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.
Outline

- Case Presentation
- Anatomy
- Definitions & Pathology
- Treatment
Case Presentation

HPI

51 y/o female Hx “right ear surgery” 11/29/2011 for cholesteatoma and meningoencephalocele

Presents 09/05/2012 w/ c/o AD non-fluctuating, non-progressive hearing loss present since initial surgery - unchanged
  ● Denies otorrhea, otalgia or vertigo

Pt had been scheduled for “2nd look” and staged OCR but had not proceeded w/ surgery
PMH/PSH

PSH

- AD T-mast CWI/Reconstruction w/o OCR w/ TM cartilage graft w/ repair of meningoencephalocele (middle fossa craniotomy)
  - AD cholesteatoma, Incus & Malleus Erosion – (removed), meningoencephalocele (middle fossa craniotomy – bone repair tegmen defect)
  - Planned – 2nd look T-plasty w/ OCR by primary surgeon – pt did not undergo procedure

- T&A, hysterectomy, appendectomy, hip

PMH

- Asthma, OSAS, DM II, HTN, Depression
Physical Exam

HENT:

AD: EAC w/o mass/lesion or abnormality, TM intact, thickened graft, unable to visualize through TM (suspect cartilage graft beneath TM), poor movement to pneumatotonscopy

AS: EAC unremarkable, TM intact, ME clear

Tuning fork: 512 Hz

- AD – BC > AC
- AS – AC > BC
- Webber lateralized right
Last audiogram May 2011 *
Speech Reception Threshold and Word Recognition: live voice
Right Ear: 40 dB HL with 100 % word recognition at 75 dB HL with 45 dB masking
Left Ear: 5 dB HL with 100 % word recognition at 40 dB HL with no dB masking
TEMPORAL SQUAMA

- Sulcus for middle temporal artery
- Articular tubercle
- Zygomatic process
- Mandibular fossa
- Petrotypanic fissure
- External acoustic meatus
- Arterial sulcus
- Superior petrosal sulcus
- Arcuate eminence
- Cochlear canalicus (External aperture)
- Jugular fossa
- Sigmoid sulcus
- Vestibular aqueduct
- Cochlear canalicus (internal aperture)
- Sigmoid process
- Styloid process

**Anatomy**

**Temporal bone**
- Tympanic
- Petrous
- Mastoid
- Squamous
- 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 canal 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 (posterior & anterior 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

- Irregular bony groove 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” 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 of temporal bone 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

- Most frequently
  - Middle ear
  - Mastoid
Cholesteatoma

- Expansile desquamated keratin debris
  - Moisture & infection/inflammation accelerate process

  - 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
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 as was the case in original definition
Congenital Cholesteatoma

Pathogenesis (3 theories)

- Failure of involution of ectodermal epithelium present during fetal development – Remark (1857), Michales (1986)
  - Anterior/superior > Posterior /superior
- Arise from embryonal rests of epithelial cells
  - Location(s)
    - Petrous pyramid, mastoid and middle ear cleft
- Metaplasia of ME mucosa
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 thru TM perf
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:
  - Ligaments
  - Folds
  - Ossicles
Common Sites of Origin - Cholesteatoma

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

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

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

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

- **Anterior**
  - Into ant pouch of von Trolsch btw TM & ant malleolar fold $\rightarrow$ ant mesotympanum (ET, etc.)
  - Superior $\rightarrow$ 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 goal**
  - 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”
Surgical Management

- No single procedure for treatment of all cases cholesteatoma

- “Controversies”
  - Endoscopic middle ear surgery - cholesteatoma
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
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 Contraindications</th>
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<td>Limited attic disease</td>
<td>Mucopurulent drainage</td>
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<td>Symptomatic retraction pocket</td>
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<td>Chronic inactive otitis media with frequent reactivation</td>
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<td>Limited atticcoantral disease</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>
<td>Canal wall–down mastoidectomy</td>
<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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Transcanal anterior 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 CWU mastoidectomy or extended to CWD procedure
Transcanal Anterior 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
Transcanal anterior atticotomy
Mastoid Surgery

- Mastoidectomy
  - Atticotomy
  - 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 with 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 with labyrinthine fistula
  - Long-standing ear disease
    - Failed 2nd 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
Canal-wall-down

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
Hearing Loss

- Conductive hearing loss common
  - Ossicular chain erosion (30%)
    - Erosion of lenticular process and/or stapes superstructure may produce 50dB conductive hearing loss (60 dB maximal)
    - Hearing loss varies despite disease extent (natural myringostapediopexy, transmission of sound through cholesteatoma sac)

- Sensorineural hearing loss
  - May indicate involvement of labyrinth

- Following surgery, 30% have further impairment of hearing d/t
  - Extent of disease present
  - Complications in healing process
  - Up to 3% total hearing loss post surgical
Labyrinthine fistula

- Incidence as high as 10% of patients
- Suspect with longstanding disease, SNHL, noise/pressure induced vertigo
- **Dx**
  - Fine cut t-bone CT (1 mm)
- Most common structure
  - horizontal canal, basal turn cochlea
- Requires CWD mastoidectomy
  - Management of matrix overlying fistula
Facial paralysis

- Requires immediate surgery when cholesteatoma involved
  - Rapid – acute infection/infected cholesteatoma
  - Slow – chronic expansion of disease

- Temporal bone CT
  - Evaluate nerve involvement
  - Most common site cholesteatoma involvement w/ VII paralysis – geniculate ganglion
    - D/t dz in anterior epitympanum
Facial Paralysis

Management

- Immediate surgical intervention
- Removal of cholesteatoma and infected material with decompression of the nerve
  - Mastoidectomy with facial recess approach for horizontal and vertical segments
  - Middle fossa approach for petrous apex
- Administration of intravenous antibiotics and high-dose steroids
- Iatrogenic injury to nerve during surgery should be immediately repaired with decompression of nerve proximal and distal to site of injury
  - Primary anastomosis → cable nerve graft → XII VII jump graft, etc.
Intracranial complications

- Complications
  - Lateral sinus thrombophlebitis, meningitis, intracranial abscess (extradural, epidural, subdural, or parenchymal), periosteal abscess
  - Potentially life-threatening
  - Incidence as high as 1%
Intracranial Complications

- Symptoms
  - 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

- Exact mechanism of pathogenesis not clear
- Knowledge of anatomy and function of middle ear
- Careful initial evaluation
- Primary goal of surgery: safe, dry ear
- Surgical strategies vary
- Complications can be life-threatening
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
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