Update on Cholesteatoma

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Introduction

- Otologic disorders long recognized as cause significant morbidity and mortality

- “Acute pain of the ear, with continued high fever, is to be dreaded, for the patient may become delirious and die.”

Hipocrates ~ 460 B.C.
Cholesteatoma

- Problem
- Anatomy
- Definitions & Pathology
- Treatment
Problem

- Cholesteatoma
  - Squamous epithelium & keratin debris trapped w/in skull base
    - Expansile process
      - Resulting in various intra-temporal and extra-temporal complications & sequela
Anatomy

Temporal bone

Tympanic
Petrουs
Mastoid
Squamouus
Zygomatic Process
Styloid process

Middle Ear Regions

- Named based on position relative to superior and inferior aspect of EAC
Epitympanum

**Location**
- Above short process malleus & horizontal facial n.

**Contents**
- Head malleus, body incus, assoc ligaments & mucosal folds
- Prussak’s Space

**Bounded laterally**
- Pars flaccida
  - Loosely organized fibrous middle layer
Epitympanum

- Communicates
  - Posteriorly
    - Via additus ad antrum → central mastoid cell tract
  - Inferiorly
    - With protympanum & mesotympanum (anterior & posterior pouch of Von Troeltsch)
Mesotympanum

- Two recesses – often not directly visible
  - **Facial recess**
    - Lateral to facial n
    - Bounded by fossa incudis superiorly & chorda tympani nerve laterally/anteriorly, facial nerve medially/posteriorly

- **Sinus tympani**
  - Lies btw:
    - Facial nerve & medial wall of mesotympanum
    - Ponticulus (superior) and subiculum (inferior)
Pro-tympanum

- Anterior portion of middle ear space
  - Location
    - Anterior to mesotympanum
  - Bordered
    - Superiorly
      - Opening for Eustachain tube
    - Anterioly/inferiorly
      - Carotid artery/bony canal

- Eustachian tube exits from superior/anterior aspect
- Communicates w/ epitympanum superiorly
Hypotympanum

- **Location**
  - Lies inferior & medial to floor of bony EAC and Mesotympanum

- Occasionally involved w/ cholesteatoma
Eustachian Tube

Function(s)

- Pressure equilibration
  - Middle ear & mastoid

- Clearance
  - Material from middle ear

- Prevention of reflux
  - Material or sound from nasopharynx
Cholesteatoma
“Cholesteatoma”

Johannes Müller (1838)

- Erroneous belief that one of primary components of tumor was fat
- “…a pearly tumor of fat...among sheets of polyhedral cells”

More appropriate name “Keratoma”

Cholesteatoma

Expansile lesion composed of:

- **Cystic**
  - Desquamated keratin center

- **Epithelial (matrix)**
  - Keratinizing stratified squamous epithelium

- **Subepithelial component (peri-matrix)**
  - Collagen, elastin, reticulin, fibroblasts, & granulation tissue in contact w/ bone
  - Secretes multiple proteolytic enzymes capable of bone destruction - ?biofilms and role of *Pseudomonas* spp.?
Cholesteatoma

- May develop anywhere within pneumatized portions of temporal bone and skull base

  - Most frequently
    - Middle ear
    - Mastoid
**Cholesteatoma**

- **Expansile**
  - Accelerated by moisture infection/inflammation

  - Erode adjacent bone
    - **Mechanical pressure effect**
    - **Biochemical elements**
      - Bacterial elements (*P. aeruginosa*), host granulation tissue & cholesteatoma produced substances
    - **Cellular osteoclast mediated enzyme activity**
      - Collagenase abundant in epidermis of cholesteatoma → stimulates osteoclast activity
Cholesteatoma

- Bone erosion by cholesteatoma
  - Leads to:
    - Destruction of mastoid trabeculae
    - Ossicular erosion
    - Labyrinthine fistula
    - Exposure of dura, facial nerve & dural venous sinus
Pathology

- Molecular models
  - Pre-neoplastic or neoplastic transformation
  - Defective wound-healing
  - Conflict btw
    - Host inflammatory response
    - Normal middle ear epithelium
    - Bacterial infection
Classification

- Congenital
- Acquired
  - Primary acquired (retraction pocket)
  - Secondary acquired (through TM perf)
Congenital Cholesteatoma

Definition (Levenson, 1989)
- White mass medial to normal TM
- Normal pars flaccida and pars tensa
- No prior history of otorrhea or perforations
- No prior otologic procedures
- Prior bouts of OM not grounds for exclusion – was case in original definition
Congenital Cholesteatoma

- Pathogenesis (4 theories)

- Implantation, Invagination/Invasion, Metaplasia, Epithelial Rests

- Failure of involution of ectodermal epithelium present during fetal development – Remark (1857), Teed (1936), Michales (1986)
  - Anterior/superior > Posterior /superior
Acquired Cholesteatoma

- **Invagination (Retraction)** – Bezold (1878, ‘89)
  - ET dysfunction & chronic “-” ME pressures →
    - Retraction of pars flaccida/pars tensa → retraction pocket
    - Normal epithelial migratory pattern altered →
    - Accumulation of keratin → enlargement of sac

- **Epithelial invasion/migration** – Bezold (1899), Habermann (1899), Karmondy (2011)
  - Epithelial in-growth through TM perforation
Acquired Cholesteatomas

- **Implantation theory**
  - Squamous epithelium implanted in ME as result of surgery, foreign body, blast injury, etc.

- **Metaplasia theory** – Wendt (1873)
  - ME mucosal epithelium transformed to keratinized stratified squamous epithelium secondary to chronic or recurrent inflammation/infection – eg. OM
Secondary Acquired Cholesteatoma

- **Basal cell hyperplasia/Papillary in-growth theory** – Lange (1925), Ruedi (1957)
  - Intact, retracted TM (secondary to poor ventilation)
  - Inflammatory reaction in Prussack’s space
  - Break in basal membrane
  - Cone-like invasion of basal cells in epithelium/cord of epithelial cells start inward proliferation as a result of inflammation
Cholesteatoma Spread

- Predictable spread along characteristic pathways influenced by anatomy
  - Ligaments
  - Folds
  - Ossicles
Common Sites of Origin – Acquired Cholesteatoma

1. Prussak’s Space
   - D/t pars flacida retraction
     - Posterior epitympanum (#1) (atticoantral)
     - Anterior epitympanum (#3)

2. Posterior mesotympanum (#2)
   - D/t pars tensa retraction (tubotympanic)
Prussack’s Space

- **Posterior**
  - Retraction along saccus medius w/ expansion posteriorly to superior incudal space → aditus & mastoid

- **Inferior**
  - Into post pouch of von Troltsch btw TM & post malleolar fold → post mesotympanum (OW, facial recess, sinus tympani)

- **Anterior**
  - Into ant pouch of von Trolsch btw TM & ant malleolar fold → ant mesotympanum (ET, etc.)
  - Superior → supratubal recess, root zygoma, middle fossa, petrous apex, etc.
Patient Evaluation

- **Pre-OP imaging – CT temporal bones**
  - Allows for evaluation of anatomy
  - May reveal evidence of extent of disease
  - Screen for asymptomatic complications
  - If suspect complications → consider contrast
Pre-OP CT Temporal Bones

Not essential for pre-OP evaluation

May/should consider for:
- Revision cases
- Chronic suppurative OM
- Suspected congenital abnormalities
- Cholesteatoma w/ SNHL, vestibular symptoms, severe pain, facial paralysis or other complication –
  - MRI of IACs w/ w/o contrast – complimentary for cases of complication/meningoencephalocele, monitoring for recurrence (DWI), etc.
Management

- Medical
  - Patients w/ unacceptable anesthesia risks
  - Aural toilet, antibiotic drops, control granulation tissue, local care/debridement
Surgical Management

- **Pre-OP counseling**
- **Primary objective**
  - “Safe, dry ear”
    - Remove/eliminate disease
      - Bone, mucosa, granulation polyps, and cholesteatoma
    - Treat/prevent complications
    - Preserve/restore/alter anatomy
      - Prevent recurrence & optimize subsequent cleaning & monitoring of ear (minimize post-op care)
- **Secondary objective**
  - Preserve or Improve hearing
    - Reconstruction of TM and ossicular system
**Pre-OP Counseling**

- **Possible adverse outcomes**
  - Facial paralysis
  - Vertigo
  - Further hearing loss
  - Tinnitus
  - CSF leak/meningitis
  - Bleeding/infection/cosmetic changes ear

- **Long-term follow-up**
  - Possible or planned additional surgeries
    - **Recurrent or residual disease**
  - Planned “Second-look”
**Recurrent and Residual Disease**

**Residual Disease**
- Disease left behind
  - Intentional or unintentional
- Un-intentional residual disease
  - Results from failure to remove all disease at initial procedure

**Recurrent Disease**
- Disease that recurs despite complete removal
  - Results from failure to alter or reconstruct anatomy of ear to facilitate cleaning of ear and prevention of recurrent disease
Surgical Management

- No single procedure for treatment of all cases cholesteatoma

- “Controversies”
  - Endoscopic middle ear surgery – cholesteatoma
  - Facial nerve monitor
Surgical Treatment

- Tympanoplasty
  - +/- atticotomy

- Intact-canal-wall (CWU/CWI/closed cavity)
  - ICW T-mast w/ w/o atticotomy, T-mast/antrotomy, T-mast reconstructed CW

- Canal-wall-down (open cavity)
  - Modified radical CWD T – mast
    - Bondy modified radical mastoidectomy procedure
  - Radical CWD T-mast
Cholesteatoma

DETERMINANTS OF OPERATIVE TECHNIQUE

Local factors
- Presence of labyrinthine or cochlear fistula
- Extent of disease
- Eustachian tube function
- Mastoid pneumatization
- Hearing status of both ears

General factors
- General medical condition
- Occupation
- Reliability

Skill and experience of surgeon
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<th>Relative Indications</th>
<th>Relative Contraindications</th>
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<td>Mucopurulent drainage</td>
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<td>Symptomatic retraction pocket</td>
<td>Radiographic evidence of significant mastoid involvement</td>
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<td>Chronic inactive otitis media with frequent reactivation</td>
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<td>Well-pneumatized mastoid</td>
<td>Extensive granulomatous disease or large cholesteatoma</td>
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<td>Clinical evidence for good eustachian function</td>
<td>Sclerotic, poorly pneumatized mastoid</td>
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<td>Ability to preserve tympanic membrane and ossicular chain anticipated</td>
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<tr>
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<td>Extensive disease</td>
<td>Disease limited to attic or antrum</td>
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<td>Poorly pneumatized mastoid</td>
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<td>Presence of complications (e.g., labyrinthine fistula)</td>
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<td>Clinical evidence of poor tubal function</td>
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<td>History of previous failure of canal wall–up mastoidectomy for chronic active otitis media</td>
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Atticotomy

Indications

- Limited retraction pocket cholesteatoma
  - Middle ear (epitympanum, mesotympanum), lateral to ossicular chain
- If extent of cholesteatoma is unknown, approach can be combined with CWI mastoidectomy or extended to CWD procedure
Atticotomy

Procedure

- Elevation of tympanomeatal flap via endaural/transcanal incision +/- removal of scutum to limits of cholesteatoma
- Removal ME adhesions & retraction pocket/cholesteatoma
- Aditus obliteration w/ muscle, fascia, cartilage or bone prior to reconstruction of the middle ear space
- Reconstruction of lateral attic wall/scutum w/ bone or cartilage is optional
  - If not done may lead to retraction disease and possible recurrence in patients w/ poor eustachian tube function
Atticotomy
Mastoid Surgery

- **Mastoidectomy**
  - **Cortical**
    - When combined w/ tympanoplasty → Intact canal wall (ICW / CWI / “Closed”/ canal wall up tympanomastoidectomy)
  - **Modified radical**
    - When combined w/ tympanoplasty → Canal wall down (CWD / “Open” / canal wall down tympanomastoidectomy)
  - **Radical**
  - **Radical w/ petrosectomy**
Intact-canal wall procedures

- Preservation of posterior EAC wall
- Single stage vs 2\textsuperscript{nd} staged procedure (6 – 12 months)

Contraindications
- Only hearing ear
  - Some authors recommend CWD only for only hearing ear
- Labyrinthine fistula
- Long-standing ear disease, ETD
  - Higher residual & recurrence rate
Intact-canal wall procedures
Intact-canal-wall (ICW)

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

- **Disadvantages**
  - Technically more difficult
  - **Recurrent disease** possible
  - Staged operation may be necessary
  - **Residual disease** harder to detect
ICW

- **Indicated**
  - In patients w/ large pneumatized mastoid and well aerated middle ear space
    - Suggests good eustachian tube function
  - Extensive TM retraction or widespread cholesteatoma

- **Contraindicated**
  - Only hearing ear
  - Patients w/ labyrinthine fistula
  - Long-standing ear disease
    - Failed 2\textsuperscript{nd} look
  - Poor eustachian tube function
Canal-wall-down-down procedures

- Prior to mid/late 1950’s, all cholesteatoma surgery was performed using CWD approach
- Exteriorize mastoid into external ear canal by taking down posterior canal wall (to level of vertical portion facial n.)
  - Modified radical mastoidectomy (classic CWD)
    - Middle ear space preserved, +/- OSCR
  - Radical mastoidectomy
    - Middle ear space eliminated, Eustachian tube orifice obliterated
Indications

- Cholesteatoma in an only hearing ear
- Significant erosion of posterior EAC wall
- Contracted mastoid
- Labyrinthine fistula
- Sclerotic mastoid w/ limited access to epitympanum
- Recurrent cholesteatoma following ICW surgery
- Prolonged ETD/poor ET function
Canal-Wall-Down

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

**Disadvantages**
- Open cavity created
  - Longer healing time
- Mastoid bowl maintenance can be lifelong problem
- Shallow middle ear space makes OCR difficult
- Dry ear precautions are necessary
Canal-wall-down
Canal-wall-down

Pitfalls

- Narrow meatus
  - Meatoplasty
    - Should be large enough to allow good aeration of mastoid cavity and permit easy visualization to facilitate postoperative care and self-cleaning

- Insufficient bone removal
  - High facial ridge
  - Overhanging edge(es)
  - Prominent mastoid tip
Canal-wall-down

Transcortical
- Simple mastoidectomy & atticoantrotomy completed from posterior cavity as in CWI (first complete CWI)
- Thinned posterior wall then removed

Transmeatal
- 1st identify Tegmen/middle fossa plate
- Enlarge EAC by following middle fossa plate posteriorly toward SDA
  - ID sigmoid sinus
- Mastoid cavity opened from ant to posterior

Meatoplasty
Reconstruction
Bondy & Modified Radical Procedure

- In pt w/ good hearing, intact OSC & TM w/ attic +/- mastoid disease
  - Cholesteatoma located laterally to chain typically epitympanum
    - Not involving middle ear space
  - Requires good Eustachian tube function and intact pars tensa
  - Good or better hearing in diseased ear

- Post EAC wall removed + mastoid (CWD)
  - Cholesteatoma marsupialized
  - OSC maintained
  - Wide meatoplasty
Radical Mastoidectomy

- Complete removal of ME (including sound transmission sys except stapes) & eradication of tubal function (+/- mastoid tip amputation)

- Indicated
  - Elderly pts w/ pre-op dead ear or non-serviceable hearing in which only goal dry/safe ear
  - Cochlear fistula
  - (Recurrent) ME cholesteatoma in deep sinus tympani
  - Cholesteatoma w/ intractable complications
  - Benign tumors of ME/mastoid w/ severe SNHL
Complications

- **Intratemporal**
  - Infection, otorrhea, bony destruction (OSC erosion, etc) petrositis, necrotizing otitis externa/osteomyelitis, facial paralysis, labyrinthitis (serous/supportive), labyrinthine fistula, cochlear fistula

- **Extratemporal**
  - Intracranial
    - Lateral sinus thrombophlebitis, meningitis, intracranial abscess (extradural, subdural, or parenchymal)
  - Extracranial
    - Subperiosteal abscess, neck (Bezold’s) abscess
Sequelae

- Hearing loss (Conductive—ossicular chain necrosis/disruption, SNHL, or mixed), recurrent cholesteatoma, & tympanosclerosis
Conclusions

- Knowledge of exact mechanism of pathogenesis not certain
- Knowledge of anatomy, physiology of the ear and pathology influence treatment
- Careful and thorough evaluation
- Primary goal of surgery: “safe, dry ear”
- Surgical strategies vary
- Complications can be life-threatening
What To Do

- Treatment should be tailored to patient
- CWD indications
  - Cholesteatoma in an only hearing ear
  - Significant erosion of posterior bony canal wall
  - History of vertigo suggesting a labyrinthine fistula
  - Recurrent cholesteatoma after CWI surgery
  - Poor eustachian tube function
  - Sclerotic mastoid w/ limited access to epitympanum
- No harm in starting CWI and converting to CWD
- Know your limitations
Bibliography