.** False

2. What should be the first step in managing epiglottitis?
- **A.** Obtain blood cultures
- **B.** Order a lateral neck radiograph
- **C.** Assess the airway
- **D.** Initiate intravenous steroids and antibiotics

3. What are the three most common signs of epiglottitis?
- **A.** Fever
- **B.** Respiratory difficulty
- **C.** "Hot potato" voice
- **D.** Drooling
- **E.** Irritability

4. What is the most common organism associated with epiglottitis?
- **A.** *Haemophilus influenzae*
- **B.** *Staphylococcus aureus*
- **C.** *Streptococcus* species
- **D.** Respiratory syncytial virus
- **E.** *Klebsiella pneumoniae*

Suggested Readings

Carey MJ. Epiglottitis in adults. Am J Emerg Med 1996; 14(4):421–424

Mathoera RB, Wever PC, van Dorsten FR, Balter SG, de Jager CP. Epiglottitis in the adult patient. Neth J Med 2008;66(9):373–377

Mayo-Smith MF, Spinale JW, Donskey CJ, Yukawa M, Li RH, Schiffman FJ. Acute epiglottitis: an 18-year experience in Rhode Island. Chest 1995;108(6):1640–1647

McEwan J, Giridharan W, Clarke RW, Shears P. Paediatric acute epiglottitis: not a disappearing entity. Int J Pediatr Otorhinolaryngol 2003;67(4):317–321

Rafei K, Lichenstein R. Airway infectious disease emergencies. Pediatr Clin North Am 2006;53(2):215–242

Shah RK, Roberson DW, Jones DT. Epiglottitis in the *Hemophilus influenzae* type B vaccine era: changing trends. Laryngoscope 2004;114(3):557–560

Sobol SE, Zapata S. Epiglottitis and croup. Otolaryngol Clin North Am 2008;41(3):551–566, ix

Wong EY, Berkowitz RG. Acute epiglottitis in adults: the Royal Melbourne Hospital experience. Aust NZ J Surg 2001;71(12):740–743

59

Deep Neck Space Abscess

Gordon H. Sun and Allen M. Seiden

History

A 4-year-old child was admitted with a 6-day history of a slowly enlarging right-sided neck mass and temperatures of 101° to 102°F. The child has a history of multiple episodes of tonsillitis, the most recent of which was treated about 2 weeks ago with amoxicillin. His other symptoms included difficulty looking from side to side, decreased appetite and oral intake, and occasional noisy breathing when agitated. An older sibling at home had symptoms of a viral upper respiratory infection about 2 weeks ago but was otherwise healthy.

This patient had no other significant medical history, had not suffered any recent insect bites or superficial skin trauma, and was otherwise up to date with immunizations. He had not been treated medically by a physician for his current symptoms, and he was not taking any medications at the time of admission.

On physical examination, the child had a temperature of around 102.5°F and appeared pale and tired but nontoxic. He avoided moving his neck while being examined. The child demonstrated occasional inspiratory stridor and a mildly hoarse cry and cough. There was a small amount of clear rhinorrhea in both nostrils. He was drooling moderately. His oral examination demonstrated very erythematous pharyngeal mucosa, 3+ erythematous tonsils without exudate, and right-sided posterior pharyngeal wall swelling. The tongue and uvula were not edematous and were both midline. No trismus was noted. Over his right posterolateral neck, there was a roughly 3 × 2 cm area of swelling, induration, and redness that was very tender to palpation. A 2 × 1 cm area within this neck mass was fluctuant, although there were no skin breaks and thus no fluid was expressible. No other cervical lymphadenopathy was appreciated.

Differential Diagnosis—Key Points

1. This child had recently been treated for acute tonsillitis and returns shortly after completing therapy with worsening symptoms. This should immediately raise suspicion that he might be developing a complication from that infection. There are at least 11 different deep neck spaces, divided by several cervical fascial planes. These planes serve as barriers to the spread of infection. Additional fascial planes attached to the hyoid bone anteriorly serve a similar function. Classification of deep neck infections is done anatomically based on the infection's relation to the hyoid bone: suprahyoid, entire neck length, and infrahyoid. Suprahyoid infections include those found in the peritonsillar, submandibular, parapharyngeal,

masticator/temporal, buccal, and parotid spaces. Full-length neck infections are found in the retropharyngeal, prevertebral, carotid, and "danger" spaces. The "danger" space is a region between the two divisions of the deep layer of the deep cervical fascia, extending from the skull base to the posterior mediastinum at the level of the diaphragm. Infrahyoid infections are found in the pretracheal (anterior tracheal) space, located between the infrahyoid strap muscles and the esophagus.

2. Demographics play an important role in determining the most likely source and microbial cause of the infection. Tonsillitis and pharyngitis are the most common sources of deep neck infections in children, whereas odontogenic infections (from poor dental hygiene) and intravenous (IV) drug abuse are the most common causes of deep neck infections in adults. In children, also consider the possibility that a previously unknown congenital neck lesion has become superinfected, such as a branchial cleft anomaly or thyroglossal duct cyst. In adults with a history of head and neck cancer, or those with significant risk factors thereof, local metastasis or recurrence of squamous cell carcinoma or other cancerous lesions may manifest as an enlarged, necrotic, superinfected cervical lymph node.
3. Numerous other causes of deep neck infections exist that are not necessarily specific to any particular demographic, such as oropharyngeal and neck trauma (from surgery or ballistic injury), iatrogenic injury to the neck (from bronchoscopy or esophagoscopy), salivary gland infection, mastoiditis, foreign body aspiration, and thyroiditis. In up to 22% of cases, no identifiable cause of the infection may be found.
4. Immunocompromise, from human immunodeficiency virus (HIV) infection, long-term steroid use, chemotherapy, diabetes mellitus, or chronic liver and kidney disease, is an important consideration as well. Patients with known HIV infection may present with recurrent neck abscesses, many of which are caused by atypical organisms such as the *Mycobacterium avium* complex. Sometimes leukopenia, rather than leukocytosis, is seen in immunosuppressed patients.
5. In this particular case, one frequent complication of tonsillitis is peritonsillar abscess. However, although this child did have poor oral intake, difficulty handling oral secretions, and fever, he did not demonstrate trismus, deviation of the uvula, or asymmetric fullness of the soft palate. Note that although fullness of the neck is often present with a deep neck space abscess, fluctuance is usually not present because of the deep location of these spaces.

◆ Test Interpretation

Initial laboratory testing should include a complete blood count with differential, serum electrolyte (renal profile) levels, and blood cultures if the patient has signs of sepsis. In this case, the patient had an elevated leukocyte count of 21,000/μL, with 80% polymorphonuclear leukocytes. Although this patient did not demonstrate any electrolyte abnormalities, in severely ill patients there may be serum hyperosmolarity.

Flexible fiberoptic laryngoscopy, including nasopharyngoscopy, is very useful in establishing the condition of the patient's airway. This patient demonstrated moderately diffuse edema of the right posterior pharyngeal wall extending down to the hypopharynx. A small amount of thin salivary secretions could be seen pooling in the pyriform sinuses. The supraglottis and glottis appeared grossly normal otherwise, and both vocal cords were appropriately mobile with phonation or crying.

Imaging studies are useful not only in confirming a diagnosis but also in localizing the infection. The gold standard imaging modality is a computed tomography (CT) contrast scan of the neck (**Fig. 59.1**). In this case, diffuse soft tissue inflammation and swelling of the retropharyngeal soft tissue on the right were seen, extending to the postauricular, occipital, and parapharyngeal tissues, with loss of well-defined tissue planes. There was mild effacement of the parapharyngeal fat and oropharyngeal soft tissues as well. Within the posterior triangle of the right neck, there was a low-attenuation region measuring roughly 3.1 × 2.2 × 1.5 cm that is rim enhancing. Scattered subcentimeter homogeneous-appearing submandibular and jugulodigastric lymph nodes can be seen. In some

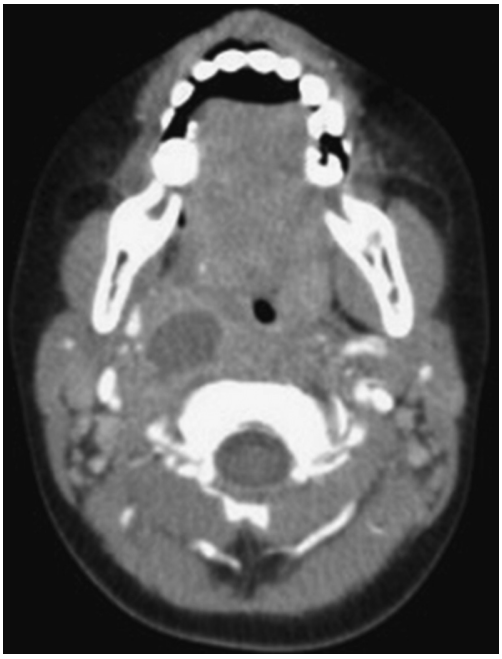

Fig. 59.1 Contrast computed tomography scan of the neck demonstrates right-sided retropharyngeal abscess.

instances, a lateral neck radiograph may be helpful as a rapidly performed tool to determine whether there is posterior pharyngeal soft tissue edema (thickening), anterior displacement of the airway, or gas bubbles in the retropharyngeal space. If there is suspicion of a (radiopaque) foreign body as the cause of the abscess, it may be identified on the radiograph. The diagnosis of retropharyngeal cellulitis or abscess approaches 90% sensitivity using the criterion of the retropharyngeal space being twice the diameter of the vertebral body at the level of C2. Suspicion of mediastinal involvement or aspiration pneumonia warrants a chest radiograph.

◆ Diagnosis

Deep neck space (retropharyngeal) abscess

◆ Medical Management

Regardless of the location of the deep neck infection, determining the status of the patient's airway remains the first priority. Certain types of deep neck infections, however, are more likely to cause airway compromise. These include retropharyngeal, parapharyngeal, and pretracheal infections; submental infections (Ludwig angina); and infections that involve multiple deep neck compartments. Symptoms such as persistent stridor, cyanosis, retractions, and oxygen desaturation may be indicative of impending airway collapse. Initial assessment of the airway should be done when the patient is first examined; this can be performed indirectly with a mirror, or via flexible fiberoptic endoscopy. Airway distress can be managed by simple observation with oxygen supplementation as needed, nasotracheal or orotracheal intubation (with or without fiberoptic guidance), cricothyrotomy, or tracheotomy. Note that certain maneuvers, such as placing the patient in the supine position or the use of excessive sedation or anesthesia, can exacerbate the patient's tenuous respiratory status and increase the chance of morbidity or mortality from loss of the patient's airway. Tracheotomy equipment should be kept at the bedside, readily available.

Once the patient's airway is secured by any of the above methods, IV fluid resuscitation and metabolic management should be started. This is especially important in patients presenting with signs of sepsis. Management of the patient's co-morbidities should be initiated as well. For example, tight glucose control in diabetic patients is important because this subset of patients is susceptible to poor wound healing and persistent, atypical infections that can easily cross fascial planes because of the patient's weakened immune system. Hyperglycemia has also been shown to increase the potency of several organisms implicated in deep neck infections.

Most deep neck infections are caused by a mixture of oropharyngeal flora, which may include *Streptococcus* spp., *Staphylococcus aureus*, *Klebsiella pneumoniae*, or *Haemophilus influenzae*. Anaerobes that have been implicated in deep neck infections include *Bacteroides fragilis*, *Peptostreptococcus*, and *Eikenella corrodens*. Previously hospitalized patients may be prone to organisms such as *Pseudomonas aeruginosa* or methicillin-resistant *S. aureus* (MRSA). Immunosuppressed patients are not only prone to infections from any of the above organisms, but they may also be afflicted with mycobacteria,

fungi, *Toxoplasma gondii*, *Pneumocystis carinii*, or other opportunistic pathogens. Therefore, appropriate empiric and culture-directed antibiotic therapy is absolutely critical in medical management of deep neck space infections.

Otherwise healthy patients may be started on ampicillin/sulbactam or a cephalosporin such as cefoxitin or ceftriaxone for broad coverage; clindamycin should be used if the patient is penicillin allergic. In immunocompromised patients or in suspected cases of pseudomonal infection, ticarcillin/clavulanate, piperacillin/tazobactam, imipenem, or clindamycin used with either ciprofloxacin or ceftazidime is recommended for empiric therapy. Suspected MRSA infections may respond well to vancomycin. These medication options may be adjusted as needed depending on local community resistance patterns. An infectious disease consultation may be worthwhile for patients who may be (or are) immunocompromised because the range of pathogens that may be causing the neck infection, and the available antibiotic armamentarium, might be unfamiliar to the otolaryngologist. Culture-guided therapy may be done early on if the infection is easily accessible to needle aspiration. This should be attempted especially if the suspected cause of the infection is atypical or otherwise unusual. Generally speaking, unless a definitive abscess has been identified on clinical examination and imaging or there is acute airway compromise or signs of sepsis, deep neck infections can be treated conservatively with IV antibiotics for 24 to 48 hours before either reimaging or surgical intervention. If the patient clinically improves and is afebrile for 48 hours, IV antibiotics may be switched to oral medications.

◆ Surgical Management

The need for surgical intervention may arise if the infection has not responded to appropriate empiric IV antibiotic coverage over 24 to 48 hours; a definite abscess or drainable fluid collection is noted on imaging studies; or if there are signs of airway obstruction, sepsis, or other impending life-threatening events secondary to the neck infection. Airway stability always remains paramount before surgery. Once the airway is secured, incision and drainage (I&D) is the mainstay of surgical therapy. Retropharyngeal abscesses, as well as a few other selected neck infections such as those in the peritonsillar space, can be accessed via a transoral approach. This involves a vertical incision through the middle of the affected posterior pharyngeal wall mucosa, followed by blunt dissection and adequate drainage of the abscess. However, most deep neck infections are more easily drained via a transcervical incision. Retropharyngeal and parapharyngeal abscesses are approached externally using a modified apron-like incision, with the vertical limb along the anterior border of the sternocleidomastoid muscle and the horizontal limb roughly through the middle of the abscess cavity. The approach involves retraction of the sternocleidomastoid laterally, identification and reflection of the carotid sheath, and ultimately blunt dissection into the abscess cavity and thorough drainage. The external approach typically requires placement of a drain.

Regardless of preexisting antibiotic use or surgical approach, specimens should be sent for Gram stain and cultures, including aerobic and anaerobic testing, plus acid-fast and fungal staining if deemed appropriate for the situation. Iodoform (antimicrobial) packing left in place in the wound and gradually removed over the next 48 to 72 hours may be beneficial.

Not only may needle aspiration be used for culture-guided therapy before definitive I&D, but occasionally it can be used for well-defined, unilocular abscesses that are not causing airway compromise. This can be done using either ultrasound or CT guidance. However, this method may result in reaccumulation of the fluid collection, which then requires a standard I&D procedure to treat completely. Open drainage remains the gold standard of treatment for this disease process.

◆ Rehabilitation and Follow-up

If an otherwise healthy patient responds to IV antibiotics only and is discharged on oral medication, follow-up should be done over the following 7 to 10 days to ensure complete resolution of the infection. Surgical patients need to be evaluated postoperatively not only to ensure that the infection was thoroughly drained, but also to verify that the wound itself is healing

appropriately and is not becoming secondarily infected. All these patients should remain on a course of oral antibiotics for at least 7 to 10 days after hospital discharge. Any signs of a recurrent neck abscess warrant continued medical therapy, reimaging, and repeat I&D if indicated. If the infection is related to a congenital neck lesion or malignancy, close follow-up should be conducted after the infection is cleared to ensure that the underlying cause of the infection is thoroughly evaluated and treated appropriately. Similarly, immunocompromised patients should be monitored closely by their primary physicians after the infection is cleared because this subset of patients is prone to recurrent neck infections.

◆ Questions

1. What is the most common source of deep neck space infection in children?
   A. Tonsillitis
   B. Odontogenic infections
   C. Trauma
   D. Mastoiditis

2. What is the most common source of deep neck space infection in adults?
   A. Tonsillitis
   B. Odontogenic infections
   C. Trauma
   D. Idiopathic

3. The "danger space" refers to that space just posterior to the retropharyngeal space and anterior to the prevertebral space. It is called this because infection involving this space can lead to which of the following?
   A. Torticollis
   B. Airway obstruction
   C. Pneumonia
   D. Mediastinitis

Selected Readings

Barzan L, Tavio M, Tirelli U, Comoretto R. Head and neck manifestations during HIV infection. J Laryngol Otol 1993;107(2):133–136

Reynolds SC, Chow AW. Life-threatening infections of the peripharyngeal and deep fascial spaces of the head and neck. Infect Dis Clin North Am 2007;21(2):557–576, viii

Roscoe DL, Hoang L. Microbiologic investigations for head and neck infections. Infect Dis Clin North Am 2007; 21(2):283–304

Tom MB, Rice DH. Presentation and management of neck abscess: a retrospective analysis. Laryngoscope 1988; 98(8 Pt 1):877–880

Vieira F, Allen SM, Stocks RM, Thompson JW. Deep neck infection. Otolaryngol Clin North Am 2008;41(3): 459–483

60

Stomatitis

Charles M. Myer IV and Allen M. Seiden

History

A 58-year-old white man presented with complaints of burning oral pain, dry mouth, and loss of taste. Five months ago, the patient was diagnosed with a right tonsillar carcinoma and subsequently had undergone a right lateral pharyngectomy and neck dissection, followed by 6 weeks of radiotherapy, completed 2 months before the current presentation. The patient received a total of 60 Gray (Gy) of radiation and did develop difficulties with sore throat after the third week. He also noted a loss of taste at that time, which had yet to show much improvement. In addition, his mouth had become quite dry, and this too had not improved following completion of radiation. The soreness seemed to resolve until around 2 weeks before presentation when the burning pain began.

He was tolerating a soft diet but described a particular sensitivity to hot and warm liquids, complaining that they produced a scorched feeling. He denied smoking or using any tobacco products since surgery. His appetite was diminished, and he had lost roughly 15 pounds from his preoperative weight. He relied on a rather old pair of dentures but noted acceptable fit.

Other medical problems included hypertension, which was well-controlled with a diuretic. No history of diabetes, heart disease, or other systemic illness was present.

Physical examination revealed a patient in no acute distress, sitting comfortably and able to handle his secretions. He appeared thin but not emaciated. At his side was a water bottle, and he was able to swallow without aspiration.

Examination of the mouth revealed parched lips and a small amount of cracking at the oral commissure bilaterally. Intraoral examination did not reveal significant edema, ulceration, or exudate. The dorsal surface of the tongue, however, had a dusky red, smooth appearance. The mucosa overlying the hard palate also appeared quite erythematous, with a sharply demarcated border that coincided with the patient's denture. The buccal mucosa was very dry but otherwise displayed normal pink coloration. The oropharynx, though dry, showed well-healed postoperative changes and was normal in color.

Differential Diagnosis—Key Points

1. Given the patient's history, it is logical to consider the impact of his recent radiation therapy. Oral mucositis from cancer therapy has increased in incidence in the past 10 years with the addition of chemotherapy along with changes in radiation protocols, including fractionation and acceleration. During curative radiotherapy, mucositis

occurs in 80% of patients. Inflammation and cell loss in the mucosal layer manifest with color changes and proceed to mucosal ulceration, which is then covered by exudative pseudomembranes. These changes typically begin after approximately 10 Gy (1000 rads) are delivered. Duration is related to mucosal stem cell loss and usually resolves within 3 to 4 weeks after completion of radiation. Symptoms consist of odynophagia, dysphagia, and thick, viscous mucus that is difficult to clear. These symptoms often cause a loss of appetite and weight loss as well as generalized fatigue and malaise. This patient described a sore throat beginning in the third week of radiation, after having around 30 Gy, and this likely did reflect a mucositis at that time. However, given the length of time since radiation and the onset of new symptoms, specifically burning pain, an alternative diagnosis should be suspected.

2. Xerostomia, or dry mouth, is an almost universal effect of radiation therapy in the head and neck in patients receiving more than 60 Gy. Radiation effects on the salivary glands begin after only 15 Gy of radiation and are somewhat reversible at the lower levels (<30 Gy). Not only is salivary flow rate diminished, but the composition of produced saliva changes as well. Adequate presence of saliva allows for healthy mucosa both through antimicrobial effects as well as protecting the integrity of the mucosal surface. Without an appropriate amount of saliva, the mucosa is at risk for irritation and infection.

3. Dysgeusia is a common occurrence during radiation therapy, with most patients experiencing a partial or complete loss of taste. Radiation results in direct damage to the taste buds, beginning at about 10 Gy, and reduces the amount and composition of saliva, which also alters taste perception. Bitter and sour flavors are more susceptible than salt and sweet sensitivity. Maximum loss generally occurs at a level of 30 Gy. Regeneration of the taste bud receptors occurs, and taste will generally return to near-normal levels by 1 year.

4. Viral-mediated processes of many etiologies can affect the oral mucosa, and differentiation between viral stomatitis and chemoradiation induced mucositis can be difficult. Herpes simplex virus (HSV), a human herpes virus (HHV), type 1 primarily and type 2 less so, are often responsible for symptomatic stomatitis. The viral infection produces cell necrosis, and the released viral particles cause a cascading infection and subsequent death of adjacent cells, forming a visible vesicle that then ulcerates. Primary HSV stomatitis affects both keratinized and nonkeratinized mucosa and is preceded by a prodrome in the affected area. The virus enters adjacent neurons, where it forms a reservoir, most often in the trigeminal ganglion. Approximately 40% of those affected undergo reactivation, which can be triggered by stress, fatigue, ultraviolet light exposure, dental injury, facial fracture, or trauma to the infected nerve itself. Secondary HSV stomatitis is often associated with a repetitive site, most commonly the vermillion border and adjacent perilabial skin.

Other viral causes of stomatitis include HHV-3, the varicella-zoster virus, which may occur primarily with a disseminated varicella infection or recurrently from a reservoir in a sensory ganglion, in a similar manner to HSV-1 infections. Herpes zoster, recurrent HHV-3, results in herpetiform vesicles progressing to pustules and ulcerations in a dermatomal pattern, preceded by a sensory prodrome.

Often human immunodeficiency virus (HIV) presents with oral lesions of different pathogenesis as immune function decreases; however, acute seroconversion following HIV infection can also produce a stomatitis. Because this patient does not exhibit any signs of mucosal vesicles or ulcerations, it is unlikely that his process is virally mediated.

5. Although unlikely in this patient, the differential of ulcerative stomatitis includes other notable causes of which the clinician should be aware. Pemphigus vulgaris and cicatricial pemphigoid are both autoimmune-mediated diseases that result in vesicular eruption with subsequent ulceration of the mucosa. Because of antibody activity directed against the epithelium adhesion molecules, pemphigus vulgaris causes separation of the superficial and basal layers. In cicatricial pemphigoid, the basement membrane is the site of cleavage.

Although the etiology is not fully understood, recurrent apthous stomatitis (RAS) is an immune-mediated ulcerative disease often presenting in younger populations. Risk factors, notable in the late onset of RAS, such as stress, trauma, specific foods, or hormonal imbalance, have been identified. Notably, the use of tobacco products decreases the likelihood of RAS. Ulcers exist in three classes: minor, major (Sutton disease), and herpetiform, classified by size and duration. Of diagnositic importance, no vesicular stage precedes the formation of the oral ulcers. RAS may be imitated by the adverse effects of medications, nonsteroidal anti-inflammatory drugs, β-blockers, and potassium channel blockers, as well as many systemic diseases and nutritional deficiencies.

6. The mucosal injury and subsequent xerostomia that chemotherapy and radiation treatment inflict predispose to opportunistic oral infections. During radiation therapy, the normal composition of oral flora can change, including increases in *Streptococcus*, *Lactobacillus*, and *Candida* spp. The normally commensal *Candida* spp., found in up to 50% of normal adults, becomes pathogenic only when there is compromised resistance of the host or competitive oral flora are reduced. Risk factors for this change include radiation therapy, immunosuppression, dental appliances, smoking, diabetes mellitus, and antibiotic use.
Symptoms of candidiasis include burning pain and taste loss, which this patient exhibits. The patient's sensation of having been scorched after consuming certain foods is typical. When the dorsal surface of the tongue is involved, depapillation generally occurs, giving the tongue a smooth appearance. With involvement of buccal or palatal mucosa, lesions are typically dusky red, flat, and irregular and often form under an ill-fitting denture. Pseudomembranous candidiasis, or thrush, is characterized by soft yellow-white plaques, similar to milk curds, which can be easily dislodged.
Candida may involve the corners of the mouth, known as angular chelitis, especially if there is a loss of vertical dimension of the jaw, seen in many head and neck oncologic resections, as well in denture use and HIV. The oral commissures display erythema and cracking, with the friable lesions often extending into adjacent skin. Combined infection with *Staphylococcus* spp. is not uncommon.

◆ Test Interpretation

The diagnosis of stomatitis and the offending cause is usually based on patient history and clinical examination and confirmed by response to therapy. More definitive confirmation may be obtained by cytologic smear, culture, or biopsy, depending on the suspected lesion.

In suspected candidiasis, scraping of the mucosal surface and subsequent staining with periodic acid-Schiff or potassium hydroxide will allow for visualization of fungal elements. Culture on Sabourand medium may be used as well; however, delaying therapy while waiting for culture results is ill advised.

◆ Diagnosis

1. Candidal stomatitis or oral candidiasis
2. Radiation-induced xerostomia
3. Hypogeusia

◆ Medical Management

All patients for whom radiotherapy will be a portion of treatment should have a thorough dental examination and undergo appropriate dental and periodontal treatment. Although there is no pharmacotherapy to circumvent mucositis, measures such as eliminating irritating factors, maintaining adequate oral hygiene and hydration, and appropriate treatment of infection can limit its clinical course.

Xerostomia impacts acute mucositis during radiation but continues to cause symptoms following completion of therapy. Whereas preventing the irradiation of salivary tissue is invaluable, given the nature of head and neck cancer, this is often not possible. Stimulating the remaining hypofunctioning tissue via gustatory or tactile sialogogues, such as acid or bitter substances, or pharmacologic sialogogues, most commonly pilocarpine, is a mainstay of treatment. A variety of adjuvant agents may be used

to clean and lubricate the oral mucosa if salivation remains inadequate. Saline, sodium bicarbonate, and hydrogen peroxide solutions may be used to help remove debris and viscous mucus. Chlorhexidine and fluoride rinses both help to prevent further dental disease. A variety of salivary substitutes also are available for use.

Taste loss is generally reversible, with recovery occurring within the first year. Specific therapy is usually not indicated, but dietary counseling for adequate oral intake can be helpful, both during therapy and in the post-treatment period.

Pharmacotherapy is effective in the treatment of oral candidiasis, with topical agents being the first choice, given low systemic absorption and effects. Nystatin oral suspension is the most widely used and may also be prescribed as an ointment in combination with triamcinolone for use in angular cheilitis and denture candidiasis. Clotrimazole oral troches or lozenges may be used in cases of nystatin intolerance; however, it can occasionally cause elevated liver enzymes. Systemic agents are indicated when disease is resistant to topical therapy or patients have risk factors for systemic infection. Azole class agents, including ketoconazole, fluconazole, and itraconazole, can be used with limited adverse effects, notably liver toxicity. Amphotericin B is rarely necessary; its use is limited by its adverse effects.

◆ Surgical Management

There is no role for surgery in the management of candidiasis or radiation-induced xerostomia.

◆ Rehabilitation and Follow-up

Although the acute fungal infection will resolve with appropriate therapy, this patient will continue to have xerostomia. As previously discussed, he will continue to be susceptible to recurring infection and dental disease. Aggressive oral hygiene, including cleansing and lubricating, as well as proper fitting of dental appliances is important in preventive care.

◆ Questions

1. The following may occur secondary to radiation therapy but are generally reversible except:
   - **A.** Dry mouth
   - **B.** Taste loss
   - **C.** Mucositis
   - **D.** Sore throat
2. Taste loss that occurs during radiation therapy is due to what?
   - **A.** Xerostomia
   - **B.** Mucositis
   - **C.** Direct injury to the taste buds
   - **D.** Overlying yeast infection
3. Stomatitis may be caused by what?
   - **A.** Medications
   - **B.** Autoimmune disease
   - **C.** Radiation therapy
   - **D.** Candidal infection
   - **E.** All of the above

Suggested Readings

Epstein JB, Polsky B. Oropharyngeal candidiasis: a review of its clinical spectrum and current therapies. Clin Ther 1998;20(1):40–57

Hairston BR, Bruce AJ, Rogers RS III. Viral diseases of the oral mucosa. Dermatol Clin 2003;21(1):17–32

Sciubba JJ. Oral mucosal lesions. In: Cummings CW, Flint PW, Harker LA, et al, eds. Otolaryngology—Head and Neck Surgery. 4th ed. Philadelphia, PA: Elsevier Mosby; 2005:1448–1492

Scully C, Gorsky M, Lozada-Nur F. The diagnosis and management of recurrent aphthous stomatitis: a consensus approach. J Am Dent Assoc 2003;134(2): 200–207

Vissink A, Burlage FR, Spijkervet FKL, Jansma J, Coppes RP. Prevention and treatment of the consequences of head and neck radiotherapy. Crit Rev Oral Biol Med 2003; 14(3):213–225

Vissink A, Jansma J, Spijkervet FK, Burlage FR, Coppes RP. Oral sequelae of head and neck radiotherapy. Crit Rev Oral Biol Med 2003;14(3):199–212

61

Cervical Lymphadenitis

Gordon H. Sun and Allen M. Seiden

History

A 2-year-old child was admitted after a left-sided neck mass noticed 2 weeks earlier failed to resolve with oral antibiotic therapy. He has had only intermittent fevers with temperatures no greater than 102°F. The neck mass had been mildly tender and red at times and currently was mildly painful and inflamed. He was otherwise asymptomatic, with no report of difficulty breathing, decreased appetite, or neck stiffness. The parents had not noticed any pits or holes within the mass, and no drainage from the mass was observed.

The patient had no significant medical history. He was born full-term, and no head and neck lesions were seen in this child at birth or immediately thereafter. There was no recent history of otitis media, tonsillitis, or pharyngitis. He had not suffered any recent insect bites or trauma to the head or neck. The family did not have any pet cats or dogs at home. The patient had not been on any trips abroad recently. The child was up to date with his immunizations. He was taking amoxicillin at the time of admission, and he had no known allergies.

On physical examination, the child had a slightly elevated temperature of 100.2°F, with otherwise stable vital signs and oxygen saturation of 99 to 100%. He appeared nontoxic and was playful during the examination. The child was not stridulous and was not coughing or drooling. He had a minimal amount of clear rhinorrhea bilaterally. His oral examination demonstrated moderately erythematous pharyngeal mucosa and 2+ tonsils without erythema or exudate. The tongue and uvula were not edematous and were both midline, and there was no posterior pharyngeal swelling. No trismus was noted. There was no facial swelling or rashes. Over his left anterolateral neck there was a roughly 3 × 1 cm area of edema and induration, without significant erythema or fluctuance. The mass was nontender to palpation. No pits or sinuses were appreciated within the mass. No thyromegaly was noted. The patient was able to move his neck in all directions without apparent difficulty during the examination.

The patient's neck mass shrank substantially after 48 hours of intravenous (IV) antibiotics were administered. He was subsequently discharged home with an additional course of oral antibiotic therapy.

Differential Diagnosis—Key Points

1. Cervical lymphoid tissue may be subdivided into three broad categories: Waldeyer ring (including the palatine tonsils and

adenoids); the lymph nodes immediately surrounding the Waldeyer ring (occipital, postauricular, preauricular, parotid, and facial nodes); and the lymphatic tissue composed of submaxillary, submental, and jugular lymph nodes. Most head and neck lymph nodes drain into the submaxillary and deep cervical lymph nodes, making these two groups the ones most commonly involved in cervical lymphadenitis.

2. The patient's age is important in narrowing the differential diagnosis of a neck mass. In adults a newly diagnosed neck mass is generally considered a sign of malignancy (metastasis) until proven otherwise. However, cervical lymphadenitis or cervical lymphadenopathy of infectious or inflammatory origin is the most common underlying cause of a pediatric neck mass, most commonly affecting children from 1 to 5 years of age. This disease process may affect other glands in the head and neck region, as in parotitis or thyroiditis or even involve superinfection of congenital neck lesions such as branchial cleft anomalies or thyroglossal duct cysts.
3. Viruses are the most frequent cause of infectious cervical lymphadenitis in both adults and children. Acute bilateral lymphadenitis is often due to viral infection, such as from Epstein-Barr virus (EBV) or human immunodeficiency virus (HIV), although *Streptococcus pyogenes* and *Mycoplasma pneumoniae* have also been known to cause infection in this manner. Group B streptococci are often seen in newborns. The most common infectious agents in acute unilateral lymphadenitis are *Staphylococcus aureus* and *S. pyogenes*. Anaerobes may be found in patients with significant dental or periodontal disease. Recent studies have suggested an increasing incidence of drug-resistant staphylococci (MRSA) causing cervical lymphadenitis in all age groups. Fungal infections of the neck from *Candida*, *Histoplasma*, or *Aspergillus* are nearly always seen only in immunocompromised patients.
4. Chronic cervical lymphadenitis is frequently seen in mycobacterial infections, toxoplasmosis, and cat-scratch disease. *Mycobacterium tuberculosis* tends to affect adults, whereas nontuberculous mycobacteria (such as *Mycobacterium avium-intracellulare*) are more frequently seen in children. Atypical mycobacterial infections tend to affect a single tonsillar or submandibular lymph node and may cause overlying skin changes such as thinned skin and discoloration. Spontaneous drainage and sinus tract formation are seen in 10% of cases. Chest radiographs are generally normal, and Mantoux purified protein derivative skin testing usually results in 5 to 9 mm of induration (mildly positive). Cat-scratch disease is caused by the gram-negative, silver-staining bacillus *Bartonella henselae* and is usually self-limited. Toxoplasmosis is caused by the protozoan *Toxoplasma gondii* and usually presents in immunocompetent patients as discrete, firm, nontender, nonsuppurative cervical lymphadenopathy. Immunocompromised patients may develop signs of central nervous system infection such as hemiparesis and dysphasia, in addition to lymphadenitis. Diagnosing toxoplasmosis requires a tissue biopsy and serologic testing.
5. Several important noninfectious causes of cervical lymphadenitis should not be overlooked during workup. Kawasaki disease, the leading cause of acquired heart disease in children of developed countries, is diagnosed if the patient has at least 5 days of high fever and four of the following five additional criteria: acute, nonpurulent cervical lymphadenopathy (usually unilateral); edema or erythema of the extremities; polymorphous exanthema; bilateral painless conjunctivitis; and oral mucosal infectious changes. Coronary artery involvement may manifest as early as within 10 days of the onset of fever. Rosai-Dorfman disease (sinus histiocytosis) manifests in children as fever, skin nodules, and massive nontender cervical lymphadenopathy similar to infectious mononucleosis (EBV infection). Sarcoidosis is an autoimmune condition most commonly seen in the second decade of life, which manifests as cervical lymphadenopathy, constitutional symptoms, and a variety of other symptoms such as cough, dyspnea, arthralgia, visual deficits, and skin rash.

Test Interpretation

In most (uncomplicated) cases of cervical lymphadenitis, determining the precise cause of the infection is unnecessary and testing should be limited. However, in acutely ill patients or those who are unresponsive to empiric antibiotic therapy, further testing may be warranted. A complete blood count with differential may be helpful because leukocytosis may be encountered in the acute phase of infection. Blood cultures and liver function testing may be useful if there is evidence of sepsis or toxicity on examination. Mantoux skin testing may help determine whether there is a mycobacterial cause of the lymphadenitis. Fine-needle aspiration (FNA) of the neck mass remains invaluable in determining the cause and establishing appropriate medical therapy. Aspirates should be sent for Gram staining, aerobic and anaerobic cultures, acid-fast staining, and mycobacterial cultures. In cases of chronic lymphadenitis or a suspicious history and examination, additional testing with fungal cultures, methenamine-silver staining for fungi, and polymerase chain reaction testing for *B. henselae* (cat-scratch disease) should be performed (**Fig. 61.1**).

If there is suspicion of abscess formation, both computed tomography (CT) scanning with IV contrast and ultrasonography are useful in delineating the extent of involvement. Ultrasonography is able to detect air-fluid levels and can distinguish between solid and cystic structures, and it is usually the first-line imaging study in children by virtue of its being noninvasive, the ease of examination, and the lack of radiation exposure. However, ultrasonography cannot penetrate bony structures, its reproducibility is variable at best, and its overall reliability is operator dependent. Abscesses are easily identifiable on CT by their rim-enhancing appearance with central low attenuation. General neck anatomy is also much easier to characterize on CT scan; CT can generally help determine whether the affected area is due to lymphadenitis or to an infected congenital anomaly such as a branchial cleft cyst.

Diagnosis

Cervical lymphadenitis

Medical Management

Most cases of acute cervical lymphadenitis are self-limiting because they result from viral infections. Empiric antibiotic coverage for *S. aureus* and *S. pyogenes* should be adequate for initial therapy. Oral therapies include amoxicillin with or without clavulanate, clindamycin, or cephalexin; good empiric IV antibiotic coverage may include ampicillin/sulbactam, clindamycin, or cefazolin. Lack of response over the next 36 to 48 hours may require reassessment of the patient's course of treatment, including changing the antibiotic regimen or obtaining

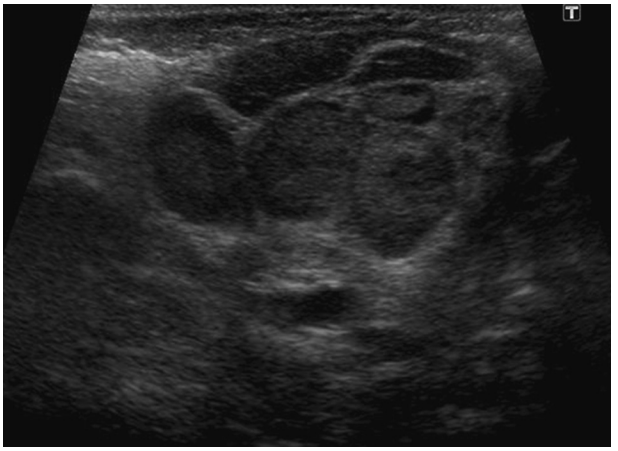

Fig. 61.1 Ultrasound of the left neck demonstrates a collection of enlarged cervical lymph nodes constituting an inflammatory mass.

imaging studies. Suspicion for atypical infections such as mycobacteria, fungi, or drug-resistant bacteria (MRSA) should warrant a change in antibiosis to cover the suspected organisms. *B. henselae* may respond well to azithromycin or trimethoprim-sulfamethoxazole in particular. Obtaining an FNA may be very helpful in guiding the selection of appropriate antibiotics.

A different approach is warranted in most noninfectious cases of cervical lymphadenitis; in many instances, management is expectant. Kawasaki disease is classically treated with aspirin and IV gamma-globulin therapy to attenuate the inflammatory response. Corticosteroids may reduce lymphadenopathy in acute flares of sarcoidosis.

◆ Surgical Management

Surgical intervention is indicated if an abscess is detected on clinical examination or imaging because routine antibiotic therapy will not eliminate the fluid collection. Simple incision and drainage are the gold standard of treatment, followed by appropriate antibiotic therapy. If nontuberculous mycobacterial infection is suspected, complete surgical excision of the affected lymph node, followed by long-term treatment with isoniazid and rifampin, is usually curative. However, in cat-scratch disease and tuberculous mycobacterial lymphadenitis (scrofula), surgical intervention may contribute to the formation of cutaneous fistulae.

◆ Rehabilitation and Follow-up

In most uncomplicated cases of cervical lymphadenitis, follow-up should be conducted only to confirm that the acute disease process has resolved. Continuous follow-up is necessary in cases where long-term antibiotic therapy is indicated, such as in mycobacterial infections. Long-term follow-up of patients with Kawasaki disease is indicated as well to monitor for signs of cardiac involvement. Immunocompromised patients should be seen on a regular basis after hospital discharge; many of these patients will be afflicted with recurrent cervical lymphadenitis and will likely need closer medical management. Any patient with a diagnosis suspicious for malignancy (on FNA or surgical biopsy of the affected lymph node) should have regular surveillance office visits, with further treatment as indicated.

Infections that required surgical intervention should also be followed up in the outpatient setting to ensure that the wound itself is healing appropriately. An infected congenital anomaly should not be surgically addressed until the acute infection has cleared.

◆ Questions

1. What is the initial step in evaluating an otherwise healthy patient who presents with tender cervical lymphadenitis?
- **A.** Complete blood count
- **B.** Chest radiography
- **C.** FNA
- **D.** Neck CT
- **E.** Empiric antibiotic therapy and closely monitor response.

2. The most common infectious agent causing acute unilateral cervical lymphadenitis is:
- **A.** Epstein-Barr virus
- **B.** *Streptococcus pyogenes*
- **C.** Histoplasmosis
- **D.** Methicillin-resistant *Staphylococcus aureus*
- **E.** *Bartonella henselae*

3. What is the best test to verify a suspected abscess complicating cervical lymphadenitis?
- **A.** Ultrasonography
- **B.** CT
- **C.** MRI
- **D.** FNA

Suggested Readings

Al-Dajani N, Wootton SH. Cervical lymphadenitis, suppurative parotitis, thyroiditis, and infected cysts. Infect Dis Clin North Am 2007;21(2):523–541

Dash GI, Kimmelman CP. Head and neck manifestations of sarcoidosis. Laryngoscope 1988;98(1):50–53

Dulin MF, Kennard TP, Leach L, Williams R. Management of cervical lymphadenitis in children. Am Fam Physician 2008;78(9):1097–1098

Freeman AF, Shulman ST. Kawasaki disease: summary of the American Heart Association guidelines. Am Fam Physician 2006;74(7):1141–1148

Hurley MC, Heran MK. Imaging studies for head and neck infections. Infect Dis Clin North Am 2007;21(2): 305–353

Montoya JG, Liesenfeld O. Toxoplasmosis. Lancet 2004; 363(9425):1965–1976

62

Zenker Diverticulum

Lee A. Zimmer

History

A 78-year-old man has a 6-month history of food getting "stuck." He reports that symptoms started with pills and solid foods sticking in the throat. He points to his neck just above the clavicle in the midline. Over the last few months, he has been having problems with frequent cough after meals and regurgitating undigested food products around 10 to 40 minutes following a meal. He denies changes in voice, odynophagia, and weight loss. He does have a history of infrequent acid reflux, which he treats with over-the-counter medications. He also was a former 1-pack-a-day smoker who quit 30 years ago.

The patient is a well-appearing 68-year-old man. Examination of the ears, nose, throat, and neck shows no evidence of tumors or ulceration. Manual compression of the neck did not result in crepitus or a gurgling sensation. The patient had no trismus and could achieve full neck extension. Flexible laryngoscopy revealed no lesions or masses in the larynx, pharynx, or hypopharynx. Mild pachydermia and posterior glottic edema with erythema were present.

Differential Diagnosis—Key Points

1. Zenker diverticulum is the top differential diagnosis in an elderly man with dysphagia and regurgitation. A history of untreated gastroesophageal reflux disease (GERD) predisposes a patient for diverticulum formation. Large diverticula often trap food products during a meal. These food products do not undergo routine breakdown from gastric enzymes. A delayed regurgitation of the pouch contents are a typical complaint.
2. Dysphagia secondary to dysfunction of the upper esophageal sphincter (cricopharyngeal dysfunction) should be considered. Individuals with a history of GERD, stroke, cranial nerve palsy, and Parkinson disease have a higher incidence of dysphagia from cricopharyngeal dysfunction.
3. Dysphagia to solids in a former smoker is a warning sign for obstructive lesions of the esophagus or mediastinum. Primary lung cancer or squamous cell carcinoma of the proximal esophagus is certainly a consideration in a tobacco user.

Test Interpretation

The patient with dysphagia and regurgitation of undigested food products should be evaluated with a barium esophagram. Barium coats the lining of the diverticulum, and a contrast-filled pouch is readily seen on radiography (**Fig. 62.1**). Irregularities within the pouch can be retained food products or rarely

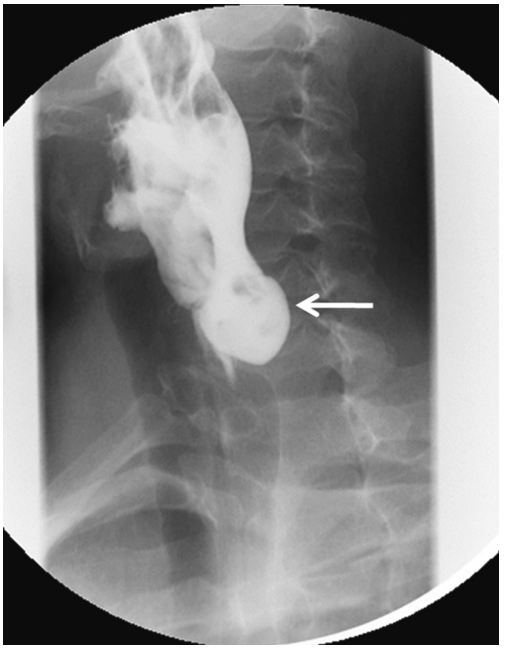

Fig. 62.1 Barium esophagram revealing a large Zenker diverticulum extending into the left side of the neck (*arrow*).

a malignancy within the diverticulum. The barium esophagram can also diagnose cricopharyngeal dysfunction. Typically a cricopharyngeal bar with transient partial obstruction is noted. Finally, a barium esophagram will often reveal mediastinal or esophageal masses compressing the proximal esophagus.

◆ Diagnosis

Zenker diverticulum

◆ Medical Management

Asymptomatic diverticula found incidentally on imaging studies can be followed up expectantly. Because most diverticula are presumed to be secondary to cricopharyngeal dysfunction in response to GERD, treatment with proton pump inhibitors is recommended. Patients should be monitored for early signs of dysphagia, such as cough, globus sensation, halitosis, and regurgitation of undigested food products. If symptoms appear, a repeat barium esophagram and surgical intervention are recommended.

◆ Surgical Management

Symptomatic Zenker diverticula of any size are treated surgically. Depending on the experience of the surgeon, an open or endoscopic approach can be performed. Diverticula smaller than 2 cm can be managed with a cricopharyngeal myotomy. With the patient under general anesthesia, a transcervical approach is used to access the cricopharyngeal muscle. A myotomy is performed in the posterior midline to prevent injury to the recurrent laryngeal nerve. Complications include esophageal perforation, unilateral vocal cord paralysis, and infection.

Larger diverticula require removal, imbrication, or pexy of the diverticulum in combination with a cricopharyngeal myotomy. The procedure begins with an esophagoscopy and the placement of packing material into the diverticulum. Packing allows for easier identification of the pouch. A transcervical approach is then performed with resection, imbrication, or pexy of the pouch. Pexy is commonly used in older, medically unstable patients to limit morbidity. Following resection, the esophagus is then primarily repaired. The cricopharyngeal myotomy is then completed, a neck drain is placed, and the neck is closed. A nasogastric tube is placed in the operating room. The nasogastric tube is removed 48 hours later after confirmation of the integrity of the esophageal repair by a barium esophagram. The patient is placed on a soft diet for 1 to 2 weeks and then advanced as tolerated to a normal diet. Complications are the same as for a cricopharyngeal myotomy.

Endoscopic techniques are also available for Zenker diverticula. The endoscopic diverticulotomy opens the diverticulum into the esophagus and completes a cricopharyngeal myotomy in the same setting. The endoscopic approach begins with a rigid esophagoscopy to evaluate the diverticulum and to rule out masses. A special rigid scope is then used to isolate the esophagus and diverticulum (**Fig. 62.2A,C**). A diverticulotomy is then

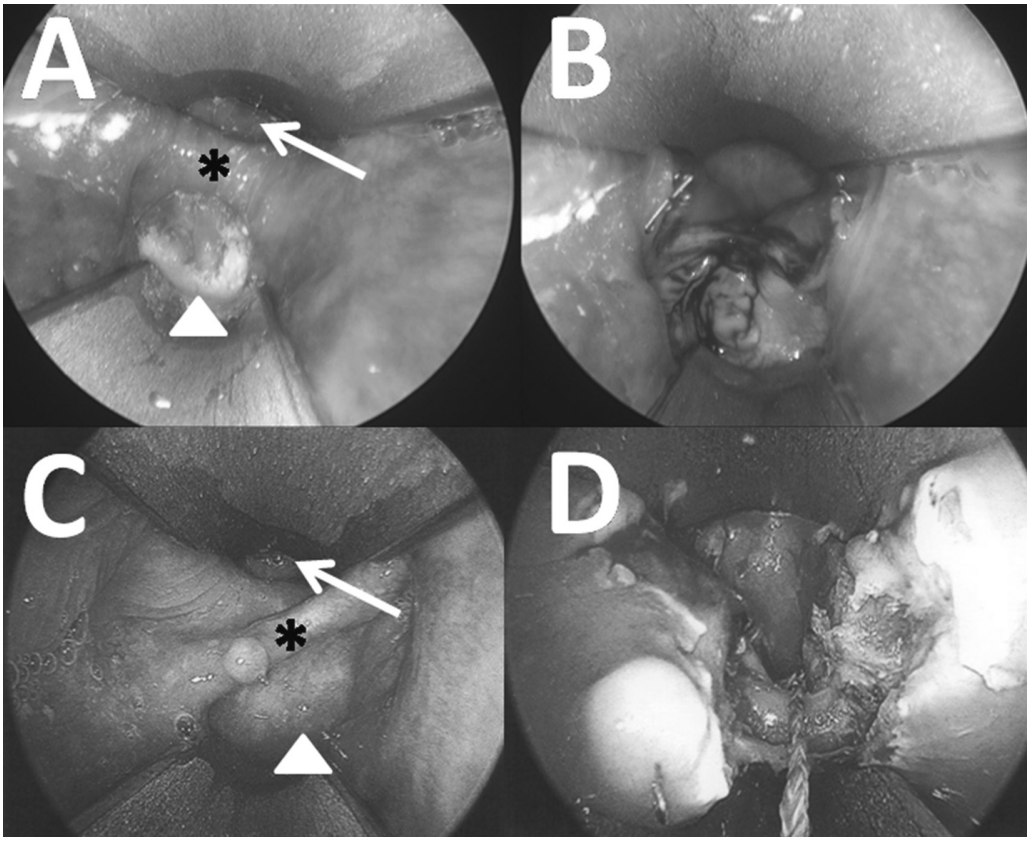

Fig. 62.2 Endoscopic views of a Zenker diverticulum. **(A)** Exposure includes the proximal esophagus (*arrow*), diverticulum (*arrowhead*), and common party wall, which includes the cricopharyngeal muscle (*asterisk*). **(B)** Using a GIA-30 stapler. **(C)** Exposure includes the proximal esophagus (*arrow*), diverticulum (*arrowhead*), and common party wall, which includes the cricopharyngeal muscle (*asterisk*). **(D)** Using the harmonic scalpel.

performed with either a CO_2 laser, endoscopic stapler (**Fig. 62.2B**), or harmonic scalpel (**Fig. 62.2D**). The endoscopic stapler can be used on diverticula larger than 2.5 cm. The advantage of the stapler includes (1) simple, efficient diverticulotomy; (2) reliable stapling and sealing of cut mucosal edges; (3) low risk of esophageal perforation or mediastinitus; and (4) outpatient surgical setting. The disadvantages of the stapling technique include the inability to perform on small diverticula (<2.5 cm) and a 5- to 10-mm mucosal ridge at the base of the pouch. The remnant edge may cause persistent or recurrent symptoms. The advantage of the harmonic scalpel or CO_2 laser is complete diverticulotomy without a remnant edge at the pouch's base. This has the potential of limiting failures seen with the stapler. Disadvantages include (1) a higher risk of perforation and mediastinitus, (2) greater risk of scarring leading to esophageal stenosis, and (3) greater length of hospital stay. Following CO_2 laser and harmonic scalpel diverticulostomy, patients are placed on antibiotic prophylaxis for 5 days and a soft diet for 1 week.

◆ Rehabilitation and Follow-up

Patients are commonly seen in the office 1 to 2 weeks after surgery. Commonly patients describe an immediate cessation of dysphagia

and regurgitation. A head and neck examination is performed to rule out neck crepitus and unusual neck pain (abscess formation). If significant neck pain is elicited, a CT neck with contrast is warranted to rule out postoperative abscess formation. Patients are then instructed to return to the office with any new complaints of dysphagia.

Questions

1. Which of the following is not true about an endoscopic Zenker diverticulotomy with a stapler?
- **A.** Simple and efficient performance of a diverticulotomy
- **B.** Reliable stapling and sealing of cut mucosal edges
- **C.** Low risk of esophageal perforation
- **D.** Greater risk of scarring leading to esophageal stenosis than an open procedure
- **E.** Inability to perform a diverticulotomy on pouches less than 2.5 cm is a disadvantage

2. Which of the following statements concerning open diverticulectomy is false?
- **A.** Rigid esophagoscopy with placement of packing in the Zenker pouch allows easier identification of the pouch during an open approach.
- **B.** Imbrication of the pouch is an effective, long-term treatment for Zenker diverticulum.
- **C.** Removal of the pouch increases the risk of esophageal stenosis in the postoperative period.
- **D.** The pexy approach excludes the need for cricopharyngeal myotomy, decreasing the length of time for surgery.

3. Diagnosis of Zenker diverticulum includes all of the following except:
- **A.** Barium esophagram
- **B.** History and physicial examination
- **C.** Modified barium esophagram
- **D.** Esophagoscopy

4. A Zenker diverticulum is present in which portion of the esophagus?
- **A.** Proximal
- **B.** Mid
- **C.** Mid and proximal
- **D.** Distal

Suggested Readings

Bonavina L, Bona D, Abraham M, Saino G, Abate E. Long-term results of endosurgical and open surgical approach for Zenker diverticulum. World J Gastroenterol 2007;13(18):2586–2589

Ferreira LE, Simmons DT, Baron TH. Zenker's diverticula: pathophysiology, clinical presentation, and flexible endoscopic management. Dis Esophagus 2008;21(1):1–8

Miller FR, Bartley J, Otto RA. The endoscopic management of Zenker diverticulum: CO_2 laser versus endoscopic stapling. Laryngoscope 2006;116(9):1608–1611

Palmer AD, Herrington HC, Rad IC, Cohen JI. Dysphagia after endoscopic repair of Zenker's diverticulum. Laryngoscope 2007;117(4):617–622

Visosky AM, Parke RB, Donovan DT. Endoscopic management of Zenker's diverticulum: factors predictive of success or failure. Ann Otol Rhinol Laryngol 2008;117(7):531–537

Wirth D, Kern B, Guenin MO, et al. Outcome and quality of life after open surgery versus endoscopic stapler-assisted esophagodiverticulostomy for Zenker's diverticulum. Dis Esophagus 2006;19(4):294–298

63

Ludwig Angina

Brandon Scott Hopkins and Allen M. Seiden

History

A 28-year-old woman arrives in the emergency department in mild distress. Her speech is garbled, she is drooling, and she complains of difficulty swallowing. On questioning, she notes that this has been worsening over the last 24 hours and points out that about 1 week ago she had an infection of her second left molar, which was treated with penicillin. She gives a history of diabetes, normally well-controlled with her insulin. She denies other medical problems and does not smoke.

On physical examination, the patient has a firm anterior neck and submental region, a firm floor of mouth, an elevated tongue, and mild trismus. She currently has no signs of respiratory distress, including no dyspnea, no retractions, no cyanosis, and an oxygen saturation of 95% on room air.

Within minutes the patient becomes more anxious, and her breathing becomes labored. The decision is made to take the patient to the operating room for an elective flexible fiberoptic intubation. After unsuccessful visualization of the larynx with the flexible scope, the patient's airway is obtained via an awake tracheostomy.

Differential Diagnosis—Key Points

1. The rapid progression of this patient's symptoms clearly suggests an inflammatory cause. Induration of the submental region and floor of mouth should raise concern for the possibility of Ludwig angina, as opposed to tonsillitis, epiglottitis, or other pharyngeal or deep neck space infection.
2. Ludwig angina is named after the German physician Wilhelm Frederick von Ludwig, who first described the condition in 1836. The word *angina* derives from the Greek word *ankhon*, which means "strangling," and aptly emphasizes the feeling patients have and the potential for airway compromise in this condition.
3. The hallmarks of Ludwig angina are dysphagia, a change in speech, a rapidly expanding cellulitis associated with submandibular and submental neck swelling that is classically very hard and tense, and floor-of-mouth firmness. Pain, fever, fetid breath, and trismus are common. The sublingual edema pushes the tongue upward and backward, leading to airway compromise. This is often an insidious process but can abruptly lead to an airway emergency.

4. Angioedema may present with similar speech difficulties, drooling, and difficulty swallowing. However, this tends to involve edema of the tongue, lips, uvula, or oropharynx. It is less traditionally associated with anterior neck or floor-of-mouth swelling, the edema tends to be more superficial and not characteristically indurated, and it is often more acute in onset. It frequently follows the use of certain medications, such as angiotensin-converting enzyme inhibitors. Management of the airway should be treated in a similar fashion.
5. Ludwig angina is essentially a compartment syndrome of the sublingual and submandibular spaces, which are enclosed by the rigid superficial layer of the deep cervical fascia. It generally begins in the sublingual space, unilaterally, with the spread of infection limited anteriorly by the mandible and inferiorly by the mylohyoid muscle. Therefore the infection extends superiorly and posteriorly, elevating the tongue and floor of mouth. Infection may also spread more posteriorly to the parapharyngeal space or inferiorly to the anterior visceral compartment of the neck. Potentially inciting causes include pharyngitis, tonsillitis, sialadenitis, tongue piercing and oral trauma, but its most common source (75–90%) is an odontogenic infection involving the second or third molars. Their roots lie below the attachment of the mylohyoid to the mandible and thus cross the sublingual and submandibular spaces. Low socioeconomic status and poor oral hygiene are also associated with this infection. Comorbid conditions predisposing to Ludwig angina have included diabetes mellitus, malnutrition, alcoholism, neutropenia, lupus erythematosus, aplastic anemia, and glomerulonephritis.
6. The bacteriology of Ludwig angina tends to be polymicrobial. Organisms cultured in Ludwig angina are often gram-positive cocci, gram-negative rods, and anaerobes, most commonly including *Streptococcus* spp., *Staphylococcus aureus, Bacteroides* spp., *Fusobacterium* spp., *Actinomyces* spp., and *Haemophilus influenzae.*

◆ Test Interpretation

Imaging with either ultrasound or computed tomography may be helpful to evaluate for areas of abscess and to assess the extent of infection (**Fig. 63.1**). Plain radiographs may also be helpful to evaluate for gas-producing organisms and mediastinal involvement

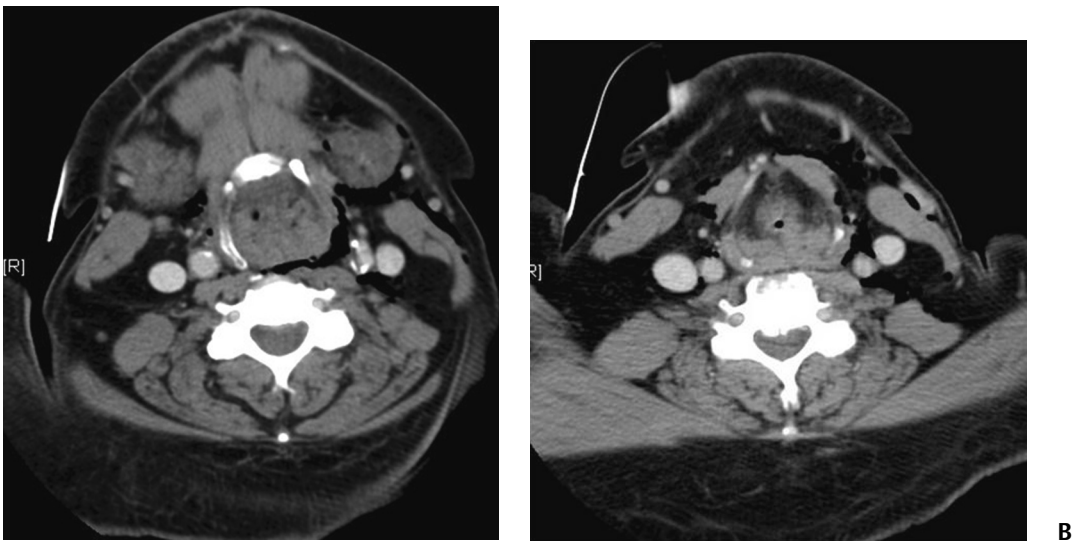

Fig. 63.1 Axial computed tomography scans in a patient with Ludwig angina demonstrate sublingual **(A)** and submandibular pre-epiglottic and supraglottic **(B)** swelling with tissue stranding, without an associated abscess.

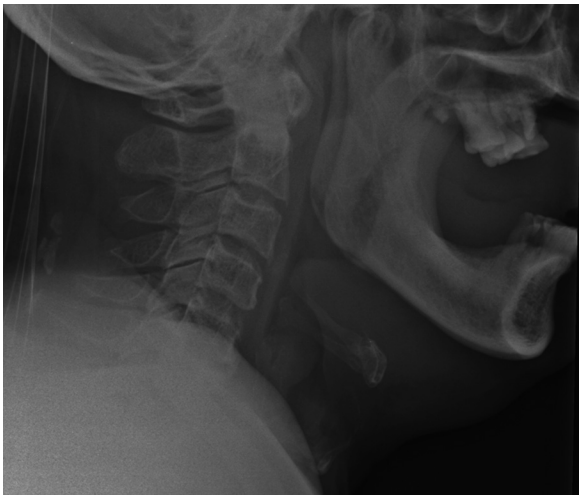

Fig. 63.2 Plain lateral neck radiograph of the same patient in Fig. 63.1 demonstrates submental and sublingual edema pushing the tongue superiorly and posteriorly.

(**Fig. 63.2**). It must be remembered that placing a Ludwig angina patient supine for a CT scan before securing their airway is risky.

◆ Diagnosis

Ludwig angina

◆ Medical Management

As mentioned, Ludwig angina should be considered an airway emergency. A safe and traditional approach is not to wait for dyspnea, cyanosis, or desaturation before obtaining an airway. This is accomplished via tracheostomy. During the past several years, however, alternatives have been safely used in the airway management of these patients. For instance, flexible fiberoptic nasal intubation has been shown to be a good choice for initial airway management, with airway control being obtained in nearly 75% of patients. The risks involved with this include abscess perforation and exacerbation of airway edema with resulting airway compromise. Choice of how to obtain an airway should be tailored to each patient and based on physician experience.

Steroids can also play a key role in the airway management of these patients by decreasing airway inflammation and edema. However, care must be taken when using steroids because their hyperglycemic effect can be a problem in the diabetic population.

Antibiotic coverage should be initiated. The overall incidence of Ludwig angina has decreased with the use of antibiotics for early odontogenic and oral infections. Antibiotics have also decreased the mortality rate from 50% in the preantibiotic era to 8% today. Broad-spectrum antibiotics should be started quickly and narrowed after cultures and sensitivities return. Common choices include penicillin with a β-lactamase inhibitor, a β-lactamase–resistant antibiotic (cefoxitin, cefuroxime, imipenem, or meropenem), an aminoglycoside, or chloramphenicol. These should be combined with anaerobic coverage by using either metronidazole or clindamycin. Intravenous antibiotic coverage should be continued until resolution begins, after which the patient can be switched to appropriate oral antibiotics.

◆ Surgical Management

The preferred method for obtaining a surgical airway is an awake tracheotomy with the patient under local anesthesia. Excessive supraglottic edema and distortion make oral and nasal intubation very difficult. An awake tracheostomy is also beneficial because you can leave the patient sitting upright and lessen the chance of airway compromise that would tend

to occur in the supine position. It has been shown that up to 33 to 75% of Ludwig angina cases require a surgical airway. In preparation for obtaining a surgical airway, the patient should be brought to an operating room, but if a patient's airway is rapidly deteriorating, time should not be wasted in transport. Lidocaine with epinephrine is infiltrated 3 to 4 cm in a horizontal skin crease halfway between the cricoid and the sternal notch. An incision can then be made with the patient sitting upright on 100% FIO_2 with minimal sedation. No sedation should be used if it is suspected that this will worsen the patient's respiratory status. A tracheostomy is then efficiently performed in the standard fashion. In particularly urgent settings, however, a cricothyrotomy should be considered.

Early in the disease process airway management is of utmost importance and usually requires intervention. However, the cellulitis may respond to antibiotic therapy. Although frank abscess usually does not occur, if the cellulitis progresses and does not show signs of resolution within 24 to 48 hours of antibiotic coverage, drainage may be needed. This typically requires multiple horizontal incisions across the submandibular and submental region, releasing so-called "dishwater" fluid rather than frank pus and revealing a woody induration. Care should be taken to avoid injury to the marginal mandibular branch of the facial nerve. Preference for drainage varies in the literature and can include suction drains, Penrose drains, or packing. If the infection has spread to one of the other deep neck spaces and an abscess does develop, this should be drained. Of note, pediatric populations seem more amenable to resolution with solely antibiotic therapy; however, surgical drainage may still be necessary if no resolution is seen within 36 hours.

Rehabilitation and Follow-up

After a patient's acute infection and airway protection have been addressed during hospitalization, follow-up as an outpatient while on oral antibiotics is important to monitor for complete resolution. Patients requiring tracheostomy should be decannulated after resolution of infection per standard protocol. Adjunctive strategies, including good oral hygiene and control of co-morbid conditions, including diabetes and immunodeficiency, are recommended.

Questions

1. What is the most significant predisposing factor associated with Ludwig angina?
- **A.** Diabetes
- **B.** Immunodeficiency
- **C.** Alcoholism
- **D.** Poor dentition

2. What is the first step in managing a patient who presents with Ludwig angina?
- **A.** Initiate antibiotics
- **B.** Extract infected molars and drain tooth abscess
- **C.** Obtain a computed tomography scan
- **D.** Secure the airway
- **E.** Bring to the operating room for incision and drainage

3. Distinguishing features of Ludwig angina include which of the following?
- **A.** Edema of the tongue
- **B.** Superficial, compressible edema of the upper neck
- **C.** Rapidly expanding tense cellulitis of the submental and sublingual regions
- **D.** Stridor

Suggested Readings

Bansal A, Miskoff J, Lis RJ. Otolaryngologic critical care. Crit Care Clin 2003;19(1):55–72

Britt JC, Josephson GD, Gross CW. Ludwig's angina in the pediatric population: report of a case and review of the literature. Int J Pediatr Otorhinolaryngol 2000;52(1):79–87

Bross-Soriano D, Arrieta-Gómez JR, Prado-Calleros H, Schimelmitz-Idi J, Jorba-Basave S. Management of Ludwig's angina with small neck incisions: 18 years experience. Otolaryngol Head Neck Surg 2004;130(6):712–717

Kremer MJ, Blair T. Ludwig angina: forewarned is forearmed. AANA J 2006;74(6):445–451

Lawson W, Reino A, Westreich R. Odontogenic infections. In: Bailey B, Johnson J, Newlands S, eds. Head & Neck Surgery—Otolaryngology. Philadelphia, PA: Lippincott Williams & Wilkins; 2006:619

Marple BF. Ludwig angina: a review of current airway management. Arch Otolaryngol Head Neck Surg 1999; 125(5):596–599

Vieira F, Allen SM, Stocks RM, Thompson JW. Deep neck infection. Otolaryngol Clin North Am 2008;41(3): 459–483

64

Sjögren Syndrome

Gordon H. Sun and Allen M. Seiden

History

A 54-year-old white woman was referred to the otolaryngology clinic for evaluation of persistent dry mouth starting 4 months ago. She stated that she has difficulty swallowing solid foods without drinking large amounts of water. The patient had also had problems with dry eyes and has been using over-the-counter moisturizing eyedrops frequently. The patient denied pain with swallowing, reflux, or aspiration. Her voice had not undergone any recent change, although on rare occasion if her mouth was particularly dry, her voice might sound mildly hoarse. She also complained of chronically cracked, chapped lips. She denied joint and muscle pain, skin rashes, or hearing loss.

Her medical history was significant only for hypertension, which was well-controlled with hydrochlorothiazide and lisinopril. Family history was significant for multiple family members with arthritis and hypertension and a maternal aunt who was hospitalized for treatment of gastric ulcers at age 44. The patient was a nonsmoker and drank alcohol only rarely. Other than antihypertensives and eye lubricant, she was not using any other medications. She reported having seasonal allergies only.

On physical examination, the patient was afebrile, alert, and fully oriented. Her conjunctivae were mildly injected, although her vision was normal and extraocular movements appeared fully intact. The nasal passages were clear, although the mucosal lining appeared dry. Oral examination demonstrated very dry oropharyngeal mucosa and patchy red and white, mildly painful lesions overlying the palate. Two dental caries were found. The parotid glands were mildly enlarged but symmetric and nontender to palpation. No cervical lymphadenopathy or thyromegaly was appreciated. Flexible nasopharyngoscopy and laryngoscopy were unremarkable. Schirmer test and a lower-lip biopsy were performed, and blood work was obtained.

Differential Diagnosis—Key Points

1. In this patient, dysphagia is most likely related primarily to the patient's xerostomia (dryness of the mouth). It is important to ascertain whether other medical problems might be contributing to dysphagia, such as an underlying neurologic disorder, gastroesophageal reflux disease, or lesions of the pharynx or esophagus.
2. Xerostomia has numerous causes. These include primary salivary gland dysfunction from granulomatous disease, human immunodeficiency virus (HIV) infection, Sjögren syndrome, or chronic sialadenitis;

dehydration; diabetes mellitus; certain drugs such as antihypertensives, antihistamines, antidepressants, antipsychotics, and diuretics; alcoholism; hypothyroidism; and radiation therapy to the head and neck. In patients at high risk for HIV infection, consider HIV testing. Minor salivary gland biopsy can help rule out primary salivary gland disorders such as Sjögren syndrome and sarcoidosis.

3. The presence of xerophthalmos (dry eyes) should raise the index of suspicion for Sjögren disease. Tear-deficient dry eye is classified by ophthalmologists into Sjögren-related and non-Sjögren tear deficiency, the latter of which is generally associated with systemic medications, graft-versus-host disease, other autoimmune processes, or HIV infection. The Schirmer test measures aqueous tear flow and can be done rapidly in the office. Less than 5 mm of wetting of the test strip after 5 minutes is associated with a clinically significant decrease in tear production. Rose Bengal dye staining of the ocular surface may be able to detect ocular abnormalities such as corneal ulcerations.
4. Sometimes, but not always, patients with Sjögren disease present with associated complaints of myalgia or arthralgia. Only about 6% of Sjögren's patients with myalgias demonstrate muscle enzyme elevation consistent with acute myositis. If present, the arthralgias are typically symmetric, although radiographically the only sign that is visible might be minimal joint-space narrowing, without apparent bony erosion.
5. Sjögren syndrome is an autoimmune disorder characterized by xerostomia and xerophthalmos caused by chronic inflammation of the salivary and lacrimal glands; it is sometimes termed the *sicca complex*. The estimated prevalence of Sjögren syndrome in the U.S. population is estimated at 1 to 3%, with a 9:1 female-to-male preponderance, making this the second most common autoimmune disease, behind rheumatoid arthritis. The average age of onset is between 40 and 60 years of age. If xerostomia and xerophthalmos occur when not in association with other autoimmune disorders, it is termed *primary* Sjögren syndrome. *Secondary* Sjögren syndrome is seen when the salivary and lacrimal glands are inflamed in conjunction with a different rheumatologic disorder, most commonly rheumatoid arthritis.
6. Sjögren syndrome affects numerous tissues and organs, including the inner ear, larynx, thyroid, sinonasal passages, lungs, gastrointestinal and hepatobiliary tracts, kidney and bladder, musculoskeletal system, central nervous system, and circulatory system. Chronic candidiasis is sometimes seen in the oral cavities of Sjögren patients. In this patient, oral candidiasis manifested as patchy leukoplakia and erythroplakia overlying the soft palate. One of the most ominous aspects of Sjögren syndrome is the increased risk of non-Hodgkin lymphoma, up to 44 times greater than that of the general population. Non-Hodgkin lymphoma of the salivary glands may present as a painless, progressively enlarging mass.

◆ Test Interpretation

Obtaining serum electrolytes and a fasting glucose level will exclude diabetes, dehydration, and diuretic use as potential causes of the patient's sicca complex. Normal thyroid-stimulating hormone levels will exclude hypothyroidism. A complete blood count may detect anemia, a nonspecific finding sometimes present in patients with Sjögren syndrome. Rheumatologic testing will help identify the disease process as primary or secondary Sjögren syndrome. Anti–SS-A and anti–SS-B are serum autoantibodies to nuclear antigens Ro and La, respectively, and are found in a substantial number of patients with primary or secondary Sjögren disease.

An autoimmune workup will generally yield the following:

Elevated erythrocyte sedimentation rate: This is a nonspecific finding.

Positive for rheumatoid factor: This test may be positive even in Sjögren patients without rheumatoid arthritis.

Positive for antinuclear antibodies: This is a nonspecific finding.

Negative for antiphospholipid antibodies

Positive for anti–SS-A/Ro antibodies

Positive for anti–SS-B/La antibodies

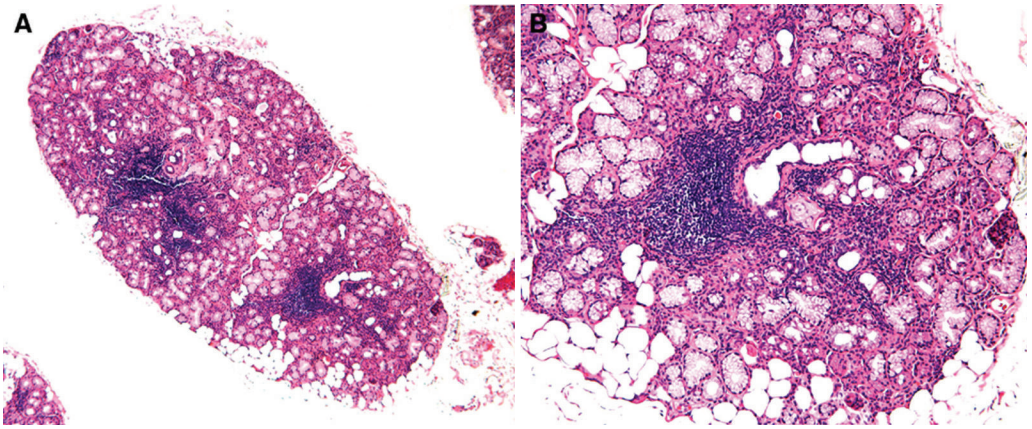

Fig. 64.1 **(A)** Minor (labial) salivary gland biopsy demonstrates multiple lymphocytic foci with intact acinar units, consistent with Sjögren syndrome (H&E staining, 5x magnification). **(B)** Minor salivary gland biopsy consistent with Sjögren syndrome (H&E staining, 10x magnification). (From Stewart CM, Bhattacharyya I, Berg K, et al. Labial salivary gland biopsies in Sjögren's syndrome: still the gold standard? Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2008;106(3):392–402.)

A minor salivary gland biopsy remains the gold standard for pathologic diagnosis of Sjögren syndrome. The classic findings are focal lymphocytic or plasma cell infiltrates as well as acinar and ductal cell destruction (**Fig. 64.1**).

◆ Diagnosis

Primary Sjögren syndrome

◆ Medical Management

The mainstays of symptomatic medical therapy for the sicca complex are artificial tears and salivary stimulants or oral lubricants. These medications are important in maintaining good oral hygiene and preventing irreversible damage to the dentition and corneas. Fluoride supplementation may have a role in preventing subgingival caries and is thus recommended. Mechanical stimulation, such as gum chewing, and sialagogues, including citrus drops and juices, can help stimulate residual saliva production. Pilocarpine was the first muscarinic M3 receptor agonist The Food and Drug Administration approved for the treatment of Sjögren-related xerostomia. The typical dose is 5 mg orally, four times daily. A more recently developed drug called cevimeline has a much higher half-life than pilocarpine, and because of its decreased affinity for M2 receptors, demonstrates a more minimal risk of cardiac side effects.

◆ Surgical Management

For the immediate treatment of Sjögren syndrome, surgery is limited to performing a minor salivary gland tissue biopsy. However, Sjögren syndrome is associated with a greatly increased risk of malignant lymphoproliferative disorders, even years after the original diagnosis is made. Up to 80% of these malignancies are ultimately diagnosed as mucosa-associated lymphoid tissue lymphomas. Parotidectomy or other major salivary gland excision may be indicated if there is a suspicious enlargement or change in the gland in question.

◆ Rehabilitation and Follow-up

Xerostomia is a lifelong problem for patients with Sjögren syndrome. Because of the possibility of severe dental and periodontal disease associated with xerostomia, close follow-up and medical management in association with a

dentist are highly recommended. An ophthalmologist should be consulted for regular eye examinations to monitor for ocular sequelae such as keratitis and corneal ulcerations. If an underlying rheumatologic disorder is diagnosed during workup, the patient's management should be coordinated with a rheumatologist. Continuous follow-up is recommended for Sjögren patients not only for routine care of sicca complex–related symptoms but also because of the substantial long-term risk for developing salivary gland or lymphoproliferative malignancies.

◆ Questions

1. Primary Sjögren syndrome is characterized by xerophthalmia and xerostomia. Secondary Sjögren syndrome has the same characteristics with the addition of which of the following?
   A. Renal failure
   B. Non-Hodgkin lymphoma
   C. Rheumatoid arthritis
   D. Candidiasis

2. Sjögren syndrome is associated with an increased risk of:
   A. Glaucoma
   B. Lymphoma
   C. Adenoid cystic carcinoma
   D. Renal failure

3. What is the most definitive test for diagnosing Sjögren syndrome?
   A. Schirmer test
   B. Anti–SS-A, anti–SS-B autoantibodies
   C. Erythrocyte sedimentation rate
   D. Lip biopsy
   E. Antinuclear antibodies

Suggested Readings

Freeman SR, Sheehan PZ, Thorpe MA, Rutka JA. Ear, nose, and throat manifestations of Sjögren's syndrome: retrospective review of a multidisciplinary clinic. J Otolaryngol 2005;34(1):20–24

Mahoney EJ, Spiegel JH. Sjögren's disease. Otolaryngol Clin North Am 2003;36(4):733–745

Roh JL, Huh J, Suh C. Primary non-Hodgkin's lymphomas of the major salivary glands. J Surg Oncol 2008;97(1):35–39

Stewart CM, Bhattacharyya I, Berg K, et al. Labial salivary gland biopsies in Sjögren's syndrome: still the gold standard? Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2008;106(3):392–402

Zintzaras E, Voulgarelis M, Moutsopoulos HM. The risk of lymphoma development in autoimmune diseases: a meta-analysis. Arch Intern Med 2005;165(20):2337–2344

XIII

Sleep

65

Snoring

David L. Steward

History

A 35-year-old man is sent in by his bed partner with complaints of snoring. He reports recent weight gain of 10 lb with worsening of his snoring. The patient denies a history of witnessed apnea or excessive daytime sleepiness. He denies insomnia and routinely sleeps 7 hours nightly. He underwent adenotonsillectomy as a child because of concerns for obstructive breathing. He has a family history of snoring. He drinks alcohol on occasion, which exacerbates his snoring. He is taking no medications and has no known medical problems.

Physical examination reveals an overweight man (body mass index 30 kg/m^2) with a neck circumference of 16.5 inches. Oropharyngeal examination demonstrates elongated and thickened soft palate and uvula, with the free edge of the soft palate visible without use of a tongue depressor or requiring phonation (modified Mallampati/Friedman tongue position I). Nasal examination is unremarkable.

Differential Diagnosis—Key Points

The primary diagnostic dilemma in a patient complaining of snoring is to exclude obstructive sleep apnea; factors increasing the likelihood of sleep apnea include obesity, neck circumference greater than 17 inches, witnessed apnea, excessive daytime sleepiness, male gender, and family history of sleep apnea. Because of the insensitivity of history and physical examination alone to exclude sleep apnea, sleep testing or polysomnography is often needed to exclude it.

The location of the source of the snoring is usually but not always the soft palate. Mouth breathing increases turbulent airflow, which worsens snoring.

Test Interpretation

Flexible fiberoptic nasopharyngoscopy is performed in the sitting and supine position to assess degree of retropalatal and possibly retroglossal pharyngeal obstruction. Volitional snoring during fiberoptic examination can reveal the site of vibration and source of the snoring.

The degree of daytime sleepiness can be assessed with the Epworth Sleepiness Scale, an eight-item questionnaire that queries the patient about the likelihood of falling asleep during the day in eight scenarios scored from 0 (none) to 3 (often). An Epworth score of 10 or more is consistent with excessive daytime sleepiness.

Polysomnography can be performed in the sleep laboratory or in the home. The laboratory provides more intensive monitoring in an

attended setting but might not as accurately reflect the patient's normal sleep patterns as home testing. Multiple night testing reduces problems of night-to-night variability but is prohibitively expensive in the sleep laboratory. Home testing is ideal for multiple-night testing, however. Apnea is defined as a cessation of breathing for 10 seconds or longer. It is deemed obstructive if associated with a respiratory effort. Hypopnea definitions vary but include partial reduction in airflow of at least 30 to 50% with an associated drop in oxygen saturation of 4% or an associated arousal or change from a deeper to lighter stage of sleep. The apnea–hypopnea index is a measure of the number of times per hour that a patient completely or partially stops breathing. An index of 5 or less is considered normal, whereas greater than that suggests the presence of sleep apnea.

Home sleep testing with a commercially available SNAP (SNAP Diagnostics, Glenview, IL) unit can provide information about the proportion of snoring that is palatal based on acoustic analysis of the snoring. This information may predict the likelihood of successful treatment of snoring with palatal procedures.

Flexible fiberoptic nasopharyngoscopy reveals palatal oscillation with volitional snoring consistent to the palatal source for the snoring. Epworth Sleepiness Scale score was 7, which is considered normal. Home sleep testing excluded sleep apnea and suggested that over 90% of the snoring was palatal.

Diagnosis

Primary snoring with primary palatal involvement

Medical Management

Behavior modification is the first line of therapy for snoring and should include weight loss when indicated. Avoidance of sedatives such as alcohol is important. Observance of good sleep hygiene with avoidance of sleep deprivation is helpful. Attempts to sleep in a lateral rather than supine position may reduce snoring for some patients. Further attempts to breathe through the nose rather than the mouth may also help.

Pharmacologic management of snoring is limited at present but may include inhaled nasal steroids for patients with turbinate hypertrophy with or without allergic rhinitis. Medical devices can be quite effective, however. Nasal dilators may help patients with nasal obstruction, especially if nighttime breathing can change from mouth to nose to improve laminar airflow and reduce palate vibration from turbulent flow.

Oral appliances that generally pull the tongue or jaw forward can be very effective but require nightly adherence to therapy. These devices can cause dental or temporomandibular joint problems and should be custom-fit to reduce these potential side effects.

Continuous positive airway pressure acts as a dilator or splint for the pharyngeal airway, which resolves snoring in most people. However, expense of the device and requirement for nightly use limit acceptance of this therapy for most patients.

Surgical Management

Surgical management of snoring can be effective for some patients, with palate treatment the most beneficial. Surgical management of snoring is almost never curative, and patients should be informed that surgical alteration of the soft palate is more likely to reduce the volume and severity of snoring than resolve it completely. Uvulopalatopharyngoplasty involves general anesthesia with resection of the uvula and advancement of the soft palate anteriorly. It is effective for snoring, but cost and pain limit utility for simple snoring. Laser-assisted and cautery-assisted palate procedures are modifications of uvulopalatopharyngoplasty that can be performed in the office with the patient under local anesthesia and can improve snoring but are still associated with significant posttreatment pain.

Less painful palate procedures for snoring have been developed that aim to stiffen rather than resect portions of the soft palate and include sclerotherapy, radiofrequency therapy, and palate implants. These procedures have been shown to be similarly effective with modest results but are generally well tolerated by patients and easy to perform in the office. Sclerotherapy, with Sotradecol (AngioDynamics,

Inc., Queensbury, NY) or alcohol, is less predictable in scarring and can result in palatal fistulas, which generally resolve spontaneously. Radiofrequency can be performed with or without temperature control with predictable results but may cause transient mucosal ulceration and requires at least two treatment sessions to optimize effectiveness. Palate implants can be accurately placed at the hard and soft palate junction but may extrude or become infected in a minority of patients. Uvulectomy, partial or complete, at the time of radiofrequency or palate implants may further improve snoring outcomes but may increase the pain and prolong healing somewhat.

Patients with severe nasal obstruction from polyps will benefit from sinonasal surgery. Patients with less severe nasal obstruction from septal deviation or turbinate hypertrophy see limited improved in snoring with nasal surgery but may benefit from improvement in nasal symptoms.

◆ Rehabilitation and Follow-up

This patient underwent temperature-controlled radiofrequency palate therapy with partial uvulectomy, and his snoring improved. He increased his exercise and reduced caloric intake and lost 15 lb, which contributed to his improvement in snoring. Continued avoidance of weight gain will be important to reduce snoring recurrence, a common problem. Re-treatment with radiofrequency or other palatal procedures would be an option if symptoms again worsen.

◆ Questions

1. Which of the following is most important to exclude before making a diagnosis of primary snoring?
   - **A.** Obesity
   - **B.** Narcolepsy
   - **C.** Obstructive sleep apnea
   - **D.** Periodic limb movement
   - **E.** Bipolar disorder
2. In the presence of heroic snoring and recent polysomnography, which of the following is consistent with primary snoring?
   - **A.** Apnea–hypopnea index less than 5
   - **B.** Apnea–hypopnea index greater than 5 and less than 15
   - **C.** Apnea–hypopnea index greater than 15 and less than 30
   - **D.** Apnea–hypopnea index greater than 30
3. Which of the following soft palate procedures offers snoring improvement with office-based treatment and low pain and patient morbidity?
   - **A.** Uvulopalatopharyngoplasty
   - **B.** Laser-assisted uvulopalatoplasty
   - **C.** Radiofrequency
   - **D.** Turbinectomy

Suggested Readings

Savage CR, Steward DL. Snoring: a critical analysis of current treatment modalities. Does anything really work? Curr Opin Otolaryngol Head Neck Surg 2007;15(3): 177–179

Friedman M, Woodson T. New techniques and approaches to sleep apnea. Oper Tech Otolaryngol Head Neck Surg 2006;17(4):215–272

66

Obstructive Sleep Apnea

David L. Steward

History

A 57-year-old man is complaining of snoring and daytime sleepiness. His wife reports witnessed apnea. His symptoms have worsened over the past few years, and he has gained weight during this period. His father snored. He denies nasal obstruction. He falls asleep immediately on lying down and sleeps 8 hours nightly but awakes feeling unrefreshed, sometimes with headache or muscle aches. He has nodded off while driving but has never had a motor vehicle accident as a result. He has systemic hypertension requiring antihypertensive therapy. He denies coronary artery disease but is on statin therapy for hyperlipidemia.

Physical examination reveals an obese man (body mass index [BMI] 35 kg/m^2) with a short, thick neck (18 inches in circumference). Oropharyngeal examination reveals a large tongue obscuring the pharynx and soft palate (modified Mallampati class or Friedman tongue position III) with dental indentations laterally. Use of a tongue depressor reveals a thickened uvula with small 1+ tonsil. Nasal examination and the remainder of his examination are unremarkable.

Differential Diagnosis—Key Points

The patient complains of snoring but has signs and symptoms of obstructive sleep apnea syndrome (OSAS). Obstructive sleep apnea (OSA) results from pharyngeal airway collapse during sleep. Confirmatory sleep testing (polysomnography) is needed to establish the presence of obstructive sleep apnea, but the diagnosis of OSAS includes the presence of symptoms of daytime sleepiness.

Symptoms of OSAS are related to sleep fragmentation from the repeated arousals necessary to reestablish airway patency. Many patients with severe OSAS never achieve the restorative deeper stages of sleep associated with REM sleep and dreaming. His greatest risk from untreated OSAS may be from motor vehicle accidents, which are increased sevenfold in these patients, and his history of falling asleep while driving is of concern.

Cardiopulmonary effects are also worrisome given his history of hypertension and hyperlipidemia. Oxyhemoglobin desaturation may relate to hypertension and cardiovascular stress from the release of norepinephrine to arouse the patient to resume breathing. Cardiopulmonary

complications are more common in obesity-hypoventilation syndrome, which can co-exist in morbidly obese patients with OSA.

Test Interpretation

Flexible fiberoptic nasopharyngoscopy in the supine position may identify areas of potential pharyngeal collapse during sleep, predominantly both retropalatal and retroglossal in most (80%) adult OSA patients. The Mueller maneuver, having the patient suck in against a closed nose and mouth, may further reveal lateral pharyngeal wall collapse. Sleep nasendoscopy has been reported to be more specific than awake fiberoptic examination, but it generally requires sedation in an endoscopy suite or operating room and is not routinely performed. The Epworth Sleepiness Questionnaire can determine the degree of daytime sleepiness present, with a score above 10 considered excessive.

Polysomnography is the gold standard for diagnosis of OSA (see snoring case). The frequency of obstructive events is related to the severity of the syndrome, reported as an apnea–hypopnea index (AHI), with 5 to 15 considered mild, 16 to 30 moderate, and greater than 30 to 40 severe. Oxyhemoglobin desaturation is reported as the proportion of time below 90% and lowest saturation recorded and is another measure of OSA. Polysomnography may identify other co-existent sleep disorders such as periodic limb movement.

His Epworth Sleepiness Score is markedly elevated at 14, consistent with excessive daytime somnolence. Fiberoptic examination reveals both retropalatal and hypopharyngeal obstruction, primarily from the base of the tongue.

His in-laboratory polysomnogram revealed an AHI of 48, with his lowest oxyhemoglobin saturation of 82% consistent with severe OSA.

Diagnosis

Severe obstructive sleep apnea

Medical Management

Behavior modification is important, especially with regard to avoiding driving while drowsy and need for significant weight loss. Pharmacologic treatment of OSAS is limited but may occasionally include the use of stimulants to treat the symptoms of daytime somnolence.

Medical devices are the mainstay of therapy for adults with OSAS, especially patients with moderate to severe disease. Continuous positive airway pressure (CPAP) and bilevel positive airway pressure serve as an airway dilator or splint effectively eliminating pharyngeal collapse and resultant OSA for almost all patients. The amount of positive pressure, reported in centimeters of water (cm H_2O), is titrated in the sleep laboratory or occasionally through self-titrating devices. Effectiveness of therapy is dependent on nightly adherence to therapy, with some patients unwilling or unable to comply with treatment. Because of the effectiveness of CPAP therapy, repeated efforts are made to optimize therapy for those having difficulty adjusting to sleeping with a mask over their nose or face.

Oral appliances designed to pull the tongue or jaw forward may be effective for some patients with mild or moderate OSA who are unable to tolerate CPAP. Custom-fit to minimize dental occlusion or temporomandibular joint symptoms is important. Repeat polysomnography with the device in place is necessary to confirm improvement in OSA.

Surgical Management

Surgical management of adult patients with OSAS is challenging. Tracheotomy can be curative but is associated with significant complications and is not accepted by most patients. It may be necessary in morbidly obese patients (BMI $>$ 40) with OSAS or obesity hypoventilation syndrome who cannot tolerate or do not benefit from CPAP.

Adenotonsillectomy is curative in most small children with OSA, but its utility in

adults is limited to young, thin patients with significant tonsil hypertrophy (3–4+).

Uvulopalatopharyngoplasty (UP3), often performed with tonsillectomy, is effective for the minority (<5%) of adult OSA patients with isolated palate obstruction and tonsil hypertrophy (Fujita type I and Friedman stage I), with success rates of 80 to 90% in this highly selected subset of patients making it an attractive first-line therapy. Success rates are more modest (40%) in unselected patients or in patients with apparent isolated palatal obstruction but without significant tonsil hypertrophy. UP3 in this group of patients is reserved for those unwilling or unable to undergo CPAP. Success rates are much worse (<5–10%) in patients with morbid obesity or and multilevel (tongue and palate) obstruction. Unfortunately, the latter group makes up around 80% of adult patients with OSAS. The primary goal of UP3 is not to shorten the soft palate but rather to advance it anteriorly to open the velopharyngeal-retropalatal airway. Preservation of all palatal musculature and posterior soft palatal mucosa will minimize the risk of development of velopharyngeal insufficiency or nasopharyngeal stenosis.

Because of the limited effectiveness of isolated palate surgery for most adult patients with OSA, multiple procedures to address hypopharyngeal collapse have been developed. The two most effective are often combined and include an anterior mandibular osteotomy with genioglossus muscle advancement along with a hyoid suspension to the mandible or thyroid cartilage. When performed in conjunction with UP3, these procedures are considered phase I surgery by the Stanford protocol and are associated with success rates of 60 to 70% of adult OSA patients.

In attempt to reduce the morbidity of genioglossus muscle advancement with mandibular osteotomy, other procedures have been developed with varying degrees of invasiveness and effectiveness. Tongue suspension with a suture secured to the mandible avoids the osteotomy and may be helpful for some patients, especially those with tongue collapse without macroglossia. Midline partial glossectomy may reduce excess tissue in the base of tongue and help some patients. Radiofrequency therapy to the tongue base can reduce tissue volume with minimal morbidity and can be performed in the office with the patient under local anesthesia, but it requires multiple treatments to be effective.

Bilateral mandibular and maxillary advancement is considered phase II surgery in the Stanford protocol and is successful in about 90% of patients who were not cured with multilevel phase I surgery described above. The acceptance of this procedure is limited by the requirement for multiple osteotomies and impact on dental occlusion. This may be considered first-line therapy, however, for OSA patients with severe midface or mandibular hypoplasia.

Bariatric surgery involving gastric bypass is effective in morbidly obese patients (BMI > 35–40) with severe OSAS who are unable to tolerate CPAP or who are unaided by it. The effectiveness of bariatric surgery for OSA depends on the weight loss achieved, often necessitating more than 100 lb in this group. Temporary tracheotomy may be necessary until adequate weight loss is achieved.

◆ Rehabilitation and Follow-up

This patient underwent nasal CPAP titrated in the laboratory to 12 cm H_2O. Repeated masks were tried until a good fit was achieved. The patient's symptoms resolved with the Epworth score of 5 on therapy. His hypertension also improved, allowing discontinuation of his antihypertensive medication. He continues to struggle with weight problems and has been unable to lose significant weight. If his symptoms recur or significant weight gain occurs, repeat CPAP titration may be required in the future.

◆ Questions

1. Which of the following tests is indicated to diagnose obstructive sleep apnea?
A. Pulmonary function test
B. Polysomnography
C. Pulse oximetry
D. Chest CT
E. Multiple sleep latency test

2. Which of the following is the most efficacious treatment for most adults with obstructive sleep apnea?
A. Continuous positive airway pressure
B. Oral appliance
C. Uvulopalatopharyngoplasty
D. Distraction osteogenesis
E. Septoplasty

3. Approximately what percentage of adult patients with obstructive sleep apnea have multilevel pharyngeal obstruction?
A. 20%
B. 40%
C. 60%
D. 80%

Suggested Readings

Coleman J. Sleep apnea, part 1. Otolaryngol Clin North Am 1998;31:979–1012

Coleman J. Sleep apnea, part II. Otolaryngol Clin North Am 1999;32:211–222

Friedman M. New techniques and approaches to sleep apnea. Oper Tech Otolaryngol Head Neck Surg 2006; 17(4):215–272

Troell RJ. Radiofrequency techniques in the treatment of sleep-disordered breathing. Otolaryngol Clin North Am 2003;36(3):473–493

XIV

Pediatric Otolaryngology

67

Airway Foreign Body

Christopher T. Wootten and Jay Paul Willging

History

A 7-year-old girl was brought to the emergency department for escalating stridor. She was initially taken to her pediatrician 2 weeks ago for noisy breathing, where she was diagnosed with asthma and given an albuterol inhaler. Despite albuterol therapy, her breathing became progressively labored. On further questioning, her mother relates the onset of stridor to a brief coughing and choking spell after the patient had been playing with a toy tea set. On examination in the emergency department, the patient demonstrated biphasic stridor. Her respirations were regular with equal breath sounds, and her skin color was pink. She became uncomfortable when placed supine, but she was in no acute distress. Posterolateral and lateral neck radiographs were obtained that showed a slightly opaque, geometrically shaped foreign body at the level of the larynx (**Figs. 67.1 and 67.2**).

Differential Diagnosis—Key Points

1. The most common causes of stridor in an infant are laryngomalacia, vocal cord paralysis, and subglottic stenosis. These congenital causes of stridor should have manifested themselves within the first year of life. Subglottic hemangiomas also cause biphasic stridor, but the onset is gradual; symptoms reach their maximum around 8 to 12 months.
2. Infectious and inflammatory processes frequently affect the pediatric airway. A prodromic upper respiratory illness that leads to stridor may indicate croup or bacterial tracheitis. More rarely, rapid-onset stridor in a toxic child is consistent with epiglottitis.
3. Foreign bodies must always be considered in the pediatric population (typically 10–24 months of age), and frequently foreign body ingestion is an unwitnessed event. Stridor is common in fixed obstructions of the larynx or trachea. Chronic esophageal foreign bodies may also cause airway compression and secondary tracheomalacia by irritating the common wall between the trachea and esophagus. The mortality rate for foreign bodies in the upper aerodigestive tract is around 2%, with laryngeal foreign bodies having a mortality rate approaching 45%.

Test Interpretation

Physical examination must assess the patient's respiratory rate, color, and degree of distress. Listening to the breath sounds will inform the location of the airway foreign body. Classically

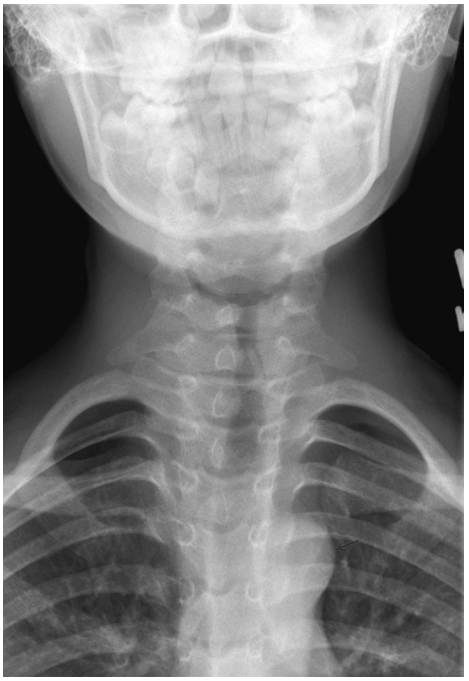

Fig. 67.1 Anteroposterior neck radiograph demonstrating a radiopaque geometric foreign body in the subglottis.

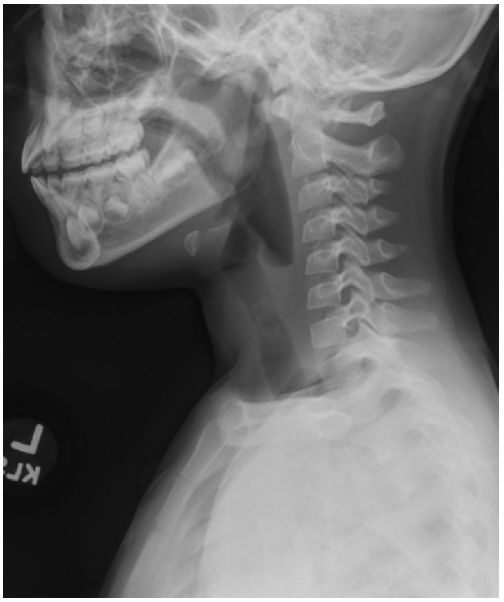

Fig. 67.2 Lateral neck radiograph demonstrating a radiopaque geometric foreign body in the subglottis.

inspiratory stridor is consistent with supraglottic pathology, but a large supraglottic foreign body will also induce gagging, retching, and a muffled voice. Laryngeal foreign bodies demonstrate biphasic stridor, whereas foreign bodies in the intrathoracic trachea cause expiratory stridor. Chest auscultation may indicate ispilateral wheezing, cough, and decreased breath sounds, but this will occur in less than 50% of the cases. A prolonged inspiratory or expiratory phase of respiration is also consistent with a distal airway foreign body. The diagnostic yield from the physical examination improves with chronicity.

In a stable patient, the initial evaluation of the airway is a high-kilovolt posteroanterior lateral neck and chest radiograph. The structural elements of the airway are outlined, and radiopaque foreign bodies may be identified. The lung fields can also be evaluated for signs of postobstructive emphysema. If a foreign body is ball-valving into a bronchus, there could be air trapping and hyperinflation of the lung field on that side, and mediastinal structures will shift away from the obstruction. Conversely, a completely obstructed bronchus may lead to absorption of the air trapped distally with widespread atelectasis. In these cases, the mediastinum will shift toward the obstruction.

When doubt arises as to the presence of a radiolucent foreign body in the distal airway, decubitus films and, rarely, airway fluoroscopy can be of assistance in determining the presence of an object in the airway. In the left lateral decubitus view, one expects the left (down) lung volume to be compressed. If the lung remains inflated, this suggests a foreign body within the left mainstem bronchus, which is obstructing free air flow. In highly selected patients, studies have been performed to integrate spiral and cine computed tomography scanning into an algorithm to diagnose radiolucent foreign bodies.

The diagnostic gold standard for aerodigestive tract foreign bodies is endoscopy. Evaluation with rigid telescopes allows proper evaluation of the entire aerodigestive system and the means of removal of any foreign body that might be present. Endoscopy also allows the physician to assess inflammation related to the foreign body and provide a prognosis for the

recovery of normal airflow dynamics once the foreign body is removed. There is no substitute for evaluation of the airway with the patient under general anesthesia anytime the question of a foreign body has been raised. The onus is on the surgeon to prove no foreign body is present in the airway.

◆ Diagnosis

Subglottic foreign body, identified by the patient as a plastic toy fragment with metallic paint

◆ Medical Management

In the emergency room, the child was able to maintain respiration without intervention, which allowed for radiographic evaluation. While in transport to the operating room, all equipment necessary to establish an artificial airway must be immediately available. Great care must be exercised to prevent undue agitation of the child, which may lead to airway compromise. When general anesthesia is induced, the airway may be unstable and mask ventilation or intubation might not be possible. A selection of appropriate ventilating bronchoscopes and suction equipment as well as an opened tracheotomy tray must be immediately available for use.

◆ Surgical Management

Because of the chronicity of the foreign body and the complex geometric shape, the ability to extract the foreign body from the larynx endoscopically was a concern. Optical foreign body forceps were used to practice on a similarly sized object before induction of anesthesia to gain familiarization with the best means of manipulating the object for removal from the larynx.

This child underwent mask induction of general anesthesia. Endoscopic evaluation of the larynx demonstrated a large, metallic-appearing foreign body lodged in the subglottis with tissue edema and granulation at its margins (**Figs. 67.3 and 67.4**). Manipulation of the foreign body with a laryngeal hook using

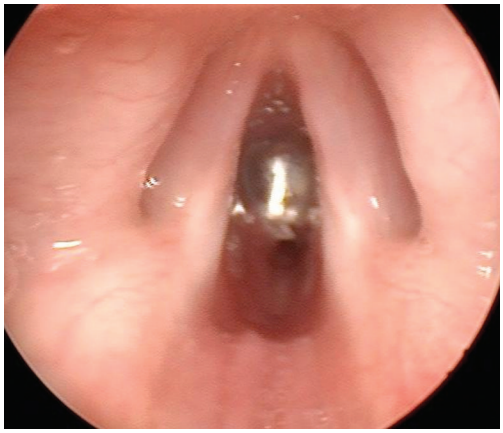

Fig. 67.3 Endoscopic view demonstrating a metallic-appearing foreign body impacted in the subglottis.

suspension microlaryngoscopy and spontaneous ventilation could not free it from the subglottis. Because of the potential of inducing laryngeal edema secondary to the manipulation of the foreign body within the larynx, a tracheotomy was performed to secure the airway. Next an anterior tracheal incision was continued superiorly, which provided access for extraction of the object from the airway. The cricoid cartilage did not need to be divided. Following removal of the foreign body (**Fig. 67.5**), bronchoscopy demonstrated no additional foreign bodies in the tracheobronchial tree. At the site of foreign body impaction, extensive granulation tissue and mucosal

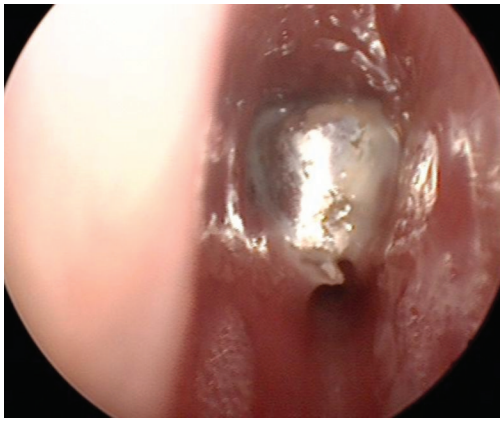

Fig. 67.4 Endoscopic view demonstrating a metallic-appearing foreign body impacted in the subglottis (the telescope is at the level of the vocal folds).

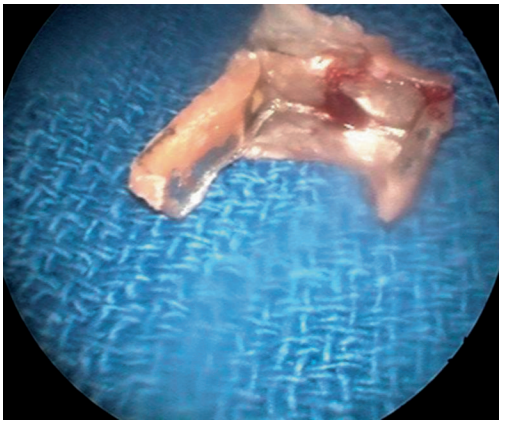

Fig. 67.5 Geometrically shaped plastic foreign body with metallic paint.

laceration were noted. An age-appropriate tracheostomy tube was inserted, and she was monitored in the hospital for several days. While capping, she demonstrated no stridor, and she was eventually decannulated. No complications developed postoperatively.

All equipment to secure the airway using a variety of methods, including orotracheal intubation, intubation with a ventilating bronchoscope, and tracheotomy must be assembled and ready for use before induction. Foreign bodies may be impacted at the level of the larynx, trachea, or bronchus. Complete airway obstruction is possible in all these areas. Extraction techniques often vary according to the site of obstruction.

Deploying optical extraction forceps through a rigid ventilating bronchoscope is the current standard of care for removing foreign bodies from the airway. A variety of graspers is available, and practicing on a copy of the suspected foreign body with different forceps before induction is helpful. Occasionally a foreign body may be impacted in a bronchus, and it might not be possible to pass the extraction forceps around the object. Fogarty embolectomy catheters are small enough to be passed alongside the foreign body. The balloon can then be inflated (carefully so as not to overdistend the bronchus), and the foreign body can then be pulled into the forceps. Of note, when a similar technique is used for extracting esophageal foreign bodies, there is a risk that the extracted foreign body may become aspirated into an unsecured airway.

Because of either the size or shape of the object or its impaction into the mucosa, occasionally a foreign body cannot be extracted through the larynx. In such situations one must not hesitate to perform a tracheotomy to gain adequate access to the airway for removal of the foreign body transtracheally. Impacted distal foreign bodies or chronic foreign bodies that have generated much granulation tissue in the airway may require a thoracotomy for their removal.

The key to successful management of airway foreign bodies is a high index of suspicion. The benefits associated with prompt diagnosis and removal of an airway foreign body outweigh the risks of endoscopy. The difficulty of extraction and the complication rates associated with airway foreign bodies increase dramatically when their duration extends beyond 72 hours. Early diagnosis and timely removal are essential to satisfactory resolution of these life-threatening events.

◆ Rehabilitation and Follow-up

When the airway problem has been adequately addressed and successfully managed, no long-term follow-up is necessary. Chronic foreign bodies induce granulation tissue and scarring, occasionally requiring surveillance endoscopy to ensure that healing is proceeding in an orderly fashion. Patient symptoms will indicate when this is necessary. Parent education is an important component of preventing recurrent airway aspirations.

◆ Questions

1. The most common causes of stridor in an infant include all of the following except:
   A. Vocal fold paralysis
   B. Laryngeal cleft
   C. Subglottic stenosis
   D. Laryngomalacia
2. A partially obstructing (ball-valving) foreign body in the left mainstem bronchus

would show which of the following on radiography?

A. Atelectatic left lung with mediastinal shift to the right

B. Hyperinflated left lung with mediastinal shift to the right

C. Compressed left lung on left lateral decubitus positioning

D. Hyperinflated left lung with mediastinal shift to the left

3. What is the diagnostic gold standard for airway foreign bodies?

A. Endoscopy

B. High-kilovolt radiography of the neck

C. History and physical examination

D. CT

Selected References

Bloom DC, Christenson TE, Manning SC, et al. Plastic laryngeal foreign bodies in children: a diagnostic challenge. Int J Pediatr Otorhinolaryngol 2005;69(5):657–662

Holinger LD. Management of sharp and penetrating foreign bodies of the upper aerodigestive tract. Ann Otol Rhinol Laryngol 1990;99(9 Pt 1):684–688

Hong SJ, Goo HW, Roh JL. Utility of spiral and cine CT scans in pediatric patients suspected of aspirating radiolucent foreign bodies. Otolaryngol Head Neck Surg 2008;138(5):576–580

Kadmon G, Stern Y, Bron-Harlev E, Nahum E, Battat E, Schonfeld T. Computerized scoring system for the diagnosis of foreign body aspiration in children. Ann Otol Rhinol Laryngol 2008;117(11):839–843

Reilly JS. Prevention of aspiration in infants and young children: federal regulations. Ann Otol Rhinol Laryngol 1990;99(4 Pt 1):273–276

Swanson KL. Airway foreign bodies: what's new? Semin Respir Crit Care Med 2004;25(4):405–411

Walner DL, Ouanounou S, Donnelly LF, Cotton RT. Utility of radiographs in the evaluation of pediatric upper airway obstruction. Ann Otol Rhinol Laryngol 1999;108(4):378–383

Zerella JT, Dimler M, McGill LC, Pippus KJ. Foreign body aspiration in children: value of radiography and complications of bronchoscopy. J Pediatr Surg 1998;33(11):1651–1654

68

Branchial Arch Anomaly

Evan J. Propst, Jay Paul Willging, and Alessandro de Alarcón

History

A 1-year-old boy presented with a right ear lesion obstructing the external auditory canal and intermittent drainage from a pit in the right cheek just inferior to the ear lobule (**Fig. 68.1**). He had no history of hearing loss, renal disease, or other systemic illness. Facial nerve function was intact and symmetric. The ipsilateral tympanic membrane and remainder of the head and neck examination were normal. Computed tomography (CT) imaging demonstrated a well-defined, low-attenuating mass anterior to the floor of the right cartilaginous external auditory canal. The mass measured 15 × 9 mm and was partially surrounded by parotid tissue (**Fig. 68.2**).

Differential Diagnosis—Key Points

1. The differential diagnosis of a pediatric neck mass can be divided into congenital and acquired lesions. Congenital masses can further be divided into midline and lateral anomalies. Acquired lesions can be benign or malignant. **Table 68.1** illustrates the differential diagnosis of pediatric neck masses.
2. Knowledge of the embryology of branchial development is essential for understanding the pathway that an anomaly will follow. The greatest head and neck embryonic development occurs in the first 8 weeks. There are six branchial arches (**Fig. 68.3**). Between these arches are clefts externally that are lined with ectoderm and pouches internally that are lined with endoderm.

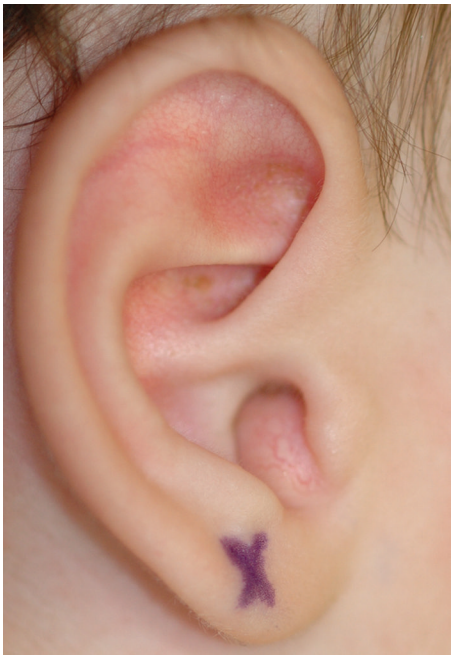

Fig. 68.1 Right ear with cystic mass obstructing the external auditory canal.

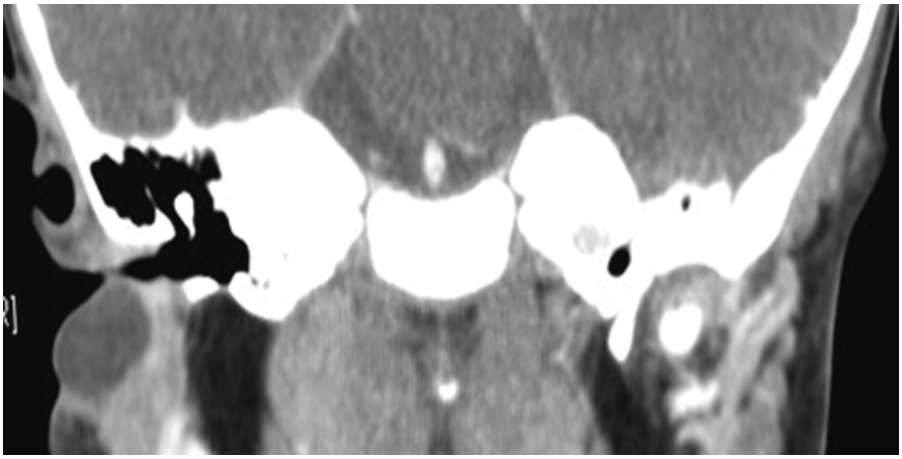

Fig. 68.2 Coronal computed tomography imaging demonstrating cystic mass inferior to external auditory canal.

The fifth branchial arch degenerates. Each arch has an associated nerve, artery, cartilaginous bar, and muscle. The sternocleidomastoid muscle (SCM) derives from the cervical somites posterior and inferior to the branchial arches (**Table 68.2**).

3. Branchial anomalies related to arch development include cysts, sinuses, and fistulae.

Table 68.1 Differential diagnosis of pediatric neck masses

Congenital	
Midline	*Lateral*
Thyroglossal duct cyst	Branchial cleft cyst
Dermoid/epidermoid cyst	External laryngocele
Teratoma	Pseudotumor of infancy (fibromatosis coli)
Plunging ranula	Thymic cyst
Lymphatic/vascular malformation	Lymphatic/vascular malformation
Hemangioma	Hemangioma
Normal thyroid gland	
Acquired	
Benign	
Infectious or inflammatory:reactive lymphadenopathy	
Sialadenitis	
Abscess	
Granulomatous (atypical mycobacteria, cat-scratch, toxoplasma, sarcoidosis)	
Neoplastic: lipoma, neurofibroma, pilomatrixoma, benign mixed tumor	
Malignant	
Lymphoma (Hodgkin, non-Hodgkin), rhabdomyosarcoma, thyroid carcinoma, salivary tumors, neuroblastoma, histiocytosis (Langerhans)	

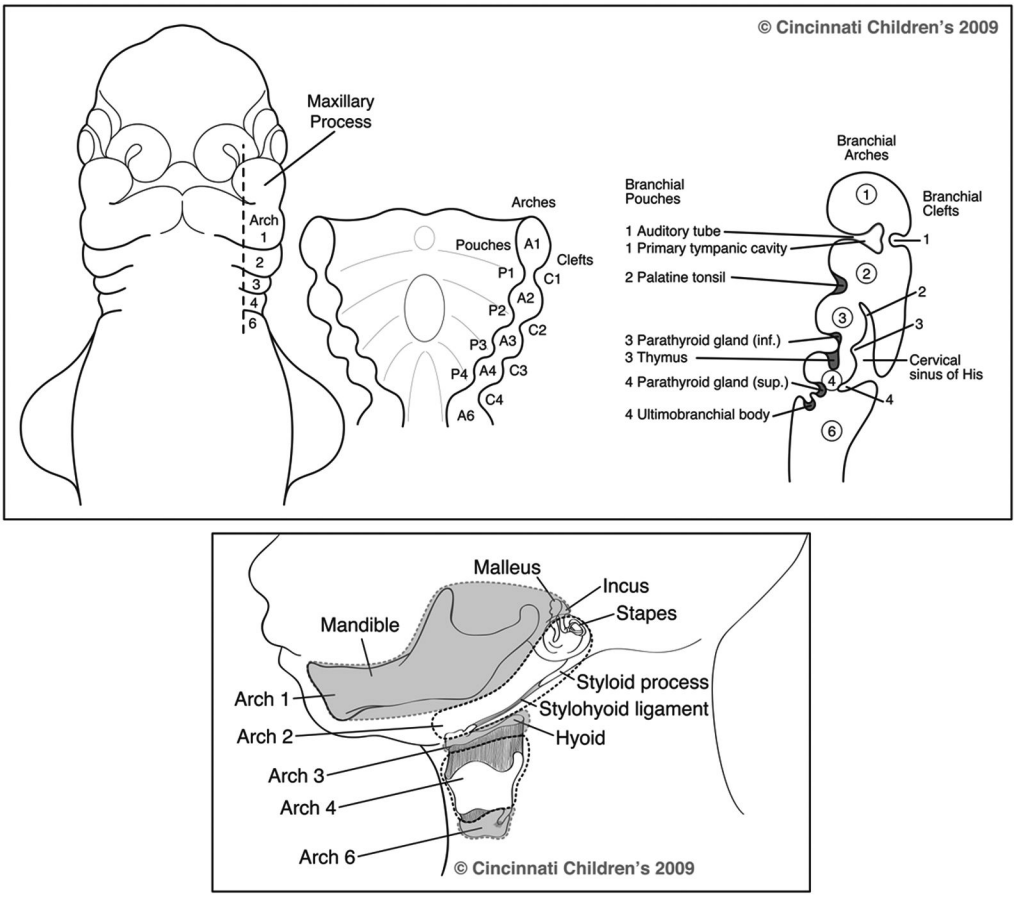

Fig. 68.3 Embryology of branchial arches, clefts, and pouches (superior) and the structures from which they derive (inferior).

Cysts are lined by mucosa or epithelium, have no external opening, and arise from embryonic rests trapped inside developing tissue. Sinuses result from the incomplete closure of branchial pouches and clefts, and they communicate with a single body surface, either the skin or pharynx. Fistulae also result from the incomplete closure of pouches or clefts; however, they communicate with two body surfaces.

4. Traditional teaching holds that the path of a branchial anomaly can be predicted based on knowledge of branchial arch embryology. Cysts, sinuses, and fistula tracts course posterior-medial-deep to other derivatives of their own branchial arch, and anterolateral and superficial to the derivatives of the arch behind them. The origin of third and fourth branchial anomalies is controversial.

First branchial anomalies account for less than 8% of all branchial arch anomalies. Work divided first branchial anomalies into types I and II based on their histology and location. The Work classification is determined postoperatively and therefore does not contribute much to preoperative planning. Type I first branchial anomalies are ectoderm-derived duplications of the membranous external auditory canal that are lined by squamous epithelium. They are usually located anterior and medial to the external auditory canal and course lateral to the facial nerve. Type II first branchial anomalies are more common than type I first branchial anomalies. They derive from ectoderm and mesoderm, are lined by

Table 68.2

Arch	Cranial Nerve	Artery	Cartilage	Muscle	Pouch
First (mandibular)	V (trigeminal)	Maxillary (degenerates)	Meckel cartilage Malleus head/neck Anterior malleolar ligament Incus body/short process Mandible Sphenomandibular ligament	Tensor tympani Tensor veli palatini Muscles of mastication Digastric anterior belly Mylohyoid	Middle ear
Second (hyoid)	VII (facial)	External carotid stapedial (degenerates)	"Reichart cartilage" Malleus manubrium Incus long process Stapes (not vestibular side of footplate) Pyramidal eminence Styloid process Hyoid lesser cornu and upper half body	Stapedius tendon Muscles of facial expression Digastric posterior belly Stylohyoid	Tonsillar fossa Palatine tonsils
Third	IX (glossopharyngeal)	Internal carotid Common carotid	Hyoid greater cornu and lower half body	Stylopharyngeus	Inferior parathyroids Thymus
Fourth	X (superior laryngeal)	Right subclavian Aortic arch	Thyroid cartilage Cuneiform	Cricothyroid membrane Inferior pharyngeal constrictors	Superior parathyroids
Sixth	X (recurrent laryngeal)	Right pulmonary Ductus arteriosus	Cricoid Arytenoid Corniculate	Intrinsic laryngeal muscles (except cricothyroid)	Parafollicular C cells of thyroid

squamous epithelium, and contain adnexae and cartilage. They usually open into the concha or external auditory canal as well as the anterior border of the SCM at the level of the mandible. Their tract can pass medial or lateral to the facial nerve. A large literature review found the course of first branchial anomalies to have a more variable relation to the facial nerve than proposed by Work. These authors found that first branchial anomalies can pass superficial, deep, or between the branches of the facial nerve. Tracts passing deep to the facial nerve were more common in women, children presenting at a young age, children with fistulae as compared with sinuses, and tracts that did not open into the external auditory canal.

Second branchial anomalies are the most common of the branchial anomalies. Their path begins along the anterior border of the SCM, courses deep to the platysma and facial nerve, deep to the external carotid artery, superficial to the internal carotid artery, deep to the posterior belly of the digastric, superficial to the glossopharyngeal and hypoglossal nerves, and enters the pharynx in the region of the tonsillar fossa. A second branchial anomaly is three times more likely to be a cyst than a sinus or fistula, and it can be located anywhere along this path. Branchio-otorenal syndrome most commonly presents with second branchial arch anomalies, preauricular pits, auricular deformities, deafness, and renal anomalies. Other findings may include facial asymmetry and palate anomalies.

Third branchial anomalies are rare. Theoretically, their path should begin along the anterior border of the SCM, course deep to the platysma and facial nerve, deep to both carotid arteries, deep to the glossopharyngeal nerve, superficial to the hypoglossal nerve, deep to the stylopharyngeus muscle, and enter the pharynx medial to the greater cornu of the hyoid bone at the apex of the pyriform sinus.

Theoretically, the path of a fourth branchial anomaly would begin low in the neck along the anterior border of the SCM, course deep to the platysma and facial nerve, deep to the common carotid artery, loop around the aortic arch on the left or the subclavian artery on the right, deep to the superior laryngeal nerve, superficial to the recurrent laryngeal nerve, deep to the inferior pharyngeal constrictor muscles, and enter the pharynx at the apex of the pyriform sinus.

A published study by James and colleagues investigated third and fourth branchial anomalies and found that most cases presented as left-sided cystic infections that became fistulae only after drainage, were closely associated with the ipsilateral thyroid lobe, and passed directly from the posteromedial surface of the thyroid through the inferior constrictor into the piriform fossa, traveling lateral and posterior to the recurrent laryngeal nerve. The tract never looped around the hypoglossal nerve or carotid arteries and never descended into the mediastinum. The authors suggested that third and fourth branchial anomalies may in fact derive from the thymopharyngeal duct, which forms during descent of the thymus during the eighth week of embryogenesis. It remains to be determined whether these branchial anomalies and thymopharyngeal remnants are three discrete entities or are in fact simply one entity derived from the thymopharyngeal duct.

◆ Test Interpretation

Radiographic evaluation of a lateral neck mass can help to confirm the diagnosis and assist with management. On CT imaging with contrast, a branchial cleft cyst will be homogeneous with low attenuation centrally and a smoothly enhancing rim. Magnetic resonance imaging will demonstrate a low T1- and high T2-weighted signal. Third and fourth branchial anomalies often have a hypodensity within the ipsilateral (usually left) thyroid lobe. Gas bubbles in the cyst or along the tract are diagnostic.

◆ Diagnosis

First branchial anomaly, Work type I

◆ Medical Management

Acutely infected branchial cleft anomalies should be treated medically with antimicrobial coverage against *Staphylococcus aureus*. Clindamycin is a lincosamide that covers *S. aureus,*

methicillin-resistant *S. aureus*, and respiratory tract anaerobes. Trimethoprim-sulfamethoxazole (Bactrim) is an effective alternative, but it should not be used in infants younger than 2 months of age. Incision and drainage may be required where there is suppuration. Surgical excision should be delayed until the infection resolves and should proceed shortly after resolution to avoid increased scarring and fibrosis that could render the surgical approach more difficult.

◆ Surgical Management

First branchial arch anomalies that swell, become infected, or drain require surgical excision. First branchial arch anomalies are located close to the facial nerve and therefore require identification of the facial nerve trunk at an early stage of dissection (**Figs. 68.4 and 68.5**). In older children and adults, the facial nerve is protected by the mastoid tip, and landmarks such as the digastric muscle, external auditory canal cartilaginous pointer, and tympanomastoid suture can be used to identify and preserve the nerve. In infants and young children, the mastoid tip is less developed because the pull of the SCM has not yet caused it to lengthen. The extratemporal facial nerve is therefore more superficial in young children and has a greater risk of being damaged. To decrease the risk of facial nerve injury, a curvilinear incision is made 2 cm below the mandible extending in a skin crease over the mastoid and ending 1 to 2 cm behind the postauricular crease. A subplatysmal flap is elevated only up to the ramus of the mandible. The facial nerve is identified in the triangular space formed by the posterior belly of the digastric muscle, the anterior border of the SCM, and the cartilaginous external auditory canal. If the mass is in this area, consideration should be given to identifying the facial nerve in the mastoid and following it out the stylomastoid foramen.

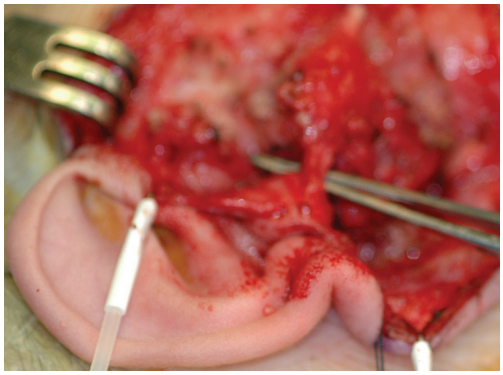

Fig. 68.4 Surgical excision of cyst using a superficial parotidectomy approach.

Controversy exists regarding the optimal age for excision of first branchial arch anomalies because of the inherent risk to the facial nerve. Waiting until the child is older to excise the anomaly makes identification and preservation of the facial nerve easier. However, observation over a long period increases the risk of infection and fibrosis, rendering surgical dissection more difficult. In general, it is

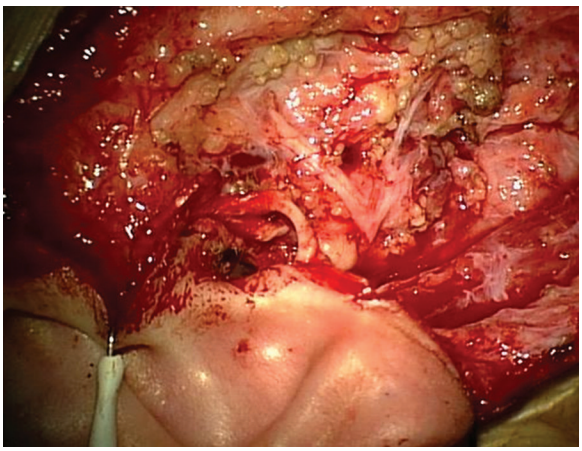

Fig. 68.5 Resection of involved external auditory canal cartilage and nearby facial nerve.

prudent to wait until the child is approximately 2 years of age, at which time the mastoid tip and facial nerve are larger. The curvilinear incision ending over the mastoid is still required because the nerve will still be quite superficial.

When the tract involves the cartilaginous external auditory canal, the cartilage and overlying skin must be excised with the tract (**Fig. 68.5**). Tracts entering the middle ear or temporal bone may be amputated with curettage of the bone if necessary. Tracts with extensive involvement of the external auditory canal may require the canal to be packed for 3 to 4 weeks to prevent subsequent stenosis.

Second branchial arch anomalies that swell, become infected, or drain also require surgical excision. The resection should include a rim of skin around the fistulous tract if present, careful dissection of the tract with a cuff of normal tissue, and removal of any cystic structures and the tract up to the tonsillar fossa mucosa. Care must be made not to injure the numerous vessels and nerves in close proximity to the tract.

Controversy exists surrounding the management of third and fourth branchial anomalies. Described methods range from cauterization of the piriform sinus to hemithyroidectomy and meticulous dissection of the tract to the piriform sinus apex. Care must be taken to preserve the recurrent laryngeal nerve.

◆ Rehabilitation and Follow-up

The rate of recurrence following resection of branchial anomalies is less than 5% when there is no history of prior infection or surgery. This rate increases to 20% when these risks are present. Permanent injury to the facial nerve is rare when first branchial anomalies are approached in a systematic fashion.

Acknowledgment We thank Jan Warren for her illustration of branchial arch embryology.

◆ Questions

1. The differential diagnosis of congenital midline neck mass includes all of the following except:
- **A.** Thyroglossal duct cyst
- **B.** Dermoid/epidermoid cyst
- **C.** Plunging ranula
- **D.** Branchial cleft cyst
- **E.** Hemangioma

2. Concerning the embryology of branchial arches, the second branchial arch gives rise to what?
- **A.** Head and neck of the malleus
- **B.** Mandible
- **C.** Long process of the incus
- **D.** Sphenomandibular ligament
- **E.** Tensor tympani tendon

3. Which of the following statements regarding branchial cleft cysts is not true?
- **A.** Second arch anomalies are the most common.
- **B.** Acutely infected cysts should receive antimicrobial treatment.
- **C.** Cysts, sinuses, and fistulae course anterolateral and superficial to other derivates of their own arch.
- **D.** Computed tomography will demonstrate a homogeneous lesion with low attenuation centrally and a smoothly enhancing rim.
- **E.** First branchial arch anomalies require early identification of the facial nerve.

Suggested Readings

D'Souza AR, Uppal HS, De R, Zeitoun H. Updating concepts of first branchial cleft defects: a literature review. Int J Pediatr Otorhinolaryngol 2002;62(2):103–109

Farrior JB, Santini H. Facial nerve identification in children. Otolaryngol Head Neck Surg 1985;93(2):173–176

Finn DG, Buchalter IH, Sarti E, Romo T, Chodosh P. First branchial cleft cysts: clinical update. Laryngoscope 1987;97(2):136–140

Graney DO, Sie KCY. Developmental anatomy. In: Cummings CW, Flint PW, Harker LA, et al, eds. Cummings

Otolaryngology Head & Neck Surgery, 4th ed. Philadelphia, PA: Elsevier Mosby; 2005:3938–3951.

James A, Stewart C, Warrick P, Tzifa C, Forte V. Branchial sinus of the piriform fossa: reappraisal of third and fourth branchial anomalies. Laryngoscope 2007; 117(11):1920–1924

Myer CM III. Congenital neck masses. In: Shumrick Da, Gluckman JL, Myerhoff WL, eds. Otolaryngology. Philadelphia, PA: WB Saunders; 1991:2535–2543.

Triglia JM, Nicollas R, Ducroz V, Koltai PJ, Garabedian EN. First branchial cleft anomalies: a study of 39 cases and a review of the literature. Arch Otolaryngol Head Neck Surg 1998;124(3):291–295

Work WP. Newer concepts of first branchial cleft defects. Laryngoscope 1972;82(9):1581–1593

69

Thyroglossal Duct Cyst

Jay Paul Willging

History

A 3-year-old black boy was noted to have a swelling in the superior neck in the midline. It came up rapidly in association with a upper respiratory infection (URI). The mass was painful to palpation with no overlying erythema. Antibiotics caused the mass to decrease in size slightly, and the tenderness resolved.

The mass was located 1 cm below the hyoid bone in the midline of the neck. It was not fluctuant to palpation, but it did feel cystic and firm. It was not freely mobile.

Differential Diagnosis—Key Points

1. Thyroglossal duct cyst. The thyroid originates in the tongue base and descends inferiorly through the neck to assume its final position in the base of the neck. Remnants of the descending tissue may persist in the anterior compartment of the neck, lying near the midline. The remnant may lie anywhere from the hyoid to the clavicles. Lingual thyroglossal duct cysts may be encountered that manifest as a tongue base mass without any visible abnormality in the neck. Antibiotics have little effect beyond correction of an acute infection within the mass. The mass tends to persist despite antibiotic treatment.
2. Dermoid. Epithelial inclusion cysts (dermoids) are often associated with developmental fusion planes. The dermoid is a benign accumulation of squamous debris. They may become infected and present with pain and swelling, or they may present as a solitary mass. In the neck, they are most commonly found above the level of the hyoid bone, a location rarely found with thyroglossal duct cysts. Antibiotics have limited effect beyond correction of an acute infection. The mass tends to persist after antibiotic treatment.
3. Lymphadenopathy. Cervical adenopathy may occur in any of the nodal chains. They are often found laterally in the neck. Midline nodes are commonly found in the submental area as well as in the base of the neck, as the paratracheal nodes. Antibiotics will often promote resolution of reactive adenopathy.
4. Branchial arch anomaly. Failure of branchial apparatus development may cause the persistence of cysts, sinuses, and fistula in the head and neck region. The location of the anomaly and its relation to the great vessels and cranial nerves are dependent on the arch from which it derives. Branchial arch anomalies tend to be located along the anterior boarder of

the sternocleidomastoid muscle, not encroaching on the midline.

5. Neoplasm. Neoplasms may develop in the neck and must always be considered in the evaluation of a child with a neck mass. In the pediatric population, around 10% of masses will be neoplasms. The tissue type would be dependent on the organ from which it derived.

Test Interpretation

1. Computed tomography (CT) imaging. The location and physical characteristic of an anterior compartment mass often suggest the diagnosis of a thyroglossal duct cyst. When this is suspected, no imaging of the mass is necessary. When there is doubt, a CT provides good visualization of the lesion and its anatomic relationships to other structures. It will show a low-density mass, but rarely the associated thyroglossal duct, which traverses the hyoid bone, to terminate in the tongue base.
   If a CT is obtained, it is important to evaluate the films to confirm the thyroid gland lies in its proper location at the base of the neck. It is possible that the only functioning thyroid tissue resides within the mass.
2. Magnetic resonance imaging (MRI). MRI will provide similar visualization of the mass and surrounding structures as a CT, but it is not the preferred test because of its higher costs and longer acquisition times. Because most patients with thyroglossal duct cysts are young, sedation is often required for MRI. Many fewer children require sedation for a CT scan.
3. Ultrasound. Ultrasound imaging will define a mass as solid or cystic. A thyroglossal duct cyst should be cystic in nature. The most important role of ultrasound is to image the base of the neck to confirm the presence of a thyroid gland. Lack of presence of a thyroid in its normal location requires further testing to identify the location of functioning thyroid tissue.
4. Thyroid scan. This radiolabeled iodine study shows concentration of the label within functioning thyroid tissue. This study has been used in the past to determine the presence of a normal thyroid in its normal location. Unless there is a question of thyroid function, this test is not required in the evaluation of a patient with a thyroglossal duct cyst.
5. Needle aspiration. This study will evacuate the cyst of its mucinous content. It is not necessary to perform this procedure routinely because cytology is not critical to the diagnosis. During active infections, an incision and drainage procedure may be necessary to allow resolution of the infection, but a needle aspiration may be used to obtain a specimen for culture and sensitivity. The noninfected cyst content is generally amber colored, thick, and tenacious.

Diagnosis

Thyroglossal duct cyst: Midline mass that is cystic, is below the hyoid bone, is not freely movable, and developed rapidly in association with a URI. An ultrasound was obtained that documented a normal-appearing thyroid gland in its normal location in the base of the neck.

Medical Management

Antibiotic treatment covering *Staphylococcus* and *Streptococcus* organisms is required for the treatment of bacterial infections involving a thyroglossal duct cyst. Unless actively infected, medical management is not required for a thyroglossal duct cyst. These are congenital structural anomalies, so surgical intervention is required to minimize recurrent infection and suppurative complications that may develop.

If a lingual thyroid is identified, often a hypothyroid state exists. In this circumstance, thyroid studies should be obtained to document that the patient is euthyroid. If a lingual thyroid is present, thyroid suppression is often necessary to suppress thyroid growth and the potential problems developing secondary to the size of the thyroid tissue within the tongue base.

Surgical Management

1. The surgical management of a thyroglossal duct cyst entails exposing the cyst through a horizontal neck incision. The cyst should be isolated, and the tract extending superiorly

should be identified. This duct should not be skeletonized; rather, some surrounding tissue should be left attached to the stalk because small extensions of the duct lumen may be present that would raise the risk of recurrence if they are transected. The duct will extend to the center of the hyoid bone. The middle section of the hyoid should be transected because there are often ramifying extensions of this duct through and around the hyoid bone. After transection of the hyoid is complete, the tract will extend into the tongue base. A core of lingual tissue should be resected along the tract to the mucosal surface of the tongue. The tract is ligated and transected, taking care not to violate the tongue base mucosa. A drain should be placed in the operative bed and the incision closed in layers.

2. Care must be exercised to identify the hyoid bone properly. In children 3 years of age and younger, the larynx remains high in the neck, with the hyoid overlying the thyroid cartilages. It is possible inadvertently to resect portions of the laryngeal framework during resection of the central portion of the hyoid.
3. A Sistrunk procedure (excision of the thyroglossal duct cyst and associated tract in continuity with the central portion of the hyoid bone) is associated with a 3 to 5% recurrence risk. If a thyroglossal duct cyst has been infected on several occasions before excision, the recurrence risk triples as the result of an increase in the number of ramifying channels that develop around the main thyroglossal duct. For this reason, the timing of surgical excision should be expedited once a history of infection within the mass has been determined.

◆ Rehabilitation and Follow-up

Wound infections tend to develop between 5 and 7 days of operation and are associated with surrounding erythema, pain at the incision site, and purulent material below the skin flaps. A recurrent thyroglossal duct cyst will generally present within the first month and will be associated with a painless swelling at the incision line, with mild surrounding erythema and a thick mucoid drainage below the skin flaps.

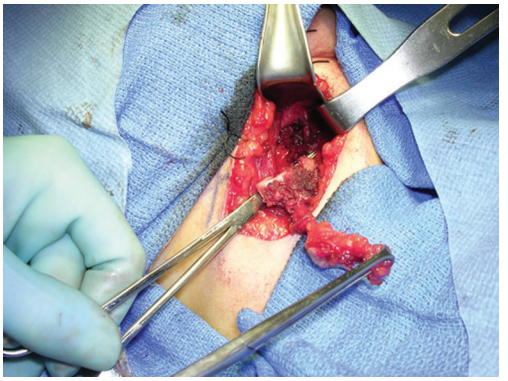

Fig. 69.1 Leaving tissue attached to the duct, resecting the central portion of the hyoid bone without extensive skeletonization, and excision of a cuff of tongue-base tissue maximizes the changes of complete excision of a thyroglossal duct cyst at the first operation.

Most recurrences are located near the incision line but may be located anywhere along the previous tract. Initial treatment could consist of incision and drainage of the area, with packing placed to allow drainage of the material and induction of fibrosis that may obliterate the residual thyroglossal duct tissue. In most cases, however, reoperation is required to remove the residual tissue.

Excision of a recurrent thyroglossal duct cyst entails excision of the original incision line and a broad excision of the material in the anterior compartment of the neck. A defined track may be seen that directs the excision to a specific area, but often no definable tract can be located; yet small pockets of mucoid material may be found in the tissues. All these areas need to be excised.

The initial operation provides the best opportunity for complete excision of the congenital thyroglossal duct cyst. Leaving tissue attached to the duct, resecting the central portion of the hyoid bone without extensive skeletonization of the structure, and excision of a cuff of tongue base tissue maximizes the changes of complete excision of the mass at the first operation (**Fig. 69.1**).

Questions

1. Imaging of the neck in a child with a thyroglossal duct cyst:
   - **A.** Is necessary to determine the diagnosis of the cervical mass
   - **B.** Is useful to identify the presence of thyroid tissue in its normal location at the base of the neck
   - **C.** Is less effective in providing needed information regarding the management of the patient than a computed tomography scan
   - **D.** Should be used only for needle localization during an aspiration procedure
2. The risk of airway complications following thyroglossal duct cyst excisions in young children:
   - **A.** Is related to the position of the hyoid bone over the thyroid cartilage of the larynx
   - **B.** Is secondary to swelling caused by excision of tissue from the tongue base
   - **C.** Is related to infections in the neck
   - **D.** Occurs less commonly than in older children
3. Excising a thyroglossal duct cyst requires:
   - **A.** Skeletonizing the tract that extends to the tongue base
   - **B.** Removing the hyoid bone after isolating it from surrounding structures
   - **C.** Removing a core of tissue from the tongue base because this was the site of origin of the mass
   - **D.** Removing the thyroglossal duct cyst and its tract along with adherent tissue, the central portion of the hyoid bone, and a core of tissue from the tongue base

Suggested Readings

Ducic Y, Chou S, Drkulec J, Ouellette H, Lamothe A. Recurrent thyroglossal duct cysts: a clinical and pathologic analysis. Int J Pediatr Otorhinolaryngol 1998; 44(1):47–50

Hewitt K, Pysher T, Park A. Management of thyroglossal duct cysts after failed Sistrunk procedure. Laryngoscope 2007;117(4):756–758

Koch BL. Cystic malformations of the neck in children. Pediatr Radiol 2005;35(5):463–477

Wootten CT, Goudy SL, Rutter MJ, Willging JP, Cotton RT. Airway injury complicating excision of thyroglossal duct cysts. Int J Pediatr Otorhinolaryngol 2009; 73(6):797–801

70

Lymphangioma

Marlene A. Soma and Jay Paul Willging

History

The otolaryngology team is consulted by the obstetric team about a 28-year-old P2G1A0 woman in her 34th week of pregnancy. She had developed polyhydramnios, and a screening ultrasound revealed a large cervicofacial mass of the fetus occupying mostly the anterior triangle of the neck with extension into the upper mediastinum. Magnetic resonance imaging (MRI) confirmed the presence of a multiloculated cystic mass with poorly defined borders that infiltrated the tongue base and floor of mouth. The lesion appeared to be displacing the fetal trachea and was causing compression of the fetal esophagus. Plans for an elective cesarean section delivery are made for the future with an otolaryngology team present in the delivery room to assist with management of the fetal airway.

The patient unfortunately enters spontaneous preterm labor before the scheduled delivery date. The otolaryngology team is called emergently to the operating room.

Differential Diagnosis—Key Points

Several key points in this history point to potential problems in the care of this child:

1. Airway obstruction at birth is a life-threatening condition and is associated with a high mortality rate, especially if there is delay in securing an airway or if there is inability to ventilate the neonate. Hypoxia, acidosis, and ischemic brain injury will result in significant morbidity, even death, in a child who may otherwise be normal. In the situation of antenatal diagnosis of fetal airway obstruction as in this case, a planned multidisciplinary approach to delivery is advocated to maximize the outcome for both the mother and child. An EXIT (ex utero intrapartum technique) procedure allows the fetus to be delivered with maintenance of the uteroplacental circulation until a fetal airway has been established. The obstetric team, two anesthesiology teams, otolaryngology, neonatology, pediatric surgery, maternal-fetal medicine specialists, an echocardiographer, and two full surgical scrub teams should be briefed and prepared for every contingency. Reliable maternal fetal circulation can be maintained for longer than 1 hour. If an experienced team of clinicians and surgeons is not available, a child might be delivered by cesarean section, maintaining the child at the level of the maternal heart, on placental support (operation on placental support [OOPS]). Reliable maternal fetal circulation can be maintained for only 5 minutes.

2. The mother and neonate have specific needs that must be met. A team dedicated to the management of each is important. Planning for the delivery should occur as soon as the lesion has been defined by imaging. The largest operating room available needs to be set up to accommodate the two teams so that each will have access to equipment without interference from the other. For the neonate, a range of appropriately sized laryngoscopes with light sources, endotracheal tubes, introducers, laryngeal mask airways, ventilating bronchoscopes, and tracheotomy instruments are essential. If a tracheotomy is required, a second surgical nurse should be available to assist with that procedure. A pediatric surgeon should also be available if concern exists for the need to obtain access to the mediastinal trachea. A pediatric anesthesiologist should be in attendance to provide for the anesthetic needs of the child.
3. The EXIT procedure differs from a routine cesarean section in that the goal is to achieve a state of uterine hypotonia with preservation of uterine volume to maintain the uteroplacental circulation. A level of fetal anesthesia without cardiac depression is also desired. In contrast, during a cesarean section, the goal is to maximize uterine tone to prevent postpartum hemorrhage and to avoid fetal anesthesia and potential respiratory depression.
4. The differential diagnosis of a congenital cystic cervicofacial mass causing extrinsic airway obstruction such as described in this case includes teratoma, lymphangioma, and arteriovenous malformation. Less common lesions causing extrinsic obstruction include branchial cleft cyst, fetal goiter, sarcoma, and neuroblastoma. The term CHAOS (congenital high airway obstruction syndrome) has been used to describe fetal upper airway obstruction of an intrinsic nature. Intrinsic airway lesions include laryngeal atresia, stenosis or web, tracheal atresia or stenosis. If there is no tracheoesophageal connection, the secretions produced by the fetal lungs cannot be excreted causing the lungs to expand and the diaphragm to flatten or invert. If the esophagus is compressed or obstructed as well, fetal swallowing is impaired and polyhydramnios may result. Regardless of the precise diagnosis of the obstructing lesion, the initial approach to the airway will be the same.
5. The approach to the compromised newborn airway must be orderly. If a neonate is spontaneously breathing, the degree of airway obstruction must be assessed to determine the need for elective intubation in the delivery room. If the airway is compromised, endotracheal intubation with an armored endotracheal tube should be attempted. If the endotracheal tube cannot be passed because of a mass compressing the lumen of the trachea (most of these can be overcome with gentle pressure on the endotracheal tube), a rigid bronchoscope can be passed into the airway to offer a method of ventilation. In some situations, traction on the mass by an assistant may help to elevate it from the airway structures. With a bronchoscope in place, a tracheotomy can then be undertaken in a controlled fashion.
6. If a bronchoscope cannot be passed, a tracheotomy must be immediately performed. This may require reflection or partial resection of a neck mass. If the cervical component of the mass precludes access to the anterior base of the neck, a thoracotomy should be performed to access the mediastinal trachea. Proper positioning of the tracheotomy between the second and third tracheal rings is important because the fetal trachea may be pulled out of the chest owing to hyperextension of the mass and neck.
7. In an EXIT procedure, depending on the length of the umbilical cord, procedures on the fetus may need to be performed on the partially delivered child (head, neck, and one arm delivered through the hysterotomy), with the child across the legs of the mother or occasionally on a Mayo table positioned at right angles to the mother. In the case described, the child was partially delivered and endotracheal intubation was secured uneventfully via direct laryngoscopy. The position of the tube was confirmed with flexible bronchoscopy. Once the child was fully delivered and stabilized, the endotracheal tube was changed to a

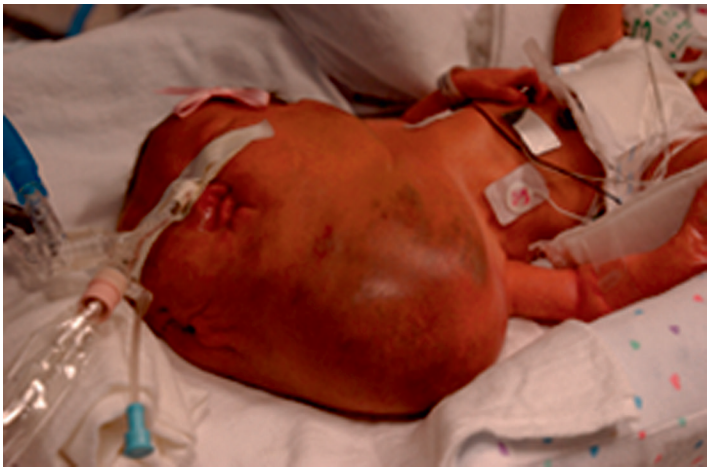

Fig. 70.1 Large bilateral cervicofacial mass in the described newborn infant, seen extending to anterior chest. Nasotracheal intubation was secured following the ex utero intraoperative technique (EXIT) procedure once the child was stabilized.

nasotracheal tube (**Fig. 70.1**) and the child was transferred to the intensive care unit.

Test Interpretation

1. Prenatal ultrasound. Because of the widespread use of prenatal ultrasound, there has been an increase in the diagnosis of fetal airway malformations. Polyhydramnios is seen in up to 40% of cases due to concomitant fetal esophageal obstruction, and it is often seen in association with large tumors. The two most common cervical fetal masses are lymphangiomas and teratomas. Lymphangiomas, also known as *lymphatic malformations*, are slow-flow anomalies of the lymphatic channels that have stable endothelium and grow commensurately with the child. They may be classified as *macrocystic*, *microcystic*, or *mixed*. On ultrasound, they typically appear as multiloculated cystic masses with poorly defined, infiltrating borders. They have preponderance for the head and neck and do not have malignant potential. Fifty percent of lesions are apparent in the neonatal period, whereas 75% present within the first year of life. Those diagnosed in early gestation tend to be associated with chromosomal abnormality. Teratomas, in contrast, are germ cell tumors composed of tissues foreign to their anatomic site (such as neural elements, cartilage, and respiratory epithelium). All three germ cell layers are represented. They may involve the floor of the mouth or the tongue and can extend into the mediastinum. On ultrasound they tend to be cystic appearing with well-defined margins, although the two lesions may be difficult to distinguish.
2. MRI. Visualization of the mass preoperatively will allow precise surgical planning and help to predict the need for airway intervention at birth (**Fig. 70.2**). Fetal MRI,

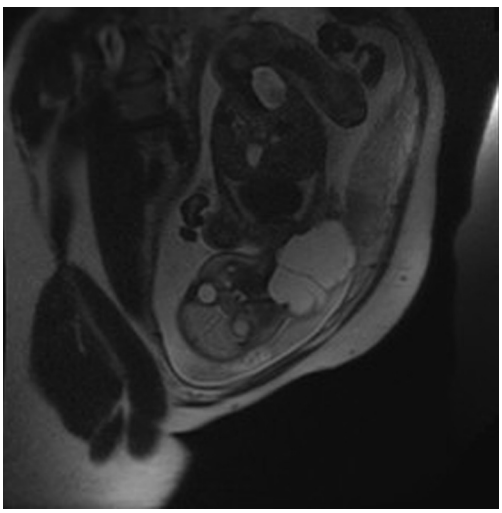

Fig. 70.2 Example of a prenatal magnetic resonance imaging scan (in a different case from the one presented here) showing a multiloculated cystic lesion arising from the cervical region of the fetus. The imaging assists assessment of the airway and planning for delivery.

as was used in the described case, has been shown to provide information on the physical characteristics, extent, vascularity, and relationship of the lesion to surrounding structures. Radiation is avoided and there have been no deleterious effects on the developing fetus. Biopsy is rarely required to differentiate the various tumors that arise in the neck. The child in this case had a lymphangioma diagnosed prenatally. MRI was also used for the neonate once the airway was stable to assist planning for resection and suitability for alternative therapies.

3. Computed tomography. This modality may provide further information about bony involvement and skeletal abnormality, but it generally plays a limited role given the inherent lack of soft tissue detail and significant exposure to unnecessary ionizing radiation.
4. Plain radiographs. Radiographic films may assist assessment of dental development in later years because cervicofacial lymphangiomas can cause skeletal distortion, malocclusion, mandibular overgrowth, and anterior open bite.

◆ Diagnosis

Lymphangioma of the cervicofacial region causing fetal airway and esophageal obstruction

◆ Medical Management

The neonate must be stabilized after the airway is secured and thoroughly evaluated for associated anomalies. Only after the child is considered stable should surgical excision of the mass be planned. Pulmonary function must be maximized preoperatively or the resection must be delayed until the neonate's pulmonary function has been optimized. Nutritional support may be required by means of a nasogastric or orogastric tube. If long-term support is required because of impaired swallowing ability, gastrostomy tube placement may be prudent.

Most children with head and neck lymphangioma are not diagnosed prenatally and do not present emergently as the child in this case. An asymptomatic neck lump is the most common complaint. For these children, the most significant complications of lymphangiomas are intralesional bleeding and infection, which often result in rapid expansion of the mass. Intravenous antibiotics are prescribed if there is an infective exacerbation but are generally not required prophylactically. Some children with limited disease are successfully managed expectantly, and there are reports of spontaneous regression in a small percentage.

Aspiration of the cysts has been described for the treatment of macrocystic lymphangiomas but often results in recurrence. Sclerotherapy has emerged in recent years as an acceptable alternative to surgery in select lesions. Under ultrasound guidance, the cysts are aspirated and then injected with a sclerosant such as hypertonic saline, sodium tetradecyl sulfate, absolute ethanol, bleomycin, or *Streptococcus pyogenes*–derived OK-432. Admission to hospital for overnight observation is recommended if there is a risk of airway swelling secondary to the manipulation of the lesion. Microcystic lesions are not suitable for sclerosant therapy.

◆ Surgical Management

The indications for surgical treatment of lymphangioma include functional limitations, intolerable symptoms, and altered aesthetics. The risk of surgery must be weighed against the risk of potential morbidity from operating in a small and distorted surgical field, especially given that this is a benign condition. Given the complex nature and presentation of lymphangioma, treatment should be individualized for each patient. Surgery may need to be staged over months to years depending on the symptoms and site of the disease.

The goal of surgery is complete resection, but this is seldom possible because lymphangiomas are often closely related to normal structures that must be preserved. The approach to the large cervicofacial lymphangioma is dictated by the extent of the lesion. Unilocular masses can be easily dissected. Infiltrative lesions are resected with the goal of maintaining function. The recurrence rate in these children is high. No vital structures should be resected with the mass because lymphangiomas are not malignancies and have no malignant potential. If the lymphangioma is infiltrating into the base of

the tongue or lateral pharyngeal wall, the potential for continued growth of the lesion and long-term airway compromise with laryngeal infiltration is high. A tracheotomy may then be required for long-term management of the patient. The blood volume of the newborn is of the order of 75 mL/kg, making the typical blood volume of a 3-kg neonate about 200 mL. Meticulous hemostasis throughout the resection is essential, but despite the best techniques, these neonates generally require intraoperative transfusions.

The airway should always be evaluated, either before or after resection of a cervicofacial lymphangioma, to determine the extent to which the airway is involved. This particular patient was successfully intubated at birth owing to the size of the mass and the potential for airway compromise. She was stabilized in the pediatric intensive care unit and underwent excision of the infiltrating lymphangioma at 1 week of age. A tracheotomy was performed because of the intense infiltration into the base of the tongue, lateral pharyngeal wall, and supraglottic larynx. Residual tumor that causes functional problems will be addressed on a pragmatic basis as she continues to grow.

In addition to ablative surgery, microcystic disease involving the tongue can be treated with radiofrequency ablation, laser, and cautery to address lesions on the surface. Tongue- reduction surgery may be helpful if bulky infiltrating disease prevents oral closure (**Fig. 70.3**). Orthodontics and orthognathic procedures may be required for facial skeletal deformities in later years.

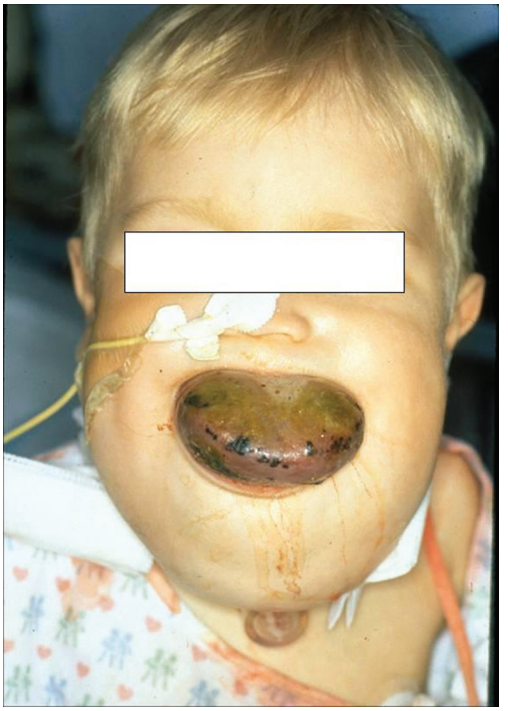

Fig. 70.3 Microcystic infiltration by lymphangioma of the tongue and floor of mouth causing airway obstruction, feeding difficulties, prevention of mouth closure, surface bleeding, and dental distortion. CO_2 laser and tongue reduction surgery were eventually required.

Rehabilitation and Follow-up

Long-term follow-up of these patients is required. Further resections are based on the location of persistent disease, the degree to which the recurrent mass becomes symptomatic for the patient, and the degree to which function is impaired. The mere presence of persistent lymphangioma does not necessitate resection. Multidisciplinary input from otolaryngology, plastic surgery, dentistry, nutritionists, and speech and language therapy is essential in the long-term management of these children.

Questions

1. Maternal-fetal circulation is best maintained via which of the following?
- **A.** EXIT (ex utero intrapartum technique) procedure
- **B.** OOPS (operation on placental support) procedure
- **C.** Cesarean section with umbilical cord left in tact
- **D.** Spontaneous vaginal delivery

2. Polyhydrmnios is a predictor of which of the following?
- **A.** Teratoma

B. Lymphangioma
C. Tracheal obstruction
D. Lack of pulmonary development

3. Management of a lymphangioma invading the tongue base and supraglottic larynx of a neonate includes which of the following?
A. En bloc resection of the mass
B. Injection of sclerosing agents into a microcystic lesion
C. Bood transfusion
D. Orthognathic procedures

Suggested Readings

Arneja JS, Gosain AK. Vascular malformations. Plast Reconstr Surg 2008;121(4):195e–206e

Dasgupta R, Adams D, Elluru R, Wentzel MS, Azizkhan RG. Noninterventional treatment of selected head and neck lymphatic malformations. J Pediatr Surg 2008; 43(5):869–873

Farrell PT. Prenatal diagnosis and intrapartum management of neck masses causing airway obstruction. Paediatr Anaesth 2004;14(1):48–52

Hartnick CJ, Rutter M, Lang F, Willging JP, Cotton RT. Congenital high airway obstruction syndrome and airway reconstruction: an evolving paradigm. Arch Otolaryngol Head Neck Surg 2002;128(5):567–570

Lim FY, Crombleholme TM, Hedrick HL, et al. Congenital high airway obstruction syndrome: natural history and management. J Pediatr Surg 2003;38(6):940–945

Marwan A, Crombleholme TM. The EXIT procedure: principles, pitfalls, and progress. Semin Pediatr Surg 2006;15(2):107–115

Mong A, Johnson AM, Kramer SS, et al. Congenital high airway obstruction syndrome: MR/US findings, effect on management, and outcome. Pediatr Radiol 2008;38(11):1171–1179

Ogamo M, Sugiyama T, Maeda Y, et al. The ex utero intrapartum treatment (EXIT) procedure in giant fetal neck masses. Fetal Diagn Ther 2005;20(3):214–218

71

Stridulous Child

Jeremy D. Prager and Charles M. Myer III

History

A 4-month-old girl is transferred to a tertiary pediatric hospital for otolaryngology consultation for a chief complaint of progressively worsening "noisy breathing" for 3 weeks. Her breathing has been worse when she is agitated. She has been feeding well and gaining weight appropriately. Changing her position does not alter her breathing. She has had no apnea, cyanosis, or sick contacts. She is an only child and is not known to have ingested anything other than formula. She has undergone no previous operations or intubations. Delivery was by cesarean section.

On physical examination, a well-developed infant is noted to be in no acute respiratory distress and comfortable in her parent's arms. Her respiratory rate is normal for age, and she is without nasal flaring or supraclavicular or subcostal retractions. Her cry is not hoarse or high pitched. Auscultation of her neck reveals a mild biphasic stridor, which becomes audible and increases in intensity when she is agitated. Inspection of the head and neck reveals a small cutaneous hemangioma on the right anterior neck skin. Flexible nasopharyngoscopy at the bedside demonstrates a normal supraglottic and glottic airway and fullness in the left subglottic region. The vocal cords are mobile. The patient's anteroposterior neck film reveals mild left subglottic narrowing. The trachea appears normal. A chest radiograph is normal without hyperinflation or mediastinal shift. Microlaryngoscopy and bronchoscopy are planned. The patient is started on a systemic steroid (e.g., prednisone 2–3 mg/kg/day).

Differential Diagnosis—Key Points

1. Airway complaints, especially those in young children, are some of the most challenging consultations for otolaryngologists. Quick assessment of the patient is required, noting the ABCs (airway, breathing, and circulation) of critical care. Within moments, it is possible to place this patient into the "not acutely ill" category.
2. This patient's symptom is progressive biphasic stridor. On physical examination and imaging there is suggestion of asymmetric narrowing in the subglottis. Incidental note has been made of a cutaneous hemangioma. No signs or symptoms of acute inflammation, such as fever to support the diagnosis of croup, epiglottitis, or bacterial tracheitis, are seen.
3. The patient's breathing was normal after birth, and nasopharyngoscopy has revealed bilateral cord motion and a normal supraglottis, ruling out vocal cord paralysis and

laryngomalacia. The patient has had no intubations, thus ruling out acquired subglottic stenosis or arytenoid dislocation. Congenital subglottic stenosis remains a possibility, as does tracheomalacia.

4. Consideration of foreign-body aspiration is obligatory in children with respiratory complaints. This patient's clinical course is not consistent with foreign-body aspiration.
5. Given the above key points, microlaryngoscopy and bronchoscopy are required to evaluate the subglottis and tracheal airway.

◆ Test Interpretation

Anteroposterior neck films can demonstrate subglottic and tracheal narrowing. The classic anteroposterior neck film is that performed for croup, in which case the subglottis is usually narrowed circumferentially and comes to a superior point (the "steeple sign"). The neck film on this patient revealed a mild narrowing of the left subglottic region and no tracheal narrowing, most consistent with a diagnosis of subglottic hemangioma.

Microlaryngoscopy and bronchoscopy confirmed a normal supraglottis and glottis. In the immediate left subglottis there was a submucosal compressible mass that obstructed about half of the patient's subglottic airway (**Fig. 71.1**). In addition, a small right submucosal compressible mass involved the undersurface of the right true vocal cord and immediate subglottis. The remainder of the tracheobronchial tree was normal.

◆ Diagnosis

Subglottic hemangioma

◆ Medical Management

Approximately half of patients with a subglottic hemangioma will have a cutaneous hemangioma. Subglottic hemangiomas are usually asymptomatic at birth. Once the hemangioma enters the proliferative phase (usually at 2 to 3 months of age), biphasic stridor is noted. The proliferative phase lasts about 1 year. The natural history of the hemangioma is typically

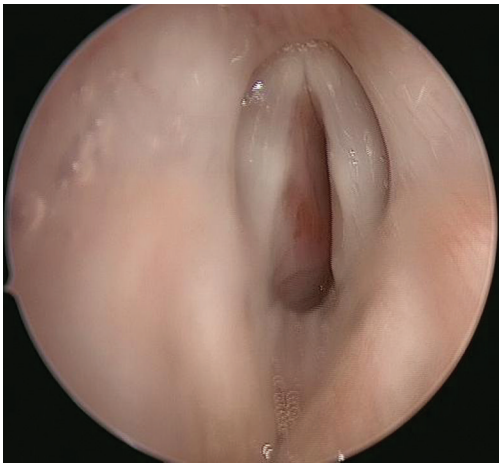

Fig. 71.1 Subglottic hemangioma.

then to involute slowly, over years. Glucose tranporter-1, an endothelial cell glucose transport protein, is overexpressed in infantile hemangiomas and may be used as a marker of disease.

Systemic steroid therapy is the mainstay of medical therapy for subglottic hemangioma. The exact mechanism of the steroid's action is not known, but in certain patients the drug causes a rapid decrease in the size of the lesion. Patients are customarily started on prednisone 2 to 3 mg/kg/day and tapered over several weeks. Some patients can be maintained on occasional steroids until the hemangioma involutes, a process that occurs slowly over several years. The benefit of this management must be weighed against the side effects of chronic steroid use, namely, endocrine abnormalities and growth suppression.

Intralesional steroid therapy may be successful in managing the hemangioma but often requires prolonged periods of intubation and multiple procedures. Interferon-α is often successful at managing the hemangioma as well but has a lengthy and serious list of side effects that include fevers, elevated liver enzymes, seizures, and spastic diplegia. It may be considered as a medical therapy of last resort.

The use of propranolol has recently been reported in the management of cutaneous and subglottic hemangiomas. If it is as successful as initial reports indicate, surgery may no longer be needed for these lesions.

Surgical Management

Tracheotomy was at one time the surgical management of choice for subglottic hemangiomas. With a tracheotomy, the airway obstruction is bypassed for as long as necessary, usually until adequate involution. The tracheotomy may be needed for years and is associated with its own not insignificant morbidity and mortality. For these reasons, tracheotomy is often used as a surgical treatment of last resort.

Endoscopic CO_2 laser resection of the hemangioma has been the preferred method of surgical treatment. Successful hemangioma management without the need for tracheotomy, prolonged intubation, or steroid therapy is often possible. This method may be ideal for small to moderate symptomatic hemangiomas in patients who are either not responsive to steroids or in whom chronic steroid therapy is not desirable. Unfortunately the CO_2 laser penetrates through the hemangioma and may cause damage to underlying and surrounding normal tissue, leading to fibrosis, scar formation, and potential glottic and subglottic stenosis. However, conservative laser treatment of limited lesions is usually successful without complications. Anecdotally, laser treatment seems more successful when used in combination with a short course of steroids and, in some instances, a short course of intubation. Several other lasers, such as the potassium-titanyl-phosphate and neodymium:yttrium-aluminum-garnet have been used as well.

Open excision of subglottic hemangioma has experienced a recent increase in popularity. The approach is similar to a cricoid split or laryngotracheoplasty. A full laryngofissure may be performed if access is difficult without splitting the thyroid cartilage. An anterior split of the airway is performed, and the hemangioma is removed with submucosal dissection. Anterior cartilage grafting may be performed at the same time. The patient may require several days to a week of postoperative intubation. This approach may bypass the need for a tracheotomy or prolonged steroid use in patients with large or circumferential hemangiomas. However, one must keep in mind the favorable natural history of this lesion when choosing this option. Creation of voice abnormalities from improper closure of the laryngofissure is a real possibility.

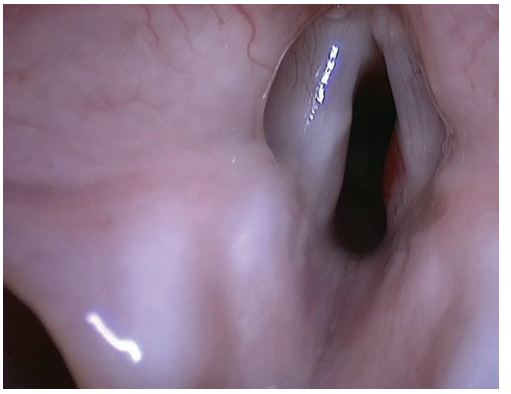

Fig. 71.2 Postoperative view with small residual right glottis-subglottic hemangioma.

This patient underwent an open hemangioma excision addressing both the left and right subglottic disease. To avoid traumatizing the right true vocal cord, a small amount of residual hemangioma was left on its undersurface. **Figure 71.2** is from microlaryngoscopy and bronchoscopy about 2 months postoperatively.

Rehabilitation and Follow-up

The natural history of subglottic hemangiomas is involution, although the timing is unpredictable. It behooves the otolaryngologist to consider surgical therapy that preserves as much normal anatomy as possible. Follow-up endoscopy is necessary to ensure that the airway remains patent and that residual hemangioma, if it exists, does not become obstructive.

Questions

1. The gradual onset of progressive biphasic stridor in a 4-month-old child suggests obstruction at what level?

A. Supraglottis
B. Glottis
C. Subglottis

D. Trachea
E. Distal airways

2. Subglottic hemangioma management may include all but which of the following?
A. Observation
B. Tracheotomy
C. Systemic steroids
D. Laser resection
E. Radiation

3. A stable 2-year-old child with current vaccinations and respiratory compromise of acute onset without viral prodrome likely has which of the following?
A. Croup
B. Epiglottitis
C. Bacterial tracheitis
D. Laryngomalacia
E. Airway foreign body

Suggested Readings

O-Lee TJ, Messner A. Subglottic hemangioma. Otolaryngol Clin North Am 2008;41(5):903–911, viii–ix

Rahbar R, Nicollas R, Roger G, et al. The biology and management of subglottic hemangioma: past, present, future. Laryngoscope 2004;114(11):1880–1891

Vijayasekaran S, White DR, Hartley BE, Rutter MJ, Elluru RG, Cotton RT. Open excision of subglottic hemangiomas to avoid tracheostomy. Arch Otolaryngol Head Neck Surg 2006;132(2):159–163

72

Juvenile Nasopharyngeal Angiofibroma

Matthew Bromwich and Jay Paul Willging

History

An 11-year-old boy was referred to a tertiary care center with a history of recurrent self-limiting epistaxis. The epistaxis was spontaneous and usually left sided. He also complained of 6 months of left-sided nasal obstruction. There was no history of trauma. He has had no change in vision or facial sensation. He denied headache.

His medical history was unremarkable. He has no allergies and takes no medication. His surgical history was significant for a previous endoscopic procedure to remove a similar mass 1 year ago.

On physical examination, obstruction of the left nasal passage was demonstrated by retained secretions. His extraocular motion was intact. He had no diplopia. His visual acuity was grossly intact. The other cranial nerves were intact. His oral cavity and oropharynx were unremarkable. Nasopharyngoscopy demonstrated a fleshy red mass in the left posterior choanae, and it filled the nasal cavity. The nasal septum was pushed to the right, and the medial maxillary wall was partially taken down (**Fig. 72.1**).

Differential Diagnosis—Key Points

The differential diagnosis of a child with a nasal mass includes nasal polyps, antrochoanal poly, nasopharyngeal angiofibroma, hemangiopericytoma, squamous papilloma, inverted papilloma, sinus mucocele, lymphoma, encephalocele, nasal glioma, teratoma, esthesioneuroblastoma,

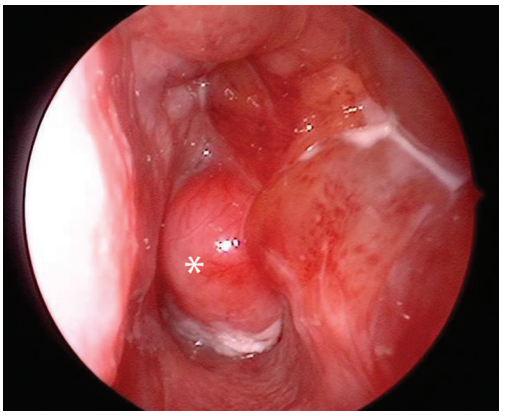

Fig. 72.1 Nasal endoscopic image of posterior nasal choanal obstruction with recurrent vascular mass.

lymphoid hyperplasia, rhabdomyosarcoma, or carcinoma.

After benign lymphoid hyperplasia (adenoid hypertrophy), juvenile nasopharyngeal angiofibroma is the most common type of benign nasopharyngeal tumor, accounting for 0.05% of all head and neck tumors. Juvenile nasopharyngeal angiofibroma (JNA) has a frequency of 1 in 5000 to 60,000 otolaryngology patients. They most commonly occur in prepubescent boys in the second decade; age range is 7 to 19 years. JNA is rare in patients older than 25 years. Some authorities advocate genetic testing if this tumor is found in a phenotypic female patient.

JNA may be secondary to a desmoplastic response of the embryonic chondrocartilage during the development of the cranial bones. The tumor is found at the superior margin of the developing sphenopalatine foramen formed by the bifurcation of the palatine bone, the horizontal ala of the vomer, and the root of the pterygoid process. From this location the tumor has easy access to the pterygopalatine fossa through the pterygomaxillary fissure. The tumor may pass into the nasopharynx, the infratemporal fossa, or through the skull base intracranially. As the tumor grows, it expands the pterygomaxillary fissure, causing anterior bowing of the posterior wall of the maxillary antrum.

A hormonal theory for development of a JNA has been suggested because of the lesion's occurrence in adolescent males. Etiology from nonchromaffin paraganglionic cells of the terminal branches of the maxillary artery has also been suggested. Deletions of chromosome 17, including regions for tumor suppressors, are present.

Patients typically present with nasal obstruction and epistaxis. As the tumor progresses, hyponasal speech and cheek and palate deformities may occur. The mass presents as a red, smooth, mucosa-covered compressible mass in the nasal cavity and nasopharynx. JNA is an aggressive benign neoplasm. Recurrence rates after treatment vary from 0 to over 50%.

◆ Test Interpretation

The plain radiographic signs thought to be classic for juvenile nasopharyngeal angiofibroma are found in **Table 72.1**. However, in most centers computed tomography (CT) will be the primary diagnostic modality.

Table 72.1 Plain-film signs of juvenile nasopharyngeal angiofibroma

Nasopharyngeal mass
Anterior bowing of the posterior wall of the maxillary antrum
Erosion of the sphenoid bone
Erosion of the hard palate
Erosion of the medial maxillary sinus
Displacement of the nasal septum

The anterior bowing of the posterior wall of the antrum seen on lateral skull film is known as the Holman-Miller sign and is pathognomonic for juvenile nasopharyngeal angiofibroma.

CT with contrast or magnetic resonance imaging (MRI) is helpful for diagnosis and staging. With these imaging studies, the extent of involvement and intracranial extension may be determined. The location and radiographic appearance of the tumor are generally diagnostic.

Biopsy is rarely needed to confirm diagnosis and should be avoided in the office because of the vascular nature of the tumor. The typical histologic appearance is an unencapsulated mass of vascular channels with a fibrous stroma. The vessel walls lack elastic fibers and have absent or incomplete smooth muscle (**Fig. 72.2**). This abnormal histology accounts for the propensity for hemorrhage with minor trauma. The cellularity of the stroma and vessels is benign. There is an abundance of mast cells found in the stroma and lack of other inflammatory cells.

Several staging systems have been created and modified to describe juvenile nasopharyngeal angiofibroma. Most are based on the extent and location of the tumor. The Sessions staging system is the most widely used for juvenile angiofibroma (**Table 72.2**).

This child's CT scan reveals a large nasopharyngeal tumor extending from the pterygomaxillary fissure into the posterior choanae. Minimal widening of the pterygomaxillary fissure is seen (**Fig. 72.3**). This CT scan demonstrates minimal anterior bowing of the posterior wall of the antrum. The Holman-Miller sign is pathognomonic for juvenile nasopharyngeal angiofibroma. The tumor also extends through the pterygomaxillary fissure into the pterygopalatine fossa and has eroded a portion

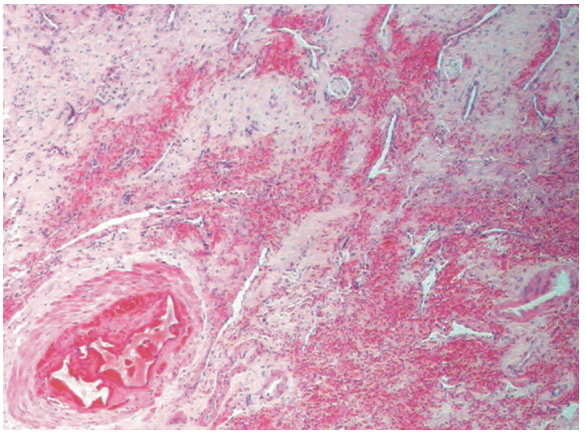

Fig. 72.2 Fixed hematoxylin and eosin histologic slide with multiple vascular clefts without muscular walls. A large vessel with a normal muscular wall is also seen.

of the pterygoid plates (**Figs. 72.3** and **72.4**). Generally, JNA does not aggressively invade intracranial structures but erodes bone and displaces the meninges. Intracranial extension is important to determine, however, because it may affect treatment planning.

◆ Diagnosis

JNA, stage IIA

◆ Medical Management

Hormonal regulation using androgen and estrogen therapy with marked reduction in tumor size has been reported in the literature. The use of hormonal therapy has been limited, however, because of hormonal side effects and a variable response to treatment.

Dramatic tumor shrinkage from low-dose external beam radiation in tumors that are intimately associated with vital structures has been reported. The time to maximal affect from radiotherapy can be as long as 12 months to 36 months. However, radiotherapy is not a benign treatment but a useful alternative when surgical excision is impractical. Radiotherapy may be of further use preoperatively when vital structures are involved to reduce tumor size in large JNA (**Table 72.2**).

◆ Surgical Management

The preferred management for JNA is complete surgical extirpation. The morbidity and potential mortality of surgery must be contemplated

Table 72.2 Staging of juvenile nasopharyngeal angiofibroma based on the system developed by Sessions et al

Stage	Description
A	Tumor limited to the posterior nares or nasopharyngeal vault
B	Tumor involving the posterior nares or nasopharyngeal vault with involvement of at least one paranasal sinus
IIA	Minimal lateral extension into the pterygomaxillary fossa
IIB	Full occupation of the pterygomaxillary fossa with or without superior erosion of orbital bones
IIIA	Erosion of the base of skull (middle cranial fossa/base of pterygoid)—minimal intracranial extension
IIIB	Extensive intracranial extension with or without extension into the cavernous sinus

Source: Sessions RB, Bryan RN, Naclerio RM, Alford BR. Radiographic staging of juvenile angiofibroma. Head Neck Surg 1981;3(4):279–283.

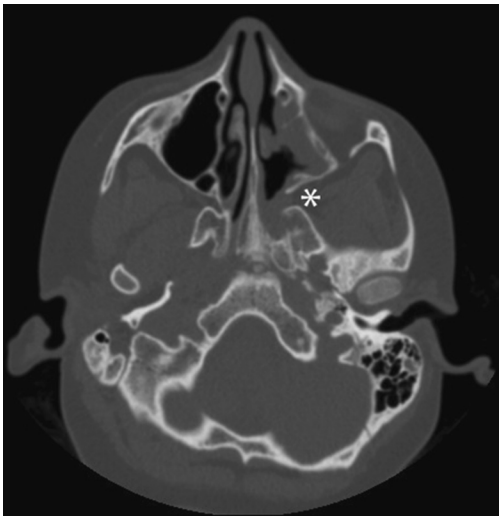

Fig. 72.3 Axial computed tomography with widened pterygomaxilliary fissure. Pterygopalatine fossa with widened pterygomaxillary fissure (*asterisk*).

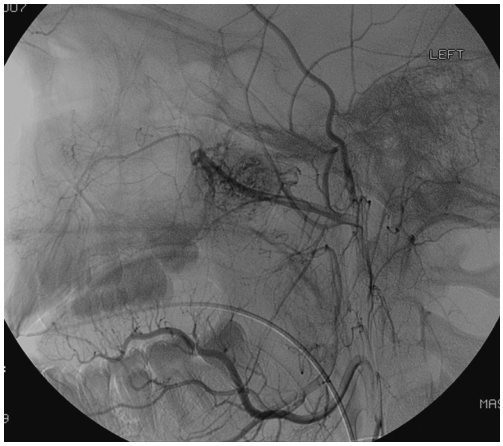

Fig. 72.5 Angiogram. Dye blush indicating vascular tumor in nasopharynx.

when planning therapy, however. Many surgical approaches have been described, including transnasal, transpalatal, transmandibular, transzygomatic, transantral, combined craniotomy and rhinotomy, and lateral rhinotomy. The lateral rhinotomy incision gives a foundation for excellent exposure. It may be combined with a lip-splitting incision and partial maxillectomy for exposure of the infratemporal fossa. Recurrences of small and medium tumors treated surgically approach zero.

Angiography is useful to delineate the vascular supply of the tumor and to allow preoperative embolization of the tumor. The main blood supply generally comes from the internal maxillary artery or the ascending pharyngeal artery (**Fig. 72.5**). Angiography and embolization should be undertaken only in preparation for surgery because revascularization occurs quickly.

Management of JNA that invades intracranially and the skull base is more controversial. The recurrence rate after surgical excision increases with the size of the tumor and its association with intracranial structures and skull-base suture lines. Erosion of the medial wall of the cavernous sinus has been reported as a poor predictor for complete excision. A recurrence rate of 32% has been found with surgical excision of larger tumors (stage IIIA or IIIB).

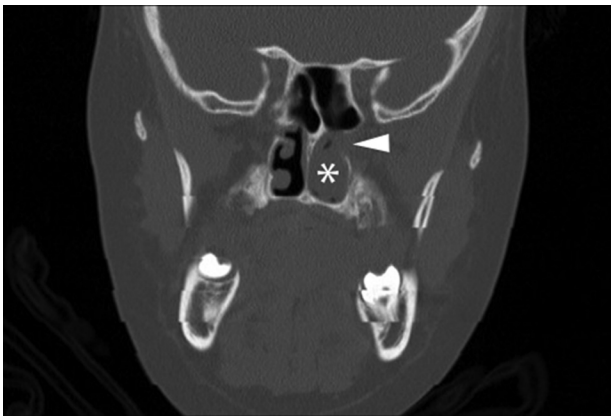

Fig. 72.4 Axial computed tomography. Mass in nasopharynx (*asterisk*). Widened pterygomaxillary fissure (*arrow head*).

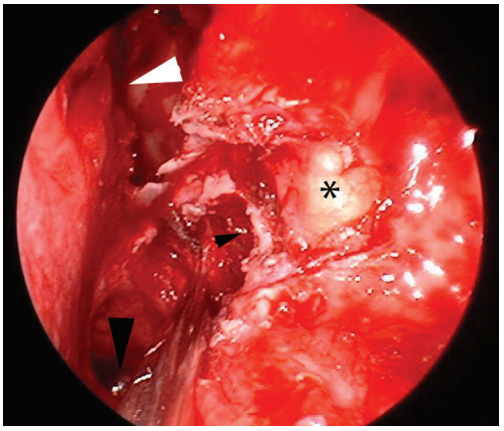

Fig. 72.6 Recurrent vascular lesion. Pterygopalatine fossa fat (*asterisk*). Sphenoid sinus (*white arrow*). Nasopharynx (*large black arrow*). Ptyergoid plates (*small black arrow*).

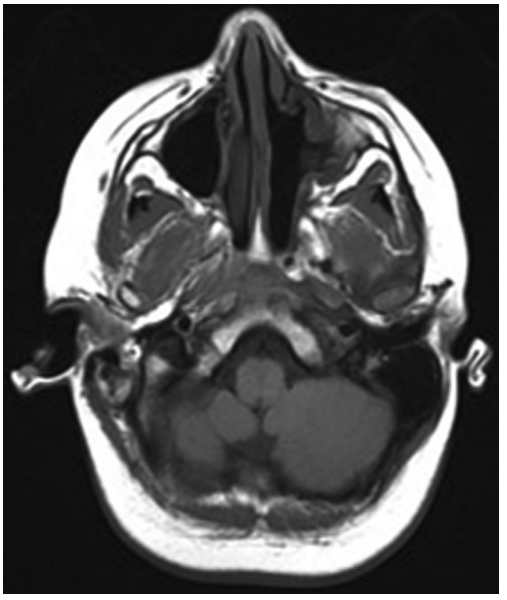

Fig. 72.7 Magnetic resonance imaging taken 6 months postoperatively.

Rehabilitation and Follow-up

This case illustrates management of a moderately sized recurrent angiofibroma. Difficult exposure and loss of endoscopic landmarks are to be expected in revision surgery. Particular care was paid in this case to remove the lesion from behind the eroded pterygoid plates, where it may have been left from previous attempts (**Fig. 72.6**). Close follow-up with potential surgical salvage or even radiotherapy must be considered if further recurrence occurs. At 6 months a repeat MRI was obtained and no evidence of recurrence was seen (**Fig. 72.7**). His symptoms have improved with a cessation of epistaxis and improved nasal airflow.

Questions

1. Girls with juvenile nasopharyngeal angiofibroma (JNA) should undergo what testing?
   - **A.** Sex hormone levels for possible hirsutism
   - **B.** Attention-deficit hyperactivity disorder levels
   - **C.** Karyotype and sex chromosome analysis for testicular feminization
   - **D.** Complete coagulation studies
   - **E.** Pregnancy test
2. The most appropriate preoperative workup for a moderately sized JNA includes which of the following?
   - **A.** CT
   - **B.** MRI
   - **C.** Angiography
   - **D.** CT, embolization
   - **E.** CT and/or MRI, blood work, embolization, group and reserve
3. Office biopsy of nasal lesions is safe and effective and can expedite diagnosis and treatment.
   - **A.** True
   - **B.** False

Suggested Readings

Beham A, Kainz J, Stammberger H, Auböck L, Beham-Schmid C. Immunohistochemical and electron microscopical characterization of stromal cells in nasopharyngeal angiofibromas. Eur Arch Otorhinolaryngol 1997;254(4): 196–199

Bremer JW, Neel HB III, DeSanto LW, Jones GC. Angiofibroma: treatment trends in 150 patients during 40 years. Laryngoscope 1986;96(12):1321–1329

Fagan JJ, Snyderman CH, Carrau RL, Janecka IP. Nasopharyngeal angiofibromas: selecting a surgical approach. Head Neck 1997;19(5):391–399

Herman P, Lot G, Chapot R, Salvan D, Huy PT. Long-term follow-up of juvenile nasopharyngeal angiofibromas: analysis of recurrences. Laryngoscope 1999;109(1): 140–147

Sessions RB, Bryan RN, Naclerio RM, Alford BR. Radiographic staging of juvenile angiofibroma. Head Neck Surg 1981;3(4):279–283

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Posttonsillectomy Hemorrhage

J. Matthew Dickson and Charles M. Myer III

History

A 5-year-old boy was evaluated for spitting up blood in the postanesthesia recovery unit 1 hour after a tonsillectomy and adenoidectomy. The procedure was done by means of electrocautery, and hemostasis was obtained using suction electrocautery. The child had just woken up and had not yet had anything to drink. He was immediately returned to the operating room, and bleeding was noted in the left tonsillar fossa. This was controlled with suction cautery, and the child was observed in hospital for 24 hours with no further bleeding. Five days later he was taken to the emergency department because of oral bleeding. The physical examination revealed an alert child with a pulse of 130 beats per minute and a blood pressure of 110/70. Examination of the oropharynx revealed no active bleeding. A clot was noted in the left tonsillar fossa. Complete blood count (CBC), prothrombin time (PT), activated partial thromboplastin time (PTT), and platelet function assay (PFA) were within normal limits. The clot was removed in the emergency department, and slight oozing was noted. Silver nitrate was used to control the bleeding. The child was admitted to the hospital for observation and discharged after 24 hours with no further bleeding. A clinic follow-up 3 weeks later revealed well-healed tonsillar fossae.

Differential Diagnosis—Key Points

1. This patient presented initially with posttonsillectomy bleeding within the first 24 hours after adenotonsillectomy. This is considered primary hemorrhage and is usually due to improper hemostasis during the primary surgery. Secondary posttonsillectomy hemorrhage is defined as any bleeding after the first 24 postoperative hours. Such bleeding usually occurs between postoperative day 5 and 10 and commonly is associated with premature separation of the granulation membrane that forms over the pharyngeal surface after tonsillectomy. The rate of primary hemorrhage generally ranges from 0.2 to 2.2% and of secondary hemorrhage from 0.1 to 3%.
2. Repeated postoperative bleeding should raise the possibility of a coagulation disorder. A thorough preoperative medical history regarding the patient and immediate family members should be obtained. Points that need to be addressed are history of easy bruising, epistaxis, oral bleeding, posttraumatic hemorrhage, excessive circumcision bleeding, postoperative or dental hemorrhage, hemarthrosis, perinatal bleeding, and recent use of any medication, especially nonsteroidal anti-inflammatory and anticoagulant medications. Systemic disorders

that might result in excessive bleeding, such as liver disease, renal disease, or hematologic disease, should be addressed as well. The preoperative physical examination may help to detect possible coagulopathies and provides additional information. The presence of petechiae might suggest vascular or platelet disorders. Mucosal and gastrointestinal hemorrhage may be associated with vascular abnormalities such as bleeding into an elbow or knee joint, which is characteristic of hemophilia A (factor VIII deficiency) or factor IX deficiency. The presence of hepatosplenomegaly may indicate a liver disorder or hemolytic neoplasm.

3. In cases of recurrent bleeding with normal coagulation studies, vascular abnormalities should be suspected and evaluation by angiography should be done.

◆ Test Interpretation

Platelet count, activated PTT, PT, and PFA are useful in the detection of inherited and acquired coagulation disorders. These tests should always be obtained preoperatively if the history or medical examination suggests a coagulation disorder. If the patient's clinical history raises the definite possibility of a specific coagulation disorder, specific tests should be performed. For example, if the patient has a history of spontaneous bruising or if the PFA-EPI and PFA-ADP are prolonged and the platelet count is normal, additional laboratory tests should be focused on the possibility of von Willebrand disease (vWD) or a primary platelet disorder. When the PFA-EPI is abnormal and the PFA-ADP is normal, aspirin-induced platelet dysfunction is most likely. Actual preoperative detection of a coagulation defect allows for vital preoperative management that improves patient care by reducing perioperative complications and may prevent a disastrous surgical outcome. The use of routine preoperative laboratory screening tests is questionable because these tests are regarded to have low positive predictive value with poor sensitivity.

Coagulation testing should be obtained following a bleeding episode and, if abnormal, hematology consultation and additional coagulation testing should be obtained. A CBC also should be obtained to assess the degree of hemorrhage and as a baseline in case further bleeding develops. In this case, laboratory analysis revealed a platelet count of 300,000/nm^3 (normal range, 135,000–466,000/nm^3), PT 12.9 seconds (normal range, 12.6–14.5 seconds), a PTT 29 seconds (normal range, 25.9–35.6 seconds), a PFA-EPI 98 seconds (normal range, 75–199), and a PFA-ADP 84 (normal range 68–112 seconds) after the secondary bleeding episode. Hematocrit was initially 35% but dropped to 30% after the second bleeding episode.

The combination of multiple bleeding episodes with normal coagulation tests should raise the suspicion of a vascular abnormality that needs to be evaluated by angiography. Angiography has a low but significant risk of morbidity and mortality. Yet, in cases of repeated postoperative bleeding, this study is essential to detect possible vascular abnormalities that might cause the repeated episodes of bleeding.

◆ Diagnosis

Posttonsillectomy hemorrhage of unknown cause

◆ Medical Management

Any coagulation disorder detected preoperatively or postoperatively should be addressed and treated as necessary. Two of the most common coagulation defects encountered are platelet dysfunction and vWD. More than 20 subtypes of vWD have been recognized to date. Some of these subtypes respond to desmopressin acetate (DDAVP) treatment. This should be evaluated by a challenge test with this drug 1 or 2 weeks before surgery. If DDAVP is to be used, treatment should be given about 1 hour before surgery at a dose of 0.3 mg/kg of body weight during a 30-minute infusion and once daily thereafter until wound healing is complete. Cryoprecipitate or factor VIII concentrates, which contain vW multimers, should be available in the event that the DDAVP fails to control hemorrhage. Other subtypes of vW are treated with exogenous vWD factor replacement in the form of cryoprecipitate or factor VIII concentrate. Drugs are the most common cause of platelet dysfunction,

and aspirin is probably the most commonly used drug known to affect platelet function. When a hemostatically normal patient who is taking aspirin requires a surgical procedure, aspirin should be discontinued for at least 2 weeks. If severe hemorrhage due to deficient platelet function is suspected, either a random donor platelet transfusion or DDAVP would be effective rapidly.

Patients with posttonsillectomy bleeding are usually admitted for observation and blood tests. An exception to this rule may be a delayed bleeding episode without signs of clot or active bleeding. This patient can be kept in an observation unit for around 4 hours while being hydrated before discharge. If the blood loss is excessive and the patient is symptomatic, blood transfusion should be considered.

◆ Surgical Management

The most important factor in management of postoperative tonsillectomy bleeding is prevention. The actual method of removal of tonsils, either by dissection or electrocautery, is probably not as important as attention to the detail of staying in the proper plane between the tonsillar capsule and its surrounding fossa. Intracapsular microdebrider tonsillectomy has shown some promise in decreasing the postoperative hemorrhage rate, but intraoperative bleeding is often increased. Newer techniques, which include bipolar radiofrequency ablation (coblation) and CO_2 laser, have not proven to decrease the incidence of postoperative bleeding. Hemostasis is generally obtained with either a suture tie or electrocautery. The operating surgeon must not terminate the procedure until she or he is absolutely confident of hemostasis. If any bleeding is noted in the mouth during emergence from anesthesia, the surgeon should not be reluctant to have the anesthesiologist deepen the plane of anesthesia so that the pharynx can be reexamined and any bleeding stopped. Although a small amount of blood-tinged saliva is acceptable in the recovery room and for the first 24 hours, any amount of bright red blood coming from the mouth or nose should alert the surgeon to the possibility of postoperative bleeding and initiate a complete examination of the oropharynx and the possibility of exploration of the area in the operating room.

Frequent swallowing and tachycardia have been reported as indirect evidence of possible postoperative bleeding. If the child has continued emesis with coffee-ground material, active pharyngeal bleeding should be suspected and the appropriate investigations and treatment should take place. If a clot is present in the tonsillar fossa during the physical examination, this should be removed because it can prevent normal tissue retraction and subsequent healing. The age and cooperativeness of the child are important in the decision-making process. In the cooperative patient, local or topical anesthesia may be sufficient to allow direct examination and removal of the clot. Hemostasis can be achieved using silver nitrate cautery or electrocautery. If this fails or if the patient is unable to tolerate care in an awake state, general anesthesia will be required to control the airway and allow the surgeon to work without distraction. The initial step in hemostasis is suctioning off any fresh clot from the tonsillar fossa. Identified bleeding sites are controlled with electrocautery or a suture tie. If these measures are not successful, suture ligature may be required to stop the bleeding. However, this must be done with extreme care. The tissue in the postoperative tonsillar fossa is fragile. Placement of sutures is often difficult because the suture tends to tear through the tissue and pull out. The surgeon then may attempt to place the suture deeper to hold more securely and may inadvertently lacerate a major vessel deep to the fossa, possibly leading to aneurysm formation. If the measures mentioned are not successful in stopping the bleeding, consideration must be given to exploration of the neck and ligation of the external carotid artery or its small distal branches. If possible, preoperative angiography may be helpful and embolization might be considered a surgical alternative or adjunct.

◆ Rehabilitation and Follow-up

If any coagulation disorder is discovered during the preoperative or postoperative period, this should be addressed and treated properly. The patient and his parents should receive a complete hematologic consultation, and the patient should be followed up periodically as necessary.

◆ Questions

1. Primary posttonsillectomy hemorrhage is defined as bleeding that occurs:
   A. Within the first 6 hours postoperatively
   B. Within the first 24 hours postoperatively
   C. Between days 1 to 3 postoperatively
   D. Between days 5 to 10 postoperatively
2. What is the most common cause of platelet dysfunction?
   A. Von Willebrand disease
   B. Medication induced
   C. Chronic liver disease
   D. Thrombotic thrombocytopenic purpura
3. Treatment of posttonsillectomy hemorrhage can include which of the following?
   A. Observation
   B. Silver nitrate cautery
   C. Suction electrocautery
   D. Suture ligature
   E. All of the above

Suggested Readings

Bolger WE, Parsons DS, Potempa L. Preoperative hemostatic assessment of the adenotonsillectomy patient. Otolaryngol Head Neck Surg 1990;103(3):396–405

Burk CD, Miller L, Handler SD, Cohen AR. Preoperative history and coagulation screening in children undergoing tonsillectomy. Pediatrics 1992;89:691–695

Lowe D, van der Meulen J, Cromwell D, et al. Key messages from the National Prospective Tonsillectomy Audit. Laryngoscope 2007;117(4):717–724

Schmidt R, Herzog A, Cook S, O'Reilly R, Deutsch E, Reilly J. Complications of tonsillectomy: a comparison of techniques. Arch Otolaryngol Head Neck Surg 2007;133(9):925–928

Windfuhr JP, Chen YS, Remmert S. Unidentified coagulation disorders in post-tonsillectomy hemorrhage. Ear Nose Throat J 2004;83(1):28–39

74

Cystic Fibrosis

Matthew Bromwich and Sally R. Shott

History

A 5-year-old girl with a history of cystic fibrosis (CF) was referred to a tertiary care center with several months of chronic sinusitis and increasing nasal congestion and drainage. She denied headache, fever, or facial pain. She has a history of good pulmonary function, but in the past several months, her forced expiratory volume in 1 second (FEV_1) had unexplainably decreased with no improvement despite aggressive medical interventions. The patient had been previously treated with courses of Augmentin, Omnicef, and Biaxin with no improvement in her nasal congestion and FEV_1.

Her medical history was significant for a prior diagnosis of CF and asthma. She has not required hospitalization or intubation. Patient reported no allergies. Medications include hypertonic saline, albuterol, Flonase, Prilosec and pancrelipase.

Her surgical history was negative.

On physical examination a large amount of inspissated mucus was present in the middle meatus. Crusted secretions were obstructing the left anterior nose. Purulent material was seen emanating from beneath the middle turbinate. Palpation of the face was positive for mild tenderness over the maxillary sinuses. Her tympanic membranes were normal, and the examination was otherwise unremarkable.

Differential Diagnosis—Key Points

Sinus radiographs are usually abnormal children with CF. Computed tomography (CT) scans have been reported to be abnormal in 100% of those evaluated.

The most likely diagnosis in this patient is chronic sinusitis with nasal polyposis. Further evaluation is needed to determine the extent of the disease and specifically to determine whether there is evidence of bony erosion from the chronic disease process.

CF is an inherited autosomal recessive disorder that results in abnormal function of the exocrine glands. Although more than 800 mutations exist, the most common is a missense mutation of the *CFTR* gene called deltaF508. The *CFTR* gene codes for an essential and ubiquitous chloride channel. Exocrine glands and ducts become obstructed throughout the body as a result of the abnormal chloride channel function. This results in the most common clinical manifestations of chronic obstructive pulmonary disease and gastrointestinal malabsorption secondary to pancreatic insufficiency.

Sinusitis is also common, if not universal, in patients with CF. Identified as a component of CF in the 1950s, Lurie first described the association of nasal polyposis and CF in 1957. The incidence of nasal polyposis in CF patients has

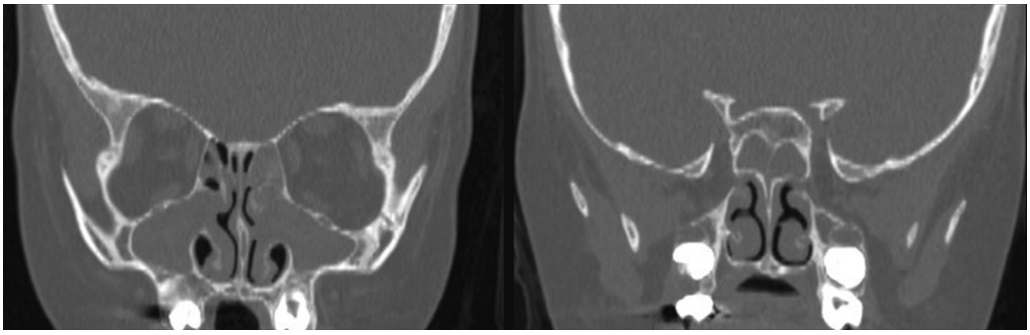

Fig. 74.1 Preoperative computed tomography in a patient with cystic fibrosis. Maxillary and sphenoid opacification.

been reported to be between 6 and 48%. The most common symptoms include nasal obstruction, rhinorrhea, and headache. Patients rarely complain of sinus symptoms specifically. Further the degree of disease is not well correlated with severity of symptoms.

◆ Test Interpretation

A CT scan of the paranasal sinuses was performed on this patient. As seen in **Fig. 74.1** there is opacification of the maxillary and sphenoid sinuses. The osteomeatal unit is also expanded. Partial opacification of the anterior/posterior and frontal sinuses is present. Edema of the inferior turbinates is also seen. No areas of bony deficiency are identified.

◆ Diagnosis

Chronic sinusitis with evidence in a patient with CF

◆ Medical Management

The effectiveness of medical management of sinusitis and nasal polyps in children with CF has been extremely disappointing. Studies evaluating the bacteriology of sinusitis in children with CF commonly identify *Pseudomonas aeruginosa*, *Staphylococcus aureus*, and *Haemophilus influenzae* as well as anaerobes. Commonly the degree of sinus disease is correlated with the severity of pulmonary problems. Prolonged courses of antibiotics and steroids, both intranasal and systemic, decongestants, and antihistamines remain the mainstay of medical management. The main goal of antibiotic therapy is to prevent, delay, or suppress formation of biofilms within the sinuses and lungs. Although only partially successful in controlling disease, it is thought that the use of antibiotics may contribute to the lower incidence of complications of sinusitis in this patient population, including orbital cellulitis and osteomyelitis. Unfortunately, however, these therapies have not been shown to affect the initial development of nasal polyps or to decrease the incidence of recurrence of the sinusitis and polyps.

The incidence of allergy in children with CF is no different from that in the general population. Other studies have shown that the pathologic nature of the nasal polyps attributable to CF is different from allergic polyps.

Hypertonic saline irrigations can be helpful. This solution, made up of water or saline, pickling salt, and baking soda, helps to mobilize the thick, tenacious nasal and sinus secretions seen in children with CF. Similarly, nasal irrigation with tobramycin has proved useful in the postoperative period.

◆ Surgical Management

Although surgical intervention has been shown to provide symptomatic relief, especially in terms of controlling the congestion

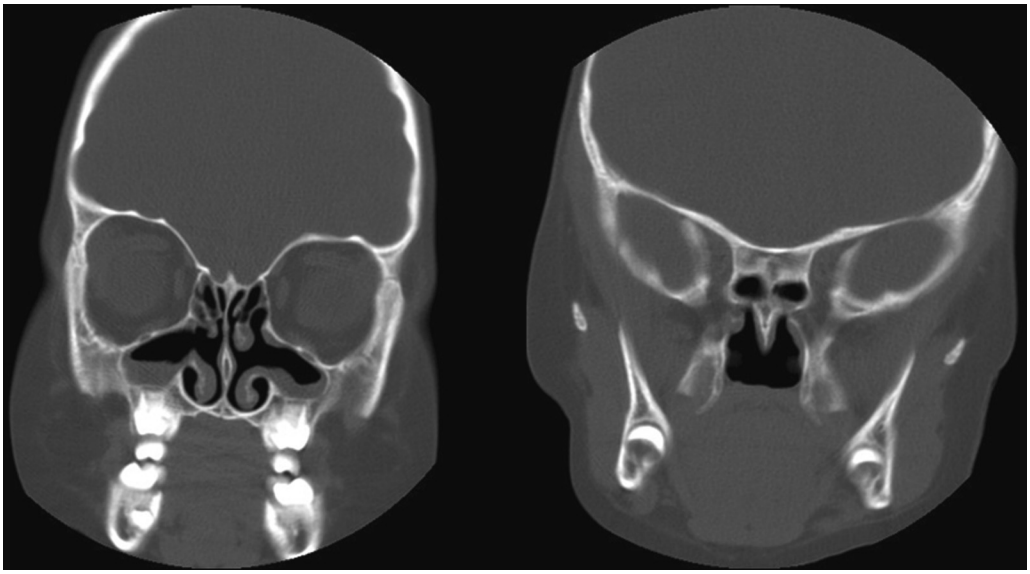

Fig. 74.2 Postoperative imaging.

and headaches associated with sinusitis and nasal polyps, the long-term effectiveness of surgery can also be disappointing. Studies evaluating the effectiveness of different surgical procedures discuss results in terms of "fewer recurrences" and "longer symptom-free" periods, but not in terms of cure rates.

However, some improvement in this regard has been made. Simple nasal polypectomy in a child with CF and nasal polyps is usually not effective. More aggressive sinus surgery has been shown to decrease the frequency of recurrences and provide longer symptom-free intervals but is rarely curative. Functional endoscopic sinus surgery (FESS) with intranasal ethmoidectomies and maxillary antrostomies with removal of polyps from the maxillary sinuses are usually required. Aggressive FESS using CT navigation and microdebrider techniques has been associated with longer symptom-free periods. Surgery is indicated only in patients with symptomatic nasal and sinus disease or when the nasal disease burden negatively impacts lung function. CT findings generally should not be used to guide timing of surgery. Ideally preoperative sedation can be avoided to enable patients to resume normal pulmonary clearance activities as soon as possible following surgery.

In this patient the first surgical intervention was performed at age 5. The CT depicted in **Fig. 74.2** shows a satisfactory early postoperative outcome. Large maxillary antrostomies, complete anterior ethmoidectomies, and bilateral sphenoidotomy were performed. The patient remained asymptomatic for only 1 year (**Fig. 74.3**).

Moss and King reported a further increase in the symptom-free period by the addition of serial antimicrobial lavage in addition to the endoscopic surgery. Placing catheters into the maxillary sinuses via the antrostomy at the time of surgery, they then treated their patients with tobramycin irrigations for 7 to 10 days postoperatively. They reported an increase in the symptom-free interval to at least 2 years.

The improvement seen with the postoperative antimicrobial irrigations stresses the need for continued medical management following surgical intervention in this ongoing, relentless disease. Hypertonic saline nasal irrigations are also quite helpful in mobilizing the tenacious secretions out of the opened sinus cavities.

Because of the chronic nature of this disease process, the need for revision surgery should be expected, and these expectations need to be openly discussed with the patient and their family. The timing of such revision surgery depends on the severity of the symptoms of nasal obstruction, headache, and facial pain and the possible effects of the purulent postnasal

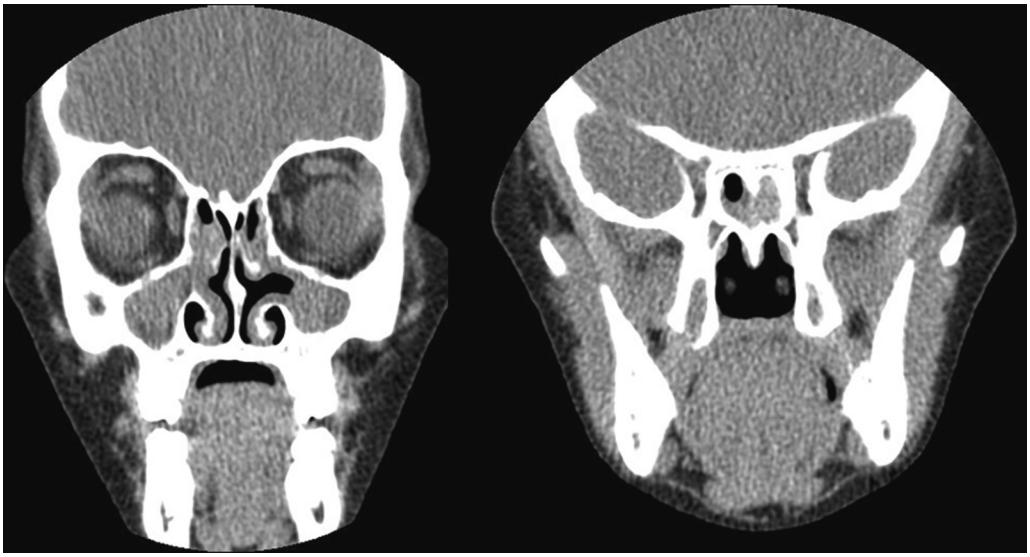

Fig. 74.3 Recurrent disease.

drainage on the pulmonary status of the patient. Exacerbations of the patient's lung disease may be linked to exacerbations of the sinusitis. At revision surgery one may find that all that is required is suctioning of the thick, pastelike secretions from the sinuses. It is also possible, however, that one will find recurrence of the extensive polyposis seen at the original surgery. Multirevision surgery in children is not without risk. Loss of landmarks from prior surgery and erosion increases the risk of surgical misadventure and injury to the skull base and eye. The use of image guidance in revision surgery is recommended.

Because of the underlying pulmonary disease, special considerations need to be taken with regard to general anesthesia and surgery. Because of the effects of CF on the child's cardiopulmonary systems, a full medical evaluation should be done before surgery by the child's pulmonologist and by the anesthesiologist. Maximization of the patient's pulmonary status before surgery is required with possible in-house preoperative antibiotics and pulmonary physiotherapy. Presurgical hydration is also important. All potential medical problems associated with CF, including pancreatic dysfunction, diabetes, and coagulation abnormalities should be considered. Communication between the surgeon, the anesthesiologist, the primary care physician, and the various specialists participating in the patient's care is imperative.

◆ Rehabilitation and Follow-up

Because of the chronic nature of sinusitis in children with CF, it is wise to encourage open communication not only with the patient and his or her family but also with the patient's pulmonologist, the most likely primary manager of the patient's care. This will provide better decision making in terms of timing of revision surgery and will keep you informed as to the pulmonary status of the patient, especially if a general anesthetic is being considered.

◆ Questions

1. What is the underlying genetic abnormality responsible for cystic fibrosis (CF)?
A. Abnormally thick secretions
B. Mutation in *CFTR* gene
C. 22q deletion
D. X chromosome translocation

2. What is the rationale behind endoscopic sinus surgery in CF patients? (Choose any that apply.)
- **A.** To preserve as much normal ciliary function as possible
- **B.** To open the sinuses as much as possible to allow for draining of dysfunctional sinuses
- **C.** To cause atrophy of the sinus air spaces
- **D.** To create a nasal airway

3. Endoscopic sinus surgery: (Choose any that apply.)
- **A.** Creates a nasal airway by removing polyps
- **B.** Reduces the patient bacterial burden of *Pseudomonas*
- **C.** Can result in an increase in the patient's FEV_1
- **D.** Can cure a patient of the sinus complications of CF
- **E.** Is contraindicated in patients with CF

Suggested Readings

Cole RR, Cotton RT. Preventing postoperative complications in the adult cystic fibrosis patient. Int J Pediatr Otorhinolaryngol 1990;18(3):263–269

Crockett DM, McGill TJ, Healy GB, Friedman EM, Salkeld LJ. Nasal and paranasal sinus surgery in children with cystic fibrosis. Ann Otol Rhinol Laryngol 1987;96(4):367–372

Cuyler JP, Monaghan AJ. Cystic fibrosis and sinusitis. J Otolaryngol 1989;18(4):173–175

Drake-Lee AB, Morgan DW. Nasal polyps and sinusitis in children with cystic fibrosis. J Laryngol Otol 1989;103(8): 753–755

Moss RB, King VV, Moss RB, King VV. Management of sinusitis in cystic fibrosis by endoscopic surgery and serial antimicrobial lavage: reduction in recurrence requiring surgery. Arch Otolaryngol Head Neck Surg 1995;121(5):566–572

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Down Syndrome

Marlene A. Soma and Sally R. Shott

History

A 6-year-old boy with Down syndrome presents to the ear, nose, and throat clinic with a 4-day history of draining ears. His mother reports that he has had nasal obstruction and rhinorrhea for many months as well. She also noted a long history of trouble breathing at night, with loud snoring, mouth breathing, and preferring to sleep in the sitting position. These symptoms improved after an adenoidectomy and tonsillectomy had been performed 18 months previously, but they had slowly returned. She was not particularly concerned about this but on further questioning admitted he was becoming more fatigued during the day and not performing his normal daytime activities as well as before.

The patient's history was significant for requiring three previous sets of pressure-equalizing (PE) tubes for chronic otitis media with effusion. He had also had multiple episodes of croup as an infant but had not had any symptoms of this for many years. The mother denied her son having any history of cardiac or pulmonary problems. She did note, however, that her son's gait had become somewhat unsteady, particularly after he was playing in the yard or rolling on the ground, and seemed to lose his balance more easily.

Physical examination revealed a mildly obese 6-year-old boy with the typical Down syndrome facies and an open-mouth breathing posture at rest. His external auditory canals were stenotic and filled with yellow mucoid discharge, making it difficult to visualize the tympanic membranes. He had poor nasal airflow, his inferior turbinates were slightly congested, and yellow rhinorrhea was evident. The oral cavity showed macroglossia, a small oropharynx, and tonsillar fossae that were well healed from his previous surgery. There was no cervical lymphadenopathy. He would not cooperate with flexible nasendoscopy or microscopy of the ears.

Differential Diagnosis—Key Points

Several key points in this history point to potential problems in the care of this child:

1. The patient has a history of chronic ear disease and eustachian tube dysfunction requiring multiple sets of PE tubes. Children with Down syndrome are more predisposed to this condition because of midface hypoplasia with nasopharyngeal abnormalities and the effects of generalized hypotonia, including the muscles of the palate, immaturity of the immune system, and adenoid hypertrophy. They also have a higher incidence of hearing loss than other children, which may have a significant impact

on their speech and language development. Chronic tympanic membrane retraction and potential for acquired cholesteatoma should also be considered in this population of patients.

2. The child has had nasal obstructive symptoms for many months. Anatomic abnormalities of the midface in Down syndrome children can cause crowding of the nasal cavities and contribute to rhinorrhea and sinusitis. Immature immunologic development may also be a factor here. Although he has had a previous adenoidectomy, it is important to consider adenoid regrowth as a potential cause, which is more common in children with Down syndrome.
3. A previous tonsillectomy and adenoidectomy had been performed on the patient. His symptoms of sleep disordered breathing improved initially but have returned. Unfortunately, because a large percentage of children with Down syndrome have airway obstruction during sleep and sleep apnea starting at a very young age, many parents frequently assume that this is "normal" for their child and do not seek treatment. Airway obstruction in children with Down syndrome can be due to multiple levels of pathology: medially displaced tonsils in the face of midface hypoplasia and a contracted oropharynx, hypotonia with oropharyngeal and hypopharyngeal collapse during sleep, and macroglossia with its associated airway obstruction. In addition, central apnea can be seen. Although removal of the tonsils and adenoids can cure the airway obstruction in typical children, up to 50% of Down syndrome children continue to have ongoing symptoms, which may be very difficult to treat. The long-term effects of obstructive sleep apnea disorders include failure to thrive, pulmonary hypertension, and behavioral issues.
4. This child presents with a history of recurrent croup. Children with Down syndrome are known to have smaller subglottic airways than the normal population and are prone to recurrent episodes of croup. This is important from an anesthesia standpoint when intubation is being considered. It is important to use the appropriate-size endotracheal tube in this child's airway.
5. The patient's history of recently noted abnormal gait, particularly after active play and rolling on the ground, may be due to some coordination problems but could also be due to atlantoaxial instability and resultant compression of the spinal cord. This is of particular importance if contemplating a surgical procedure where the patient's neck will be manipulated.

◆ Test Interpretation

1. Cervical spine films. Before any surgical procedure in which manipulation of the neck is required, particularly in view of the patient's history of change in gait, cervical spine films should be done. Cervical spine abnormalities seen in Down syndrome include atlantoaxial instability, abnormal congenital fusion of the vertebral bodies, degenerative changes in the C2–3 and C3–4 cervical interspaces, and spinal cord compression. Hyperextension, in the presence of atlantoaxial instability, can cause compression of the spinal cord. Lateral neck radiographs in the extension, flexion, and neutral position should be performed. If there is any question of abnormality, as in this patient's case, a computed tomography scan evaluation is also helpful. The cervical spine film of the patient described in this case was normal.
2. Airway films. With this patient's history of recurrent croup, airway films may suggest some subglottic narrowing. Special precautions, particularly in terms of the size of endotracheal tubes used and documentation of an air leak around the endotracheal tube, should be taken at the time of any surgery. The nasopharynx can also be assessed with these radiographs, especially when the patient will not tolerate a flexible endoscopic examination. In this case, the subglottic airway appeared narrow, and review of his previous surgical records showed he required an endotracheal tube two sizes smaller than usual for his age. Significant adenoid regrowth obstructing the nasopharynx was also identified.
3. Chest radiography. With this patient's long history of upper airway obstruction, a chest radiograph would be helpful to rule

out signs of cor pulmonale secondary to the chronic upper airway obstruction. His radiograph was normal.

4. Metabolic studies. Patients with Down syndrome have a higher incidence of hypothyroidism. Again, this would be important to know if general anesthesia is considered and may be contributing to the history of fatigue. If not done previously, these laboratory studies should be done. Thyroid studies were within normal limits for this child.
5. Audiology. The current guidelines for audiologic testing in children with Down syndrome is for screening at birth followed by tests every 6 months up to age 3, then annual testing after 3 years of age. Depending on the child's developmental status and cooperation, ear-specific pure-tone audiometry provides superior information compared with behavioral audiometry. A baseline study was performed at the initial visit and then is repeated after treatment of the otorrhea. He had evidence of a mild conductive hearing loss bilaterally on this occasion.
6. Polysomnogram. A formal polysomnogram is indicated at this point for this patient's treatment course to evaluate objectively the severity of the obstruction and to characterize the type. Children with Down syndrome in particular have multiple potential sources of airway obstruction. They may improve after a tonsillectomy and adenoidectomy, but a high percentage may continue. In addition, a sleep study would help to rule out central apnea. The need for further airway support, such as continuous positive airway pressure (CPAP) or oxygen supplementation, could then be determined. In this case, an elevated obstructed apnea hypopnea index with hypoxemia, hypercarbia, and sleep fragmentation confirmed ongoing obstructive sleep apnea. A trial of CPAP was not tolerated.
7. Cine-sleep magnetic resonance imaging. This test can be used to define the level(s) of residual obstruction. It provides a high-resolution dynamic assessment of the airway without subjecting the child to ionizing radiation. Performed under light sedation, anesthesia, or when the child is sleeping, consecutive fast gradient echo sequence images are taken to capture periods of snoring, obstruction, or oxygen desaturation and are then displayed in a cine format. (**Fig. 75.1**). Multiple levels of the airway can be assessed at the same time. Treatment can be tailored for the individual, directed toward the site of the obstruction. In this patient, adenoid regrowth was confirmed, and relative macroglossia with lingual tonsillar hypertrophy was also found to be contributing to ongoing upper airway obstruction (**Fig. 75.2**).

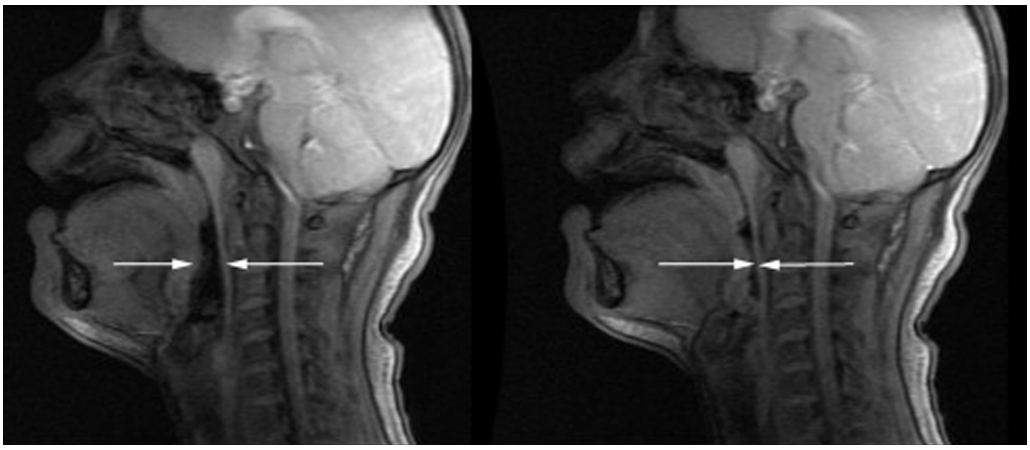


Fig. 75.1 Cine magnetic resonance imaging demonstrating dynamic airway obstruction in a patient with Down syndrome.

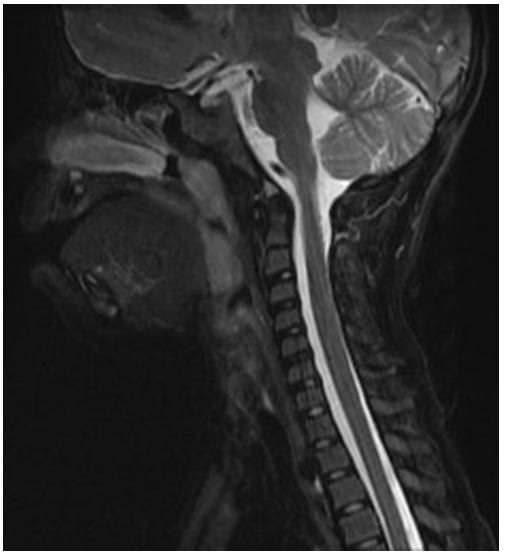


Fig. 75.2 Sagittal magnetic resonance imaging scan showing prominent adenoid regrowth and lingual tonsillar hypertrophy.

Diagnosis

PE tube otorrhea, adenoid regrowth, and persistent obstructive sleep apnea attributable to lingual tonsillar hypertrophy in boy with Down syndrome

Medical Management

The patient has multiple concurrent problems. The external auditory canals should be cleaned to allow examination of the tympanic membranes and a swab collected for microbiological analysis. This may be difficult in the office setting owing to behavioral issues and stenotic ear canals. If the child is systemically unwell and has other symptoms of an upper respiratory tract infection, oral antibiotics may be warranted. Topical antibiotics also play a role.

Adenoid regrowth has been identified as a cause for the nasal obstruction and discharge as well as contributing to the airway obstruction. Topical nasal saline drops may provide symptom relief, and oral antibiotics given for the ears will likely help treat any infective component of adenoiditis before undertaking further treatment that will require general anesthesia.

Surgical Management

In the described case, surgical management is likely to be required to allow ear toilet and otologic microscopy, to treat the adenoid regrowth (revision adenoidectomy), and to address the lingual tonsillar hypertrophy (lingual tonsillectomy). Given that the child has Down syndrome, special precautions must be taken during the surgical procedure to avoid hyperextension of the neck. The surgery is performed with the patient in the neutral position. No shoulder rolls are used, and the head is kept in as horizontal a position as possible.

With this patient's history of recurrent croup, at the time of intubation, a smaller tube than would be expected for the patient's age and size should be used. The appropriateness of the tube chosen should be confirmed with the presence of an air leak around the endotracheal tube.

Lingual tonsillectomy has been described using cautery, the microdebrider, and coblation techniques. A Crowe-Davis gag may be used to provide exposure or, as in this case, a handheld Lindholm laryngoscope.

Postoperatively, patients may have a higher rate of complications than children without Down syndrome because of multiple sources of airway obstruction combined with hypotonia and central apnea. There is a higher incidence of postoperative airway obstruction that will need to be monitored closely, ideally in a high-dependency setting.

Rehabilitation and Follow-up

The patient was given oral and topical antibiotics for the ears in the clinic. Examination under anesthesia revealed patent PE tubes with no evidence of retraction or cholesteatoma. The discharge improved after the medical treatment and revision adenoidectomy. Repeat audiogram showed improvement in hearing thresholds.

The patient's stay in hospital was uneventful, and he was discharged on the first postoperative day tolerating intake well. He was seen in the clinic, and a follow-up polysomnogram was arranged several months later. This showed improvement in all parameters. The patient should be followed up in the future to monitor for the development of recurrent problems.

Questions

1. The manifestations of Down syndrome include all of the following except:
   - **A.** Midface hypoplasia
   - **B.** Epicanthic skin folds
   - **C.** Generalized hypertonia
   - **D.** Macroglossia
   - **E.** Stenosis of the external auditory canal
2. Airway obstruction in Down syndrome children is commonly due to any of the following except:
   - **A.** Adenoid hypertrophy
   - **B.** Lingual tonsil hypertrophy
   - **C.** Central apnea
   - **D.** Vocal cord palsy
   - **E.** Subglottic stenosis
3. Important considerations when intubating a child with Down syndrome include which of the following?
   - **A.** Using an endotracheal tube that is larger than expected
   - **B.** Using an endotracheal tube that is smaller than expected
   - **C.** Using an endotracheal tube that is appropriate for the child's age
   - **D.** Gentle hyperextension of the neck to improve airway visualization
   - **E.** Using a cuffed endotracheal tube

Suggested Readings

Donnelly LF, Shott SR, LaRose CR, Chini BA, Amin RS. Causes of persistent obstructive sleep apnea despite previous tonsillectomy and adenoidectomy in children with down syndrome as depicted on static and dynamic cine MRI. AJR Am J Roentgenol 2004;183(1):175–181

Fricke BL, Donnelly LF, Shott SR, et al. Comparison of lingual tonsil size as depicted on MR imaging between children with obstructive sleep apnea despite previous tonsillectomy and adenoidectomy and normal controls. Pediatr Radiol 2006;36(6):518–523

Goldstein NA, Armfield DR, Kingsley LA, Borland LM, Allen GC, Post JC. Postoperative complications after tonsillectomy and adenoidectomy in children with Down syndrome. Arch Otolaryngol Head Neck Surg 1998;124(2):171–176

Jacobs IN, Gray RF, Todd NW. Upper airway obstruction in children with Down syndrome. Arch Otolaryngol Head Neck Surg 1996;122(9):945–950

Kanamori G, Witter M, Brown J, Williams-Smith L. Otolaryngologic manifestations of Down syndrome. Otolaryngol Clin North Am 2000;33(6):1285–1292

Merrell JA, Shott SR Sr. OSAS in Down syndrome: T&A versus T&A plus lateral pharyngoplasty. Int J Pediatr Otorhinolaryngol 2007;71(8):1197–1203

Mitchell RB, Call E, Kelly J. Ear, nose and throat disorders in children with Down syndrome. Laryngoscope 2003;113(2):259–263

Mitchell RB, Call E, Kelly J. Diagnosis and therapy for airway obstruction in children with Down syndrome. Arch Otolaryngol Head Neck Surg 2003;129(6):642–645

Shott SR. Down syndrome: common otolaryngologic manifestations. Am J Med Genet C Semin Med Genet 2006;142C(3):131–140

Shott SR, Amin R, Chini B, Heubi C, Hotze S, Akers R. Obstructive sleep apnea: Should all children with Down syndrome be tested? Arch Otolaryngol Head Neck Surg 2006;132(4):432–436

Shott SR, Donnelly LF. Cine magnetic resonance imaging: evaluation of persistent airway obstruction after tonsil and adenoidectomy in children with Down syndrome. Laryngoscope 2004;114(10):1724–1729

76

Recurrent Respiratory Papillomatosis

Jeremy D. Prager and Sally R. Shott

History

A 3-year-old boy was taken to his primary care physician with a history of continuous and progressively worsening hoarseness over several months. His history was negative for respiratory complaints with the exception of viral upper respiratory infections, which were normal in course and character. His history was not consistent with vocal abuse or overuse. He did not have any dysphagia and was gaining weight appropriately. His birth history was normal. He was the first child of a young family in a nonsmoking household without pets. His review of systems was negative. Several rounds of antibiotics failed to alleviate the hoarseness, and the patient was referred to an otolaryngologist.

Physical examination in the office revealed a well-appearing child in no acute distress. Examination of the ears, nose, oral cavity, oropharynx, and neck was normal. The child was not retracting. His voice was moderately hoarse, and a mild amount of inspiratory stridor could be heard when the patient was breathing rapidly and deeply. A flexible nasopharyngoscopy revealed a warty, exophytic mass on the left true and false cord, extending to the anterior commissure.

Based on the history and physical examination, the patient was taken to the operating room for a microlaryngoscopy and bronchoscopy with biopsy.

Differential Diagnosis—Key Points

1. Hoarseness is never "normal." It is a symptom that indicates that the apposing vocal cord surfaces have been altered. Hoarseness is, however, a common complaint. The differential diagnosis of hoarseness is lengthy, ranging from inflammation secondary to common viral upper respiratory infections and laryngopharyngeal reflux to more complex disorders, including nerve deficits and neoplastic processes.
2. Hoarseness secondary to viral upper respiratory infection is accompanied by other complaints consistent with the disease process. It is self-limiting. Hoarseness caused by allergic or irritant inflammation will be temporally related to exposure and may be accompanied by other symptoms as well.
3. Laryngopharyngeal reflux results in laryngeal inflammation, which can cause hoarseness as a symptom. Often these children are identifiable by findings on the history and physical examination, including flexible nasopharyngoscopy. This type of hoarseness may be chronic and recurrent, occurring

more consistently in the morning. A course of antireflux therapy may be tried and the patient's presumed diagnosis confirmed by his or her response to therapy. Additional pH probe and impedence studies may be warranted.

4. Constant or progressive hoarseness is commonly due to a vocal cord lesion. The differential diagnosis of a vocal cord lesion differs from that in the adult population. The most common lesions are vocal cord nodules. These lesions are most consistent with vocal abuse. Symptoms can be expected to fluctuate somewhat. Other lesions include neoplastic processes, of which laryngeal papilloma is the most common. Less common lesions include hemangiomas, benign and malignant mesenchymal tumors, and squamous cell carcinoma. Biopsy is required for diagnostic confirmation in cases of neoplasia.
5. Impaired vocal cord mobility may cause hoarseness. This will likely be recognized on flexible nasopharyngoscopy. If impaired vocal cord mobility is a congenital problem or one acquired as an infant (e.g., postcardiac surgery), it is likely to have been identified earlier than 3 years of age. Acquired recurrent laryngeal nerve dysfunction may also be caused by metabolic disorders, nutritional deficiency, heavy metal poisoning, and as part of a postinfectious polyneuropathy. Recurrent laryngeal nerve paresis may result from an overinflated or high-riding endotracheal tube cuff. Vocal cord mobility problems may also cause trauma to the cricoarytenoid joint. causing arytenoid dislocation as can occur during traumatic intubation or blunt trauma to the external neck.

◆ Test Interpretation

Microlaryngoscopy and bronchoscopy were performed with the patient breathing spontaneously under intravenous sedation. Oxygenated air was insufflated into the upper airway with an endotracheal tube placed in the low oropharynx, out of the view of the laryngoscope. In this fashion, endotracheal intubation and excessive instrumentation of the glottis and trachea were avoided. At our institution, photodocumentation is performed at several levels of the airway.

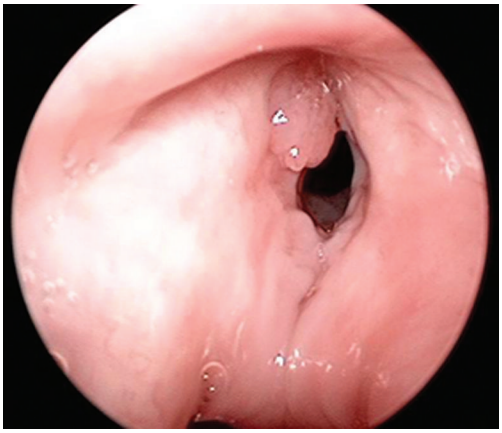


Fig. 76.1 Recurrent respiratory papillomatosis lesion on left true and false vocal folds.

Microlaryngoscopy revealed a discrete focus of exophytic tissue emanating from the surface of the left true and false vocal folds, extending from midcord region to the anterior commissure (**Fig. 76.1**). The surgeon performed an excisional biopsy with a cup forcep followed by removal of all gross disease, sparing the anterior commissure with a microdebrider (**Fig. 76.2**). Histopathologic evaluation revealed polypoid frondlike projections of connective tissue covered by stratified squamous epithelium without evidence of dysplasia (**Fig. 76.3**). Viral typing of the lesion revealed human papillomavirus (HPV) type 6.

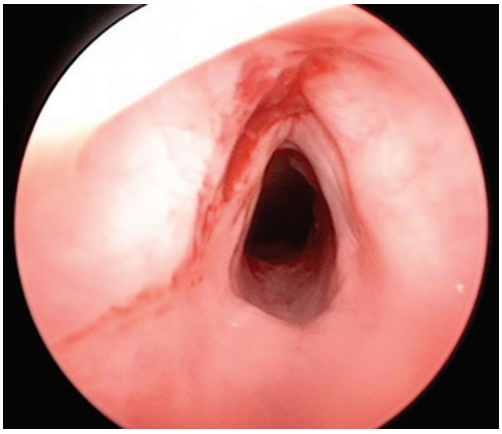


Fig. 76.2 Patient in Fig. 76.1 after microdebrider removal of gross papillomas.

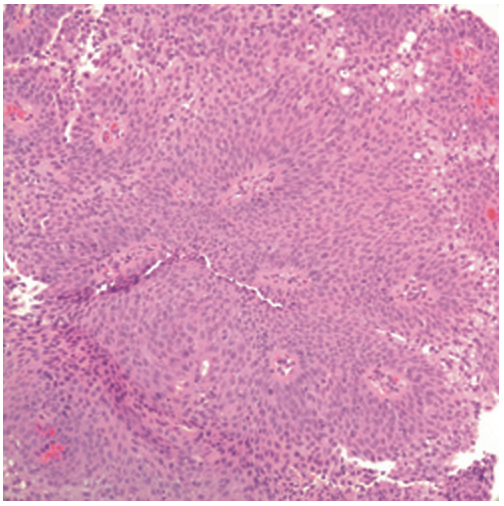


Fig. 76.3 Medium-power hematoxylin and eosin histopathology slide of a papilloma. "End-on" view of papillomas with central fibrovascular cores. (Courtesy of Dr. Rachel Sheridan, Department of Pathology, Cincinnati Children's Hospital Medical Center.)

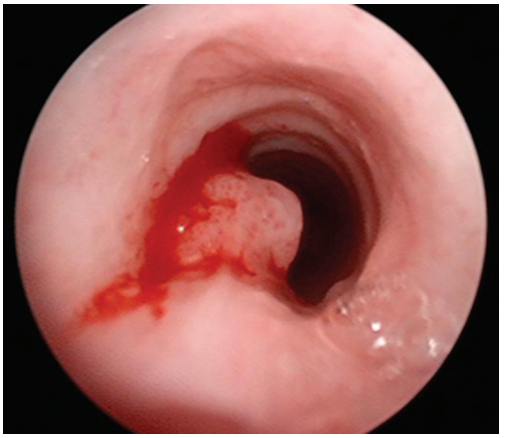


Fig. 76.4 Patient in Figs. 76.1 and 76.2 with evidence of tracheal papilloma partially obstructing the airway.

◆ Diagnosis

Laryngeal papillomatosis. Given that it is the nature of this disease process to recur and present in multiple locations within the respiratory tract, it is often clinically referred to as recurrent respiratory papillomatosis (RRP).

◆ Medical Management

RRP is a chronic disease most commonly caused by HPV types 6 and 11. HPV is a double-stranded DNA virus in the Papovaviridae family. Infection with HPV type 11 is associated with a more aggressive disease course. RRP affects both children and adults. The disease is more aggressive in the pediatric population, and younger children have more aggressive disease than do older children. The incidence is around 4 per 100,000 children. Pediatric patients average nearly 20 surgical interventions. The method of viral transmission is unclear, although vertical transmission during birth is presumed to account for most cases.

The course of RRP is variable and difficult to predict. The course may consist of spontaneous resolution, stable disease with occasional fluctuations in severity, and aggressive disease requiring frequent surgical intervention and adjuvant therapy. Aggressive RRP may spread to more distant sites within the upper aerodigestive tract, including the oral cavity, tracheobronchial tree, and esophagus (**Fig. 76.4**).

By its nature, RRP is both a medical disease in that the epithelium is infected with a virus and a surgical disease in that it can cause voice or respiratory compromise. Surgical management is the main method of treatment. Medical therapy is reserved for disease requiring four or more surgical interventions within a year and is used as an adjuvant to surgery.

Interferon-α therapy usually consists of daily treatment for 6 months followed by thrice weekly treatment. The dose can be weaned, but evidence suggests that longer courses of treatment (on the order of several years) yield better results. There is conflicting evidence regarding success rates with this therapy. Certain patients experience a relapse when therapy is stopped. Common side effects include acute reactions such as fever and flulike symptoms. Chronic reactions include a decrease in growth rate, elevation of liver enzymes, leukopenia, seizures, and spastic diplegia. Interferon-α may be more effective on HPV type 6 than on type 11.

Cidofovir, another antiviral, is the most frequently used adjuvant therapy in pediatric RRP. Originally designed and approved by the Food and Drug Administration as an agent for cytomegalovirus retinitis, evidence supporting the use of cidofovir in RRP is limited to reports

of case series. Studies with similar levels of evidence have failed to show significant benefit. Animal studies have demonstrated carcinogenic potential, an aspect of therapy that must be reviewed in informed consent. Other reported side effects of cidofovir include rash, headache, nephrotoxicity, and neutropenia (more commonly in doses used for cytomegalovirus retinitis), although it should be noted that no systemic side effects have been reported when cidofovir is given intralesionally.

Multiple other agents have been investigated. None has shown convincing benefit. Emerging therapy is focused on HPV vaccines, including the quadrivalent HPV vaccine. Targeting HPV types 6, 11, 16, and 18, this vaccine may prevent the maternal infection and the subsequent vertical transmission.

◆ Surgical Management

Surgical management begins with microlaryngoscopy and bronchoscopy with biopsy. After histopathologic confirmation of disease, surgical management consists of papilloma removal for unacceptable voice or airway compromise. In the setting of disease that requires surgical intervention, yearly biopsy is recommended to evaluate for dysplastic change. Malignant transformation is a rare occurrence.

There are multiple methods of surgical treatment for RRP, including cold steel techniques, CO_2 laser, and powered microdebrider. Recent literature supports the use of the microdebrider as a safe and cost-effective alternative to the CO_2 laser. Each technique has its own advantages and disadvantages. Surgeon preference and comfort often dictate the choice of method. Any surgical procedure should have as its goal the removal of gross disease with preservation of normal structures since latent viral infection exists in surrounding tissue and eradication of this infection is not surgically possible.

◆ Rehabilitation and Follow-up

Yearly biopsy of recurrent lesions is recommended. Aggressive lesions that are rapidly growing or spreading should be biopsied more frequently. The physician may consider an immunodeficiency workup. In addition, controlling laryngopharyngeal reflux is recommended.

◆ Questions

1. What is the most commonly used medical treatment of laryngeal papilloma?
   - **A.** Interferon
   - **B.** Cidofovir
   - **C.** Photodynamic therapy
   - **D.** Viral vaccine
   - **E.** Topical steroid
2. A 4-year-old boy with progressive hoarseness is most likely to have which of the following?
   - **A.** Vocal cord paralysis
   - **B.** Laryngopharyngeal reflux
   - **C.** Vocal cord nodules
   - **D.** Recurrent respiratory papillomatosis
   - **E.** Arytenoid dislocation
3. Proper management of recurrent respiratory papillomatosis includes all but which of the following?
   - **A.** Preservation of normal structures during surgery
   - **B.** Laser masks and suction to minimize aerosolization of virus
   - **C.** Consideration of adjunctive therapy for aggressive disease
   - **D.** Frequent biopsy for aggressive disease
   - **E.** Early tracheotomy

Suggested Readings

Broekema FI, Dikkers FG. Side-effects of cidofovir in the treatment of recurrent respiratory papillomatosis. Eur Arch Otorhinolaryngol 2008;265(8):871–879

Derkay CS, Wiatrak B. Recurrent respiratory papillomatosis: a review. Laryngoscope 2008;118(7):1236–1247

Gerein V, Rastorguev E, Gerein J, Jecker P, Pfister H. Use of interferon-alpha in recurrent respiratory papillomatosis: 20-year follow-up. Ann Otol Rhinol Laryngol 2005; 114(6):463–471

Pasquale K, Wiatrak B, Woolley A, Lewis L. Microdebrider versus CO_2 laser removal of recurrent respiratory papillomas: a prospective analysis. Laryngoscope 2003; 113(1):139–143

XV

Trauma

77

Mandibular Fracture

David B. Hom

History

A 27-year-old man, involved in an altercation, was kicked to the jaw and anterior neck 2 hours previously. He did not lose consciousness. Since then, he complains of malocclusion and difficulty opening his mouth. He notes some neck discomfort and hoarseness but has full range of motion of his neck. On physical examination he has shifting of his jaw to the left and premature contact on his left posterior second molars (teeth #15 and #18). He also has numbness of his right anterior chin. The tympanic membrane is intact with no hemotympanum.

Differential Diagnosis—Key Points

By history and examination, several clinical entities must be considered with this presentation:

1. All patients with head and neck trauma warrant cervical spine evaluation to rule out cervical spine injury, which include cervical spine radiographs and clinical examination. Even in the emergent setting when the clinical situation dictates against complete cervical spine radiographs, the neck must be immobilized and protected from any out-of-axis movements until appropriate cervical injury is ruled out. In some instances low cervical spine injury can be missed despite performance of full cervical radiographic series because of the difficulty of radiographically imaging this area in an injured patient.
2. Laryngotracheal injury must be ruled out in patients who have cervical trauma with symptoms of tenderness at the laryngeal tracheal region, hoarseness, or dyspnea, Serious types of laryngotracheal injuries are fractures of the laryngeal or cricoid cartilages and laryngotracheal separation. Laryngeal fractures may present with anterior neck ecchymosis, subcutaneous crepitance (from free air dissecting out into the soft tissue planes), or crepitance with palpation of the laryngeal fracture. Fiberoptic flexible scope examinations may show edema, ecchymosis, or distortion of the internal anatomy. Hoarseness may be present. In laryngotracheal separation, the trachea is torn away from the larynx. In this instance, the patient may initially still have a good airway, but the airway will progressively decrease, requiring a tracheotomy.
3. Significant blunt cervical trauma can result in vascular involvement. This injury would include intimal disruptions of the carotid.
4. Subjective malocclusion after mandibular trauma increases suspicion of a mandible fracture. Patients with normal dentition can detect small alterations in occlusion

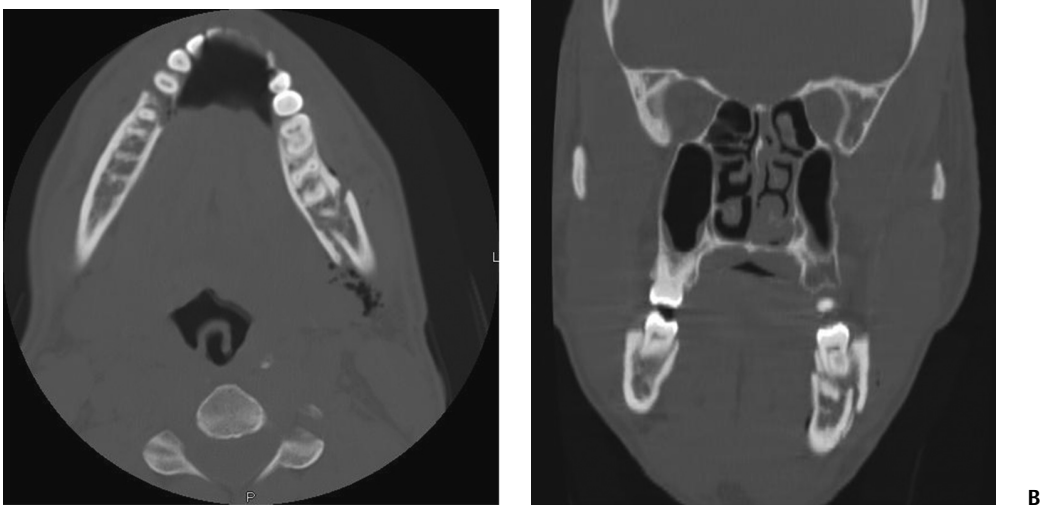
that are smaller than 1 mm. Other causes for malocclusion would be a maxillary fracture, isolated dental injury, and soft tissue injury with muscle spasms or preexisting malocclusion. In addition, numbness of the chin gives evidence that the inferior alveolar nerve is involved with the mandible fracture.

5. If the patient has missing or fractured teeth, suspicion for aspiration of a fractured tooth should be considered and a chest radiograph performed.
6. Most mandible fractures occur in pairs. When two fractures occur on the mandible, the mandibular bone is frequently drawn posteriorly by the attached lingual musculature. In addition, the suprahyoid musculature loses its stabilizing insertion points, which could result in a narrowed airway and inability to clear secretions. In this instance, airway obstruction can occur, which should be treated as an immediate priority.

Test Interpretation

The physical examination will reinforce suspicion that a mandible fracture is present. By placing firm manual posterior pressure on the chin and bimanual testing for mandibular mobility, the patient is usually able to point to the areas of mandible fracture involvement. For any patient with indirect cervicofacial trauma, cervical spine injury needs to be ruled out. If the patient has pain to his neck, anterior-posterior and lateral plain films of the cervical spine are required. If the radiologist is unable to clear the cervical spine, the neck collar should be left in place until a spinal injury has been ruled out. If air is in the neck spaces and plain films are not definitive enough to rule out a cervical spine injury, then an axial computed tomography (CT) scan of the cervical spine will be required.

Once the potential life-threatening injuries (airway, hemodynamic, spinal, laryngotracheal, and vascular) have been ruled out, the facial injuries can be evaluated. A CT scan including axial cuts and coronal cuts (if the cervical spine is cleared) can provide a good radiographic assessment of the mandible (**Fig. 77.1**). In addition a panoramic film can be helpful and can often be performed in conjunction with a CT scan of the head and cervical spine. If a CT scan or orthopantomogram is unobtainable, then plain radiographic views of the mandible (posterior-anterior, right and left lateral oblique, and a reverse Towne) can be helpful and are inexpensive methods for obtaining a mandible fracture assessment.



A B

Fig. 77.1 (A,B) Computed tomography scan of the mandible (axial and coronal) shows left displaced mandibular angle and right body fractures.

Diagnosis

Left mandibular angle and right body fractures

Medical Management

Medical management is directed toward (1) maintenance of an appropriate airway (especially in bilateral subcondylar, angle or body fractures), (2) preventing an infection, (3) pain control, and (4) nutritional support. Prevention of infection involves early initiation of antibiotics that provide coverage for intraoral microorganisms because most mandibular fractures have lacerations extending through oral mucosa. Chlorhexidine mouth rinse may be beneficial. Mandibular pain can be significant. Moderate doses of narcotic analgesics are often required to control pain adequately, but narcotics should be used carefully because of the potential of compromising the airway. If required, pulse oximetry monitoring and close observation may be required. In addition, consideration should be given to providing fluid hydration and nutritional supplementation because patients may have markedly decreased oral intake.

Surgical Management

Successful surgical treatment is anatomic reduction and immobilization of fractured segments to allow for appropriate bone union and establishing premorbid occlusion. Bilateral subcondylar fractures are the most likely mandible fractures that can cause airway obstruction, and a secure airway is the top priority. The definitive repair is performed depending on the location and comminution of the fractures, degree of bony displacement, size of the mandible, presence and condition of dentition, and associated facial fractures. Treatment goals are to establish the patient's premorbid occlusion in three planes of space (anteroposterior, vertical, mediolateral positioning), and accurate condylar repositioning to facilitate optimal mandibular rotation and translation. In addition to occlusion, examining the dental cusps may reveal articular facets that represent areas of dental wear. Dental occlusion is usually established with arch bars and intermaxillary fixation. Next the fractures are exposed through either intraoral or external incisions and the bony segments are reduced and rigidly fixated with mandibular plates and screws.

If there is any question of bony stability, the patient can be left in intermaxillary fixation for 2 to 4 weeks postoperatively to maximize bony union. If the patient has a history of seizure disorder or excessive alcohol overuse, one is very reluctant to leave the patient in intermaxillary fixation because of concern about aspiration during an emesis episode.

Rehabilitation and Follow-up

Follow-up includes optimizing occlusion and bony union while at the same time ensuring proper nutrition until healing is complete. Postoperative radiographs can be done to confirm bony reduction and plate placement. Changes in occlusion should he treated when first identified and aided with elastic rubber bands to guide the bite. If the patient's teeth are wired into maxillary mandibular fixation postoperatively, the patient should always carry a wire cutter to cut these wires if the patient becomes nauseated or before emesis should occur.

After maxillary mandibular fixation, jaw rehabilitation should be started with mandible range-of-motion exercises for at least several months. For subcondylar fractures, jaw exercises (shifting jaw side to side, mouth opening while touching tongue tip to the roof of the mouth, moving the jaw forward) twenty times in a row, four to six times a day are important for rehabilitation.

Questions

1. When two fractures occur on the mandible, the mandible is frequently drawn:
- **A.** Posteriorly by the lingual musculature
- **B.** Laterally by the masseter muscles
- **C.** Anteriorly by the geniohyoid muscles
- **D.** Obliquely by the medial pterygoid muscles
- **E.** Inferiorly by the platysmal muscles

2. To establish optimal dental occlusion during maxillary mandibular fixation, what teeth take the most precedence?
   - **A.** Dental-wear facets of the teeth
   - **B.** First molar contact of the upper and lower teeth
   - **C.** Canine orientation of the upper and lower teeth
   - **D.** Incisor teeth relationship
   - **E.** Premolar contact relationship of the upper and lower teeth
3. What condition would be contraindicated for keeping a patient in maxillary mandibular fixation postoperatively?
   - **A.** Previous orthodontia
   - **B.** History of seizure disorder
   - **C.** History of the being immunocompromised
   - **D.** History of coagulopathy
   - **E.** Decreased mental status
4. What mandible fracture has the highest likelihood of causing airway obstruction?
   - **A.** Bilateral angle fractures
   - **B.** Bilateral symphyseal fractures
   - **C.** A left the symphyseal fracture with an angle fracture
   - **D.** Bilateral ramus fractures
   - **E.** Bilateral subcondylar fractures

Suggested Readings

Ellis E III, Walker LR. Treatment of mandibular angle fractures using one noncompression miniplate. J Oral Maxillofac Surg 1996;54(7):864–871, discussion 871–872

Jewett BS, Shockley WW, Rutledge R. External laryngeal trauma analysis of 392 patients. Arch Otolaryngol Head Neck Surg 1999;125(8):877–880

Kellman R. The cervical spine in maxillofacial trauma: assessment and airway management. Otolaryngol Clin North Am 1991;24(1):1–13

Levy FE, Smith RW, Odland RM, Marentette LJ. Monocortical miniplate fixation of mandibular angle fractures. Arch Otolaryngol Head Neck Surg 1991;117(2):149–154

Valentino J, Marentette LJ. Supplemental maxillomandibular fixation with miniplate osteosynthesis. Otolaryngol Head Neck Surg 1995;112(2):215–220

78

Naso-Orbito-Ethmoid Fractures

David B. Hom

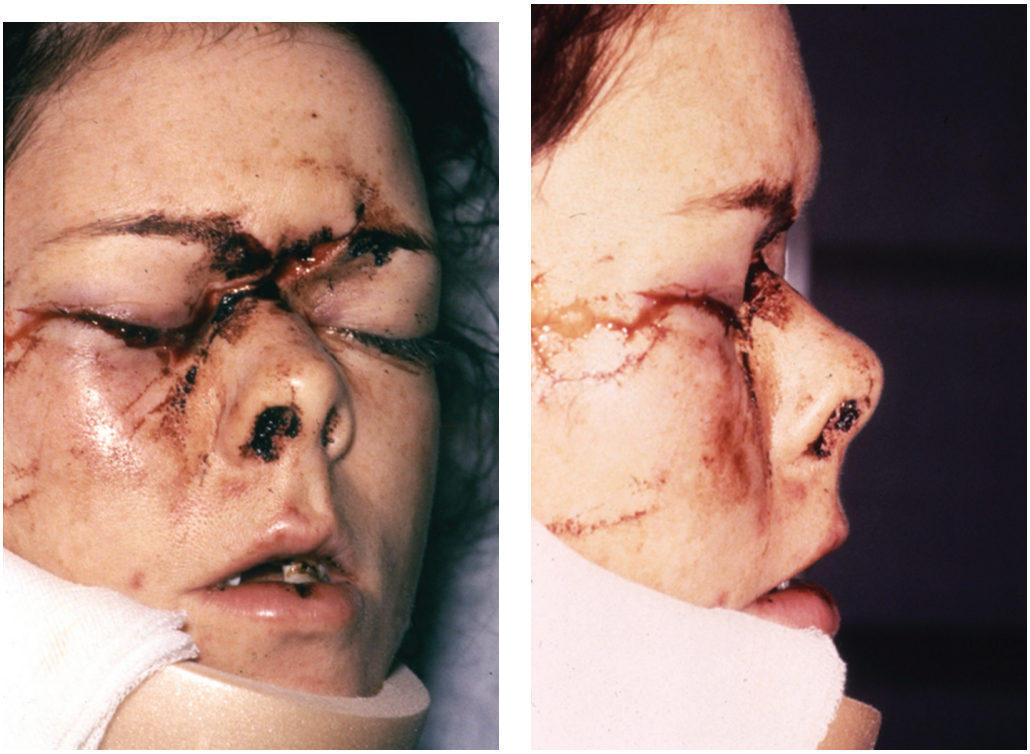
History

A 34-year-old woman involved in a motor vehicle accident struck her upper face to the dashboard with brief loss of consciousness. The steering wheel and windshield were broken with extensive damage to her car. She was transported by ambulance to the emergency department.

On physical examination, she was alert and oriented ×4. Her vital signs were stable. Her C-collar was in place, and two intravenous lines were in place. A 3-cm laceration was over her superior nasal dorsum. The nasal dorsum was obviously depressed with the nasal bones visible through the open wound. With gentle manual manipulation, loose crepitance was evident on the nasal pyramid. No septal hematoma and no evidence of cerebrospinal fluid (CSF) leak was evident. Her eye examination showed normal extraocular movements, and her vision was intact to finger counting. Her pupils were equal and reactive to light. Her intercanthal distance was 42 mm. Her facial buttresses other than her central naso-orbital region were intact. Her cervical spine was later cleared, and her chest radiograph was clear. The remaining physical examination was benign (**Fig. 78.1**).

Differential Diagnosis—Key Points

1. With any facial injury, a complete systemic trauma evaluation is required. The overall status of the patient must be evaluated and take priority (airway, breathing, circulation, cervical status, intracranial status). Only after these are steps are stabilized can obvious facial injuries be addressed. In some instances, the facial fractures may need to be addressed in a later setting, depending on the medical status.
2. The face is examined to evaluate the extent of the fractures while being cognizant of possible injury to surrounding structures, including the orbits and globe, the frontal lobe, the frontal sinus, the medial canthi, and the lacrimal system. Any evidence of CSF leak must be ruled out.
3. Patients with naso-orbito-ethmoid (NOE) fractures may have up to 67% of ocular injuries, with 20 to 25% of these serious and 3% resulting in blindness.
4. Classic clinical findings supporting naso-orbital injury include the following:
   - An intercanthal distance greater than one half the interpupillary distance
   - An intercanthal distance greater than 35 mm



A B

Fig. 78.1 Patient before repair.

- Medial canthus rounding
- Epiphora resulting from injury to the nasolacrimal duct system or a displaced punctum
- Enophthalmos
- Dystopia

5. The clinical diagnosis of NOE fractures might not be obvious because of significant soft tissue swelling masking the underlying skeletal injury of the central midface. Classic nasal findings include a loss of height of the nasal dorsum with superior rotation of the nasal tip (the pig-nose deformity). At times, NOE fractures will be detected only by imaging studies.
6. Complex NOE fractures can be the most challenging facial fractures to repair. Achieving the same pretrauma nasal appearance and medial canthal position can be difficult to attain, and the patient should be informed of this fact.
7. Bimanual examination by placing a mosquito clamp in the nose and digitally palpating the medical canthal on the same side can help determine whether free mobility is present at the naso-orbital ethmoid area. In the past, the eyelid traction test (pulling the lower eyelid laterally away from the nose) has been described as the classic sign for traumatic telecanthus; however, this finding may be evident only in severe cases.

◆ Test Interpretation

Fine-cut coronal and axial computed tomography (CT) scans (1.5-mm slices) of the face are helpful to delineate the fractures and their displacement when the patient is stabilized (**Fig. 78.2**).

◆ Diagnosis

Impacted naso-orbito-ethmoid complex fracture

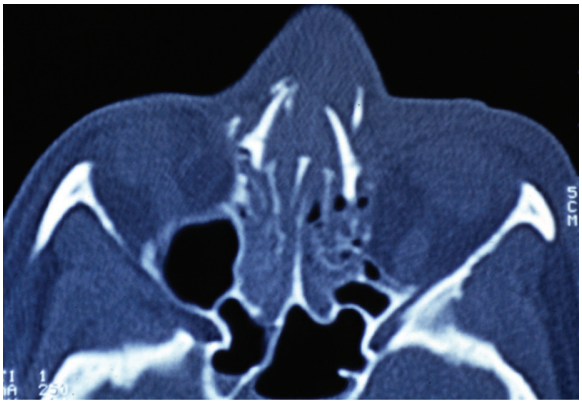


Fig. 78.2 Computed tomography scan (axial view) of the face shows the lacrimal crests bilaterally splayed apart with a traumatic telecanthus.

◆ Medical Management

Most CSF leaks associated with undisplaced fractures isolated to the ethmoid roof will resolve spontaneously after proper fracture reduction. They are managed expectantly by elevating the head of bed, avoiding Valsalva maneuvers, prescribing stool softeners and antitussives, avoiding nose blowing, and observing for spontaneous closure of the leak. However, more extensive comminution of the frontobasilary region associated with dural tears, depressed bony segments, and frontal lobe injury will require neurosurgical involvement. Antibiotic coverage is controversial during this period.

Ocular trauma can often be managed expectantly. Hyphema, corneal abrasions, and subconjunctival hemorrhage require no surgical intervention. Orbital trauma with nondisplaced fractures that do not involve the extraocular muscles usually require no surgical intervention. Retrobulbar hemorrhage may result in blindness if not treated emergently (usually within 90 minutes of loss of vision). Initial management should consist of lateral canthotomy with cantholysis and the administration of steroids and possibly mannitol and acetazolamide to reduce intraocular pressure. Ophthalmologic consultation should be obtained emergently and to follow ocular status.

◆ Surgical Management

After defining the extent of the fractures with axial and coronal CT images, planning involves obtaining adequate exposure for fixation. Although the exact method used may vary among surgeons, once the displaced bony fragments have been adequately reduced, traction of the medial canthi should be applied in the medial, posterior, and superior direction.

Approaches include the following:

- Through the traumatic laceration (if present)
- Open-sky or butterfly approach H-type incision (transverse nasion with bilateral Lynch incisions)
- Bicoronal

A bicoronal approach provides the widest exposure with the best cosmetic result in a subperiosteal plane. When exposing the medial orbital rims, an attempt should be made to locate the medial canthal tendon attachment and not to strip the bony insertions of the medial canthal tendon off the lacrimal crest. Inferior NOE exposure can be achieved by a lower eyelid incision.

The most important surgical step in treating NOE fractures is correcting the traumatic telecanthus. This step involves proper reduction and internal fixation of the medial orbital bone bearing the medial canthal tendon. Depending on the degree of the fracture, optimal stabilization requires one or more of the following:

1. Junctional fixation
2. Interfragmentary fixation
3. Transnasal wiring

Junctional fixation involves attaching the central medial orbital bony fragment (a single piece or reconstructed bone from multiple

pieces) to stable adjacent bone at the frontal bone, inferior orbital rim, and nasal piriform with miniplates.

Interfragmentary fixation can be used to fuse comminuted medial orbital bone pieces together using wires or miniplates. Transnasal wiring is used to reduce and secure laterally displaced central fragments. For splayed NOE fractures, transnasal wiring (28 or 30 gauge) is required to bring the medial canthi together to best correct the traumatic telecanthus. Transnasal wiring is often the most challenging surgical step.

In most cases the medial canthi are still attached to the lacrimal bone. However, if the medial canthus is avulsed off the bone or if the medial canthal-bearing bone fragment is too small to work with for fixation, medial canthal reconstruction is required. One favored technique described by Raveh identifies the medial canthus and secures it with braided wire (32-gauge stainless steel), which is then passed transnasally through a point slightly posterior and superior to the upper end of the lacrimal fossa. The wire is then secured to a screw in the frontal bone at the supraorbital rim. If the medial canthi are detached bilaterally, a separate wire is passed for each medial canthus. If significant bone loss has occurred at the medial orbital rims to support the medial canthus, bone grafting using larger bone fragments or split calvarial bone can be used to contour the defect. In this case the canthal tendon should be passed through a drilled hole in the bone graft and fixed using the transnasal wiring technique with the bone graft secure with miniplates.

Other surgical options are transnasal intercanthal wiring over plates to maintain canthal position and support of the comminuted nasal bones. The sublabial facial degloving may be required for inferior exposure.

Primary repair of traumatic telecanthus is much easier to perform than secondary correction of residual telecanthus because of soft tissue scarring and retraction of the displaced bone fragments over time. After scarring has occurred, repair of residual telecanthus becomes very difficult because of the soft tissue contraction.

In treating the fractured nasal septum, in most instances, a conservative approach is initially taken. Septal hematomas should be drained, and the dislocated septum can be repositioned using closed manipulation. Many surgeons feel that open septoplasty procedures in the acute setting of severe septal injury are not indicated because the mucosal edema and severe comminutions make proper reduction and fixation very difficult. At a later date any residual deformity can be addressed on a delayed basis.

Nasal dorsal reconstruction is often undertreated in managing NOE fracture. Left untreated without adequate nasal dorsal support, retrodisplacement and splaying of the nasal bones, cartilaginous dorsal saddling, decreased nasal length, and an upturned nasal tip can result. If bony support has been lost, but the cartilaginous skeleton, nasal tip, and septum are intact, open reduction and internal fixation of the displaced nasal bone will frequently result in acceptable contour. However, if the cartilaginous skeletal support is lacking, restoring the bony and cartilaginous nasal framework should be considered along with augmentation.

Augmentation materials for the nasal dorsum include using outer calvarial bone grafts, rib cartilage, or iliac crest bone. In the acute setting, outer calvarial bone grafts as a cantilever secured to the frontonasal junction with plate are usually used because the donor site is already in the field.

Isolated medial wall fractures (<50% area involved) do not usually result in enophthalmos, and thus they often do not require reconstruction. Indications for medial orbital wall repair would be (1) obvious enophthalmos or diplopia caused by herniation of orbital contents in the medial wall defect, (2) medial rectus muscle entrapment, and (3) large defects of the medial orbital in association with defects in the orbital floor or roof.

If an obvious laceration or tear of the lacrimal drainage system is evident, then the lacrimal duct can be probed and stented with Silastic tubing, if possible.

◆ Rehabilitation and Follow-up

Later nasal surgery with dorsal grafting may be required to restore nasal contour. Patients must also be followed up for frontal sinus mucocele if the fractures involve the frontal sinus

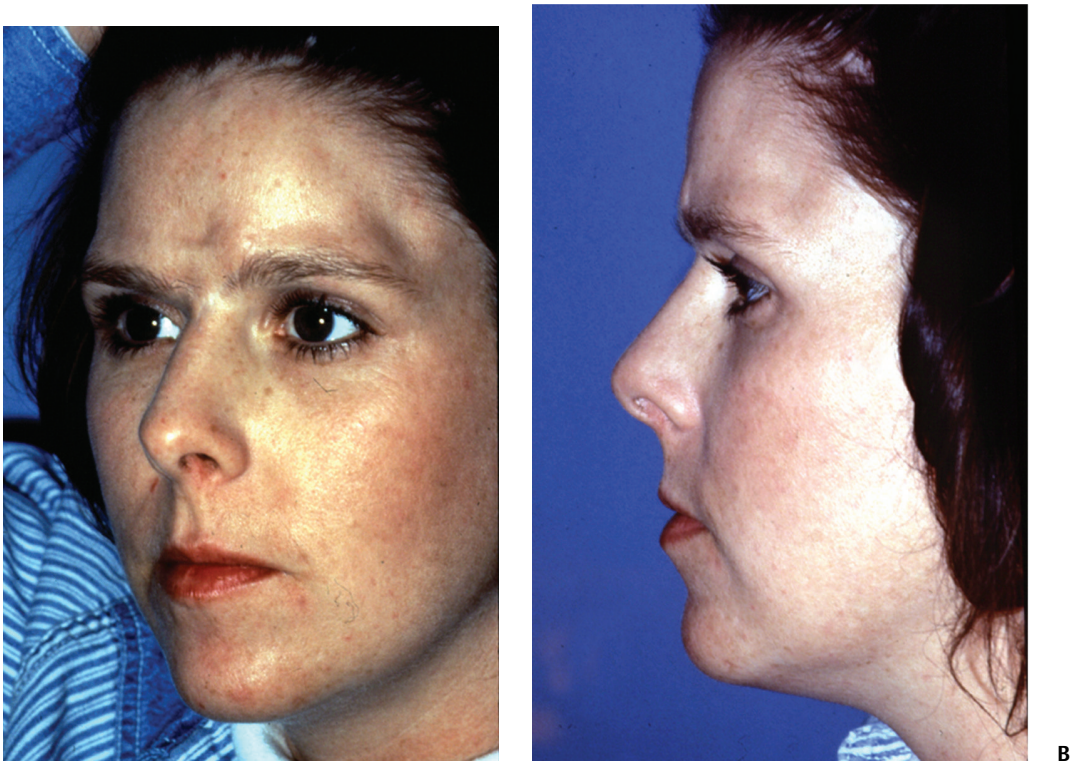


Fig. 78.3 Patient after naso-orbito-ethmoid fracture repair.

or nasofrontal duct. A dacryocystorhinostomy done 3 to 6 months after injury may be indicated for persistent epiphora secondary to lacrimal drainage system obstruction (**Fig. 78.3**).

Questions

1. What physical finding is most characteristic for a naso-orbito-ethmoid complex fracture?
   - **A.** Proptosis
   - **B.** Bilateral periorbital ecchymosis
   - **C.** Epistaxis
   - **D.** Intercanthal distance of more than 35 mm
   - **E.** Enophthalmos
2. An accurate way to determine whether an NOE fracture is present acutely on physical examination is to:
   - **A.** Place a mosquito clamp in the nose and digitally manipulate the medial canthal area
   - **B.** Manually push the nasal bones medially to determine stability
   - **C.** Have the patient in the Valsalva position to check for evidence of proptosis
   - **D.** Measure the intercanthal distance if it is greater than 28 mm
   - **E.** Perform rigid in-nasal endoscopy looking for bone disruption at the lamina papyracea
3. Most CSF leaks with nondisplaced fractures isolated to ethmoid roof will:
   - **A.** Spontaneously resolve after fracture reduction
   - **B.** Require placement of fascia to seal the leak
   - **C.** Require endoscopic evaluation to determine the size of CSF leak
   - **D.** Require antibiotic antibiotics to prevent meningitis
   - **E.** Be likely associated with orbital emphysema

Suggested Readings

Frodel JL Jr. Management of the nasal dorsum in central facial injuries: indications for calvarial bone grafting. Arch Otolaryngol Head Neck Surg 1995;121(3): 307–312

Markowitz BL, Manson PN, Sargent L, et al. Management of the medial canthal tendon in nasoethmoid orbital fractures: the importance of the central fragment in classification and treatment. Plast Reconstr Surg 1991; 87(5):843–853

Raveh J, Laedrach K, Vuillemin T, Zingg M. Management of combined frontonaso-orbital/skull base fractures and telecanthus in 355 cases. Arch Otolaryngol Head Neck Surg 1992;118(6):605–614

79

Frontal Sinus Fracture

David B. Hom

History

An unbelted 34-year-old woman was involved in a motor vehicle accident in which she struck her forehead to the windshield with loss of consciousness. The patient was unconscious with no gross focal neurologic deficits, and her hemodynamic status was stable. No clear rhinorrhea was evident. Neurosurgical consultation and head computed tomography (CT) scan evaluation showed anterior and posterior table commuted fractures with pneumocephalus. No evidence of intracranial parenchymal injuries or of intracranial bleeding was seen. The lateral cervical spine film was clear, and she had no other bodily injuries. The patient was emergently taken to the operating room by neurosurgery to rule out intracranial injuries and for her severe frontal sinus fractures. The otolaryngology service was consulted while the patient was being transported to the operating room to participate in the frontal sinus fracture repair.

Differential Diagnosis—Key Points

1. In a patient with multiple trauma, the overall status must be emergently evaluated and be the top priority (airway, breathing, circulation, cervical status, intracranial status). Only after these steps are stabilized can facial injuries be addressed. In some instances, facial fractures may need to be addressed in a later setting, depending on the medical status. In this instance, the major goal of the neurosurgical team was to evaluate for intracranial injuries and to address her comminuted frontal sinus fractures with the assistance of otolaryngology.
2. It is optimal to obtain a CT of the face (axial and coronal views) in patients with significant head trauma correlating it to a complete facial exam. In other instances, due to the patient's changing clinical status, the otolaryngology service may be contacted while the patient is being expeditiously transported to the operating suite by the trauma surgical services.
3. In a patient who has multiple major body traumas, it is imperative to have a clear communication with all the involved surgical teams to approach the patient in a staged coordinated manner to prevent lengthy procedures by multiple services at one sitting if the patient's status is precarious.
4. For any head trauma, the possibility of cervical spine injury must be ruled out with cervical spine radiographs and clinical examination. However, if the patient is unconscious, the clinical assessment of a cervical spine cannot be performed, and frequently the patient is left in a cervical collar because an occult cervical injury may still

be present. Even in the emergent setting when the clinical situation dictates against complete cervical spine radiographs, the neck must be immobilized and protected from any out-of-axis movements until appropriate cervical injury is ruled out. In some instances low cervical spine injury can be missed despite performance of full cervical radiographic series owing to the difficulty of radiographically imaging this area.

5. In depressed anterior frontal sinus wall fractures, soft tissue edema can mask a contour deformity in the acute phase. Thus, a CT scan is required. High-resolution CT scan (axial and coronal images using 1.5-mm cuts) is very helpful in delineating the extent of bony displacement. Associated findings with frontal sinus fractures are central nervous system (CNS) injuries and cerebrospinal fluid (CSF) leaks. As in this case, injury to the posterior frontal sinus wall significantly increases the risk of dural tears (70% of displaced posterior walls have CSF leaks). Other possible CNS injures includes brain contusion, subdural hematoma, pneumocephalus, and frontal lobe laceration.

Test Interpretation

The CT image (**Fig. 79.1**) shows anterior and posterior frontal sinus fractures, which are comminuted and depressed with pneumocephalus and a right orbital roof fracture. The lateral cervical spine radiographs did not have any evidence of cervical injury.

Diagnosis

1. Multiple facial lacerations
2. Depressed frontal sinus fracture involving the anterior and posterior table
3. Orbital roof fracture
4. Pneumocephalus

Medical Management

Prophylactic antibiotics (broad-spectrum) should be considered preoperatively because this is a clean contaminated wound. Given the nature of the displaced posterior table fracture, a dural tear with subsequent CSF leak is highly likely. This possibility must be kept in mind during the initial management of the patient and surgical exploration of the frontal sinus.

Because the patient is young and healthy with no other bodily injury, no other medical issues besides stabilization and monitoring for the intracranial injury appear to affect the medical management immediately. In the operating room, her eye status was evaluated and cleared by the ophthalmology team because of the orbital roof fracture.

Surgical Management

After the life-threatening factors from multiple injuries have been stabilized, a plan for surgical treatment of the frontal sinus injury can be performed. Many times, a CT scan of the head might not give appropriate image resolution, and thus a finer-cut facial CT scan (1.5-mm axial and coronal views) may be helpful. If the

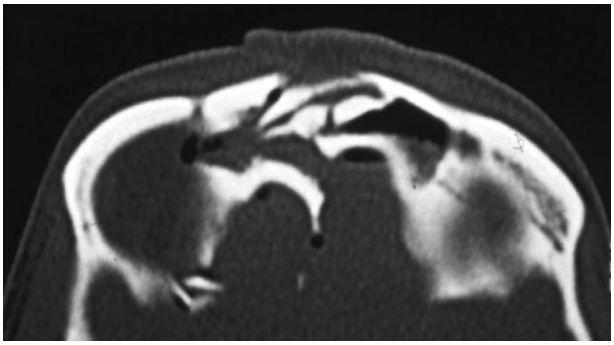


Fig. 79.1 Computed tomography image of anterior and posterior frontal sinus fractures.

patient is in a cervical collar, the coronal CT views will need to be reconstructed to prevent manipulation of the cervical spine.

The surgical treatment of frontal sinus fractures is based on whether the anterior or posterior table fracture is involved. If only the anterior table is fractured, with the nasofrontal duct uninjured, the surgical repair is done primarily to restore the contour of the forehead. In these cases, the depressed bony segment can be raised with bony fixation by several approaches (coronal, prior open laceration, transverse forehead incision, or endoscopically). At times bony fixation might not be needed if the reduced bone segment can maintain its correct anatomic position. If severe comminution is present, titanium mesh can be used to stabilize the bone pieces. If there is significant missing bone, an outer calvarial bone graft can be placed to restore contour of the bony gap. If neurosurgery performs a frontal craniotomy, an inner-table calvarial bone graft can be obtained from the bicortical bone window on the table ex vivo before it is placed back to its original site.

For posterior table fractures, which are not comminuted or significantly displaced, many surgeons remove all the intrasinus mucosa, plug the nasofrontal ducts with fascia, and obliterate the sinus with abdominal fat. This procedure is performed to reduce the risk of mucocele formation from trapped mucosa between the bony fracture edges. Frontal sinus mucoceles may take many years to grow. In the setting of an intact anterior table with a posterior table fracture, an osteoplastic flap can be done for surgical exploration and exposure of the frontal sinus.

Only one patent nasal frontal duct is required for the frontal sinus to function. This patency can be tested by placing water dyed with methylene blue into the frontal sinus; by placing dried pledgets intranasally, one can examine for blue dye on the nasal pledgets. If needed, the midline frontal sinus septa may need to be burred down to ensure full communication of the frontal sinus cavity to the remaining nasofrontal duct. In instances when the nasofrontal duct has been disrupted by a transverse fracture, frontal sinus obliteration should be considered.

For comminuted posterior table fracture, a cranialization procedure with neurosurgery should be considered. This procedure involves removing the posterior table of the frontal sinus with complete removal of all the frontal sinus mucosa and plugging the nasofrontal duct. In this case, a cranialization procedure with neurosurgery was performed and no dural tears were found.

Rehabilitation and Follow-up

This patient may need care at a rehabilitation facility for her closed head injury.

In regard to frontal sinus injury, subsequent mucocele formation can still develop over years. Thus, if a patient should have signs of future chronic sinus discomfort or infection, follow-up CT scans would be indicated. If a persistent air-fluid level in the frontal sinus is evident, it may suggest a CSF leak at the posterior frontal sinus wall or decreased function of the nasofrontal duct.

Questions

1. A 34-year-old man involved in an altercation has multiple comminuted posterior table frontal sinus fractures. What is the best treatment?
- **A.** Cranialization
- **B.** Obliteration with abdominal fat
- **C.** Open reduction and internal fixation of comminuted pieces
- **D.** Placement of titanium mesh
- **E.** Performing a Reidel procedure

2. A 24-year-old woman involved in a motor vehicle accident has an isolated depressed anterior table frontal sinus fracture. What is the best treatment?
- **A.** Cranialization
- **B.** Obliteration with abdominal fat
- **C.** Open reduction and internal fixation of depressed bone
- **D.** Placement of titanium mesh
- **E.** Performing a Reidel procedure

3. A 35-year-old man involved in a motor vehicle accident has transverse fracture through both of his frontal nasal ducts. What is the best treatment?
A. Cranialization
B. Obliteration with abdominal fat
C. Open reduction and internal fixation of depressed bone
D. Placement of titanium mesh
E. Performing a Reidel procedure

Suggested Readings

Davidson JS, Birdsell DC. Cervical spine injury in patients with facial skeletal trauma. J Trauma 1989;29(9): 1276–1278

Donald PJ. Frontal sinus ablation by cranialization: report of 21 cases. Arch Otolaryngol 1982;108(3): 142–146

Ducic Y, Hom DB. Reconstruction of frontal sinus fractures. In: Rengachary S, Benzel E, eds. Calvarial and Dural Reconstruction. Park Ridge, IL: American Association of Neurological Surgeons; 1998:102–116.

Stanley RB Jr. Management of frontal sinus fractures. Facial Plast Surg 1988;5(3):231–235

Wilson BC, Davidson B, Corey JP, Haydon RC III. Comparison of complications following frontal sinus fractures managed with exploration with or without obliteration over 10 years. Laryngoscope 1988;98(5):516–520

80

Auricular Avulsion

David B. Hom

History

A 32-year-old man involved in a motor vehicle accident goes to the emergency department 3 hours after injury for a partial avulsion of the left ear (**Fig. 80.1**). The amputated ear tissue was wrapped in a moist washcloth surrounded by ice (**Fig. 80.2**). The patient is alert and oriented ×4 and has no evidence of other serious injuries; his cervical spine was cleared.

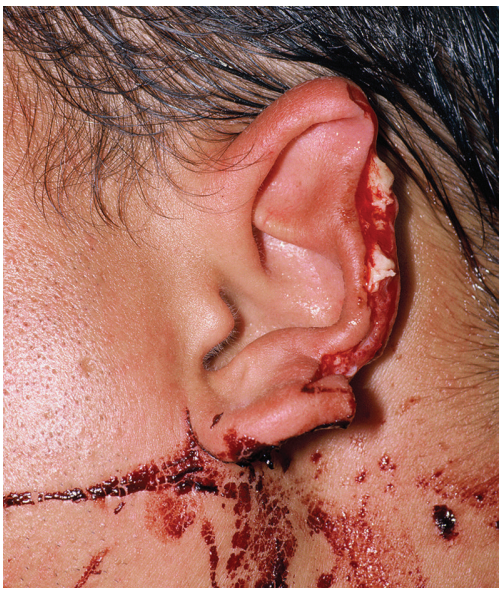


Fig. 80.1 Avulsed left ear 3 hours after injury.

Differential Diagnosis—Key Points

1. With any head injury, a complete systemic trauma evaluation is required. The overall status of the patient must be evaluated and take priority (airway, breathing, circulation, cervical status, intracranial status). Only after these steps are stabilized, can obvious auricular injuries can be addressed.
2. Excessive cautery or clamping and ligation of vessels on the remaining auricle should be avoided to maximize revascularization of the reattached avulsed piece.
3. The auricle should be examined, irrigated copiously, and any foreign bodies removed.
4. The hearing, tympanic membrane, and external auditory canal should be examined. The function of the facial nerve should be checked.

Test Interpretation

Based on the physical examination, the diagnosis is clear. If blood loss is a concern, a hemoglobin test can be done.

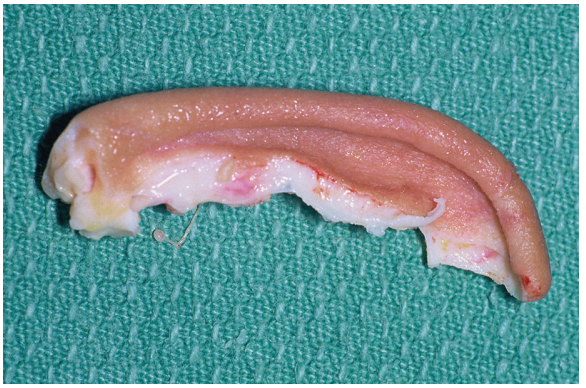


Fig. 80.2 Avulsed skin cartilage segment comprising of the helix and scaphoid fossa.

Diagnosis

Traumatic partial auricular avulsion with most of the helix and scaphoid fossa of the auricle completely amputated

Medical Management

To maximize salvage of the avulsed composite skin cartilage tissue before reaching the operating room, one should wrap it with an isotonic saline-soaked gauze contained in a plastic bag in an ice cooler with surrounding ice to decrease the tissue metabolic demands. Steroids are controversial but may be helpful.

If the torn auricular tissue is still attached to the ear, the pedicle should be preserved because a very small pedicle can help the tissue survive owing to the dense vascular arcade of the auricle.

Surgical Management

Many methods to preserve auricular cartilage have been described. Because the auricle tissue is completely avulsed, the following are possible treatment options:

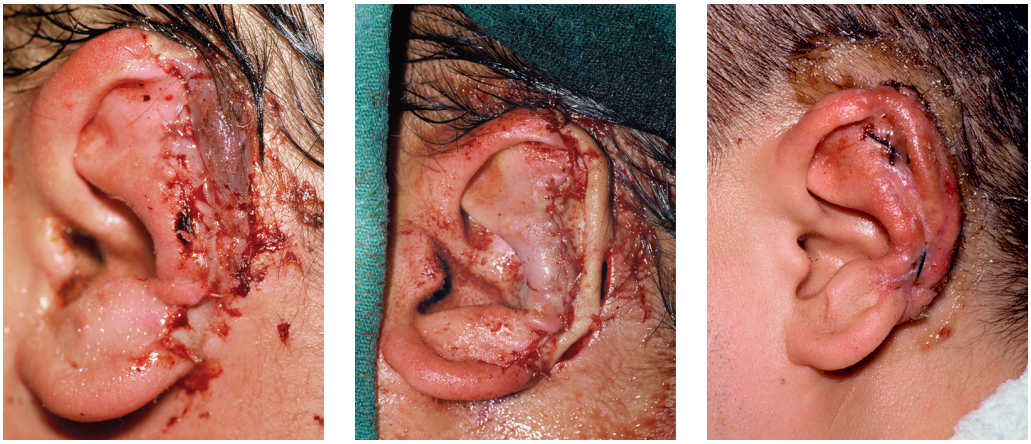
1. Partial burying and fenestrating the cartilage with reattachment (Baudet principle)
2. Delayed staged reconstruction
3. Osseo-integrated alloplastic reconstruction
4. Microvascular anastomosis (not for this case, because the vessels of the avulsed piece are too small to anastomose)

Postauricular skin and local scalp advancement flaps can be used to cover the bare cartilage when it is attached to its original recipient site. Using this technique, the overlying auricular skin is dermabraded to remove the epidermis. The cartilage is then buried by creating a subcutaneous pocket and postauricular skin or scalp is advanced over the auricle. Depending on the amount of postauricular skin available and its elasticity, hair-bearing scalp may be used. After 2 to 3 months, the cartilage is usually revascularized and is freed from the postauricular skin and covered using skin grafts.

Cartilage that did not survive can be replaced using rib or contralateral conchal cartilage. A temporoparietal fascial (TPF) flap may be used to cover the cartilage with a skin graft after the auricular skin is removed from the cartilage, with the cartilage placed in its normal anatomic position. The thin, flexible TPF flap is draped over the cartilaginous frame and covered with a skin graft.

Another useful method to restore a detached auricle (Baudet principle) is removing the posterior auricle skin, making fenestrated windows through the cartilage to increase exposure of the anterior skin surface area to its underlying vascularized bed and reattaching the amputated piece. (**Fig. 80.3**).

Two to three months later, the ear is brought out, and a split-thickness skin graft is placed in back of the auricle. If more lateralization of the auricle is needed, a wedge of cartilage can be placed behind the auricle and an anterior based facial flap can be placed over the wedged cartilage so that the overlying skin graft will take.



A–C

Fig. 80.3 (A–C) Reattachment of avulsed skin cartilage segment using the Baudet principle described in the text.

If microvascular anastomosis is considered for a larger auricle amputation, an adequate artery and vein will need to be identified in the auricle and recipient site. If the auricle survives, this provides a superior result requiring limited revisions. Many surgeons using microvascular anastomosis or free flaps use systemic anticoagulation in the operative and postoperative period with some combination of dextran, heparin, or aspirin used at various doses and combinations. The role of hyperbaric oxygen is controversial. Although hyperbaric oxygen may help survival of amputated body parts during the initial phases of revascularization, it does not take the place of meticulous surgical technique and will not rescue devitalized tissue. Its use should be considered but must be weighed against its potential complications.

If venous congestion becomes apparent, medical-grade leeches *(Hirudo medicinalis)* may be helpful in decongesting the flap. In addition to removing blood, leeches secrete hirudin, which acts as an anticoagulant to the congested tissue. If leeches are used, cotton should be placed in the ear canal to prevent the leeches from entering. In addition, the patient should be monitored for a dropping hematocrit if leeches are used. Leeches may transmit *Aeroinons hydrophileantibiotic*, thus, trimethoprim-sulfamethoxazole or ciprofloxacin should be instituted.

If the ear canal is transected, a pediatric chest tube stent or packing can be placed to minimize stenosis for several months.

Another option for total auricular reconstruction in a later setting is osseo-integrated implants for mounting a prosthetic ear. A prosthetic ear is fashioned around clips that attach to the abutments of the implants.

◆ Rehabilitation and Follow-up

Options for reattaching an auricle may require multiple stages and prolonged follow-up. With surgical reconstruction, it is difficult to match the untraumatized contralateral ear, and the patient must be made aware of this difficulty. This is why the patient's input at the time of the initial trauma is important to help guide the surgeon to the best outcome for that specific individual.

◆ Questions

1. A 24-year-old man sustained a traumatic partial auricular avulsion injury from a knife wound 2 hours ago involving the upper two thirds of the anti-helix and scaphoid fossa. What is the best method for treatment?

A. Microvascular anastomosis

B. Osteo-integrated alloplastic reconstruction

C. Delayed auricular reconstruction discarding the avulsed piece
D. Burying and fenestrating the avulsed cartilage
E. Placing a rib graft to the auricle

2. A 28-year-old man who is 4 hours away sustains an avulsion injury involving the upper third of his left auricle. To maximize salvage from this avulsed piece, what should the patient be instructed to do with the piece during transport?
A. Wrap it in isotonic saline–soaked gauze, and place it in a cooler surrounded with ice.
B. Wrap it in hypertonic saline–soaked gauze and place it in a cooler surrounded with ice.
C. Wrap it in isotonic saline–soaked gauze and place it in a thermos with warm water.
D. Immerse it in dry ice.
E. Wrap it in a dry gauze covered with antibiotic ointment.

3. A 30-year-old man underwent a first-stage attachment of his avulsed ear cartilage involving the upper third of his ear. The second stage of reconstruction is planned with a large area of bare cartilage present. What is the best cosmetic method for reconstruction?
A. To use a temporoparietal fascial flap with a skin graft
B. To cover with radial skin free flap
C. To cover with cadaveric freeze-dried dermis
D. To cover with a superior scalp flap
E. To cover with inferior-based cervical flap

Suggested Readings

Brent BD. Reconstruction of the auricle. In: Mathes SJ, ed. Plastic Surgery. 2nd ed. Philadelphia, PA: Saunders Elsevier; 2006:633–698

de Chalain T, Jones G. Replantation of the avulsed pinna: 100 percent survival with a single arterial anastomosis and substitution of leeches for a venous anastomosis. Plast Reconstr Surg 1995;95(7):1275–1279

Destro MW, Speranzini MB. Total reconstruction of the auricle after traumatic amputation. Plast Reconstr Surg 1994;94(6):859–864

Frodel JL Jr, Barth P, Wagner J. Salvage of partial facial soft tissue avulsions with medicinal leeches. Otolaryngol Head Neck Surg 2004;131(6):934–939

Wilkes GH, Wolfaardt JF. Osseointegrated alloplastic versus autogenous ear reconstruction: criteria for treatment selection. Plast Reconstr Surg 1994;93(5):967–979

81

Penetrating Neck Trauma

David B. Hom

History

A 16-year-old female passenger was broadsided and ejected from her car. A sharp metallic fence rod impaled her left upper cervical region (zone II). She was seen emergently at an outside hospital and was found to be hemodynamically stable (blood pressure 170/84, pulse rate 107) with spontaneous respirations, 14 per minute. She is unconscious. Her cervical spine is clear. No active bleeding was present from the 4-cm left neck wound.

On physical examination, she was in no respiratory distress and had no stridor. Only a right carotid pulse could be palpated. Two 14-gauge intravenous (IV) lines were started. No other injuries were present. The wound was not probed.

At the outside facility, an emergent tracheotomy was done and the carotid artery appeared not to be exposed in this wound (**Fig. 81.1**).

Differential Diagnosis—Key Points

Any patient who has sustained trauma initially should be evaluated according to the Advanced Life Support System protocol. The airway is managed first and may require tracheotomy

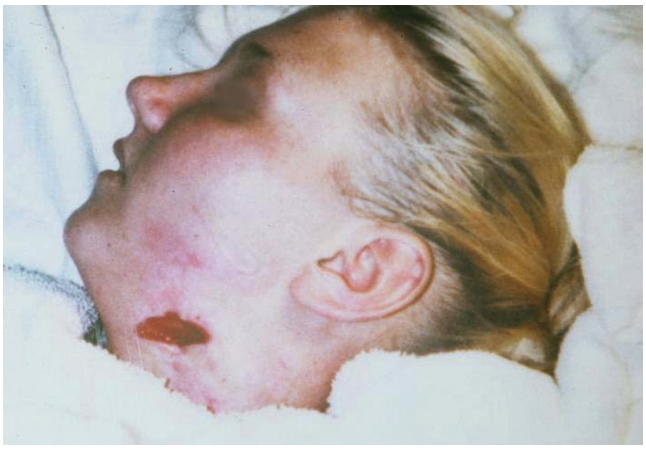


Fig. 81.1 Penetrating neck wound entering zone II.

with the patient under local anesthesia to establish a stable airway for laryngotracheal injuries. Large-bore IVs are then started and the patient's fluid resuscitated as needed.

The neck is divided into three zones for trauma evaluation. *Zone* I is from the clavicle to the cricoid cartilage; *zone* II is from the cricoid to the angle of mandible; and *zone* III is from the mandible angle to the skull base.

If any immediate life-threatening signs or symptoms from penetrating neck trauma (i.e., massive bleeding, expanding hematoma, nonexpanding hematoma in the presence of hemodynamic instability, hemomediastinum, hemothorax, systemic shock not responding to fluid resuscitation, increasing subcutaneous emphysema, major hemoptysis) are evident, emergent neck exploration is warranted.

In the stable patient, the treatment choices remain controversial. Important parameters to consider are the zone that is involved (I, II, or III), mechanism of injury, and velocity of the projectile. These parameters are helpful to determine whether injury to vital structures has occurred.

Currently many trauma centers manage stable patients with non–life-threatening penetrating neck trauma with selective exploration if the following is available: on-site angiography, flexible and rigid endoscopy, and close physical examining monitoring.

High-velocity projectiles (>610 m/s) cause a significantly greater amount of damage than low-velocity projectiles. Zone I and zone III injuries usually require arteriography on all stable patents. Stable patients with zone II injuries may require angiography. Because hemorrhage is the leading cause of death for penetrating neck injuries, hemodynamic and neurologic status should always be monitored closely for at least 48 to 72 hours.

For blunt neck trauma, the laryngeal, cervical, vascular, and digestive structures can be injured. Symptoms can manifest in a delayed fashion and are easily underdiagnosed. Thus, in patients who have had significant blunt neck trauma, careful observation should be undertaken to monitor for progressive signs and symptoms of laryngeal, digestive, cervical, and vascular injury.

In Zone III injuries, exposure can be exceedingly difficult because the carotid follows its course medial to the mandible toward the skull base.

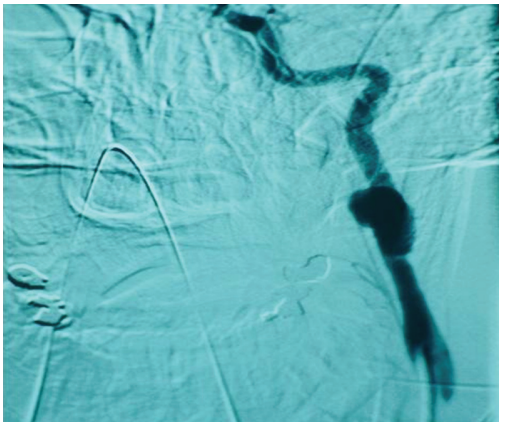


Fig. 81.2 Angiogram showing a 3-cm traumatic aneurysm of the internal carotid several centimeters from the wound site at the level of C1. It was not actively bleeding on angiogram.

◆ Test Interpretation

- Hemoglobin 10 g/dL / hematocrit 31%. Chemistry panel, prothrombin time, partial thromboplastin time were within normal limits
- Chest radiography—no pneumothorax

Angiogram had been performed before surgery and revealed a 3-cm traumatic aneurysm of the internal carotid several centimeters from the wound site at the level of C1. It was not actively bleeding on the angiogram. Computed tomography (CT) scan did not show any larynx or tracheal fracture or foreign bodies. Barium swallow was normal (**Fig. 81.2**).

◆ Diagnosis

1. Zone II penetrating neck injury
2. Traumatic aneurysm of left internal carotid at level of C1

◆ Medical Management

Initial management of patients with penetrating neck injuries should follow the basic tenents of trauma care. This emergent management requires (1) establishing an airway, (2) maintaining blood perfusion, and (3) defining the severity of the wound. In the emergency

department, the airway can be established by intubation, cricothyroidotomy, or tracheotomy. Direct transcervical tracheal intubation is safer than oral or nasal intubation when the oral cavity, pharynx, or larynx are traumatized and filled with blood. In the setting of a gunshot wound, it may be difficult to evaluate the cervical spine fully until the airway is controlled. Multiple blind intubation attempts will risk enlarging a lacerated piriform sinus wound and extending it iatrogenically into the mediastinum. Similarly extending the neck in an injured trachea can distract apart the proximal and distal segments, thus enlarging a tracheal tear. The airway must be established and the hemodynamic status stabilized before transporting the patient to the angiography suite.

Large-bore IV lines are placed, even when the patient is not hypotensive, so that fluids can be rapidly introduced if needed. The IV lines should be placed on the opposite side of the injury because of the possibility of venous lacerations proximal to the IV. The patient can be placed in the Trendelenburg position to minimize the potential of air embolism.

Under no circumstances should a penetrating neck wound be probed because clot dislodgement and uncontrollable bleeding can occur.

Every patient with significant neck trauma should have routine anterior and lateral neck and chest radiographs. If a significant pneumothorax is identified by a radiograph or physical examination, a chest tube should be inserted emergently.

In the rare circumstance of an exsanguinating oral hemorrhage, a tracheotomy must be performed immediately and the pharynx must be packed to tamponade the major vessel hemorrhage.

Once the airway is secure, angiography is the most urgently performed diagnostic study because hemorrhage can be life threatening. A positive angiogram may mandate an immediate trip to the operating room, but evaluation of the upper digestive tract in the radiology suite may be useful if time and the patient's condition permit. Zone I and zone III injuries usually require routine preoperative arteriography on stable patients because their surgical approach is more difficult than zone II injuries. In addition, when wounds involve both sides of the neck with zone I and zone III injuries, four-vessel angiography (bilateral carotid and vertebral arteries) should be considered in stable but symptomatic patients.

◆ Surgical Management

In this case, an angiogram had been performed before surgery revealing a 3-cm aneurysm of the internal carotid several centimeters from the wound site at the level of C1. It was not actively bleeding. Follow-up CT scan did not show any laryngofracture or abscess. She was then transferred to our facility and followed by neurosurgery.

A direct laryngoscopy revealed no evidence of mucosa injury, foreign bodies, or hematoma. The wound was 4 cm wide below the posterior digastric muscles but not entering the pharynx. The carotid artery was grossly 3 to 4 cm away from this neck wound, and the wound was left open to heal by secondary intention. A repeat angiogram was performed, and the patient was followed up serially over a 2-day course to determine whether the traumatic aneurysm was enlarging.

The choices are exploring the wound surgically and having neurosurgery repair the carotid artery versus embolizing the internal carotid to treat the traumatic carotid aneurysm. The decision was made to embolize the internal carotid with balloons and monitor the patient closely. The patient tolerated the procedures well with no neurologic sequelae.

If the patient is taken to the operating room for neck exploration, a wide-apron incision can be made from the mastoid tip to the midline of the neck at the cricoid level for definitive exploration.

Possible injuries from neck trauma are described below.

Vascular Injuries

Most vascular injuries manifest within 48 hours. Zone I vascular perforation requires thoracic surgery for which a mediastinotomy extension or a formal lateral thoracotomy may be needed. Zone III injuries at the skull base may be temporarily controlled with pressure; however, access to the injury may require a mandibulotomy for accessibility.

All veins in the neck can be safely ligated to control hemorrhage; if both internal jugular

veins are interrupted by the injury, an attempt to repair can be done. External carotid artery injuries are easily managed by suture ligation because collateral circulation is good. Common carotid or internal carotid injury in zone II is explored once the diagnosis is made with attempts for vascular repair. End-to-end anastomosis or autogenous grafting is recommended when stenosis is evident by arteriography. Ligation of the common or internal carotid injuries is generally reserved for irreparable injuries and in patients who are in a profound coma state with bilateral fixed and dilated pupils. Delayed complications from unrepaired vascular injuries include aneurysm formation, dissecting aneurysm, and arteriovenous fistulas.

Interventional radiologists have used angiographic techniques to treat vascular injury. In many instances, embolization procedures can help control arterial disruption. For arterial injuries in zone III, transcatheter arterial embolization can be an effective modality to obtain hemostasis. Penetrating injuries in zone III can have multiple vascular injuries involving the internal carotid artery, internal maxillary artery, and external carotid artery. In areas of difficult vascular access at the skull base, detachable balloons or steel coils can be placed for carotid occlusion.

Control of hemorrhage from vertebral artery penetration can be very difficult owing to its location in the bony canal within the cervical spine. Because vertebral artery injuries often result in both neurologic and hemodynamic sequelae, they are frequently associated with higher morbidity and mortality rates. Surgical vascular repair in zone III can be complex, so endovascular treatment approaches are very helpful.

Laryngotracheal Injuries

Laryngeal mucosal lacerations from penetrating injury ideally should be repaired early (within 24 hours) because the time elapsed before repair can affect airway, scarring, and voice quality.

Significant glottic and supraglottic lacerations and displaced cartilage fractures need surgical approximation. Endoscopy and CT will differentiate between the patients that need only observation (small laceration, shallow laceration, nondisplaced fracture) and those that require a thyrotomy or open fracture reduction and mucosal approximation. A soft laryngeal stent may be needed for badly macerated mucosa.

In severe cases, a low tracheotomy with the patient under local anesthesia is advocated. Tracheal injuries may be repaired primarily using 3-0 monofilament sutures that go through the cartilage rings but do not pass into the tracheal lumen. For minor, incomplete lacerations, tracheotomy might not be required. Suprahyoid-releasing incisions may be required for additional length.

Simple tracheal lacerations that do not detach a tracheal ring or encroach on the airway can be repaired without a tracheotomy. More severe disruptions (gunshot wound directly to the trachea) imply more soft tissue injury, and a 6-week tracheotomy either below or through the tracheal injury is the safest procedure. Later the stenosis may require sleeve resection, but if the stenosis is soft, a T-tube tracheotomy tube can often manage it.

Digestive Tract Injuries

To rule out possible esophageal perforation, most radiologists recommend a Gastrografin swallow initially since barium extravasation can radiographically distort soft-tissue planes and is more toxic. A barium swallow should follow a negative Gastrografin study if suspicion remains high.

Missed esophageal tears represent most of the delayed injuries and can progress to mediastinitis and increased mortality. Some centers claim that flexible esophagoscopy can be used to circumvent the need for general anesthesia during rigid endoscopy; however, flexible endoscopy can miss perforations near the cricopharyngeus and hypopharynx.

Some trauma centers recommend neck exploration for patients who have air in the soft tissues of the neck despite yielding normal endoscopy results. To rule out pharyngeal and esophageal injuries during the surgical exploration, a nasogastric tube can be gently pulled up to the level of the neck and methylene blue infused through the nasogastric tube to help localize the injury site.

By using the combination of flexible endoscopy and rigid esophagoscopy to examine the entire cervical and upper thoracic esophagus, reportedly no perforations have been missed.

If a pharyngeal perforation is still strongly suspected but unconfirmed, the patient is given no food and is observed for several days. Fever, tachycardia, or widening of the mediastinum on serial chest radiographs requires a repeat endoscopy or a neck exploration should be considered.

When an esophageal injury is found early, management involves a two-layer closure with wound irrigation, debridement, and adequate drainage. After repair of the mucosa perforation, a muscle flap may be interposed over the esophageal suture line for further protection. If an extensive esophageal injury is present, it may necessitate a lateral cervical esophagostomy and later definitive repair.

Many surgeons perform direct laryngoscopy, bronchoscopy, and rigid esophagoscopy with the patient under anesthesia for penetrating injuries of the neck when air is seen in the soft tissues or if hemoptysis, hematemesis, or other suspicious clinical findings are present. Direct laryngoscopy and rigid bronchoscopy can be combined with flexible airway examination to recognize and stent a lacerated trachea temporarily. In the setting of a cervical spine fracture, rigid esophagoscopy might need to be omitted.

Even if the clinical examination is benign, follow-up examination at least three times every 24-hour shift for several days is needed to monitor vital signs, and the neck examination change is needed. Odynophagia, subcutaneous emphysema, hematemesis (or blood from the nasogastric aspirate), and fever are the most common findings for esophageal injury. A patient with negative physical examinations, normal radiographs, and normal endoscopies will most likely have a negative neck exploration, and no significant injury will be discovered.

◆ Rehabilitation and Follow-up

This patient gradually became more alert with no neurologic deficits. Over a 6-month course, neurologically she fully recovered and was back in high school (**Fig. 81.3**).

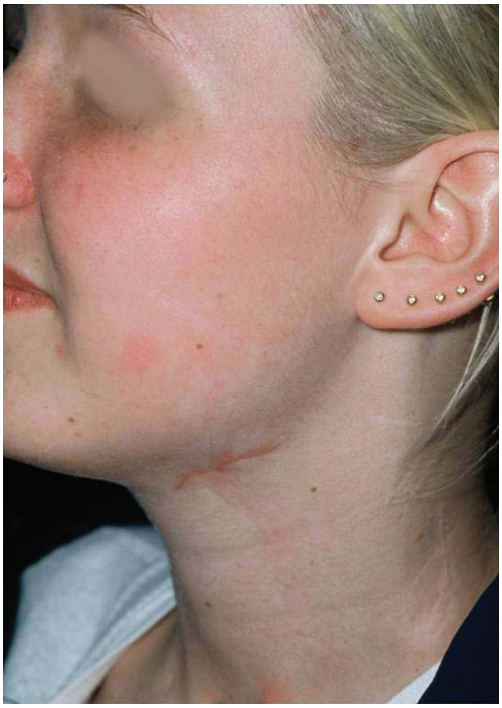


Fig. 81.3 Patient postoperatively.

1. Zone II is the most common. Zone I injuries are the most life threatening, with up to 66% mortality if the common carotid artery is injured.
2. Undiagnosed esophageal injuries can result in severe sequelae. Some reports have stated 20% mortality with delayed esophageal perforation diagnosis.
3. If severe esophageal injury is present, creating controlled fistula may help to control the infection and salivary flow.
4. For carotid injuries that require ligation of the common or internal carotid system, there is a 30% stroke rate.
5. Follow-up appointments should be in 1 week to remove sutures and assess any changes and then again at 1, 3, and 6 months to evaluate for any neurologic changes suggestive of a progressive vascular injury and thromboembolic events.

Questions

1. A 22-year-old man sustained a gunshot wound from a handgun resulting in an entrance wound posterior to the left mandibular angle. He has no active bleeding and no neurologic changes. He is alert and oriented, and his airway is stable. His cranial nerves are fully intact. What would be the next step?
   - **A.** Request a neurosurgery consult
   - **B.** Observe for 24 hours
   - **C.** Order an angiogram
   - **D.** Order a barium swallow
   - **E.** Perform an elective tracheotomy
2. What penetrating neck injury region is most difficult for surgical excess?
   - **A.** Zone I
   - **B.** Zone II
   - **C.** Zone IV
   - **D.** Level A
   - **E.** Type 5
3. A 27-year-old man sustained a stab wound 4 cm below his left mandibular angle. What region of his neck is involved?
   - **A.** Zone I
   - **B.** Zone II
   - **C.** Zone III
   - **D.** Zone IV
   - **E.** Zone V
4. A 32-year-old woman sustained a bow and arrow injury entering her left midneck in the posterior triangle region. Her vitals are stable with no active bleeding. She has no signs of neurologic injuries or hemodynamic compromise. She does complain of significant dysphagia with no airway difficulties. What is the step to rule out an esophageal perforation?
   - **A.** A computed tomography scan with intravenous contrast
   - **B.** A barium swallow
   - **C.** A direct laryngoscopy
   - **D.** A flexible laryngoscopy
   - **E.** A gastrografin swallow

Suggested Readings

Hom DB, Maisel R. Penetrating and blunt neck injuries. In: Flint P, Haughey B, Richardson M, et al, eds. Otolaryngology—Head and Neck Surgery. 5th ed. Philadelphia, PA: Mosby; 2008

Kountakis S, Rafie J, Ghorayeb B, Stiernberg C. Gunshot wounds to the head and neck. Arch Otolaryngol 1992;118:592–597

Sclafani AP, Sclafani SJ. Angiography and transcatheter arterial embolization of vascular injuries of the face and neck. Laryngoscope 1996;106(2 Pt 1):168–173

Stassen NA, Hoth JJ, Scott MJ, et al. Laryngotracheal injuries: does injury mechanism matter? Am Surg 2004;70(6):522–525

82

Orbital Blowout Fracture

Paul A. Brannan and David B. Hom

History

The patient is a 45-year-old man involved in a motor vehicle collision who presents with double vision and pain with eye movements. He reported that he was struck in the left periorbital area. He denied loss of consciousness. Subjectively, he had no difficulty seeing out of the eye but noted that his cheek was numb on the left side. He was otherwise healthy, with no medical or ocular history.

Physical examination showed a healthy-appearing man with periorbital ecchymosis. Pupil examination and vision were normal in both eyes. Significant diplopia was present in upgaze, and he complained of mild pain with eye movements. On the left, motility was limited in upgaze only and enophthalmos was present. The left eye also appeared lower compared with the right (**Figs. 82.1** and **82.2**).

Differential Diagnosis—Key Points

1. When evaluating patients with orbital and facial trauma, the more serious injuries need to be ruled out first, such as airway,

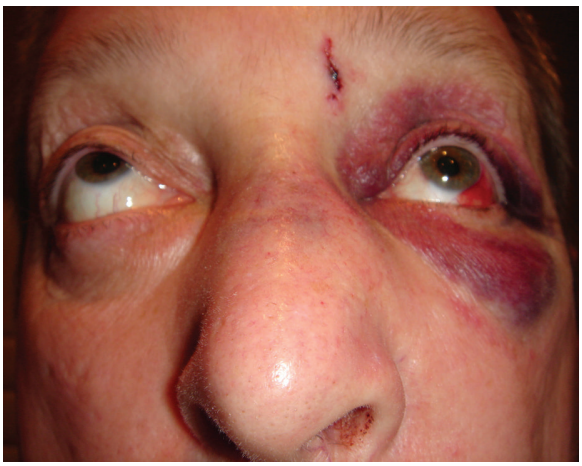


Fig. 82.1 A 45-year-old man with restriction of superior gaze following left periorbital trauma.

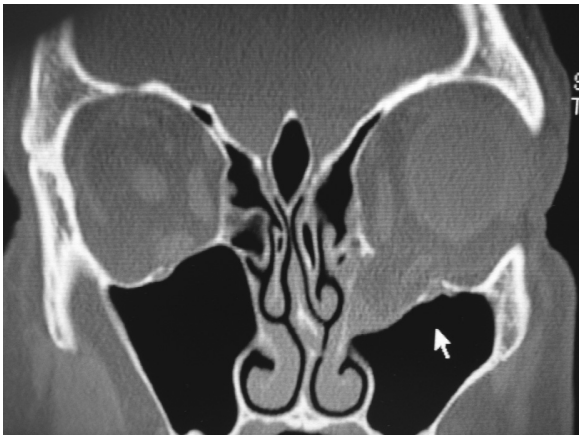


Fig. 82.2 Coronal computed tomography scan showing left orbital floor fracture and prolapse of orbital soft tissue.

breathing, circulation intracranial injury, and vision-threatening problems. The patient had no loss of consciousness with the injury and had a normal neurologic examination. The other major concern would be ruling out an injury to the globe.

2. The entire injury appears to be localized to the periorbital area, and the extent of the injury cannot be fully determined on external appearance alone. The motility examination indicates that extraocular muscles may be impinged or paretic, possibly from an orbital fracture. Also, the globe is sunken down and back relative to the right eye, which is consistent with a significant floor fracture. It is important to consider the surrounding facial structures for injury when an orbital fracture is present, namely, a zygomatic complex fracture or other facial fractures. The orbital rims should be palpated for tenderness and step-offs, as should the malar eminence. Any malpositions or depressions in comparison to the normal side by the supra vertex and submental views should be noted.
3. Cheek and lip numbness can also indicate that an orbital floor fracture is present given that the infraorbital nerve runs along the floor. It is often involved and may be the only sign of a fracture on the examination. These should be evaluated and documented in trauma patients.
4. A significant eye injury occurs in one third of patients with orbital fractures, and a dilated fundoscopic examination by an ophthalmologist is recommended.

◆ Test Interpretation

A vital diagnostic test is a thorough ophthalmic evaluation to rule out globe injury. Corneal abrasions, hyphema, lens dislocation, retinal detachment, and rupture of the globe are among the injuries that should be ruled out with dilated examination. Delay in diagnosing severe eye problems may result in loss of vision; thus ophthalmologic consultation in a timely fashion is recommended.

High-resolution computed tomography (CT) imaging (axial and coronal views) provides the best information for evaluating orbital bones in relation to the soft tissues and orbital fat. When fractures are identified, herniation of orbital contents into the surrounding sinuses can lead to extraocular muscle entrapment within or below the bony fragments. In this case there was an isolated orbital floor blowout fracture and the surrounding bones were intact, but the inferior rectus muscle was adjacent to the edge of the fracture.

An important part of the examination in patients with orbital fractures is forced duction testing. If it is unclear whether the muscle is entrapped, the insertion of the muscle can be grasped with a fine forceps with the aid of a topical anesthetic, and the globe is rotated. Severe restriction indicates that the muscle is entrapped and needs to be released surgically. This patient had limited upgaze but normal forced duction testing, and thus the limited motility was likely as a result of edema and hemorrhage.

Hertel exophthalmometry is another important examination that can be attempted, but it

may be difficult in the traumatic setting; this measures the outward protrusion of both eyeballs relative to each other and determines whether relative enophthalmos or exophthalmos is present. The instrument is placed on the lateral orbital rims, so if they are not intact from a large tripod fracture or there is too much discomfort, the test is not useful. An easy alternative is to have the patient tilt his chin up and the globes can be viewed from below, giving a gross assessment of the eye positions. This patient had 3 mm of enophthalmos at presentation.

The findings of periorbital ecchymosis, edema, restricted eye motility, cheek numbness, and radiographic evidence of an orbital floor fracture are all diagnostic of a blowout fracture. The compressive forces of the blow to the inferior orbital rim are transmitted posteriorly and the weakest portion of the orbit, the floor, or medial orbital wall is fractured. There was no muscle entrapment in this patient, and the limited eye motility was due to edema and hemorrhage within the orbital tissues.

A distinct form of blowout fracture is the medial orbital wall blowout. Medial rectus incarceration can rarely occur in these cases, and the diagnosis is made similar to diagnosis of floor fractures. When there is entrapment, horizontal motility is limited in both directions to some degree, depending on where the muscle is incarcerated.

◆ Diagnosis

Left orbital blowout fracture without entrapment

◆ Medical Management

Most orbital floor and medial wall fractures do not require surgical intervention, and if surgery is needed, it is rarely on an urgent basis. One exception to this is the "trapdoor fracture" that is seen in the pediatric population. The bones of children are thick and elastic and tend to snap back into position after being fractured. Thus the orbital contents are compressed into the maxillary sinus when a fracture occurs and the bone transiently bends downward but can return to its normal position, leaving orbital contents trapped below the bone. When the inferior rectus is incarcerated in such a fracture, this is an emergent situation. Delay in releasing the muscle from the fracture will lead to ischemia and fibrosis of the muscle, resulting in permanent problems with motility. Early repair of these fractures is indicated to provide the best chance of preserving the function of the inferior rectus.

Other cases of blowout fractures (with the exceptions of large fractures, muscle entrapment, or enophthalmos >3 mm) can be observed for resolution of the edema and motility problems. Ophthalmic consultation is required to rule out injury to the globe because 33% of orbital fractures are associated with a significant eye injury. The patient should be instructed not to blow his nose to prevent orbital emphysema and should not take medications containing blood thinners in case surgery will be required. If double vision and pain are present, a 1-week course of oral prednisolone (1 mg/kg/day) can be considered to hasten resolution of the edema and abnormal eye movement. Patients should be monitored closely to ensure that the motility is improving and that they are not developing enophthalmos.

Patients with smaller fractures, no enophthalmos, and normal forced duction testing can usually be observed without surgery. However, late enophthalmos from fat atrophy and fibrosis can develop over the first month as the edema from the trauma resolves and may require surgery. Most authorities believe a delayed repair can achieve acceptable results with regard to eye position.

◆ Surgical Management

Surgery is indicated if enophthalmos or muscle entrapment is present. The orbital floor is explored (either through a transconjunctival or an external approach) and the orbital soft tissues are carefully pulled out of the fracture. Forced duction testing should be done before and after surgery to ensure that release of the tissues has been accomplished. Once the entire fracture is exposed, the orbital floor needs to be resurfaced. Historically, autogenous bone grafts were used, but this adds significant surgical and recovery time. Alloplastic materials are commonly used and readily available, virtually eliminating bone

grafts. These include nylon, porous polyethylene, and titanium sheets, which can be cut to the proper size. Care should be taken not to entrap orbital tissues between the posterior part of the implant and the bone, and forced ductions should be rechecked after the implant is placed.

Following surgery, the patient is usually admitted for 23 hours of observation with frequent vision checks, head elevation, and iced dressings. Orbital hemorrhage is the most worrisome complication that may occur in this period. It manifests by pain, proptosis, and inability to move the eye. The hematoma develops in a closed space (the orbit) and can compress the optic nerve and blood supply to the eyeball. This is a surgical emergency that may require opening of the wounds, canthotomy and cantholysis of the lateral canthal tendon, drainage of any hematomas, and control of any active bleeding.

Long-term complications include undercorrection of the enophthalmos. If more surgery needs to be done, one should wait 6 months for adequate resolution of the edema and healing before considering augmenting the floor with additional implants or repositioning of the implant. Persistent muscle entrapment may also occur and can be confirmed with forced duction testing and CT. If entrapment is present, exploration of the floor with release is indicated. If the muscle is only paretic or scarred from the trauma, one should wait at least 6 months before considering muscle surgery to allow for spontaneous improvement.

Rehabilitation and Follow-up

Patients should be monitored closely for the first 2 weeks following surgery. Eye motility and eye position should be measured until most of the edema has resolved and the patient has an adequate result.

Questions

1. The following are signs of an orbital blowout fracture except which one?
- **A.** Double vision and pain with eye movement
- **B.** Cheek and lip numbness
- **C.** Enophthalmos or gross globe displacement
- **D.** Blurry vision
- **E.** Radiographic evidence of a fracture

2. Which is not a surgical indication for orbital floor fracture repair?
- **A.** Entrapment of an extraocular muscle
- **B.** Fracture of greater than 50% of the floor
- **C.** Enophthalmos of 4 mm
- **D.** Double vision from orbital hemorrhage adjacent to the fracture
- **E.** Gross inferior placement of the globe into the maxillary sinus

3. What is the best way to confirm that extraocular muscle entrapment is present?
- **A.** A dilated examination of the eye
- **B.** Computed tomographic imaging alone
- **C.** Magnetic resonance imaging alone
- **D.** Restriction on forced ductions combined with imaging
- **E.** Enophthalmos and blurry vision

4. What is the goal of orbital floor fracture repair?
- **A.** Release of entrapped contents, improve globe position, and resurfacing the floor
- **B.** Resolve double vision
- **C.** Release the infraorbital nerve to improve cheek and lip sensation
- **D.** Improve visual acuity
- **E.** Precise repositioning of the bones without using implants

5. When is an orbital blowout fracture considered a surgical emergency in a child?
- **A.** The floor is partially shattered into fragments.
- **B.** The inferior rectus muscle is entrapped behind bone.
- **C.** More than 3 mm of enophthalmos is present.
- **D.** Double vision is present.
- **E.** The medial or lateral walls are fractured as well.

Suggested Readings

Brannan PA, Kersten RC, Kulwin DR. Isolated medial orbital wall fractures with medial rectus muscle incarceration. Ophthal Plast Reconstr Surg 2006;22(3):178–183

Cole P, Boyd V, Banerji S, Hollier LH Jr. Comprehensive management of orbital fractures. Plast Reconstr Surg 2007;120(7, Suppl 2):57S–63S

Garibaldi DC, Iliff NT, Grant MP, Merbs SL. Use of porous polyethylene with embedded titanium in orbital reconstruction: a review of 106 patients. Ophthal Plast Reconstr Surg 2007;23(6):439–444

Lelli GJ Jr, Milite J, Maher E. Orbital floor fractures: evaluation, indications, approach, and pearls from an ophthalmologist's perspective. Facial Plast Surg 2007;23(3): 190–199

Moore CC, Bromwich M, Roth K, Matic DB. Endoscopic anatomy of the orbital floor and maxillary sinus. J Craniofac Surg 2008;19(1):271–276

Nam SB, Bae YC, Moon JS, Kang YS. Analysis of the postoperative outcome in 405 cases of orbital fracture using 2 synthetic orbital implants. Ann Plast Surg 2006;56(3): 263–267

Ozturk S, Sengezer M, Isik S, Turegun M, Deveci M, Cil Y. Long-term outcomes of ultra-thin porous polyethylene implants used for reconstruction of orbital floor defects. J Craniofac Surg 2005;16(6):973–977

Parbhu KC, Galler KE, Li C, Mawn LA. Underestimation of soft tissue entrapment by computed tomography in orbital floor fractures in the pediatric population. Ophthalmology 2008;115(9):1620–1625

Rinna C, Ungari C, Saltarel A, Cassoni A, Reale G. Orbital floor restoration. J Craniofac Surg 2005;16(6):968–972

Tuncer S, Yavuzer R, Kandal S, et al. Reconstruction of traumatic orbital floor fractures with resorbable mesh plate. J Craniofac Surg 2007;18(3):598–605

XVI

Facial Plastic and Reconstructive Surgery

83

Prominauris and Otoplasty

Gresham T. Richter and David B. Hom

History

A 5-year-old boy has a strong family history of large, protruding ears. Like his family members, he was noted to have prominent ears shortly after birth. His parents had been informed that his ears would ultimately normalize with time. However, increasing projection of his ears has become a center of attention. His parents are now anxious about further teasing, as suffered by other family members at school and the impact on his self-esteem as he grows. Otherwise, he has been a healthy and happy child. His physical examination is unremarkable except for obvious underdevelopment of the antihelical folds of his ears, which contributes the lateral projection of each helix (**Fig. 83.1**). The auriculocephalic angle measures about 45 degrees bilaterally. Prominent deep conchal bowls are also present.

Differential Diagnosis—Key Points

The external ear growth continues into early childhood as the auricle reaches 85% of adult size by 3 years of age. Near-complete development is achieved by 5 or 6 years of age and is considered the appropriate age for repair, especially because this age marks the onset of self-recognition and school matriculation.

Specific anatomic landmarks and criteria have been established to help define the normal-appearing external ear (**Fig. 83.2**). Normal measurements are based on accepted averages for the size and shape of the ear as well as the location and projection of the ear on the head

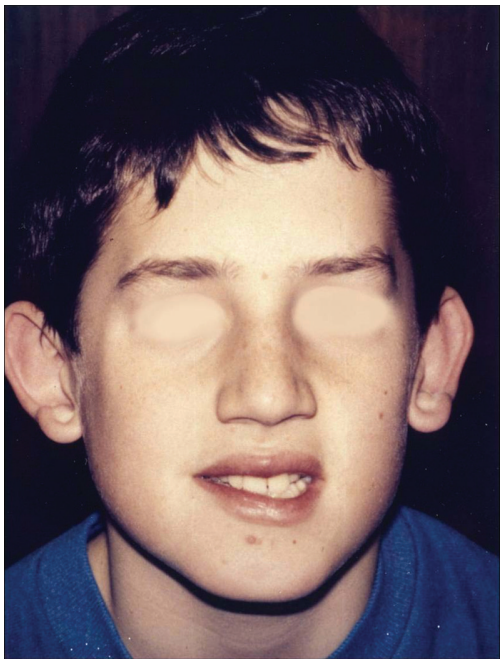


Fig. 83.1 The patient.

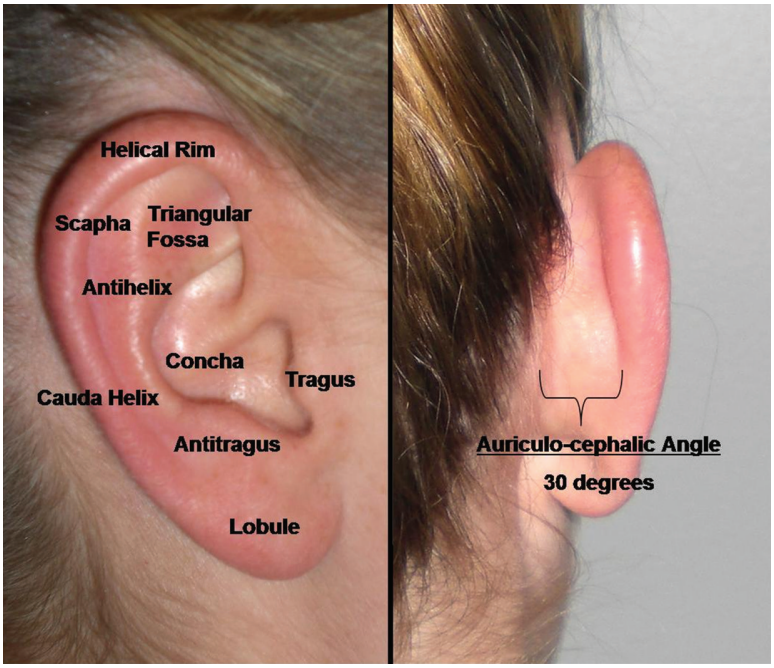


Fig. 83.2 Normal auricle and landmarks in an adult.

(**Table 83.1**). Even though prominauris is often obvious, several key features determine the presence or absence of correctable prominent ears. The physical examination is the key diagnostic element in assessment of prominent ears.

Although most individuals have slight asymmetry from one auricle to the other, cosmetically detectable auricular deformities occur in 3 to 5% of the population. *Prominauris*, or ear protrusion, is defined by the abnormal projection of the ear from the mastoid cortex. This is the most common ear deformity and is frequently the result of unfurling or incomplete development of the antihelical and superior crural folds. A broad, deep conchal bowl can also contribute to the abnormal appearance of the ear. This is another common auricular deformity that may occur alone or in combination with other abnormalities. Thus surgical correction requires proper recognition of each abnormality of the pinna and concha in persons with atypical ears. Each ear must be examined separately because the deformity may differ from one ear to the other. Specific terms help to describe frequently encountered external ear deformities.

Table 83.1 Normal external ear parameters

Vertical length	55–60 mm
Width	5% of height (30–35 mm)
Vertical axis	Angled 20% posteriorly
Auriculomastoid angle	20–35 degrees
Location	
Superior pinna	Level with lateral brow
External auditory canal	Level with inferior orbital rim
Helical-mastoid distances	
Superior rim	18–20 mm
Middle (external canal)	14–16 mm
Inferior (cauda helix)	10–12 mm

- The *protruding ear* has a normal shape and size but with unfurling of the superior crus and antihelix giving lateral protrusion. Present at birth, these abnormalities become more conspicuous with increasing age.
- The *lop ear* is an abnormal ear with helical cartilage that is soft and lacks strength to maintain its structure. The ear subsequently hangs limp, causing it to fold at the upper pole and resemble the appearance of a rabbit's floppy ear. Faulty development of the helix, scapha, and antihelix are attributed to this

deformity. Sometimes the ear will appear smaller than normal.

- The *cup ear* consists of both the *lop* and the *protruding* ear. A deep concave concha cavum is the distinguishing characteristic. In addition, a poorly developed upper pole and antihelix are combined with a short helix and cupped lobule. The vertical height of the auricle is often smaller than normal with the auricular cartilage frequently thicker and cartilage remodeling more challenging.

Nonetheless, auricular deformities are best determined based on the degree of deviation from the shape, size, and location of a normal-appearing ear (abbreviated in **Table 83.1**). Key diagnostic and treatment guidelines for prominent ears are listed below.

1. Ninety-eight percent of normal ears reveal an auriculocephalic angle, defined as the angle between the midpoint of the lateral helix to the mastoid bone, between 20 and 35 degrees. The cosmetically idea angle thus averages 30 degrees. Angles exceeding 40 degrees are generally considered abnormal (**Fig. 83.3**).
2. Normally the fossa triangularis of the pinna faces lateral rather than anterior. The long axis of the pinna averages 55 mm from lobule to the dome. The short axis averages 34 mm from the tragus to the helix.
3. The normal distance between the helix and mastoid averages 10 to 25 mm. This distance changes along the height of the pinna as it normally broadens from the cauda helix (inferior) to the superior pole. The distance from the mastoid bone to the cauda helix approximates 10 to 12 mm; at the midpoint of the ear, 15 to 18 mm at the level of external auditory canal, and at the superior pole 18 to 20 mm. These distances are greater in the prominent ear
4. The normal auricle has a vertical axis directed around 20 degrees posterior. The average vertical height of the auricle is 6 cm. The width is about 55% of the length. The superior aspect of the auricle is usually level with the brow.
5. Other significant auricular landmark deformities in the prominent ear include a poor antihelical fold, an overdeveloped concha, an abnormally formed scapha, and an obvious lack of superior and inferior crus surrounding the fossa triangularis.

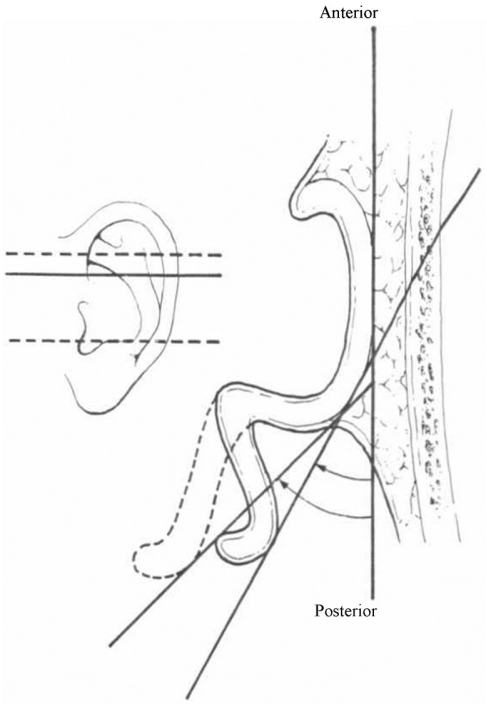


Fig. 83.3 Top view of left auricle depicting auriculocephalic angle.

Test Interpretation

No specific diagnostic modality, beyond physical examination with auricular measurements, is necessary in the evaluation of patients with simple prominauris. Accompanying deformities of the external canal, such as stenosis or atresia, warrant computed tomographic imaging and audiologic evaluation of the deeper structures arising from the first and second branchial arches.

Before any intervention is done on the prominent ear, photographic documentation of the deformity is paramount. Four views are recommended for preoperative assessment, including a posterior full-head view, an anterior face view, a direct close-up of the involved ears, and a lateral head view using the Frankfort horizontal line. Coagulation testing should be performed on individuals with a family history of bleeding disorders before surgery because

hematomas are the most common and dreaded common complication of otoplasty.

◆ Diagnosis

Prominauris, or prominent ears

◆ Medical Management

For a patient 5 years old, no effective medical therapy is available for correcting the anatomic deformity of prominent ears. However, psychosocial support and intervention are occasionally necessary in cases accompanied by the emotional consequences of teasing and harassment.

◆ Surgical Management

The goal of otoplasty is to achieve normal range of contour, shape, size, and symmetry. Ideally, the surgical results should last the lifetime of the patient. Depending on the abnormality, several surgical techniques are available. Sometimes more than one technique will be used. Otoplasty should result in restoration of normal auriculocephalic angle, reduction of an oversized conchal bowl, and return of the natural helical and antihelical curves. It is of utmost importance that the helical rim remains gentle without pinching or unusual breaks. Previously designed surgical tools to achieve these results include strategic suture placement, carefully designed cartilage scoring, and tempered amounts of cartilage trimming or contouring. Surgeon preference and training often direct the techniques used. Nonetheless, each ear needs to be addressed separately with the patient under the same anesthetic and the objective to arrive at symmetry between the two ears. Because the auricle may still be growing for up to 5 years, surgical correction is best delayed until the age of 5.

Perioperative Preparation

Otoplasty for young children is best performed under general anesthesia. Older children (teenage) and adults may undergo the procedure with light intravenous sedation and a total auricular block. Preoperative photographs need to be closely examined and a general surgical design should be created for both ears before surgery. Examination of the natural curves of a normal-appearing ear preceding the repair is essential for proper design.

Creation of an antihelix frequently uses Mustarde suture techniques. Placed correctly, these sutures provide a smooth contour to the new antihelix while securing the helical rim slightly posterior to its native angle. Three to four horizontal mattress sutures are generally secured between the fossa triangularis and scapha and the concha and scapha. Some surgeons use 25-gauge needles to indicate the antihelix location before suture placement.

Light-weight (4.0) permanent suture is ideal for correction of the antihelix. Prolene, nylon, and Mersilene have been used with successful long-term maintenance of the repair. A telephone-ear deformity can result from overcorrection of the central suture relative to the others.

Scoring of the anterior or posterior surface of the helical cartilage has also been used with Mustarde suture techniques to prevent late reprotrusion of the auricle. In older patients, because of increased resistance and calcification of the auricular cartilage, scoring or weakening of the anterior surface can be performed through anterior stab skin incisions or a small skin flap.

Conchal Correction (Cup-Ear Deformity)

Correction of the enlarged concha is often necessary in patients with protruberant ears. The posterior conchal cavum can be shaved tangentially and setback sutures (4–0 Prolene or nylon) can be placed to the mastoid periosteum.

◆ Rehabilitation and Follow-up

Proper dressing of the ears is critical for postoperative success of otoplasty. Following completion of each ear, cotton balls soaked with mineral oil or Vaseline should be secured in the concavities of the scaphoid fossa, fossa triangularis, and concha cavum. At the end of the case, the ears are wrapped with a standard mastoid dressing. For 12 to 24 hours a mastoid dressing is left, followed by a headband over

the top third of the auricles for 1 month to maintain its new position (**Fig. 83.4**).

Major complications following otoplasty include (1) infection (cellulitis, chondritis), (2) hematoma, (3) inadequate correction, (4) telephone-ear deformity, (5) hypertrophic scar or keloid, (6) cartilage buckling, and (7) suture bowstringing. Cellulitis and chondritis are feared complications because they can lead to long-lasting and difficult-to-correct deformities of the ear. Infections are often heralded with late pain out of proportion to the examination. Coverage for *Pseudomonas* is imperative. Any evidence of abscess or hematoma formation should be addressed urgently with incision and drainage. Repeat debridements may be necessary, and antibiotic coverage should also be used.

The most common complication of otoplasty is undercorrection of the original auricular deformity. Revision surgery is best withheld until 1 year from the original surgery to achieve the ideal cosmetic results. Buckling occurs when sutures are placed too far apart.

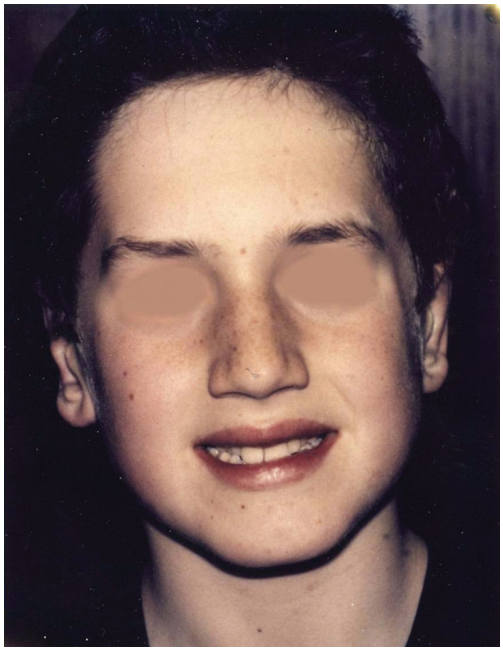


Fig. 83.4 Patient postoperatively (7 years later).

Questions

1. Faulty development of which auricular structures contributes to the appearance of a "lop" ear deformity?
- **A.** Concha
- **B.** Superior crus and helical rim
- **C.** Tragus and lobule
- **D.** Scaphae, antihelix, and helix
- **E.** Temporal insertion of the helix

2. Which measurement is important to determine before operating on the protruding ear?
- **A.** Auriculotemporal angle
- **B.** Cephalic-vertical distance
- **C.** Auriculocephalic angle
- **D.** Helical-mastoid distance
- **E.** All of the above

3. A 4-year-old boy has an abnormal ear that is drawing the attention of his parents and others. You examine the ear, which reveals a normal shape but an unfurling of the superior crus and a distance between the mastoid and helix of 45 mm. What technique is appropriate for repairing the ear, and what is the ideal distance between the mastoid and helix?
- **A.** Furnus technique, 35 mm
- **B.** Mustarde technique, 8 mm
- **C.** Converse approach, 31 mm
- **D.** Mustarde technique, 20 mm
- **E.** Furnus technique, 15 mm

4. When performing otoplasty, what is the ideal age for the procedure and the ideal auriculomastoid angle?
- **A.** 2 years, 35 degrees
- **B.** 3 years, 15 degrees
- **C.** 5 years, 40 degrees
- **D.** 3 years, 20 degrees
- **E.** 5 years, 30 degrees

5. What is the most common complication of otoplasty?
- **A.** Overcorrection
- **B.** Undercorrection
- **C.** Failure
- **D.** Hematoma
- **E.** Infection

Suggested Readings

Adamson PA, Litner JA. Otoplasty technique. Facial Plast Surg Clin North Am 2006;14(2):79–87

Becker DG, Lai SS, Wise JB, Steiger JD. Analysis in otoplasty. Facial Plast Surg Clin North Am 2006;14(2):63–71, v

Converse JM, Nigro A, Wilson FA, et al. A technique for surgical correction of lop ears. Plast Reconstr Surg 1955; 15:411–418

Furnas DW. Complications of surgery of the external ear. Clin Plast Surg 1990;17(2):305–318

Mustarde JC. The correction of prominent ears using simple mattress sutures. Br J Plast Surg 1963;16:170–178

84

Cleft Palate

Jay Paul Willging

History

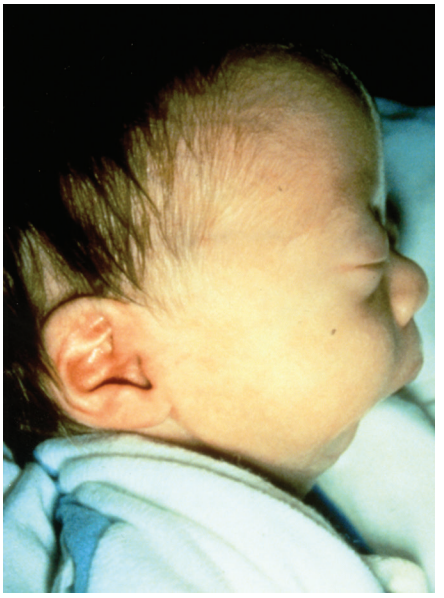
A 38-week-old infant was delivered vaginally with initial Apgar scores of 8 and 10, and he developed intermittent respiratory problems in the delivery room. Desaturations occurred when the child was breathing quietly while in a supine position. On admission of the neonate to the newborn nursery, a physical examination demonstrated a large U-shaped cleft of the palate, mandibular hypoplasia, and glossoptosis. With the child in a side-lying position, saturations were maintained at 98% with no retractions. Feedings were difficult, with frequent choking episodes. Cleft palate feeders were used along with positioning to improve the efficiency of feeding and minimize desaturations.

Over 1 week, a sleep study was obtained that showed no airway obstruction when the child was either prone or on his side. A genetics consultation was obtained that confirmed the diagnosis of Pierre Robin sequence, with no associated syndrome. Feeding abilities improved. Appropriate weight gain was documented over the week. The child was discharged home with close follow-up, and no additional problems developed.

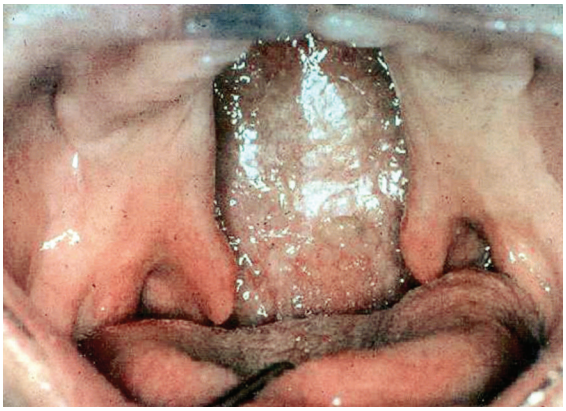
The palate was repaired when the boy was 9 months of age. Ventilation tubes were inserted for a history of chronic otitis media. Mandibular growth continued. He was followed up by the craniofacial anomalies team over the years for coordinated orthodontic and craniofacial care. No additional treatment was required for the retrognathic mandible. Normal resonance was achieved following his palatoplasty, which reconstructed the velum and restored normal velopharyngeal sphincter function (**Fig. 84.1**).

Differential Diagnosis—Key Points

1. The embryologic sequence involved in the normal closure of the palate includes (1) an intrinsic force that enables the palatal shelves to change from a vertical to a horizontal orientation, (2) migration of the tongue in an anteroinferior direction away from the shelves, and (3) palatal fusion. The configuration of the skull base and the size and migration of the tongue and mandible may be responsible for the development of an isolated cleft of the palate.
2. Cleft palate is associated with Pierre Robin sequence. The sequence consists of (1) mandibular hypoplasia, (2) glossoptosis, and (3) midline cleft of the palate. The sequence usually occurs as an isolated triad of abnormalities but occasionally constitutes part of more complex entities such as trisomy 18 or Stickler syndrome. The cause of cleft palate in Pierre Robin sequence is



A



B

Fig. 84.1 The patient showed the typical features of Pierre Robin sequence. Note the severely retrodisplaced mandible **(A)** and the palatal clefting **(B)**.

thought to be from retropositioning or underdevelopment of the mandible, leading to upward displacement of the tongue and causing arrest of the developing palatal shelves, eventually leading to cleft palate. Airway obstruction in patients with Pierre Robin sequence may be life threatening.

3. A cleft palate should alert the clinician to the possibility of other anomalies or associated syndromes. A genetic consultation is advisable.
4. The incidence of clefts of the lip, palate, or both represent one of the most frequently occurring congenital deformities. The frequency is higher in whites (1.34 per 1000 births) than in blacks (0.41 per 1000 births). The chances for a future offspring to have an isolate cleft palate are (1) one sibling has a cleft with no parent with a cleft, 2%; (2) one sibling has a cleft with one parent with a cleft, 17%; and (3) no sibling with a cleft with one parent with a cleft, 7%.
5. Children with a cleft palate are best managed by a team approach to ensure coordinated care. Timing of surgical interventions is often dependent on the achievement of particular milestones: tooth eruption, palatal expansion, and language development.
6. Feeding difficulties exist in nearly all infants with cleft palate. The cleft defect prevents sealing of the oral cavity from the nasopharynx, resulting in an inability of the infant to develop adequate negative pressure required for sucking. Unless specific intervention through the use of adaptable feeding devices is done, these infants fail to gain weight appropriately.
7. Over 80% of patients with cleft palate will develop otitis media with middle ear effusion before cleft repair. Eustachian tube dysfunction is the cause of middle ear effusions in this patient population.

◆ Test Interpretation

In addition to a complete physical examination, no specific laboratory or other diagnostic tests are needed to make this diagnosis. Some genetic testing may be obtained if other findings on physical examination warrant suspicion of a syndrome.

◆ Diagnosis

Pierre Robin sequence with cleft palate

◆ Medical Management

Airway issues are a major concern in this patient population. They tend to stratify into three distinct groups: (1) no problem with their

airway or feeding ability, (2) intermittent airway and feeding problems, (3) severe airway issues. Severe airway problems need immediate intervention. Endotracheal intubation can alleviate the problem but can be very difficult because of the retrognathia that is present. A laryngeal mask airway may temporize the situation until definitive treatment can be undertaken. Patients with intermittent problems should be monitored with a sleep study to define the severity of the problem. Feeding issues are exaggerated by airway problems. Supplemental oxygen with feedings or when sleeping may allow time for continued growth and development to occur such that the problem spontaneously resolves. This management option requires close monitoring to ensure that improvements are occurring. Children with no airway or feeding issues are followed up expectantly.

Feeding is an immediate concern following the birth of an infant with a cleft deformity. It is essential that the parents be taught how to feed the infant. It is necessary to deliver the food to the posterior surface of the tongue and allow the infant to initiate swallowing at that time. Many commercially available nipples of various shapes and sizes can be used to accomplish this goal. Breastfeeding is difficult without the use of prosthetic feeding devices. The primary caregiver is taught to feed the child slowly and allow for frequent burping throughout the feeding.

Genetic evaluation is important to ensure that the clefting is not secondary to an underlying syndrome. Patients with retrognathia associated with a syndrome tend to have worse airway symptoms than those without a syndrome. The airway interventions required for the syndromal patients need to be more aggressive to alleviate the airway obstruction observed than in those patients without an underlying syndrome. Results of genetic evaluations affect treatment approaches.

Hearing and middle ear function must be evaluated in patients with a cleft because of the high incidence of middle ear disease. Acute otitis and chronic middle ear effusions are common. Ventilation tube insertion is associated with a high incidence of otorrhea when performed before the palate is repaired. Hearing screens are helpful in ensuring that no intervention is required.

Velopharyngeal insufficiency occurs when the velum is unable to seal the nasopharynx from the oropharynx during connected speech. An adequate evaluation of resonance cannot be reliably performed until the child has developed connected speech. This is generally around the age of 3 years. If velopharyngeal insufficiency is present, early intervention is associated with the best results as compensatory speech patterns are not given time to develop.

Dental hygiene is important in children with a cleft palate. The potential for future orthodontic intervention warrants developing good dental hygiene habits in childhood to optimize the ability to correct any maxillary crowding or other malocclusion problems that should develop.

Regular follow-up with a craniofacial anomalies team ensures coordinated management of the medical, surgical, and psychosocial problems that commonly develop in children with a cleft.

◆ Surgical Management

The ideal method for management of airway obstruction secondary to retrognathia or glossoptosis is controversial. Conservative measures need to be considered before moving to surgical interventions. Tracheotomy is the most conservative method of bypassing the area of obstruction. In select patients, mandibular distraction may be appropriate to avoid tracheotomy. Distraction may be used as a means of allowing decannulation at an earlier time compared with waiting for natural growth and development to occur to clear the airway. Other surgical interventions have their proponents and include hyoid suspension with lip adhesion.

Surgical closure of the palate should occur when the tissue on the palatal shelves is sufficient to close the defect in the palate easily. Generally waiting until the child is between 9 and 12 months of age permits reliable flaps to be transposed. Early repair (before the development of speech patterns) aids in overall speech and language development.

Management of chronic otitis media with ventilation tube insertion should be considered to ensure normal hearing and maximize speech and language development. Tube otorrhea rates decrease once the palate has been

repaired because the repair also improves eustachian tube function.

Rehabilitation and Follow-up

Long-term follow-up of children with Pierre Robin sequence is required to ensure adequate midface development, occlusion, and speech and language development. Regular evaluations by the craniofacial anomalies team ensure timely interventions to stem problems developing over time. Speech pathologists can identify resonance problems and recommend interventions before the child develops speech strategies to compensate for the difficulty with abnormal resonance occurring because of the cleft palate.

Questions

1. The presence of an isolated cleft of the secondary palate is generally associated with all of the following except:
- **A.** Mental retardation
- **B.** Velopharyngeal insufficiency
- **C.** Middle ear effusion
- **D.** Feeding problems
- **E.** Nasal regurgitation

2. Retrognathia causes feeding problems secondary to which of the following?
- **A.** Impairment of normal lateralization movements of the tongue
- **B.** Abnormal shape of the tongue
- **C.** Interference with normal respiratory patterns during feeding
- **D.** Obstruction of the esophageal inlet by the tongue base
- **E.** Impaired hypopharyngeal muscle contraction due to the position of the tongue base

3. Pierre Robin sequence:
- **A.** Classically refers to the triad of a unilateral cleft of the secondary palate, glossoptosis, and mandibular hypoplasia
- **B.** Associated with myopia, flattened facial appearance, joint problems, and hearing loss is classified as Stickler syndrome
- **C.** Is always associated with airway obstruction
- **D.** Requires mandibular distraction to establish proper class I occlusion in adulthood
- **E.** Can be identified on genetic fluorescent in situ hybridization studies

Suggested Readings

Kummer AW. Cleft Palate and Craniofacial Anomalies: Effects on Speech and Resonance. Florence, KY: Cengage Learning; 2007

Losee JE, Kirschner RE. Comprehensive Cleft Care. New York, NY: McGraw-Hill; 2008

Meyer AC, Lidsky ME, Sampson DE, Lander TA, Liu M, Sidman JD. Airway interventions in children with Pierre Robin Sequence. Otolaryngol Head Neck Surg 2008; 138(6):782–787

85

Nasal Obstruction from a Nasal Septal Deflection

Christopher R. Savage and David B. Hom

History

A 35-year-old man presents to the clinic complaining of a 10-year history of left nasal blockage. He is not bothered by his nasal appearance. He has a small bony hump on his nose and an amorphous nasal tip. His lower nose is bent slightly to the left. He complains of persistent difficulty breathing through the left side of his nose. He does not have any environmental allergies. In the past, he has tried decongestants and nasal steroid sprays for 3 months given to him by his primary care physician without any benefit; he quit using these 6 months ago. He states that his goal with surgery is to improve his breathing on the left side. He denies any nasal trauma and previous nasal surgery. He is currently married and has been an electrician for the past 10 years. He does not smoke or have any nasal allergy symptoms. He does not currently use any over-the-counter intranasal decongestants and is taking no medications. He denies recurrent sinus infections.

On physical examination, his facial skin is moderately thick but not very oily. His chin appears to be in proper anatomic position. He has adequate nasal tip projection, and his nasal tip rotation is 95 degrees. The dorsum of his nose has a small dorsal bony hump at the rhinion and slight leftward deviation. The tip is not ptotic and recoils nicely on removal of fingertip pressure. The nasal tip appears widened and slightly bulbous. He has no alar retraction. Intranasal examination reveals pink mucosa and slightly hypertrophied left inferior turbinate. By visual inspection and palpation, the anterior septum deviates significantly to the left, almost touching the inferior turbinate anteriorly. Administration of topical decongestant slightly reduces the inferior turbinate size, but it does not significantly improve left nasal airflow. Lifting the internal and external nasal valves with cotton-tipped applicators only provides minimal improvement in airflow. Nasal endoscopy shows no intranasal polyps, synechiae, or adenoid hypertrophy.

Differential Diagnosis—Key Points

1. The fact that this patient has constant left nasal obstruction, refractory to topical therapy, along with a deviated nasal septum indicates a left anatomic nasal obstruction. In addition, topical decongestant made little difference in inferior turbinate size,

therefore reducing the likelihood that the obstruction is solely from inferior turbinate hypertrophy.
2. The patient is realistic about his goals and does not expect life-altering changes to occur.
3. It is important to inquire about other nasal surgeries or trauma to determine the expected amount of scar tissue and help with surgical planning.
4. The patient is a nonsmoker with no obvious environmental allergens contributing to his nasal obstruction.

Test Interpretation

The diagnosis is made almost exclusively on the basis of the history and physical examination. Laboratory or radiologic testing is of little benefit.

Diagnosis

Septonasal deflection causing nasal obstruction can be from congenital or acquired causes. Because the nose consists of several different structures (nasal bones, maxillary crest, upper and lower lateral cartilages, nasal septum, and turbinates), a deflection of any one of these components can lead to significant airflow problems.

The general areas of nasal obstruction are the external nasal valve (septum, lower lateral cartilage) and internal nasal valve (bordered by the upper lateral cartilage, nasal septum, pyriform aperture, and inferior turbinate).

Nasal airflow is dependent on three major parameters: septal position, orientation of the nasal bones and cartilages, and turbinate size. To treat nasal obstruction optimally, all three of these factors need to be addressed independently and as a unit in relation to each other. The internal nasal valve is usually the narrowest channel through which inspired air passes. Improving nasal breathing may require increasing the cross-sectional diameter of the nasal valves and nasal laminar airflow. Poiseuille's law describes that a small increase in the radius of the internal nasal passage will improve the nasal airflow and decrease resistance to the fourth power. Specifically, this formula is as follows:

$$\text{Rate of airflow (volume/sec)} = [\text{radius}^4 \times \text{pressure gradient} / (\text{viscosity} \times \text{length})] \times \text{constant}.$$

Thus, small internal nasal corrections can significantly improve nasal airflow.

In addition, if the external nose is a contributing factor for nasal obstruction, it can also have a component of dynamic valvular collapse (narrowing on inspiration) or static valvular collapse (being narrow in the noninspired state).

Using cotton-tipped swabs to lift the specific weak areas of nasal valve collapse can help determine what sites could be further supported while asking the patient what maneuvers improve the nasal breathing. This patient has no evidence of nasal valve collapse on physical examination.

In this case, the patient has a caudally deflected septum and some enlargement of his turbinates, which are the major contributors in causing his nasal obstruction.

Medical Management

Medical therapy will offer limited benefit in this patient because of the anatomic obstruction of his caudally defected septum. Therefore, surgical intervention is the best option to improve the nasal breathing. However, nasal obstruction can be mitigated with nasal steroid sprays, mast cell inhibitors, antihistamines, commercial nasal support devices, and environmental allergy immunotherapy if such therapies are indicated.

Surgical Management

Nasal surgery can be divided into two components: correction of nasal obstruction and correction of cosmetic deformity. Often these goals can be accomplished during the same procedure. In this particular case, the patient is interested only in improving his nasal breathing. He has a caudal left septal deviation contributing to his nasal obstruction.

In this case, a hemitransfixion incision can be initially made with a likely transformation into a full transfixation incision for adequate exposure and freeing both sides of the anterior septal mucosa flaps with partial-thickness scoring on the concave side to decrease the memory of the caudal septum bend. If the deviation is near or along the maxillary crest, a "swinging-door" technique can be performed. This technique is useful for treating a caudal septal strut that is deviated off the premaxillary crest. The deviated cartilage is incised along the floor of the nose after bilateral mucoperichondrial flaps are elevated. This allows for the remaining cartilage to be brought to the midline by placing a figure-of-eight suture of 3–0 chromic between the cartilage and periosteum of the maxillary crest to secure it to the midline.

If morselization and scoring of the septal cartilage as well as bringing the septal strut to the midline are not adequate, a septal onlay baton graft can be placed alongside the bent septum. With this technique, posterior septal cartilage or ethmoid bone is harvested to cover the anterior deformity. After drilling holes in the graft on the back table, the graft is placed against the deformed septum and secured with absorbable sutures thus stenting to septum.

If the septum remains significantly defected, an external approach will be required to provide maximum exposure to all the nasal components. Through an external approach, the upper lateral cartilages can be detached from the dorsal septum and extended spreader grafts can be placed to straighten the caudally deflected septum. In addition, the caudal septum can be placed between the medial crura acting as a strut.

If severe caudal septum deflection is evident, the deflected caudal septum can be completely excised, tailored ex vivo, and placed back in a better position along with extended spreader grafts to it in proper position.

When completing a septoplasty, at least a 1-cm dorsal and caudal strut should be present at the end of the case to maintain adequate nasal dorsal and caudal septal support.

For internal nasal valve narrowing, spreader grafts could be used in addition to inferior turbinate reduction using submucosal shaving, radiofrequency ablation, or electrocautery. Alar Batten grafts, suture suspension techniques, and lateral crura struts could also be used to help open the external nasal valve.

A running quilting suture is then placed to coapt the septal mucosa on the septum with or without an internal splint. The intranasal incisions are then reapproximated. If any undermining was done under the nasal dorsal nasal skin, the nose is taped and an external splint is applied to obliterate any loose empty subcutaneous empty space.

◆ Rehabilitation and Follow-up

The external and internal nasal splints are removed in 1 week. Edema and some ecchymosis are expected and may persist up to 1 year, especially from an external approach. Crusting is removed with peroxide on cotton-tipped swabs, and antibiotic ointment is applied. Daily saline nasal spray is used for several months to minimize crusting and aid in healing.

◆ Questions

1. All of the following are components of the internal nasal valve except:
- **A.** Distal end of the upper lateral cartilage
- **B.** Caudal septum
- **C.** Inferior turbinate
- **D.** Middle turbinate
- **E.** Pyriform aperture

2. The rate of airflow through the nasal opening is most affected by which of the following?
- **A.** Pressure gradient
- **B.** Length of passage through which the air is flowing
- **C.** Radius

D. Viscosity
E. Gravity

3. When performing a septoplasty, what is the least width of caudal septal cartilage that should be left behind?
A. 1 mm
B. 10 mm
C. 7 mm
D. 5 mm
E. 20 mm

Suggested Readings

Arden RL, Coleman GB. Nasal obstruction. In: Mathog RH, Arden RL, Marks SC, eds. Trauma of the Nose and Paranasal Sinuses. New York, NY: Thieme; 1995:171–203

Ballert JA, Park SS. Functional rhinoplasty: treatment of the dysfunctional nasal sidewall. Facial Plast Surg 2006;22(1):49–54

Ducic Y, Hilger P. Surgical correction of the deviated septum. In: Murakami C, ed. Functional Reconstructive Rhinoplasty. Facial Plast Surg Clin North Am.1999;7: 319–331

Kridel RW, Scott BA, Foda HM. The tongue-in-groove technique in septorhinoplasty: a 10-year experience. Arch Facial Plast Surg 1999;1(4):246–256, discussion 257–258

86

Pollybeak Nasal Deformity

Christopher R. Savage and David B. Hom

History

A 47-year-old woman states that she would like improvement of the appearance of her nose on lateral profile (**Fig. 86.1**). She states that she had a nasal surgery 4 years ago by another physician, and she is pleased that he improved her nasal breathing; but she feels like her nose is beaklike on side profile. She can breathe through both sides without difficulty. She is in good health and does not smoke. She is currently married and works as a full-time

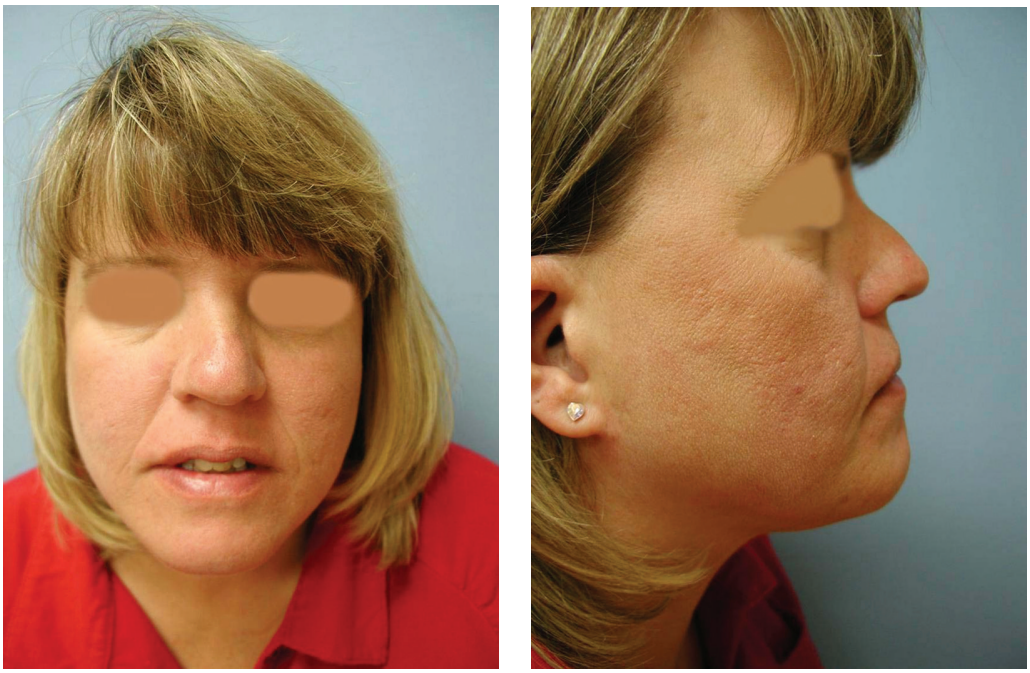


Fig. 86.1 Frontal **(A)** and lateral **(B)** views of a 47-year-old woman interested in rhinoplasty.

elementary teacher. She is pleasant, realistic, and seeks improvement of her lateral nasal profile. She has been considering a second nasal surgery over the past 3 years. Her motivation for the nasal surgery is self-improvement.

On physical examination she is a Fitzpatrick type 2 skin type. The facial skin appears to be thick with many sebaceous units. On anterior view, the middle and lower nose is deviated to the left. Her nasal tip is amorphous, being widened and bulbous. On lateral view, her chin appears to be prognathic. Her nasal tip projection is adequate and is not ptotic. Her nasal tip rotation is about 105 degrees. Her nasion has adequate projection. The dorsum of her nose does reveal a convexity of the cartilaginous dorsum and supratip region. Palpation of her nasal tip effects good recoil on removal of fingertip pressure, but it is lower than the supratip region. She also has decreased columellar show near the columella labial junction. There is no alar retraction. Intranasal examination reveals pink mucosa with normal-sized inferior turbinates. A well-healed previous marginal incision scar is present. The septum is midline. Opening the internal and external nasal valve with cotton-tipped applicators offers no changes in nasal breathing.

◆ Differential Diagnosis—Key Points

1. The patient is realistic about her goals and does not expect life-altering changes to occur.
2. The abnormal tip–supratip relationship in this patient is referred to as a pollybeak deformity. This deformity is usually secondary to a rhinoplasty leading to postoperative fullness of the supratip region.
3. The patient also has a deviated middle and lower nose with an amorphous nasal tip.
4. The good recoil of the nasal tip shows adequate strength of underlying support structures.
5. It is important to inspect the face in its entirety and not just the nose to determine overall symmetry and the relationship of the chin, cheeks, eyes, and forehead. In addition, skin thickness, pigmentation, and the degree of sebaceous units should be noted. All of these can influence surgical outcome.

◆ Test Interpretation

The diagnosis is made almost exclusively on the basis of the history and physical examination. Laboratory or radiologic testing is of little benefit.

◆ Diagnosis

Pollybeak deformity refers to the fullness of the supratip causing a convexity of the nasal dorsum with a low lying tip, giving the appearance of a parrot's beak. There are several causes for pollybeak deformity, including loss of tip support, overresection of the bony dorsum, underresection of the cartilaginous hump, excessive supratip soft tissue, and supratip scar formation. Soft tissue scar formation can occur because there is thicker skin in supratip regions, which causes slower contraction allowing for scar formation to form in the potential supratip dead space. This patient had an excessive cartilaginous dorsum and soft tissue fullness at the supratip region. She was not interested in surgery to alter her chin profile.

◆ Medical Management

Medical therapy offers some benefit in the early several month postoperative period. Small quantities (less than 0.1 mL) of dilute steroid (triamcinolone 10 mg/mL) injected in the subcutaneous plane (not intradermal) can help reduce early supratip edema and scar formation. Nightly paper taping over the supratip region up to 6 weeks may also help in redraping the nasal skin. In this case, it would be too late to institute steroid injections from the first nasal surgery. However, 1 month after the revision nasal surgery, if supratip edema persists, dilute steroid injection can be considered.

◆ Surgical Management

Nasal surgery can be divided into three components depending on the cause of the pollybeak; the best approach, regardless of cause, is an open approach to provided maximum exposure to all the nasal components. This is performed first by doing bilateral marginal incisions and then incising the columella with

an inverted V incision, extended to connect with the marginal incisions. Iris scissors are then used to separate the columellar skin off of the medial crura of the lower lateral cartilages. The elevation is then continued along the nasal tip and dorsum until the nasal bones are visualized.

If the pollybeak is from soft tissue only, conservative resection of this scar and subcutaneous tissue is performed and the skin is then redraped. A dermal stitch can be used to help keep the skin in place and prevent further dead space. A high mucosal transfixion suture can also be placed to help reduce this dead space. If the pollybeak is from underresected cartilage, then the extra cartilage is resected and a columellar strut can be placed to help maintain tip support. If the pollybeak deformity is from overresection on the bony dorsum, then onlay grafting can be implemented. This is especially helpful for a low-lying nasion.

If additional nasal tip projection is required, a tip graft (Peck graft, $9 \times 4 \times 1.5$ mm thick cartilage) can be placed on top of the superior aspect of the domes to increase nasal tip projection to minimize the supratip fullness. A columella baton can be placed at the anterior columella to increase the columella show at the columella labial junction in this patient. The nasal skin is then reapproximated using interrupted sutures, cleaned, and then taped. Taping and an external nasal splint are then applied.

To address her deviated nose, one would consider middle and lateral osteotomies, a right spreader graft, and additional narrowing of the nasal tip with a cephalic trim and dome-binding sutures.

◆ Rehabilitation and Follow-up

The patient returns to the clinic in 1 week for removal of the dressing. Edema and some ecchymosis are expected for up to 1 year. Intranasal crusting is removed with half-strength peroxide. Antibiotic ointment is applied. Nightly paper taping over the supratip region up to 6 weeks may also help in redraping the nasal skin. The patient is instructed on proper incision care and is scheduled to return in a month. At this point, one can occasionally consider giving a dilute steroid (triamcinolone) injection (less than 0.1 mL, 10 mg/mL) in the subcutaneous space if needed.

◆ Questions

1. Which of the following are causes for pollybeak nasal deformity?
   - **A.** Loss of tip support
   - **B.** Overresection of the bony dorsum
   - **C.** Underresection of the cartilaginous hump
   - **D.** Excessive supratip soft tissue
   - **E.** All of the above
2. Soon after rhinoplasty, if pollybeak deformity is occurring, which of the following medical therapy can be instituted?
   - **A.** Subcutaneous steroid injection
   - **B.** Nightly paper taping
   - **C.** Antihistamines
   - **D.** Both A and B
   - **E.** None of the above
3. Which the following facts taken during a patient's history is a warning that the patient might not be an optimal candidate for cosmetic nasal surgery?
   - **A.** She has been considering the procedure for several years.
   - **B.** She is happily married.
   - **C.** She believes the procedure will help her get a job promotion.
   - **D.** She is very realistic about her expectations postprocedure.

Suggested Readings

Hanasono MM, Kridel RW, Pastorek NJ, Glasgold MJ, Koch RJ. Correction of the soft tissue pollybeak using triamcinolone injection. Arch Facial Plast Surg 2002;4(1):26–30

Johnson CM, Toriumi DM. Open Structure Rhinoplasty. Philadelphia, PA: WB Saunders; 1990:516

Papel ID, Frodel J, Holt GR, et al. Facial Plastic and Reconstructive Surgery, 2nd ed. New York, NY: Thieme; 2002

Sheen JH, Sheen AP. Problems in secondary rhinoplasty. In: Sheen JH, ed., Aesthetic Rhinoplasty. 2nd ed. St. Louis, MO: Mosby-Year Book; 1987:1135–1407.

Tardy ME Jr, Kron TK, Younger R, Key M. The cartilaginous pollybeak: etiology, prevention, and treatment. Facial Plast Surg 1989;6(2):113–120

87

Eyelid Ptosis Rejuvenation

Paul A. Brannan and David B. Hom

History

A 55-year-old woman presents with "heavy eyes" and occasional frontal headaches. Her friends often tell her she looks sleepy and they cannot see her eyes because they look "droopy." She must manually lift her eyebrows to see better; another surgeon has offered her an endoscopic brow lift. She is seeking another opinion.

Further questioning reveals that her headaches occur toward the end of the day, especially after prolonged reading, but review of systems is otherwise normal. She has worn contact lenses for longer than 20 years and has not had eye surgery. Examination shows a healthy-appearing woman with bilateral upper blepharoptosis and the top half of her pupils are covered by the eyelid margin. Levator function is normal, and there is no variability of the eyelid height. She has mild temporal brow ptosis. (**Fig. 87.1**).

Differential Diagnosis—Key Points

Individuals presenting with "heavy" or "droopy" eyelids need to be fully evaluated to accurately determine the cause for the eyelid abnormality. Simple eyelid tightening or brow-lifting procedures may not always be indicated, and multiple systemic conditions can cause eyelid malpositions.

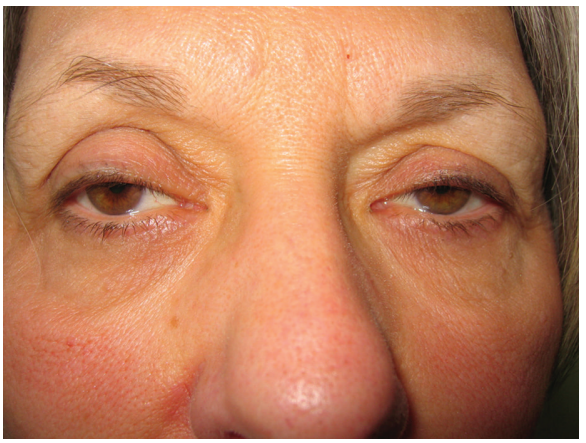


Fig. 87.1 Preoperative photo of the patient.

Common terms used to describe the eyelid are listed below to clarify this confusing terminology:

- *Blepharochalasis* refers to any excess tissues of the eyelids and is a broad term. However, this should be reserved for a rare variant of angioneurotic edema, which presents as recurrent eyelid swelling in young women.
- *Dermatochalasis* is the most appropriate term used to describe excess upper-eyelid skin that occurs with age.
- *Blepharoptosis*, or *ptosis* of the upper eyelid, describes the abnormal downward positioning of the eyelid. When the eyelid margin hangs over the pupil, this constricts the superior visual field.
- *Pseudoptosis* refers to the false appearance of blepharoptosis, in which the excess upper lid and brow tissues hang over the eyelid margin.
- Lid retraction occurs when the eyelid is open too far and the sclera can be seen between the cornea and the eyelid margin. This can result in inability to close the eye (*lagophthalmos*) and exposure keratopathy.

Several systemic conditions can cause eyelid abnormalities:

- Hyperthyroidism in Graves disease can cause upper and lower eyelid retraction and proptosis of the globes. Hypothyroidism causes periorbital edema and lower eyelid bags, referred to as *festoons*.
- Allergies can cause intermittent periorbital edema and erythema as well as dermatitis of the eyelids.
- Blepharoptosis has several causes, from congenital ptosis to acquired forms. Acquired ptosis can be caused by muscular abnormalities such as ophthalmoplegia, oculopharyngeal dystrophy, and myasthenia gravis. Neurologic problems such as oculomotor palsy and Horner syndrome can cause ptosis. Mechanical ptosis results from eyelid masses that increase the weight of the eyelid.
- The most common cause of blepharoptosis is from involutional changes that occur with age, causing disinsertion or dehiscence of the levator tendon from the tarsal plate. These patients tend to have high, indistinct eyelid creases and normal levator muscle function.
- Brow ptosis is another cause of redundant upper eyelid tissues. When the brow tissues lose their elasticity, they descend below the superior orbital rims and gravity pulls the brow downward. This can cause pseudoptosis of the eyelid with peripheral hooding, and patients overuse the frontalis muscle, which can result in frontal headaches. Deep rhytides of the forehead are usually present.

If there is any question about a systemic condition causing blepharoptosis, medical or neurologic consultation is required before surgery is performed.

Test Interpretation

In general, patients a undergoing eyelid surgery should have a thorough ophthalmic examination.

The major concern in this case is determining the cause of her eyelid heaviness, including blepharoptosis, dermatochalasis, and brow ptosis. Blepharoptosis is the most impressive finding on her examination, and at least half of the pupil is covered by the eyelid. She also has a high crease and normal levator function, indicating that this is likely an involutional process. The margin-reflex distance is used to measure the height of the eyelid, which is usually 4 mm. It is measured by shining a light at the cornea while the patient looks straight ahead and measuring the distance of the light reflex to the upper margin.

The brow should be relaxed and its position relative to the orbital rim noted. Upper eyelid skin should be measured as well, and when excess skin drapes over the margin, this constricts the superior visual field. This patient has mild temporal brow ptosis and excess upper skin but not enough to cause visual symptoms.

The levator function of the eyelid needs to be measured by fixating the brow, having the patient look down, then up. The amount of the excursion approximates the levator function and is normal at 12 to 16 mm.

Tear production can be evaluated if the patient has any signs or symptoms of dry-eye syndrome. This can be performed using Schirmer strips. These strips are placed in the fornices and act like a wick over the lower eyelid margin. The eye should produce at

least 10 mm within 5 minutes to be normal. Patients with dry eyes must be counseled that their symptoms will likely worsen following any eyelid surgery, but topical lubricants are usually adequate in providing relief.

Formal visual-field testing using point sources of light in the peripheral visual fields can also establish loss of vision from droopy or excess eyelid tissues. The extent of the peripheral vision is measured with the eyelids in the relaxed position and then taped into normal anatomic position. The tests are compared, and greater than 12 degrees of improvement is considered significant.

Diagnosis

Bilateral involutional blepharoptosis, mild dermatochalasis, mild temporal brow ptosis

Medical Management

There is no role for medical management of the involutional eyelid changes. If dry eye is present, frequent artificial tear use and nightly lubricants can help in the early postoperative period.

Surgical Management

This patient would benefit from bilateral blepharoptosis repair. One could consider cosmetic blepharoplasty and a temporal brow lift to augment her appearance at the same time. Ptosis repair can be performed in several ways, with the anterior approach levator advancement. A small incision in the eyelid crease or through a standard blepharoplasty incision gains access to the levator tendon. A horizontal mattress suture is passed through the tarsal plate and the levator tendon, adjusting the height as needed. The surgery is done with light sedation and a local anesthetic so the patient can cooperate to achieve the best surgical result. Overcorrection, undercorrection, and lid notching are the most common side effects.

There are many ways to correct the brow ptosis, including both external and internal approaches. External approaches involve removal of tissue, and scarring is more evident (direct, midforehead, pretrichial, coronal approaches). Internal approaches, either endoscopic or through a blepharoplasty incision, leave less visible scars but can be more labor intensive. These have become more popular recently, and various bone fixation devices have been created to hold the brow tissue up until the periosteum reattaches in a more superior position. The possible complications of brow procedures are scarring, numbness, visual changes, and damage to the frontalis branch of the seventh cranial nerve (**Fig. 87.2**).

Rehabilitation and Follow-up

Patients are followed up postoperatively for adequate wound healing and eye-surface lubrication. Major time of recovery is usually within 14 days.

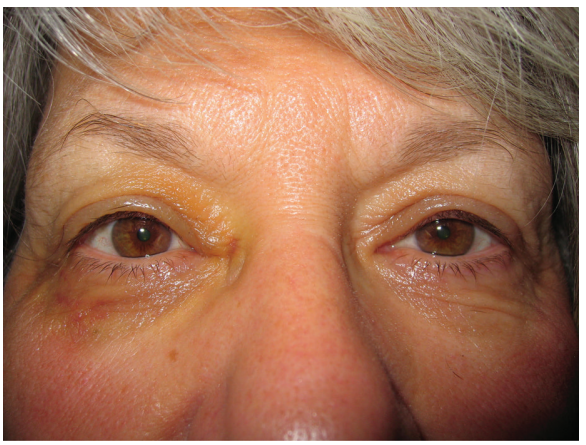


Fig. 87.2 Postoperative photo of the patient.

Questions

1. Droopy eyelids can result from all of the following except:
   - **A.** Dermatochalasis, or excess upper eyelid skin
   - **B.** Blepharoptosis
   - **C.** Eyebrow ptosis, or pseudoptosis
   - **D.** Eyelid retraction from Graves disease
   - **E.** Eyebrow ptosis and dermatochalasis combined
2. Which systemic condition is least likely to cause changes of the eyelids?
   - **A.** Chronic allergies
   - **B.** Myasthenia gravis
   - **C.** Graves hyperthyroidism
   - **D.** Diabetes mellitus
   - **E.** Horner syndrome
3. In the preoperative assessment of the patient requesting eyelid rejuvenation, which is the least important consideration?
   - **A.** Amount of upper eyelid skin
   - **B.** Height of the upper eyelid margin relative to the pupil
   - **C.** History of dry-eye syndrome and tear production
   - **D.** History of active thyroid eye disease
   - **E.** Patient's age

Suggested Readings

Ahmad SM, Della Rocca RC. Blepharoptosis: evaluation, techniques, and complications. Facial Plast Surg 2007; 23(3):203–215

Graf RM, Tolazzi AR, Mansur AE, Teixeira V. Endoscopic periosteal brow lift: evaluation and follow-up of eyebrow height. Plast Reconstr Surg 2008;121(2):609–616

Moskowitz BK, Patel AD, Pearson JM. Aesthetic and functional management of eyelid and orbital reconstruction. Facial Plast Surg 2008;24(1):69–77

Pereira LS, Hwang TN, Kersten RC, Ray K, McCulley TJ. Levator superioris muscle function in involutional blepharoptosis. Am J Ophthalmol 2008;145(6):1095–1098

Rohrich RJ, Coberly DM, Fagien S, Stuzin JM. Current concepts in aesthetic upper blepharoplasty. Plast Reconstr Surg 2004;113(3):32e–42e

Tabatabai N, Spinelli HM. Limited incision nonendoscopic brow lift. Plast Reconstr Surg 2007;119(5): 1563–1570

Viana GA, Sant'Anna AE, Righetti F, Osaki M. Floppy eyelid syndrome. Plast Reconstr Surg 2008;121(5): 333e–334e

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Postoperative Complication from Eyelid Surgery

Paul A. Brannan and David B. Hom

History

The patient is a 55-year-old man who has just undergone bilateral lower lid blepharoplasty. He suffered a severe coughing episode in the recovery area and experienced sudden pain behind both eyes. He complained of double vision and difficulty seeing. When questioned about his medications, he admitted that he had not taken his antihypertensive this morning and had taken multiple aspirin tablets for a headache over the past few days.

Examination revealed a man in moderate distress, blood pressure 180/95, with a pulse of 115. He had diffuse periorbital ecchymoses and gross proptosis. Motility was limited in all directions, and he was able to count fingers with both eyes.

Differential Diagnosis—Key Points

1. This is a vision-threatening emergency that needs to be addressed quickly to avoid loss of vision.
2. Orbital hemorrhage is the most likely diagnosis given the history of recent eyelid surgery, ecchymosis, decreased vision, pain, and proptosis. An acute glaucoma attack would cause pain and decreased vision but would not manifest with ecchymosis or proptosis. A corneal abrasion would also cause pain and blurry vision only. Retinal detachment is unlikely and would result only in diminished vision.
3. The intraocular eye pressure and a funduscopic examination of the retinal veins and arteries should also be checked if possible. These require specialized equipment and an ophthalmologist to perform them correctly; however, the urgency of the situation may not allow for this.

Test Interpretation

No specific test should be performed in this urgent situation other than ophthalmic consultation.

Diagnosis

Postoperative orbital hemorrhage with an orbital hematoma

Medical Management

Management of an orbital hemorrhage requires a correct, timely diagnosis and a complete understanding of the process. Intraocular pressure is elevated when a significant hematoma is present. However, this pressure increase is directly related to orbital pressure and is not a problem intrinsic to the eyeball. Management of this problem should not be directed at lowering the eye pressure with topical or systemic medications. The goal is to lower intraorbital pressure by increasing orbital volume. This can be accomplished by several surgical methods.

In the presence of a post-blepharoplasty patient with an orbital hemorrhage, the following steps are recommended:

1. Call for immediate ophthalmic consultation.
2. If a tarsorrhaphy suture is present, remove it.
3. Obtain some form of vision, either with a handheld near card or small print. If these are not available, counting fingers vision is adequate.
4. Elevate the head of the bed and remove the eyelid sutures. This may allow an egress of blood and clots. Obvious clots should be carefully removed.
5. Palpate the globe to see whether the pressure has been relieved and recheck the vision.
6. Dexamethasone 0.1 to 0.5 mg/kg can be given to reduce optic nerve edema.
7. If hypertension is present, this should be treated immediately with intravenous antihypertensives.

Surgical Management

If the orbit remains tight and vision is diminished, a lateral canthotomy and cantholysis should be performed. By detaching the lateral canthal tendon from the lateral orbital wall, the orbit can expand anteriorly 4 to 5 mm and decompress. This is done by placing one blade of the scissors behind the tendon and one side on the skin and making a cut down to the marginal tubercle of the zygoma. The inferior tendon is then lysed by rotating the scissors 90 degrees downward and releasing the tendon remnants.

If significant improvement does not occur, the patient should be taken back to the operating room as soon as possible. The wounds should be spread open carefully to release any discrete hematomas. However, this is unusual and the blood is often diffusely infiltrated within the orbital tissues, making drainage unlikely. Vision-threatening hematomas may require orbital bony decompression. This is a more involved procedure requiring general anesthesia, and the medial wall and orbital floor are removed. This allows expansion of the orbital contents into the ethmoid and maxillary sinuses and is quite effective. However, the surgery takes 45 to 60 minutes to perform, and returning to the operating room under general anesthesia adds additional time that can lead to irreversible vision loss.

Rehabilitation and Follow-up

The patient should be monitored closely until the orbital examination improves and stabilizes. Head elevation, iced saline gauze, and bed rest assist in the first 48 hours after surgery. If corneal exposure becomes an issue because of the proptosis or chemosis, aggressive lubrication is required until this subsides. The canthotomy and cantholysis can be repaired when the periorbital edema decreases enough to allow easy repair without tension on the eyelid.

Questions

1. In the post-blepharoplasty patient with a significant orbital hemorrhage and optic nerve compromise, which intervention is not indicated?

A. Open the surgical wounds and cauterize active bleeding
B. Start systemic antihypertensives and corticosteroids

C. Start topical ophthalmic antihypertensives to decrease intraocular pressure
D. Perform canthotomy and cantholysis to decrease orbital pressure
E. Evacuate if hematomas are present

2. When an orbital hemorrhage is suspected in the postoperative period, all the following should be performed except:
A. Check vision
B. Assess extraocular motility
C. Grossly assess proptosis
D. Check pupils for an afferent defect
E. Place a tarsorrhaphy suture to compress the orbit

3. Signs of an orbital hemorrhage include all the following except:
A. Orbital pain
B. Decreased motility
C. Decreased intraocular pressure
D. Decreased vision
E. Ecchymosis

Suggested Readings

Cruz AA, Andó A, Monteiro CA, Elias J Jr. Delayed retrobulbar hematoma after blepharoplasty. Ophthal Plast Reconstr Surg 2001;17(2):126–130

Erdogmus S, Govsa F. The arterial anatomy of the eyelid: importance for reconstructive and aesthetic surgery. J Plast Reconstr Aesthet Surg 2007;60(3):241–245

Gausas RE. Complications of blepharoplasty. Facial Plast Surg 1999;15(3):243–253

Hass AN, Penne RB, Stefanyszyn MA, Flanagan JC. Incidence of postblepharoplasty orbital hemorrhage and associated visual loss. Ophthal Plast Reconstr Surg 2004; 20(6):426–432

Joshi AS, Janjanin S, Tanna N, Geist C, Lindsey WH, Lindsey C. Does suture material and technique really matter? Lessons learned from 800 consecutive blepharoplasties. Laryngoscope 2007;117(6):981–984

Morax S, Touitou V. Complications of blepharoplasty. Orbit 2006;25(4):303–318

Schlenker JD, Slavin SA. Proptosis immediately following blepharoplasty due to an inferior rectus intramuscular hematoma. Ann Plast Surg 2006;56(4):437–438

Teng CC, Reddy S, Wong JJ, Lisman RD. Retrobulbar hemorrhage nine days after cosmetic blepharoplasty resulting in permanent visual loss. Ophthal Plast Reconstr Surg 2006;22(5):388–389

Wolfort FG, Vaughan TE, Wolfort SF, Nevarre DR. Retrobulbar hematoma and blepharoplasty. Plast Reconstr Surg 1999;104(7):2154–2162

Answers

Chapter 1

1. C
2. C
3. C

Chapter 2

1. B
2. A
3. C

Chapter 3

1. C
2. D
3. B

Chapter 4

1. C
2. D
3. A

Chapter 5

1. Sinus tympani
2. Canal wall down approach
3. Primary acquired

Chapter 6

1. Medical management with oral fluoride, air-conduction hearing aid, and bone-anchored hearing aid. Stapes surgery would not be recommended because of the risk of chorda tympani nerve injury, affecting his occupation.
2. 2000 Hz
3. In this patient, computed tomography scan is appropriate to rule out cholesteatoma and tympanosclerosis as opposed to otosclerosis. However, exploratory tympanotomy would be the gold standard for diagnosis.

Chapter 7

1. Surgical resection
2. Cranial nerves VII to XII
3. 4%

Chapter 8

1. Infectious (viral)
2. MRI with contrast
3. Systemic, transtympanic

Chapter 9

1. Aminoglycoside use, labyrinthectomy, and vestibular neurectomy
2. Migraine-associated vestibulopathy
3. Low

Chapter 10

1. A
2. C
3. B
4. B

Chapter 11

1. False (and an area of controversy)
2. CT
3. False. The goal is to stop the vertiginous symptoms and prevent further loss of hearing.

Chapter 12

1. Scala media, scala vestibuli, scala tympani
2. Wound-related complications (infection, wound breakdown)
3. Labyrinthitis ossificans

Chapter 13

1. Beta-2 transferrin
2. Computed tomography. Magnetic resonance imaging can be performed secondarily to assess for the presence of an encephalocele.
3. False; this brain tissue is not functional and can be resected with bipolar forceps.

Chapter 14

1. False—because of the risk of life-threatening complications, including hemorrhage or stroke. Radiologic imaging studies (particularly computed tomography) are the ideal modality for diagnosis.
2. Obese women of childbearing age.
3. Surgery and radiation therapy

Chapter 15

1. Translabyrinthine approach
2. Retrosigmoid and middle cranial fossa
3. Cranial nerve V (as in this patient with a diminished corneal reflex)

Chapter 16

1. C
2. D
3. D
4. A

Chapter 17

1. Longitudinal
2. Over 90%
3. Implantation of external auditory canal skin

Chapter 18

1. A
2. C
3. A
4. D

Chapter 19

1. True
2. Meatal foramen (entrance to labyrinthine segment)
3. Ocular complications (e.g., exposure keratitis, which can lead to blindness if left untreated). Artificial tears and eye ointment must be used liberally. Ophthalmologic consultation is warranted if the patient is at a high risk for ocular complications (e.g., anesthetic cornea caused by trigeminal nerve dysfunction).

Chapter 20

1. Local anesthetic related
2. Electroneuronography and electromyography
3. False

Chapter 21

1. C
2. B
3. D
4. E
5. B

Chapter 22

1. C
2. D
3. E

Chapter 23

1. B
2. E
3. D

Chapter 24

1. E
2. D
3. E

Chapter 25

1. C
2. C
3. True

Chapter 26

1. D
2. E
3. D

Chapter 27

1. C
2. E
3. A

Chapter 28

1. C. Explanation: Hemoptysis is commonly seen in patients who have recurrent or persistent head and neck cancer. This is not a common symptom in laryngeal chondronecrosis.
2. E. Explanation: A–D are all useful interventions in the management of chondronecrosis of the larynx.
3. E. Explanation: A–D are all endoscopic features seen in laryngeal chondronecrosis.

Chapter 29

1. A
2. D
3. A

Chapter 30

1. A
2. C
3. C

Chapter 31

1. B
2. C
3. B

Chapter 32

1. B
2. A
3. B

Chapter 33

1. A
2. C
3. C

Chapter 34

1. B
2. E
3. B

Chapter 35

1. C
2. D
3. D

Chapter 36

1. Pleomorphic adenoma
2. Prestyloid space
3. Transcervical

Chapter 37

1. C
2. A
3. D

Chapter 38

1. A
2. D
3. D

Chapter 39

1. C
2. B
3. E
4. C

Chapter 40

1. E
2. C
3. C
4. A

Chapter 41

1. D
2. B
3. D

Chapter 42

1. B
2. D
3. C
4. C

Chapter 43

1. C
2. E
3. C

Chapter 44

1. C
2. B
3. D
4. E

Chapter 45

1. F
2. C
3. D

Chapter 46

1. B
2. B
3. E
4. C

Chapter 47

1. C
2. B
3. A

Chapter 48

1. C
2. C
3. A

Chapter 49

1. E
2. A
3. B

Chapter 50

1. A
2. C
3. B
4. D

Chapter 51

1. B
2. A
3. C
4. C

Chapter 52

1. A
2. B
3. D

Chapter 53

1. D
2. E
3. C
4. C
5. C

Chapter 54

1. A
2. B
3. E

Chapter 55

1. C
2. A
3. E

Chapter 56

1. C
2. C
3. C

Chapter 57

1. B
2. B
3. C

Chapter 58

1. B
2. C
3. A, B, and E
4. A

Chapter 59

1. A
2. B
3. D

Chapter 60

1. A
2. C
3. E

Chapter 61

1. E
2. B
3. B

Chapter 62

1. D
2. D
3. D
4. A

Chapter 63

1. D
2. D
3. C

Chapter 64

1. C
2. B
3. D

Chapter 65

1. C
2. A
3. C

Chapter 66

1. B
2. A
3. D

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