

TITLE: Cholesteatoma

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Introduction

Cholesteatomas are expanding lesions of the temporal bone that are composed of a stratified squamous epithelial outer lining and a desquamated keratin center. The matrix is composed of fully differentiated squamous epithelium resting on a connective tissue matrix. Cholesteatomas were named by Johannes Mueller in 1838 with the original erroneous belief that one of the primary components of the tumor was fat. Cholesteatomas may develop anywhere within pneumatized portions of the temporal bone, with the most frequent locations being the middle ear and the mastoid. This tumor formation can lead to many sequelae including infection, otorrhea, bone destruction, hearing loss, facial nerve paresis or paralysis, labyrinthine fistula, as well as intracranial complications including epidural and subdural abscesses, parenchymal brain abscesses, meningitis, and thrombophlebitis of the dural venous sinuses. The destructive nature of cholesteatomas can be attributed to the outer layer of granulation tissue that secretes multiple enzymes which come into contact with bone.

Types of Cholesteatoma

Cholesteatomas can be classified as one of two different types: congenital and acquired. Congenital cholesteatomas are believed to arise from embryonal inclusions or rests of epithelial cells. This classification of cholesteatoma presents behind an intact tympanic membrane, without continuity to the external ear canal and in the absence of etiological factors such as tympanic membrane perforation and a history of ear infections. Modification of the definition of congenital cholesteatoma by Levenson, et. al. in 1989, established a set of criteria for the definition of congenital cholesteatoma in the middle ear. These criteria included, (1) a white mass medial to a normal tympanic membrane, (2) a normal pars flaccida and pars tensa, (3) no prior history of otorrhea or perforations, and (4) no prior otologic procedures. In addition, (5) prior bouts of otitis media were not grounds for exclusion as was the case in the original definition. Levenson's study revealed that the mean age at presentation was 4.5 years with a male preponderance of 3:1. Two thirds of the cases were confined to the anterior superior quadrant of the middle ear. The pathogenesis of congenital cholesteatomas has been theorized by many physicians and researchers, yet the underlying etiology remains unclear. Two

prominent theories include the failure of the involution of ectodermal epithelial thickening that is present during fetal development in proximity to the geniculate ganglion and metaplasia of the middle ear mucosa.

Acquired cholesteatomas are subdivided into primary acquired and secondary acquired cholesteatoma. Several pathogenic mechanisms have been proposed to explain the formation of acquired cholesteatomas, with no single process being accepted as the mechanism for the development of all cases. The common factor of all acquired cholesteatomas is that the keratinizing squamous epithelium has grown beyond its normal limits. Primary acquired cholesteatomas ultimately form due to an underlying Eustachian tube dysfunction that causes retraction of the pars flaccida. Eustachian tube dysfunction directly results in poor aeration of the epitympanic space. This draws the pars flaccida medially on top of the malleus neck, forming a retraction pocket. Once a retraction pocket develops, the normal migratory pattern of the tympanic membrane epithelium is altered, enhancing the potential accumulation of keratin. As keratin accumulates, the sac that has formed will slowly enlarge.

The pathogenesis of secondary acquired cholesteatomas is attempted to be explained by several theories: the implantation theory, the metaplasia theory, and the epithelial invasion theory. The implantation theory proposes that squamous epithelium becomes implanted into the middle ear as a result of surgery, a foreign body, or a blast injury. The metaplasia theory proposes that desquamated epithelium is transformed to keratinized stratified squamous epithelium secondary to chronic or recurrent otitis media. This theory is not believed to be an explanation for a significant cause of cholesteatoma formation in humans. The mechanism behind the epithelial invasion theory is that whenever there is a permanent perforation of the tympanic membrane, the squamous epithelium starts migrating along the perforation edge and may continue medially along the undersurface of the drum destroying the columnar epithelium. It has been proposed that this process is triggered by lingering, chronic infection within the tympanic cavity. Papillary ingrowth refers to the development of cholesteatoma arising from an intact pars flaccida. It is theorized that an inflammatory reaction in Prussack's space, likely secondary to poor ventilation in this area, may cause a break in the basal membrane allowing a cord of epithelial cells to start their proliferation inwards.

Anatomy

Cholesteatoma growth patterns are predictable in that they are channeled along characteristic pathways by ligaments, folds, and ossicles. The most common locations from which cholesteatoma arise are the posterior epitympanum, the posterior mesotympanum and the anterior epitympanum. A basic knowledge of the anatomy of this area provides a foundation for understanding the disease progression and concepts for surgical management.

The middle ear can be divided into three compartments: the mesotympanum, hypotympanum, and epitympanum. The boundary that defines these areas is the external auditory canal. The epitympanum is superior and medial to the superior aspect of the external auditory canal. The hypotympanum is inferior and medial to the inferior aspect of the external auditory canal. The mesotympanum is medial to the external auditory canal with its inferior and superior boundaries defined by the inferior and superior aspect of the external auditory canal respectively.

The mesotympanum contains the stapes, long process of the incus, handle of the malleus and the oval and round windows. The eustachian tube exits from the anterior aspect of the mesotympanum. Two recesses extend posteriorly from the mesotympanum that are often not visible directly- the facial recess and sinus tympani. The facial recess and sinus tympani, are the most common location for cholesteatoma persistence after chronic ear surgery. The facial recess is lateral to the facial nerve, bounded by the fossa incudis superiorly and the chorda tympani nerve laterally. This recess may be directly accessed through a posterior approach via the mastoid (posterior tympanotomy or facial recess approach). The sinus tympani lies between the facial nerve and the medial wall of the mesotympanum and is very difficult to access surgically.

The epitympanum lies above the level of the short process of the malleus, containing the head of the malleus, body of the incus, and their associated ligaments and mucosal folds. The annular ligament sends off fibrous bands from the anterior and posterior tympanic spines that meet at the neck of the malleus. The dehiscent area in the tympanic bone, known as the notch of Rivinus, lies above these bands. The dense fibers that form the middle layer of the pars tensa do not extend to the pars flaccida. The lack of this structural support predisposes Shrapnell's membrane to retraction when negative middle ear pressure is present secondary to Eustachian tube dysfunction.

Cholesteatomas of the epitympanum start in Prussack's space between the pars flaccida and neck of the malleus with the upper boundary being the lateral malleolar fold. The most common spread patterns of cholesteatomas from Prussack's space are through the posterior epitympanum, posterior mesotympanum and anterior epitympanum. The most common spread pattern of the three is the posterior epitympanic route where the cholesteatoma spreads to the superior incudal space lateral to the body of the incus potentially gaining access to the mastoid through the aditus ad antrum. The second most common is the inferior route, through the posterior pouch of von Troeltsch. This pouch lies between the tympanic membrane and the posterior malleolar fold. Spread via this route allows cholesteatoma to gain access to the regions of the stapes, round window, sinus tympani and facial recess. Anterior epitympanic cholesteatomas form anterior to the malleus head. They may be easily overlooked during tympanomastoidectomy if the area is not explored. Facial nerve dysfunction may occur with these lesions, which can also gain access to the supratubal recess of the middle ear via the anterior pouch of von Troeltsch.

As previously described, the hypotympanum is the portion of the middle ear that lies inferior and medial to the floor of the bony ear canal. It is an irregular bony groove that is seldom involved by cholesteatoma. Occasionally, the jugular bulb may be dehiscent in this area.

Prevention of Cholesteatoma Formation

When a patient evaluated in clinic is noted to have a retraction pocket, the otolaryngologist must recognize that this manifestation is due to Eustachian tube dysfunction and that the condition precedes the development of acquired cholesteatoma. As a result, a long-term tympanostomy tube should be placed to resolve the negative middle ear pressure. This intervention may allow the tympanic membrane to revert to a neutral position. If the retraction pocket is adherent to the ossicles or folds or if it has been present for an extended period of time, the retraction pocket will persist. If the retraction pocket persists, surgical exploration may be

indicated.

Patient Evaluation

As always, the initial patient evaluation should include a thorough history. A detailed otologic history should be obtained in order to elicit the early symptoms of cholesteatoma including hearing loss, otorrhea, otalgia, nasal obstruction, tinnitus and vertigo. A previous history of middle ear disease, such as chronic otitis media and/or tympanic membrane perforation may be revealed. Progressive unilateral hearing loss with a chronic foul smelling otorrhea should raise suspicion.

In addition to a thorough head and neck examination, the otologic examination should be meticulous and complete. Otomicroscopy is of the utmost importance in evaluating the presence of cholesteatoma and extent of disease. The ear should be thoroughly cleaned of otorrhea and debris with cotton-tipped applicators or suction. A retraction pocket may be seen, often in the attic and posterosuperior quadrant of the tympanic membrane. Accumulation of squamous debris may occur within the pocket. Granulation tissue may arise from the diseased infected bone of the scutum or posterior bony wall. When extensive, a polyp may protrude through an attic defect. Extreme caution should be used with polyp removal as it may be adherent to important underlying structures such as the ossicles or facial nerve. Pneumatic otoscopy should be performed in every patient with a cholesteatoma. A positive fistula (pneumatic otoscopy will result in nystagmus and vertigo) response suggests erosion of the semicircular canals or cochlea. Cultures should be obtained with wet, infected ears. Topical and/or oral antibiotics should be administered in these cases.

Pure tone audiometry with air and bone conduction, speech reception thresholds, and word recognition usually reveal a conductive hearing loss in the affected ear. The degree of conductive loss will vary considerably depending on the extent of disease. A moderate conductive deficit in excess of 40 dB indicates ossicular discontinuity, usually from erosion of the long process of the incus or capitulum of the stapes. A mild conductive deafness may be present with extensive disease if the cholesteatoma sac transmits sound directly to the stapes or footplate. Audiometry results should always be correlated with the 512Hz tuning fork exam. Tympanometry results will vary and may suggest decreased compliance or perforation of the tympanic membrane.

Preoperative imaging with computed tomographies (CTs) of the temporal bones (2mm - section without contrast in axial and coronal planes) allows for evaluation of anatomy, which may reveal evidence of the extent of the disease as well as screen for asymptomatic complications. Although a temporal bone CT is not essential for preoperative evaluation, they should be obtained for revision cases due to altered landmarks from previous surgery, for patients with complications of chronic suppurative otitis media, suspected congenital abnormalities, or cases of cholesteatoma in which sensorineural hearing loss, vestibular symptoms, or other evidence of complications exist.

Preoperative counseling is an absolute necessity prior to surgery. The primary objective of surgery is a safe dry ear which is accomplished by treating all supervening complications, removing diseased bone, mucosa, granulation polyps, and cholesteatoma while preserving as

much normal anatomy as possible. Improvement of hearing is a secondary goal. Possible adverse outcomes must be discussed including facial paralysis, vertigo, further hearing loss, and tinnitus. The patient should understand that long-term follow-up will be necessary and that they may need additional surgeries.

Surgical Management

Cholesteatoma is treated surgically with a primary goal of total eradication of cholesteatoma to obtain a safe and dry ear. The second objective is restoration or maintaining the functional capacity of hearing. The third objective is to maintain a normal anatomic appearance of the ear if possible. The surgical procedure to be used should be designed for each individual case according to the extent of disease. More extensive disease will usually dictate a more aggressive surgical approach.

Canal-Wall-Down (CWD) Procedure

Prior to the advent of the tympanoplasty, all cholesteatoma surgery was performed using a CWD approach. This procedure involves taking down the posterior canal wall to the level of the vertical facial nerve and exteriorizing the mastoid into the external ear canal. The epitympanum is obliterated with removal of the scutum, head of the malleus and incus. A classic CWD operation is the modified radical mastoidectomy in which the middle ear space is preserved. The radical mastoidectomy is a CWD operation in which the middle ear space is eliminated and the eustachian tube is plugged. Meatoplasty should be large enough to allow good aeration of the mastoid cavity and permit easy visualization to facilitate postoperative care and self cleaning. The indications for this as an initial approach are cholesteatoma in an only hearing ear, significant erosion of the posterior bony canal wall, history of vertigo suggesting a labyrinthine fistula, recurrent cholesteatoma after canal-wall-up surgery, poor eustachian tube function, and a sclerotic mastoid with limited access to the epitympanum.

The advantages of the CWD procedure are that residual disease is easily detected, recurrent disease is rare, and the facial recess is exteriorized. The major disadvantage of this procedure is the open cavity and that mastoid bowl maintenance can be a lifelong problem. Healing takes longer in open cavities and the middle ear is shallow and difficult to reconstruct. Dry ear precautions are essential.

Canal-Wall-Up (CWU) Procedure

The CWU procedure was developed to avoid the problems and maintenance necessary when CWD procedures are performed. CWU consists of preservation of the posterior bony external auditory canal wall during simple mastoidectomy with or without a posterior tympanotomy. A staged procedure is often necessary with a scheduled second look operation at 6 to 18 months for removal of residual cholesteatoma and ossicular chain reconstruction if necessary. The procedure should be adapted to the extent of disease as well as the skill of the otologist. This approach may be indicated in patients with a large pneumatized mastoid and a well aerated middle ear space, suggesting good eustachian tube function. CWU procedures are contraindicated in only hearing ears or in patients with a labyrinthine fistula, long-standing ear disease, or poor eustachian tube function.

The advantages of CWU compared with CWD mastoidectomies are more rapid healing time, easier long-term care, no dry ear precautions, and hearing aids are easier to fit and wear if they are needed. The disadvantages associated with this procedure are the difficulty of technique leading to longer operative time, residual disease is more difficult to detect, retraction pockets leading to recurrent disease are possible, and staged operations are often necessary.

Transcanal Anterior Atticotomy

A transcanal anterior atticotomy is indicated for limited cholesteatoma involving the middle ear, ossicular chain, and epitympanum. If the extent of the cholesteatoma is unknown, this approach can be combined with a CWU mastoidectomy or extended to a CWD procedure. The atticotomy involves elevation of a tympanomeatal flap via an endaural incision with removal of the scutum to the limits of the cholesteatoma. After removal of the disease, the aditus is obliterated with muscle, fascia, cartilage or bone prior to reconstruction of the middle ear space. Some advocate reconstruction of the lateral attic wall with bone or cartilage, however, this may lead to retraction disease and possible recurrence in patients with poor eustachian tube function.

Bondy Modified Radical Mastoidectomy

Although rarely used today, the Bondy procedure is a useful for specific types of cholesteatoma. It is indicated for attic and mastoid cholesteatoma that does not involve the middle ear space and is lateral to the ossicles. Preferably, the mastoid should be poorly developed for creation of a small cavity. The eustachian tube function should be adequate, with an intact pars tensa and aerated middle ear space. The Bondy procedure is performed like the modern modified radical mastoidectomy with the exception that the middle ear space is not entered.

Complications

The expansion of cholesteatomas combined with the propensity of infection result in numerous complications that include ossicular chain destruction, exposure of the membranous labyrinth, exposure of the facial nerve and dura, and infection of the mastoid and intracranial spaces.

Conductive hearing loss is a common complication of cholesteatoma as ossicular chain erosion occurs in 30% of cases. Erosion of the lenticular process and or stapes superstructure may produce a conductive hearing loss as high as 50dB. However, hearing loss may vary with the development of a natural myringostapediopexy or transmission of sound through a cholesteatoma sac to the stapes or footplate. This results in less of a conductive hearing loss. The ossicular chain should always be assumed to be intact. Evidence of sensorineural hearing loss may indicate involvement of the labyrinth. Following surgery, 3% of operated ears have further impairment permanently due to the extent of the disease present or due to complications in the healing process. Patients should be counseled that there is a possibility of total loss of hearing in the operated ear. Also, with two-staged operations, the hearing will be worse after the first operation.

A labyrinthine fistula may occur in as many as 10% of patients with chronic ear infection

due to cholesteatoma. A fistula should be suspected in a patient with longstanding disease with sensorineural hearing loss and/or vertigo induced by noise or pressure changes in the middle ear. Absence of a positive fistula test does not rule out this complication. Fine cut CT of the temporal bone should be obtained if this condition is suspected. The most common site of a labyrinthine fistula is the horizontal semicircular canal, although the basal turn of the cochlea is also at risk. The procedure of choice with this complication is a modified radical mastoidectomy, as discussed previously. Management of the matrix overlying the fistula depends on the infection status of the ear, the degree of hearing loss in the affected and nonaffected ear, the size and location of the fistula, and the surgeon's experience. In an only hearing ear, matrix should be left intact over the fistula. Matrix should also be left over extensive fistulae of the vestibule or cochlea if hearing is normal. Matrix can be removed in a relatively dry, uninfected ear with a normal hearing opposite ear, and the fistula covered with bone pate or fascia.

Facial paralysis in patients with cholesteatoma requires immediate surgery. The paralysis may develop acutely secondary to infection or slowly from chronic expansion of the cholesteatoma. A temporal bone CT should be obtained to help localize the nerve involvement. The most common site of facial nerve involvement is at the geniculate ganglion due to disease in the anterior epitympanum. A simple mastoidectomy with a facial recess approach will expose the tympanic and mastoid portions of the facial nerve, while a middle fossa approach is required when there is petrous apex involvement. Removal of the cholesteatoma and infected material with decompression of the nerve usually results in the recovery of facial nerve function. Administration of intravenous antibiotics and high-dose steroids are also helpful. Iatrogenic injury to the nerve during surgery should be immediately repaired with decompression of the nerve proximal and distal to the site of injury.

Intracranial complications of cholesteatoma are potentially life-threatening. Infections such as a periosteal abscess, lateral sinus thrombosis and intracranial abscess occur in less than 1% of all cholesteatomas. Findings suggesting an impending intracranial complication include suppurative malodorous otorrhea, usually with chronic headache, pain and/or fever. The presence of mental status changes with nuchal rigidity or cranial neuropathies warrant neurosurgical consultation with urgent intervention. Epidural abscess, subdural empyema, meningitis and cerebral abscesses should be treated immediately prior to definitive otologic management of ear disease.

A brain hernia presents at a revision surgery as a meningoencephalocele or an encephalocele. It results from a defect in the tegmen tympani or tegmen mastoideum due to trauma from the drill at the time of the original surgery. This condition must be carefully inspected and repaired at the time of surgery.

Conclusion

The pathogenesis of cholesteatoma remains uncertain. The identification and behavior of the disease, however, is well described. For successful management of cholesteatomas, it is essential to possess a basic knowledge of the important anatomic and functional characteristics of the middle ear. Careful and thorough evaluations are the key to the early diagnosis and treatment of the disease. Early diagnosis and treatment can prevent complications and preserve

hearing. Treatment of cholesteatoma is surgical with the primary goal to eradicate disease and provide a safe and dry ear. Surgical approaches must be customized to each patient depending on the extent of their disease. The surgeon must be aware of the serious and potentially life-threatening complications of cholesteatomas.

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Portions of this paper and presentation were taken directly from the September 18, 2002 Grand Rounds presentation by Michael Underbrink and Arun Gadre entitled Cholesteatoma.

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