Cholesteatoma-Pathogenesis and Surgical Management

Grand Rounds Presentation
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Introduction

- **Cholesteatoma (keratoma)** - essentially an accumulation of skin in ME/mastoid
- Insidious nature
- Variable symptoms depending on extent and location of disease
- Primarily a surgical disease
- High rate of recidivistic disease
- Long-term follow-up essential
Pathology and Classification

- Non-neoplastic accumulation of keratinizing stratified squamous epithelium with desquamated keratin debris
- Subepithelial fibroconnective tissue
- Granulation tissue
- Bone destruction possible
- Elaboration of collagenase and other inflammatory mediators
Pathology and Classification

- Congenital cholesteatoma
- Acquired cholesteatoma
- Canal cholesteatoma
Congenital Cholesteatoma

- Cholesteatoma sac medial to an intact tympanic membrane
- Normal pars flaccida and tensa
- No h/o TM perforation or otorrhea
- No h/o otologic trauma or surgery
- H/o prior episodes of OM does not preclude its presence
Acquired Cholesteatoma

- Usually found in posterosuperior quadrant of TM with asso. retraction pocket or perforation
- Primary acquired cholesteatoma asso. with pre-existing retraction pocket
- Secondary acquired cholesteatoma arises in setting of persistent TM perforation
Canal Cholesteatoma

- Found lateral to TM
- Idiopathic, post-traumatic, and iatrogenic variants
- Must be distinguished from keratosis obturans
Eustachian Tube Dysfunction

- Important in pathogenesis of middle ear disease and cholesteatoma
- Essential role in recurrent disease and surgical failure
- Preoperative clinical assessment of tubal patency mandatory
- Tubal function and ME aeration particularly important in postoperative hearing results
Pathogenesis

- Migratory nature of TM epithelium and cholesteatoma
- Iatrogenic implantation
- Invasion of squamous epithelium
- Invagination theory
- Basal cell proliferation
- Metaplasia
- Embryonic squamous epithelial cell rests
Anatomic Considerations

- Tympanic cavity derived from endodermally-lined first branchial pouch
- Characteristic pathways of disease spread
- Attic or epitympanum-Prussack’s space
- Posterior mesotympanum-facial recess and sinus tympani
Evaluation

- History-long h/o ear complaints
- Physical examination-otomicroscopy
- Audiology-CHL
- Imaging-assessment of mastoid disease, surgical road map, revision cases, sensorineural hearing loss, vestibular symptoms
Management

- **Surgical disease**
- **Patient age (I.e. pediatric)**
  - Cholesteatoma generally considered more aggressive
- **Primary goal is eradication of disease**
  - With hearing preservation or improvement secondary
- **Final therapeutic decisions often made at surgery**
Non-surgical Management

- Office management of limited disease in elderly patients with comorbidities
- Topical antibiotic preparations including those containing steroids sometimes useful preoperatively
Surgical Management

- No consensus regarding optimal surgical strategy
- Principal controversy concerning intact canal wall vs. canal wall down mastoidectomy
- Therapy must be individualized on case-by-case basis
Preoperative Patient Counseling

- Surgical goals
- Risks of surgery including facial paralysis, tinnitus, vertigo, worsening of hearing
- Possible need for staged procedure
- Chronic nature of disease process with need for long-term follow-up
- Routine aural toilet if mastoid bowl created
Tympanostomy Tube Insertion

- Alleviation of early TM retraction in setting of ETD
- Arrest pathologic process prior to irreversible changes such as atelectasis, deep retraction pocket formation, TM perforation, or cholesteatoma formation
- Assist in maintenance of ME aeration after tympanoplasty or tympanomastoidectomy
**Tympanomeatal Flap/Tympanoplasty**

- Smaller congenital cholesteatomas of involving TM or ME
- Acquired cholesteatomas limited to mesotympanum
Intact Canal Wall Mastoidectomy

- Preservation of posterior canal wall during simple mastoidectomy with or without posterior tympanotomy (facial recess approach)
- Cholesteatomas of attic, antrum, post. mesotympanum with adequate ME and mastoid aeration
- Staging necessary with ME mucosal abnormalities, ossicular erosion, residual disease
Canal Wall Down Mastoidectomy

- Removal of post. canal wall to level of vertical facial nerve
- Creation of mastoid cavity with exteriorization of mastoid into EAC
- Scutum removed with obliteration of epitympanum and removal of malleus head and incus
- MRM ME space maintained while radical mastoid eliminates ME space and obliterates eustachian tube
Canal Wall Down
Mastoidectomy

- Surgery in an only-hearing ear
- Poor anesthetic risk
- Poor pt compliance with unreliable F/U
- Poor tubal function and ME aeration
- Sclerotic mastoid
- Extensive canal wall defect
- Labyrinthine fistula
- Meatoplasty and mastoid obliteration
Atticotomy

- Removal of scutum
- Limited attic disease
- Scutal reconstruction with autologous cartilage
Bondy Procedure

- Removal of scutum and posterior canal wall with preservation of ossicles and ME space

- Larger attic cholesteatomas lateral to ossicles in pt with sclerotic mastoid
Intact Canal Wall Advantages

- More rapid healing
- Easier long-term postoperative care
- No water precautions necessary (particularly important in children)
- More options available for hearing aid, if necessary
Intact Canal Wall Disadvantages

- Epitympanum/mastoid not accessible to postop inspection
- Supratubal space not easily accessible unless malleus head and incus removed
- Both residual and recurrent disease more likely
- Greater number of procedures usually required for disease eradication
Canal Wall Down Advantages

- Easy detection of residual disease
- Recurrent cholesteatoma rare
- Fewer procedures necessary for eradication of disease
Canal Wall Down Disadvantages

- Longer healing time
- Special cavity care often necessary for proper healing
- Periodic cleaning necessary
- Accumulation of debris may occur with increased risk of infection
- Water precautions necessary
Results of Therapy

- Rosenberg et al. examined variables with regard to residual-recurrent disease (retrospective)
  - 232 children with cholesteatoma (244 ears)
  - Ossicular erosion asso. with residual-recurrent disease (necessitates 2nd look)
  - Recidivism 61% at 6 years (necessitates long-term F/U)
Dodson et al. examined cases of 66 children with cholesteatoma (73 ears) retrospectively with ave. F/U 37.7 mos.

- ICW-41% recidivism and CWD-12% recidivism
- Postop SRT less than 30 dB in 75% of ICW and 72% of CWD
- Prefer ICW with 2nd stage
Results of Therapy

- Hirsch et al. retro. reviewed 164 cases of ped. chol. (116 avail. for 5 year F/U)
- Majority of pts required CWD procedure
- Recidivism 11% for tympanoplasty, 19% for ICW, 5% for MRM, and 0% for radical mastoid
- Also reported fewer revisions and better hearing results with CWD
Complications

- Conductive hearing loss
- Labyrinthine fistula
- Facial nerve paresis or paralysis
- Intratemporal or intracranial complications
- Encephalocele
Conclusions

- Exact pathogenesis not entirely clear
- Important anatomic considerations in management
- Eradication of disease primary goal
- No universally accepted surgical strategy
- High rate of recidivism with long-term F/U essential
- Maintain vigilance for complications