

**TITLE: Cholesteatoma**

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## **I. Introduction**

Cholesteatoma is an abnormal accumulation of keratin-producing squamous epithelium in the middle ear, epitympanum, mastoid or petrous apex. It has been further defined as a three dimensional epidermoid structure exhibiting independent growth, replacing middle ear mucosa, and resorbing underlying bone. Although it is not a neoplastic lesion, it can be insidious and potentially dangerous to the patient. The term "cholesteatoma" was first used by Johannes Müller in 1838 to describe a true neoplasm he thought was "a pearly tumor of fat...among sheets of polyhedral cells". In fact, the cholesteatoma appears histologically as a benign keratinizing squamous cell cyst made up of three components, i.e. the cystic content, the matrix and the perimatrix. The cystic content is composed of fully-differentiated anucleate keratin squames. The matrix contains the keratinizing squamous epithelium lining a cyst-like structure. The perimatrix or lamina propria is the peripheral part of the cholesteatoma consisting of granulation tissue, which may contain cholesterol crystals. The perimatrix layer is in contact with bone, and it is this granulation tissue, which produces various proteolytic enzymes that may result in bone destruction.

## **II. Classification and pathogenesis**

Cholesteatoma may be classified according to presumed etiology into two general categories: congenital and acquired. Acquired cholesteatomas can be further divided into primary and secondary acquired. Congenital cholesteatomas are thought to arise from embryonal inclusions or rests of epithelial cells. It refers to cholesteatomas present 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. They can be further classified according to location within the temporal bone (the petrous pyramid, mastoid and middle ear cleft). Levenson, et. al., established a set of criteria for the definition of congenital cholesteatoma in the middle ear. These 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. In their study (over 40 cases), 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.

Several pathogenic mechanisms have been produced to explain the development of acquired cholesteatomas. No single process is accepted as the mechanism for the development of all cases. However, in all types the keratinizing squamous epithelium has spread beyond its normal limits. With primary acquired cholesteatomas, the cause is due to underlying Eustachian tube dysfunction resulting in retraction of the pars flaccida. The problem becomes poor aeration of the epitympanic space which draws the pars flaccida medially on top of the malleus neck. Once a retraction pocket develops, the normal migratory pattern of the tympanic membrane epithelium is altered, encouraging the accumulation of keratin. If not addressed, the sac slowly enlarges to and around the ossicles, the attic walls, etc. The following theories explain secondary acquired cholesteatoma pathogenesis. The implantation theory proposes that squamous epithelium becomes implanted into the middle ear as a result of surgery, foreign body (ventilating tubes), or blast injury. The metaplasia theory explains that as a result of chronic or recurrent otitis media the low cuboidal epithelium of the middle ear becomes transformed to a keratinized stratified squamous epithelium, similar to other parts of the body (nose, sinuses, bronchi) in response to chronic irritation or infection. The mechanism behind the epithelial invasion or migration 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 (Shrapnell's membrane). 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.

### **III. Anatomic Considerations**

Cholesteatomas enlarge with fairly typical patterns of growth. The most common locations from which cholesteatoma arise are the posterior epitympanum, the posterior mesotympanum and the anterior epitympanum. Cholesteatomas are channeled along characteristic pathways by surrounding mucosal folds, the middle ear ossicles, and their suspensory ligaments. 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 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 impossible to visualize directly. These spaces, the facial recess and sinus tympani, are the most common location for cholesteatoma persistence after chronic ear surgery. The sinus tympani lies between the facial nerve and the medial wall of the mesotympanum and is very difficult to access surgically. The facial recess is lateral to the facial nerve, bounded by the fossa incudis superiorly and the chorda tympani nerve laterally. It may be directly accessed via a posterior approach, through the mastoid (posterior tympanotomy or facial recess approach). The hypotympanum is the portion of the middle ear that lies below 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.

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 in the face of negative middle ear pressure.

Epitympanic cholesteatomas start in Prussack's space between the pars flaccida and neck of the malleus with the upper boundary of the lateral malleolar fold. The most common locations of spread of cholesteatomas from Prussack's space are via the posterior epitympanum, posterior mesotympanum and anterior epitympanum, in that order. The most common 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, thought 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 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.

#### **IV. Evaluation**

**History** - A careful otologic history should be obtained in order to elicit the early symptoms of cholesteatoma. The most common presenting symptoms are 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 evident. Progressive unilateral hearing loss with a chronic foul smelling otorrhea should raise suspicion.

**Physical Examination** - In addition to a thorough head and neck examination, particular attention should be paid to the otologic exam. Otomicroscopy is most

important in evaluating the presence of cholesteatoma and extent of disease. The ear should be thoroughly cleaned of otorrhea and debris. A retraction pocket may be seen, often in the attic or posterosuperior quadrant of the TM. Accumulation of squamous debris may occur within the pocket. Granulation tissue may be 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 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.

Audiology - 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.

Imaging - Preoperative imaging with CT of the temporal bones allows pre-operative imaging of anatomy, some evidence of the extent of the disease and a screen for asymptomatic complications. However, it has not gained wide acceptance as an essential aid to planning surgery in uncomplicated cases of cholesteatoma. Temporal bone CT 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.

## **V. Management**

Cholesteatoma is a surgical disease for which the primary, universally accepted goal is total eradication of cholesteatoma to obtain a safe, dry ear. The second objective is restoration or maintaining the functional capacity of the ear, the hearing. The third objective is to maintain a normal anatomic appearance of the ear if possible. Management of complications when they arise takes priority over other objectives. The surgical procedure to be used should be designed for each individual case according to the pathology present. The extent of disease often will determine the aggressiveness of the surgical approach.

As with any surgical procedure, preoperative counseling is mandatory. Surgical goals, risks of surgery (facial paralysis, vertigo, tinnitus, hearing loss), possibility of

staged procedure, need for long-term follow-up and routine aural toilet if necessary should be reviewed in detail with the patient.

Medical management, including aggressive aural toilet, powder applications, and office local care may exteriorize and safely decompress the accumulating keratin debris. This may be a valid management strategy for patients in whom anesthesia poses an unacceptable risk. Such management is not recommended in children. Preoperatively, it is very important to eliminate drainage and any acute inflammatory changes. This will reduce troublesome intraoperative bleeding and help with the delineation of irreversible disease that must be removed from preservable structures.

Retraction pockets: Tympanostomy tube insertion for ventilation of the middle ear may alleviate early tympanic membrane retraction associated with eustachian tube dysfunction. A long term ventilation tube is often necessary. If the pocket persists despite tube placement, surgical exploration is indicated. Alternatively, excision of the pocket at the time of tympanostomy tube placement with or without tympanoplasty has been described to prevent development of cholesteatoma and ossicular discontinuity.

### **Canal-wall-down (CWD) procedures**

Prior to the advent of tympanoplasty techniques, all cholesteatoma surgery was of this type. These procedures involve taking down the posterior canal wall to the vertical facial nerve, 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 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:

- 1) cholesteatoma in an only hearing ear
- 2) significant erosion of the posterior bony canal wall
- 3) history of vertigo suggesting a labyrinthine fistula
- 4) recurrent cholesteatoma after ICW surgery with poor eustachian tube function
- 5) 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. Also, dry ear precautions are necessary.

### **Intact-canal-wall (ICW) procedure**

This procedure was developed to avoid cavity problems altogether. It 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 12 months for removal of residual cholesteatoma and ossicular chain reconstruction. 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. Intact canal wall procedures are contraindicated in only hearing ears or in the patient with a labyrinthine fistula, long-standing ear disease, or poor eustachian tube function.

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

### **Transcanal anterior atticotomy**

This procedure 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 an intact canal wall 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, this is a useful procedure 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.

## **VI. Complications of cholesteatoma**

Conductive hearing loss is a common complication of cholesteatoma as ossicular chain erosion occurs in as many as 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 myringostapediopexy or transmission of sound through a cholesteatoma sac to the stapes or footplate. 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. Patient's should be counseled that on occasion there is a total loss of hearing in the operated ear, and with two-staged operations, the hearing will be worse after the 1st operation.

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. The most common site is the horizontal semicircular canal, although the basal turn of the cochlea is also at risk. The procedure of choice with this complication has been the modified radical mastoidectomy, as discussed previously. Management of the matrix overlying the fistula depends on the infection status of the ear, degree of hearing loss in the affected and nonaffected ear, size and location of the fistula and 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 following infection or slowly from chronic expansion of the cholesteatoma. A CT of the temporal bone is obtained which helps localize the involvement. The most common site is the geniculate ganglion from disease in the anterior epitympanum.

A simple mastoidectomy with facial recess approach will expose the tympanic and mastoid portions of the facial nerve, while a middle fossa approach is required with involvement of the petrous apex. Removal of cholesteatoma and infected material with decompression of the nerve usually suffice. 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 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 chronic with 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.

## Conclusions

The exact mechanism or pathogenesis of cholesteatoma is not clearly identified, however, neither the aggressiveness of the disease nor the description of its key elements are debated. For successful management of the disease, it is essential to possess a basic knowledge of the important anatomic and functional characteristics of the middle ear. Careful and thorough evaluation are keys to the early diagnosis and treatment of the disease, which may prevent complications and preserve hearing. Cholesteatoma is a surgical disease with the primary goal to eradicate disease and provide a safe, dry ear. The surgical strategies, however, vary greatly depending on the extent of disease and surgeon's experience. The surgeon must be aware of the serious and potentially life-threatening complications of cholesteatomas.

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