Cholesteatoma

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Introduction

- Keratin-producing squamous epithelium in the middle ear, mastoid or petrous apex
- Johannes Müller (1838) coined the term “a pearly tumor of fat...among sheets of polyhedral cells”
- Exhibits independent growth, replaces mucosa, resorbs bone
Introduction

- Histologically made up of:
  - Cystic content – anucleate keratin squames
  - Matrix – keratinizing squamous epithelium
  - Perimatrix – granulation tissue in contact with bone (produces proteolytic enzymes)
Pathology

Molecular models

- Preneoplastic transformation events
- Defective wound-healing process
- Collision between host inflammatory response, normal middle ear epithelium, and bacterial infection
Preneoplastic transformation

- Hyperproliferative keratinocytes
  - Increased proliferation
  - Decreased terminal differentiation
- Expression of epithelial markers in the basal and suprabasal layers (cytokeratins – 10, 13, 16, filaggrin, involucrin); confirm they arise from pars flaccida and overlying EAC skin
- High expression of epidermal growth factor receptor, transforming growth factor
- Upregulation of p53
Defective wound healing

- Chronic inflammatory response around matrix (granulation/perimatrix)
- Infiltration of activated T-cells and macrophages
- Production of cytokines (TGF, TNF, IL-1, IL-2, FGF, PDGF)
- Causes increased migration and invasion of cholesteatoma epithelium and fibroblasts
Host response vs. bacteria

- Bacterial related antigens producing host inflammatory response may stimulate the migrating epithelium’s uncoordinated proliferation
- Granulation induces invasion of keratinocytes
- Granulation – contains proteases, acid phosphatases, bone resorption proteins, osteoclast-activating factors, prostaglandins
- Keratin implanted into mouse calvaria was shown by Chole, et. al., to activate osteoclasts and produce a localized inflammatory bone remodeling similar to cholesteatomas
Classification

- Congenital
- Acquired
  - Primary acquired (retraction pocket)
  - Secondary acquired
Pathogenesis

- Congenital
  - Arise from embryonal rests of epithelial cells
  - Location (petrous pyramid, mastoid and middle ear cleft)
- Levenson criteria
  - White mass medial to normal TM
  - Normal pars flaccida and tensa
  - No history of otorrhea or perforations
  - No prior otologic procedures
  - Prior bouts of otitis media not grounds for exclusion
Congenital cholesteatoma
Pathogenesis

- Primary acquired
  - Eustachian tube dysfunction
  - Poor aeration of the epitympanic space
  - Retraction of the pars flaccida
  - Normal migratory pattern altered
  - Accumulation of keratin, enlargement of sac
Primary acquired cholesteatoma

**ACQUIRED CHOLESTEATOMA**

- Eardrum intact
- Cholesteatoma has eroded through bone of ear canal exposing the bones of hearing
Pathogenesis

Secondary acquired

- Implantation – surgery, foreign body, blast injury
- Metaplasia – transformation of cuboidal epithelium to squamous epithelium from chronic infection
- Invasion/Migration – medial migration along permanent perforation of TM
- Papillary ingrowth – intact pars flaccida, inflammation in Prussack’s space, break in the basal membrane, cords of epithelium migrate inward
Anatomic Considerations

- Mesotympanum
  - Facial recess
  - Sinus tympani
- Hypotympanum
- Epitympanum
Anatomic Considerations

- Epitympanum
  - Above short process of malleus
  - Contains head of malleus, body of incus and associated ligaments and mucosal folds
  - Pars flaccida lacks support from a fibrous middle layer
Anatomic Considerations

- Epitympanic cholesteatoma patterns of spread from Prussack’s space
  - Posterior epitympanum
  - Posterior mesotympanum
  - Anterior epitympanum
Cholesteatoma spread

- Posterior epitympanum – through superior incudal space to mastoid antrum
Cholesteatoma spread

- Posterior mesotympanum – inferiorly through posterior pouch of von Troeltsch to stapes, round window, sinus tympani and facial recess
Cholesteatoma spread

- Anterior epitympanum – anterior to head of malleus, may gain access to supratubal recess via anterior pouch of von Troeltsch
Evaluation

- **History**
  - Hearing loss, otorrhea, otalgia, tinnitus, vertigo and nasal obstruction
  - Previous history of chronic otitis media, tympanic membrane perforation or otologic surgery
  - Progressive unilateral hearing loss with chronic fetid otorrhea suspicious
Evaluation

- Physical Examination
  - Otomicroscopy
  - Posterosuperior retraction pocket with squam
  - Granulation from diseased bone
  - Aural polyps
  - Pneumatic otoscopy – positive fistula response suggests erosion into labyrinth
  - Cultures should be obtained in infected ears
Evaluation

- **Audiology** – usually conductive loss, may vary greatly; confirm with tuning forks
- **Imaging**
  - CT temporal bone – definitely obtain for revision cases, complications of chronic suppurative otitis media, sensorineural hearing loss, vestibular symptoms, other complications of cholesteatoma
Management

- Surgical disease with definite objectives:
  - Removal of disease for safe, dry ear
  - Restore or maintain functional capacity of ear, i.e., hearing
  - Maintain normal anatomy if possible
  - Management of complications takes priority
- Each case treated individually according to extent/location of disease
- Preoperative counseling
Management

- Medical
  - Aural toilet, local care,
  - patients with unacceptable anesthesia risks
- Preventive
  - Tympanostomy tube for early retraction pockets
  - Surgical exploration for persistence
Surgical Management

- Canal-wall-down procedures
- Intact-canal-wall procedure
- Transcanal anterior atticotomy
- Bondy modified radical procedure
Canal-wall-down-down procedures

- Exteriorizing mastoid into external ear canal by taking posterior canal wall down
- Modified radical mastoidectomy – middle ear space preserved
- Radical mastoidectomy – middle ear space eliminated, Eustachian tube orifice obliterated
Canal-wall-down indications

- Cholesteatoma in an only hearing ear
- Significant erosion of posterior wall
- Labyrinthine fistula
- Limited access to epitympanum from sclerotic mastoid
- Recurrent cholesteatoma following ICW surgery with ETD
Canal-wall-down indications
Canal-wall-down

- Advantages
  - Residual disease easy to detect
  - Rare recurrence
  - Facial recess exteriorized

- Disadvantages
  - Open cavity, lifetime maintenance
  - Longer healing time
  - Middle ear shallow (difficult OCR)
  - Water precautions necessary
Intact-canal wall procedure

- Preservation of posterior wall
- With/without posterior tympanotomy
- 2nd staged procedure (6 – 12 months)
- Contraindications
  - Only hearing ears
  - Labyrinthine fistula
  - Long-standing ear disease, ETD
Intact-canal wall procedure
Intact-canal-wall

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 often necessary
- Residual disease harder to detect
Transcanal anterior atticotomy

- Limited cholesteatoma (middle ear, ossicular chain, epitympanum)
- Endaural incision to raise flap
- Removal of scutum around cholesteatoma
- Aditus obliterated
- Reconstruction of lateral attic wall optional
Transcanal anterior atticotomy
Bondy modified radical procedure

- Attic and mastoid disease
- Lateral to ossicles, not involving middle ear space
- Cholesteatoma marsupialized
- Requires good Eustachian tube function and intact pars tensa
Complications of cholesteatoma

- Hearing loss
- Labyrinthine fistula
- Facial paralysis
- Intracranial complications
Hearing loss

- Conductive hearing loss common
- Ossicular chain erosion – 30%
- Severity of loss varies despite extent of disease
- SNHL – may indicate labyrinth involved
- Surgical complication rates – 3% (can be total hearing loss)
Labyrinthine fistula

- Up to 10% of patients
- Suspect with longstanding disease, SNHL, induced vertigo
- CT should be obtained
- Most common structure – horizontal canal
- Requires CWD mastoidectomy
- Management of matrix overlying fistula
Facial paralysis

- With cholesteatoma requires immediate surgery
- Rapid – infected cholesteatoma
- Slow – chronic expansion of disease
- CT localizes involved portion
- Most common site – geniculate ganglion
Facial paralysis

Management

- Mastoidectomy with facial recess approach for horizontal and vertical segments
- Middle cranial fossa for petrous apex
- Remove cholesteatoma and infected debris
- IV antibiotics and steroids helpful
- Iatrogenic injury repaired immediately
Intracranial complications

- Potentially life threatening
- Periosteal abscess, lateral sinus thrombosis, intracranial/epidural abscess, meningitis
- Less than 1% of all patients
- Suppurative otorrhea, chronic headache, pain, fever – impending intracranial complication
- Mental status changes, nuchal rigidity, cranial neuropathies require neurosurgical consult
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