Cholesteatoma

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Overview

- Definition
- Classification and Theories
- Anatomic Considerations
- Patient Evaluation
- Management
- Complications
Definition
Cholesteatoma

- Named by Johannes Mueller in 1838
  - Erroneous belief that one of the primary components of the tumor was fat
  - “a pearly tumor of fat…among sheets of polyhedral cells”

- More appropriate name has been suggested to be keratoma to describe tumor composition
Cholesteatoma

- Expanding lesion of the temporal bone composed of
  - Cystic content: desquamated keratin center
  - Matrix: keratinizing stratified squamous epithelium
  - Perimatrix: granulation tissue that secretes multiple proteolytic enzymes capable of bone destruction

- May develop anywhere within pneumatized portions of the temporal bone

Most frequent locations:
- Middle ear space
- Mastoid
Classification and Theories
Cholesteatoma Classification

- Congenital
- Acquired
  - Primary
  - Secondary

Cholesteatoma Formation

- Multiple theories proposed regarding etiology behind tumor formation
- Proposed mechanisms remain theories
Congenital Cholesteatoma

- **Definition (Levenson, 1989)**
  - White mass medial to normal tympanic membrane
  - Normal pars flaccida and pars tensa
  - No prior history of otorrhea or perforations
  - No prior otologic procedures
  - Prior bouts of otitis media were not grounds for exclusion as was the case in original definition
Congenital Cholesteatoma

- Pathogenesis theories
  - Failure of involution of ectodermal epithelial thickening that is present during fetal development in proximity to geniculate ganglion
  - Metaplasia of the middle ear mucosa
Congenital Cholesteatoma

Anterosuperior quadrant > Posterosuperior quadrant
Congenital Cholesteatoma

Large congenital cholesteatoma

ossicular erosion

cholesteatoma
Acquired Cholesteatomas

- Common factor: keratinizing squamous epithelium has grown beyond its normal limits
Primary Acquired Cholesteatomas

- Ultimately form due to underlying Eustachian tube dysfunction that causes retraction of pars flaccida
  - Results in poor aeration of epitympanic space which draws pars flaccida medially on top of malleus neck, forming retraction pocket
  - Normal migratory pattern of the tympanic membrane epithelium altered by retraction pocket
  - Enhances potential accumulation of keratin
Primary Acquired Cholesteatomas

Pars flaccida retraction

Pars tensa retraction
Secondary Acquired Cholesteatomas

- **Implantation theory**
  Squamous epithelium implanted in the middle ear as a result of surgery, foreign body, blast injury, etc.

- **Metaplasia theory**
  Desquamated epithelium is transformed to keratinized stratified squamous epithelium secondary to chronic or recurrent otitis media

- **Epithelial invasion theory**
  Squamous epithelium migrates along perforation edge medially along undersurface of tympanic membrane destroying the columnar epithelium

- **Papillary ingrowth theory**
  Inflammatory reaction in Prussack’s space with an intact pars flaccida (likely secondary to poor ventilation) may cause break in basal membrane allowing cord of epithelial cells to start inward proliferation
Anatomy
Middle Ear Regions

- Named based on position relative to superior and inferior aspect of external auditory canal (EAC)
  - Epitympanum: superior to superior limit of EAC
  - Mesotympanum: bound superiorly by superior limit of EAC and inferiorly by inferior limit of EAC
  - Hypotympanum: inferior to inferior limit of EAC
Middle Ear Regions

- Attic (Epitympanum)
- Incus
- Tympanic Membrane
- Oval Window
- Round Window
- Stapes
Epitympanum

- Lies above the level of the short process of the malleus

- Contents:
  - Head of the malleus
  - Body of the incus
  - Associated ligaments and mucosal folds
Mesotympanum

- **Contents:**
  - Stapes
  - Long process of the incus
  - Handle of the malleus
  - Oval and round windows

- **Eustachian tube exits from the anterior aspect**

- **Two recesses extend posteriorly that are often not visible directly**
  - Facial recess
    - Lateral to facial nerve
    - Bounded by the fossa incudis superiorly
    - Bounded by the chorda tympani nerve laterally
  - Sinus tympani
    - Lies between the facial nerve and the medial wall of the mesotympanum
Hypotympanum

- Lies inferior and medial to the floor of the bony ear canal
- Irregular bony groove that is seldom involved by cholesteatoma
Anatomic Considerations

- Annular ligament sends fibrous bands from anterior and posterior tympanic spines that meet at neck of malleus and make up middle layer of pars tensa.
- Dehiscent area in the tympanic bone (Notch of Rivinus) lies above fibrous bands.
- Dense fibers that form middle layer of pars tensa do not extend to pars flaccida.
  - Lack of structural support predisposes Shrapnell’s membrane to retract when negative middle ear pressure is present secondary to Eustachian tube dysfunction.
Anatomic Considerations

- Cholesteatomas of epitympanum start in Prussack’s space between pars flaccida and neck of malleus with upper boundary being the lateral malleal fold
Cholesteatoma Spread

- Predictable in that they are channeled along characteristic pathways by:
  - Ligaments
  - Folds
  - Ossicles
Common Sites of Cholesteatoma

Origin

- Posterior epitympanum
- Posterior mesotympanum
- Anterior epitympanum
Cholesteatoma Spread

- Posterior epitympanic cholesteatoma passing through superior incudal space and aditus ad antrum
Cholesteatoma Spread

- Posterior mesotympanic cholesteatoma invading the sinus tympani and facial recess
Cholesteatoma Spread

- Anterior epitympanic cholesteatoma with extension to geniculate ganglion
Patient Evaluation
Patient Evaluation

- **History**
  - **Detailed otologic history**
    - Hearing loss
    - Otorrhea
    - Otalgia
    - Nasal obstruction
    - Tinnitus
    - Vertigo
  - **Previous history of middle ear disease**
    - Chronic otitis media
    - Tympanic membrane perforation
    - Prior surgery
Patient Evaluation

- Head and neck examination
- Otologic examination
  - Otomicroscopy is essential in evaluating the extent of disease
  - Clean ear thoroughly of otorrhea and debris with cotton-tipped applicators or suction
  - Culture wet, infected ears and treat with topical and/or oral antibiotics
  - Pneumatic otoscopy should be performed in every patient with cholesteatoma
    - Positive fistula (pneumatic otoscopy will result in nystagmus and vertigo) response suggests erosion of the semicircular canals or cochlea
Patient Evaluation

- **Hearing evaluation to assess for conductive hearing loss**
  - Pure tone audiometry with air and bone conduction
  - Speech reception thresholds
  - Word recognition

- **512Hz tuning fork exam**
  - Always correlate with audiometry results

- **Tympanometry**
  - May suggest decreased compliance or TM perforation
Patient Evaluation

- Degree of conductive loss will vary considerably depending on the extent of disease
  - 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
  - Mild conductive deafness may be present with extensive disease if cholesteatoma sac transmits sound directly to stapes or footplate
Patient Evaluation

- Preoperative imaging with computed tomographies (CTs) of temporal bones (2mm - section without contrast in axial and coronal planes)
  - Allows for evaluation of anatomy
  - May reveal evidence of the extent of the disease
  - Screen for asymptomatic complications
Patient Evaluation

- CT is not essential for preoperative evaluation
- Should be obtained for:
  - Revision cases due to altered landmarks from previous surgery
  - Chronic suppurative otitis media
  - Suspected congenital abnormalities
  - Cases of cholesteatoma in which sensorineural hearing loss, vestibular symptoms, or other complication evidence exists
Patient Evaluation

- Preoperative counseling is an absolute necessity prior to surgery
- 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
  - Preserving as much normal anatomy as possible
- Improvement of hearing is a secondary goal
Patient Evaluation

- Possible adverse outcomes must be discussed
  - Facial paralysis
  - Vertigo
  - Further hearing loss
  - Tinnitus

- Patient should understand that long-term follow-up will be necessary and that they may need additional surgeries
Cholesteatoma Management
Preventative Management

- Tympanostomy tube for early retraction pockets
- Surgical exploration for retraction persistence
Cholesteatoma Management

- Treated surgically with primary goal of total eradication of cholesteatoma to obtain a safe and dry ear
- Patients with unacceptable risk of anesthesia need local care
**Surgical Management**

- Canal-wall-down procedures (CWD)
- Canal-wall-up procedure (CWU)
- Transcanal anterior atticotomy
- Bondy modified radical procedure
Canal-Wall-Down

- Prior to the advent of the tympanoplasty, all cholesteatoma surgery was performed using CWD approach

- Procedure involves:
  - Taking down posterior canal wall to level of vertical facial nerve
  - Exteriorizing the mastoid into external auditory canal

- Epitympanum is obliterated with removal of scutum, head of malleus and incus
Canal-Wall-Down

- Classic CWD operation is the modified radical mastoidectomy in which middle ear space is preserved.

- Radical mastoidectomy is CWD operation in which:
  - Middle ear space is eliminated
  - Eustachian tube is plugged

- Meatoplasty should be large enough to allow good aeration of mastoid cavity and permit easy visualization to facilitate postoperative care and self cleaning.
Canal-Wall-Down

Indications for CWD approach:

- 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
- Sclerotic mastoid with limited access to epitympanum
Canal-Wall-Down

- **Advantages:**
  - Residual disease is easily detected
  - Recurrent disease is rare
  - Facial recess is exteriorized

- **Disadvantages:**
  - Open cavity created
    - Takes longer to heal
  - Mastoid bowl maintenance can be a lifelong problem
  - Shallow middle ear space makes OCR difficult
  - Dry ear precautions are essential
Canal-Wall-Down

- Posterior canal wall
- Fossa incisus
- Zygomatic root
- Mastoid tip
- Sigmoid sinus
- Horizontal semicircular canal
- Sinodural angle
- Tegmen
Canal-Wall-Down
Canal-Wall-Down
Canal-Wall-Down

Articular surface

Anterior buttress

Stapes

Tegmen
Canal-Wall-Down

- Eustachian tube
- Pyramidal process
- Cochleariform process
- Facial ridge
- Horizontal semicircular canal
Canal-Wall-Up

- CWU procedure developed to avoid problems and maintenance necessary with CWD procedures
- CWU consists of preservation of posterior bony external auditory canal wall during simple mastoidectomy with or without a posterior tympanotomy
- Staged procedure often necessary with a scheduled second look operation at 6 to 18 months for:
  - Removal of residual cholesteatoma
  - Ossicular chain reconstruction if necessary
- Procedure should be adapted to extent of disease as well as skill of otologist
Canal-Wall-Up

- CWU may be indicated in patients with large pneumatized mastoid and well aerated middle ear space
  - Suggests good eustachian tube function
- CWU procedures are contraindicated in:
  - Only hearing ear
  - Patients with labyrinthine fistula
  - Long-standing ear disease
  - Poor eustachian tube function
Canal-Wall-Up

- **Advantages:**
  - Rapid healing time
  - Easier long-term care
  - Hearing aids easier to fit
  - No water precautions

- **Disadvantages:**
  - Technically more difficult
  - Staged operation often necessary
  - Recurrent disease possible
  - Residual disease harder to detect
Canal-Wall-Up
Transcanal Anterior Atticotomy

- Indicated for limited cholesteatoma involving middle ear, ossicular chain, and epitympanum
- If extent of the cholesteatoma is unknown approach can be combined with CWU mastoidectomy or extended to CWD procedure
Transcanal Anterior Atticotomy

- Procedure involves:
  - Elevation of tympanomeatal flap via endaural incision with removal of scutum to limits of the cholesteatoma
  - Aditus obliteration with muscle, fascia, cartilage or bone prior to reconstruction of the middle ear space
  - Reconstruction of lateral attic wall with bone or cartilage is optional
    - May lead to retraction disease and possible recurrence in patients with poor eustachian tube function
Transcanal Anterior Atticotomy
Bondy Modified Radical Procedure

- Useful for attic and mastoid cholesteatoma that does not involve middle ear space and lateral to ossicles
- Like modern modified radical mastoidectomy with exception that middle ear space is not entered
- Mastoid should be poorly developed for creation of a small cavity
- Eustachian tube function should be adequate with intact pars tensa and aerated middle ear space.
- Rarely used today
Canal Wall Status

- In 2002 Shohet et. al. reports recidivism rates with CWU as high as
  - Adults = 36%
  - Children = 67%
- Approximately 30% of cases for cholesteatoma will be CWD
  - House Ear Institute 1982
  - St. Joseph’s Hospital in Phoenix, AZ 2003
Canal Wall Status

- Syms et al. examined surgical approach to manage cholesteatoma- retrospective review of 486 cases in 2003
  - 68.5% CWU: 31.5% CWD
  - CWU had “second look operation”
    - Residual cholesteatoma found in 26.9% second procedures
    - Residual cholesteatoma found in 2.7% third procedures
Canal Wall Status

- Kos et. al. examined long-term implications of CWD- retrospective review in 2004
  - 259 cases with follow-up range of 1 to 24 years (mean = 7 years)
  - Recurrent cholesteatoma in 6.1%
  - TM perforation in 7.3%
  - Dry, self-cleaning ears in 95%
  - Otorrhea in 5%
Canal Wall Status

- **Kos et. al. (continued)**
  - Hearing threshold
    - Improved in 30.7%
    - Unchanged in 41.3%
    - Decreased in 28%
  - Sensorineural hearing loss greater than 60dB in 2 patients
  - Facial paralysis in 1 patient
  - Vertigo in 4 patients
In 2005 Gantz et. al. reported 130 cases of canal wall reconstruction tympanomastoidectomy with mastoid obliteration

- No evidence of recurrence = 98.5%
- Recurrence treated with CWD (1.5%)
- Second look ossiculoplasty in 78%
- Post-operative wound infection was 14.3% for first 42 patients
  - Decreased rate to 4.5% in last 88 patients with 2 days of post-operative IV antibiotics
Novel Techniques

- Canal Wall Reconstruction technique
  - Complete cortical mastoidectomy with opening of facial recess and removal of incus and malleus head
  - Posterior canal wall skin elevated, annulus elevated
  - Microsagittal saw used to cut posterior canal wall
  - Cholesteatoma removed
  - Posterior canal wall bone replaced
    - Cortical bone chips used to block attic and mastoid from tympanum
    - Bone pate’ holds bone chips in place
Novel Techniques
Novel Techniques

- In 2005, Godinho et. al. reported 42 cases of Canal Wall Window (CWW) performed for pediatric cholesteatoma
  - CWW compared to CWU and CWD
    - CWW converted to CWD in 14%
    - Dry ear results
      - CWW = 94%, CWD = 92%, CWU = 90%
    - Recidivation rate at 1 year
      - CWW = 19.5%, CWD = 0%, CWU = 7.7%
    - CWW provided similar post-operative hearing to CWU, rather than increased air-bone gap seen with CWD
Novel Techniques

- **CWW effective for posterior superior cholesteatoma**
  - Increases visibility
  - Conduit for instrument manipulation

- **CWW technique**
  - CWU mastoidectomy performed
  - Lateral epitympanotomy
  - Slit drilled from lateral cortex to medial aspect of canal wall at annulus
Novel Techniques
Novel Techniques

- **CWW reconstruction**
  - Tragal cartilage placed in defect after scoring proximal perichondrium and cartilage
  - Cortical bone graft may be used as an alternative
Novel Techniques
Complications
Cholesteatoma Sequelae

- Infection
- Otorrhea
- Bone destruction
- Hearing loss
- Facial nerve paresis or paralysis
- Labyrinthine fistula
- Intracranial complications

Complications are caused by expansion and infection
Hearing Loss

- **Conductive hearing loss: ossicular chain erosion (30%)**
  - Erosion of lenticular process and/or stapes superstructure may produce 50dB conductive hearing loss
  - Hearing loss varies despite disease extent (natural myringostapediopexy, transmission of sound through cholesteatoma sac)

- **Sensorineural hearing loss: involvement of labyrinth**

- Following surgery, 30% have further impairment due to:
  - Extent of disease present
  - Complications in healing process
Labyrinthine Fistula

- Incidence: as high as 10%
- Symptom: Sensorineural hearing loss and/or vertigo induced by noise or pressure change
- Common site: horizontal semicircular canal, basal turn of cochlea
- Diagnosis: Fine cut temporal bone CT (1mm)
- Management: modified radical mastoidectomy with management of matrix overlying fistula
Facial Paralysis

- May develop:
  - Acutely secondary to infection
  - Slowly from chronic expansion of cholesteatoma
- Temporal bone CT: localize the nerve involvement
- Most common site: geniculate ganglion due to disease in the anterior epitympanum
- Management: Needs immediate surgery
  - Removal of cholesteatoma and infected material with decompression of the nerve (mastoidectomy, middle fossa approach)
  - Administration of intravenous antibiotics and high-dose steroids
  - Iatrogenic injury to the nerve during surgery should be immediately repaired with decompression of nerve proximal and distal to site of injury
Intracranial Complications

- Potentially life-threatening
- Incidence: as high as 1%
- Complications
  - Periosteal abscess
  - Lateral sinus thrombosis
  - Intracranial abscess
  - Meningitis
- Symptom:
  - Suppurative malodorous otorrhea
  - Chronic headache
  - Fever
  - Otalgia
**Intracranial Complications**

**Management:**

- 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

- Pathogenesis of cholesteatoma remains uncertain
- Essential to possess basic knowledge of the important anatomic and functional characteristics of the middle ear for successful management of cholesteatomas
- Careful and thorough evaluations are the key to early diagnosis and treatment
- Treatment is surgical with primary goal to eradicate disease and provide a safe and dry ear
- Surgical approaches must be customized to each patient depending on extent of disease
- Surgeon must be aware of serious and potentially life-threatening complications of cholesteatomas
Bibliography

- Portions of this paper and presentation were taken directly from the June 18, 2003 Grand Rounds presentation by Michael Underbrink and Arun Gadre entitled Cholesteatoma.
What To Do

- Treatment should be tailored to case
- **Absolute CWD indications**
  - 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
  - Sclerotic mastoid with limited access to epitympanum
- **No harm in starting CWU and converting to CWD as necessary**
- **Know your surgical limitations**
Quiz
Quiz

1. What space lies between the pars flaccida and #13?
   - A. Prussak’s space
   - B. Anterior pouch of Von Troeltsch
   - C. Posterior pouch of Von Troeltsch
   - D. The final frontier
2. The primary component of cholesteatoma is fat, as discovered by Johannes Mueller in 1838.

A. True

B. False
Quiz

3. What 2 major groups are cholesteatomas classified into (pick 2 letters)?

- A. Acquired
- B. Infected
- C. Expansive
- D. Congenital
4. Possible adverse outcomes of surgical treatment of cholesteatomas are

- A. Facial paralysis
- B. Vertigo
- C. Further hearing loss
- D. Tinnitus
- E. All of the above
5. CT Scans must be obtained prior to any surgery for cholesteatoma

- A. True
- B. False
- C. It depends on who your faculty is
Quiz

6. CWU procedures are contraindicated in

A. Only hearing ears
B. Patients with a labyrinthine fistula
C. Long-standing ear disease
D. Poor eustachian tube function
E. All of the above
7. Surgical treatment of cholesteatoma has a primary goal of hearing restoration

- A. True
- B. False
8. Surgical approaches must be customized to each patient depending on extent of disease

- A. True
- B. False
Quiz Bonus

What is this a picture of?

- A. Cat eye
- B. Snake skin
- C. Tympanic membrane stained with osmium to show keratin patches