Cholesteatoma

- Author: Peter S Roland, MD; Chief Editor: Arlen D Meyers, MD, MBA more...

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Practice Essentials

A cholesteatoma consists of squamous epithelium that is trapped within the skull base and that can erode and destroy important structures within the temporal bone. Its potential for causing central nervous system (CNS) complications (eg, brain abscess, meningitis) makes it a potentially fatal lesion. See the image below.

A large cholesteatoma. No landmarks are visible, which typically is the case with advanced cholesteatomas.

Generally, the following 3 types of cholesteatoma are identified:

- Congenital cholesteatoma
- Primary acquired cholesteatoma
- Secondary acquired cholesteatoma

Signs and symptoms

The hallmark symptom of a cholesteatoma is a painless otorrhea, either unremitting or frequently recurrent. Other symptoms include the following:

- Conductive hearing loss
- Dizziness: Relatively uncommon
- Drainage and granulation tissue in the ear canal and middle ear: Unresponsive to antimicrobial therapy

Occasionally, cholesteatoma initially presents with symptoms of CNS complications, including the following:

- Sigmoid sinus thrombosis
- Epidural abscess
- Meningitis

Unlike other cholesteatomas, the congenital type is usually identified behind an intact, normal-appearing tympanic membrane. The child often has no history of recurrent suppurative ear disease, previous otologic surgery, or tympanic membrane perforation.[1, 2, 3, 4]

See Clinical Presentation for more detail.

Diagnosis

No laboratory tests or incisional biopsies are generally necessary for the diagnosis of cholesteatomas, because the diagnosis can be made based on physical examination and radiologic findings.

Computed tomography (CT) scanning is the diagnostic imaging modality of choice for these lesions, owing to its ability to detect subtle bony defects.

Histologically, surgically removed cholesteatoma specimens demonstrate typical squamous epithelium. The histology is indistinguishable from that of sebaceous cysts or keratomas removed from any other portion of the body.

Audiometry should be performed prior to surgery whenever possible. Air and bone conduction, the speech reception threshold, and speech discrimination scores should all be determined within a few weeks of the proposed operative procedure.

Magnetic resonance imaging (MRI) is used when very specific problems, such as the following, are suspected[5]:

- Dural involvement or invasion
- Subdural or epidural abscess
- Brain herniation into the mastoid cavity
- Inflammation of the membranous labyrinth or facial nerve
- Sigmoid sinus thrombosis
- Meningitis

See Workup for more detail.

Management

Virtually all cholesteatomas should be excised. The only absolute contraindications to the surgical removal of cholesteatomas are medical in nature.

Canal wall-down tympanomastoidectomy

In the canal wall–down (open) procedure, the posterior canal wall is removed. A large meatoplasty is created to allow adequate air circulation into the cavity that arises from the operation. Canal wall–down operations have the highest probability of permanently ridding patients of cholesteatomas.

Canal wall-up tympanomastoidectomy

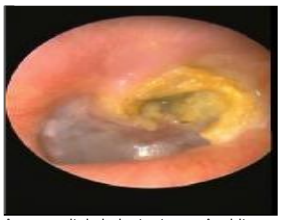
In the canal wall–up (closed) procedure, the canal wall is preserved. Canal wall–up procedures have the advantage of maintaining a normal appearance, but the risk of persistent or recurrent cholesteatomas is higher than in the canal wall–down operation.

See Treatment for more detail.

Background

A cholesteatoma consists of squamous epithelium that is trapped within the skull base that can erode and destroy important structures within the temporal bone. Its potential for causing central nervous system (CNS) complications (eg, brain abscess, meningitis) makes it a potentially fatal lesion (see the images below). (See Etiology and Pathophysiology and Prognosis.)

Epitympanic (attic) cholesteatoma. This is a typical primary acquired cholesteatoma in its earliest stages.



A congenital cholesteatoma. A white mass can be seen behind an intact drum.

Throughout the early half of the 20th century, cholesteatomas were managed by exteriorization. The mastoid air cells were exenterated, the posterior wall of the external auditory canal was removed, and the opening into the resulting cavity was enlarged to ensure adequate air exchange and to make visual inspection simple.

During the 1950s and 1960s, the House Otologic Group developed a new approach. The group attempted to leave the basic underlying anatomic structure of the ear and temporal bone intact, principally by preserving the canal wall. These aggressive attempts to conserve the normal anatomy of the ear created great controversy. Surgeons tended to align themselves with either the old "canal wall–down" (ie, open cavity) procedure[6] or with the new "canal wall –up" (ie, closed cavity) method.[7]

Most otologic surgeons have now migrated to an intermediate position, with the majority of those in the United States now performing both techniques and basing the selection of the open or closed cavity procedure on the individual circumstances of each patient. (See Workup and Treatment.)

Etiology and Pathophysiology

A cholesteatoma consists of squamous epithelium that is trapped in the skull base. Squamous epithelium trapped within the temporal bone (ie, middle ear or mastoid) can expand only at the expense of the bone that surrounds and contains it. Consequently, a growing cholesteatoma can erode and destroy important structures within the temporal bone.

Cholesteatomas cause bony erosion by either of the following mechanisms:

- Pressure effects produce bony remodeling, as occurs normally throughout the entire skeleton when pressure is applied consistently over time
- Enzymatic activity at the margin of the cholesteatoma enhances osteoclastic activity, which greatly increases the speed of bone erosion; osteolytic enzymes appear to increase when a cholesteatoma becomes infected

Occasionally, a cholesteatoma escapes the confines of the temporal bone and skull base. Extratemporal complications can occur in the neck, CNS, or both. When a cholesteatoma within the cranium grows large enough to produce mass effect, brain dysfunction can develop.

Generally, the following 3 types of cholesteatoma are identified: congenital, primary acquired, and secondary acquired.

Congenital cholesteatoma

Congenital cholesteatomas arise as a consequence of squamous epithelium trapped within the temporal bone during embryogenesis. The typical congenital cholesteatoma is found in the anterior mesotympanum or in the perieustachian tube area. They are identified most commonly in early childhood (6 mo to 5 y).[8]

As they expand, congenital cholesteatomas can obstruct the Eustachian tube and produce chronic middle ear fluid and conductive hearing loss. They can also expand posteriorly to encase the ossicular chain and, by this mechanism, produce conductive hearing loss.

Unlike other cholesteatomas, the congenital type is usually identified behind an intact, normal-appearing tympanic membrane. The child often has no history of recurrent suppurative ear disease, previous otologic surgery, or tympanic membrane perforation.[1, 2, 3, 4]

Primary acquired cholesteatoma

A primary acquired cholesteatoma results from tympanic membrane retraction. The classic case develops from progressively deeper medial retraction of the pars flaccida into the epitympanum (attic). As this process continues, the lateral wall of the epitympanum (the scutum) is slowly eroded, producing a defect in the lateral wall of the epitympanum that slowly expands.

The tympanic membrane continues retracting medially until it passes over the heads of the ossicles and into the posterior epitympanum. Ossicular destruction is common. If the cholesteatoma pokes posteriorly into the aditus ad antrum and the mastoid itself, erosion of the tegmen mastoideum, with exposure of the dura and/or erosion of the lateral semicircular canal with deafness and vertigo, may result.[9]

A second type of primary acquired cholesteatoma arises when the posterior quadrant of the tympanic membrane is retracted into the posterior middle ear. The drum initially adheres to the long process of the incus. As retraction continues medially and posteriorly, squamous epithelium envelops the superstructure of the stapes and retracts into the sinus tympani.

Primary cholesteatomas arising from the posterior tympanic membrane are likely to produce facial nerve exposure (and occasionally paralysis) and destruction of the stapedial superstructure. Surgical removal from the sinus tympani may be extremely challenging.

Secondary acquired cholesteatoma

Secondary acquired cholesteatomas result directly from an injury to the tympanic membrane. This injury can be a perforation caused by acute otitis media or trauma, or it may be due to surgical manipulation of the drum.

A procedure as simple as the insertion of tympanostomy tubes could implant squamous epithelium into the middle ear, ultimately producing a cholesteatoma. Posterior marginal perforations are the most likely to result in cholesteatoma formation. Although central perforations are considered unlikely to produce cholesteatomas, they do so occasionally. Any deep retraction pocket can lead to cholesteatoma formation if the retraction pocket becomes deep enough to trap desquamated epithelium.[10, 11]

Epidemiology

The incidence of cholesteatomas in the United States is unknown, but they are a relatively common reason for otologic surgery (approximately weekly in tertiary otologic practices). Death from intracranial complications of cholesteatoma is now uncommon, with the change being attributable to earlier recognition, timely surgical intervention, and supportive antibiotic therapy. Cholesteatomas remain a relatively common cause of permanent, moderate conductive hearing loss in children and adults.

Prognosis

Eliminating a cholesteatoma is almost always possible; however, multiple operations may be required. Because surgery is generally successful, complications from uncontrolled cholesteatoma growth are now relatively uncommon.

Canal wall–down (open cavity) tympanomastoidectomy offers a very low rate of recurrence or persistence of cholesteatomas. Reoperation for cholesteatomas occurs in 5% of patients or less. This compares quite favorably to the 20-40% recurrence rates associated with closed-cavity techniques (canal wall–up).[6]

Nonetheless, because the ossicular chain and/or tympanic membrane cannot always be completely restored to normal, cholesteatomas remain a relatively common cause of permanent, moderate conductive hearing loss.

Mortality

Death from intracranial complications of cholesteatoma is now uncommon, with the change being attributable to earlier recognition, timely surgical intervention, and supportive antibiotic therapy.

Contributor Information and Disclosures

Author
Peter S Roland, MD Professor, Department of Neurological Surgery, Professor and Chairman, Department of Otolaryngology-Head and Neck Surgery, Director, Clinical Center for Auditory, Vestibular, and Facial Nerve Disorders, Chief of Pediatric Otology, University of Texas Southwestern Medical Center; Chief of Pediatric Otology, Children's Medical Center of Dallas; President of Medical Staff, Parkland Memorial Hospital; Adjunct Professor of Communicative Disorders, School of Behavioral and Brain Sciences, Chief of Medical Service, Callier Center for Communicative Disorders, University of Texas School of Human Development

Peter S Roland, MD is a member of the following medical societies: Alpha Omega Alpha, American Academy of Otolaryngic Allergy, American Academy of Otolaryngology-Head and Neck Surgery, American Auditory Society, American Laryngological Rhinological and Otological Society, American Neurotology Society, American Otological Society, North American Skull Base Society, and Society of University Otolaryngologists-Head and Neck Surgeons

Disclosure: Alcon Labs Honoraria Consulting; Advanced Bionics Honoraria Board membership; Cochlear Corp Honoraria Board membership; Med El Corp travel grants Consulting

Chief Editor
Arlen D Meyers, MD, MBA Professor of Otolaryngology, Dentistry, and Engineering, University of Colorado School of Medicine

Arlen D Meyers, MD, MBA is a member of the following medical societies: American Academy of Facial Plastic and Reconstructive Surgery, American Academy of Otolaryngology-Head and Neck Surgery, and American Head and Neck Society

Disclosure: Allergy Solutions, Inc None Board membership; RxRevu Honoraria Chief Medical Editor; Medvoy

Salary Founder and President; Corvectra Consulting fee Senior Medical Advisor; Cerescan Ownership interest Consulting; Essiahealth Consulting fee Advisor; Carespan Advisor; Covidien Consulting fee Consulting

Additional Contributors
The author is grateful for the expert help of Pam Henderson in the preparation of this manuscript.

Additional Contributors

Gerard J Gianoli, MD Clinical Associate Professor, Department of Otolaryngology-Head and Neck Surgery, Tulane University School of Medicine; Vice President, The Ear and Balance Institute; Chief Executive Officer, Ponchartrain Surgery Center

Gerard J Gianoli, MD is a member of the following medical societies: American Academy of Otolaryngology-Head and Neck Surgery, American College of Surgeons, American Neurotology Society, American Otological Society, Society of University Otolaryngologists-Head and Neck Surgeons, and Triological Society

Disclosure: Vesticon, Inc. None Board membership

John E McClay, MD Associate Professor of Pediatric Otolaryngology, Department of Otolaryngology-Head and Neck Surgery, Children's Hospital of Dallas, University of Texas Southwestern Medical School

John E McClay, MD is a member of the following medical societies: American Academy of Otolaryngic Allergy, American Academy of Otolaryngology-Head and Neck Surgery, American College of Surgeons, and American Medical Association

Disclosure: Nothing to disclose.

Paul D Petry, DO, FACOP, FAAP Consulting Staff, Freeman Pediatric Care, Freeman Health System

Paul D Petry, DO, FACOP, FAAP is a member of the following medical societies: American Academy of Osteopathy, American Academy of Pediatrics, American College of Osteopathic Pediatricians, and American Osteopathic Association

Disclosure: Nothing to disclose.

Jack A Shohet, MD President, Shohet Ear Associates Medical Group, Inc; Associate Clinical Professor, Department of Otolaryngology-Head and Neck Surgery, University of California, Irvine, School of Medicine

Jack A Shohet, MD is a member of the following medical societies: American Academy of Otolaryngology-Head and Neck Surgery, American Medical Association, American Neurotology Society, American Tinnitus Association, and California Medical Association

Disclosure: Nothing to disclose.

Francisco Talavera, PharmD, PhD Adjunct Assistant Professor, University of Nebraska Medical Center College of Pharmacy; Editor-in-Chief, Medscape Drug Reference

Disclosure: Medscape Salary Employment

Mary L Windle, PharmD Adjunct Associate Professor, University of Nebraska Medical Center College of Pharmacy; Editor-in-Chief, Medscape Drug Reference

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