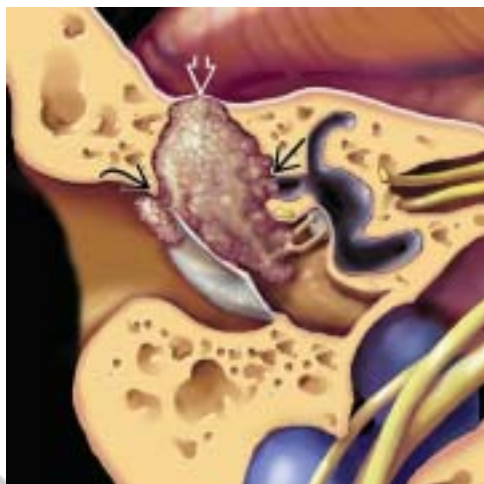


Acq Cholesteatoma, Pars Flaccida



Acquired pars flaccida cholesteatoma. Coronal drawing of right T-bone shows a large cholesteatoma welling up from a pars flaccida perforation, amputating the scutum (curved arrow), thinning the tegmen tympani (open arrow) and eroding through the cortex of the lateral semicircular canal (arrow).

Key Facts

- Synonyms: "Attic" or "Prussak's space" cholesteatoma
- Definition: Exfoliated keratin within stratified squamous epithelium; begins in Prussak's space
- Pars flaccida portion of tympanic membrane (TM), also known as Shrapnell's membrane, is small superior portion of TM
 - Pars flaccida retraction pocket & perforation predispose to cholesteatoma formation in Prussak's space
- Middle ear mass in posterolateral attic, displacing ossicles medially
- Most common type of cholesteatoma; 80% of all acquired cholesteatomas

Imaging Findings

General Features

- Best imaging clue: Mass in Prussak's space with ossicles deviated medially combined with scutum or lateral middle ear wall erosion

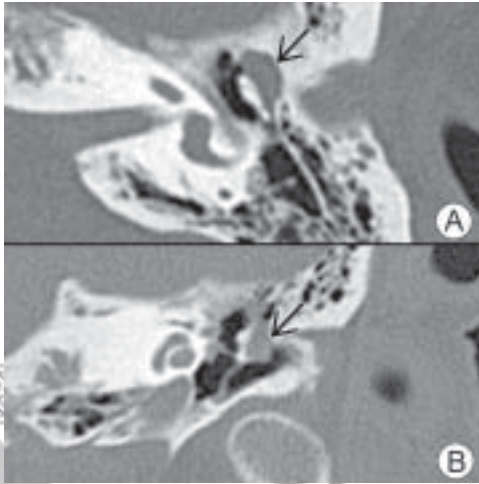
CT Findings

- Soft tissue mass in Prussak's space (medial to attic wall, lateral to head of malleus & body of incus, above lateral malleolar ligament)
 - May extend posterolateral to aditus ad antrum, into mastoid antrum
- Inferior extension to posterior middle ear recesses may occur in children
- Scutum erosion is common
- May widen aditus, or remodel lateral attic wall
- Ossicular erosion occurs 70% of time, with long process of incus most commonly eroded
 - Incus body & malleus head may also be eroded

MR Findings

- T1 & T2: Middle ear mass, isointense to Cerebrospinal Fluid (CSF)
- T1 C+: Low signal intensity mass, nonenhancing except for peripheral rim of granulation tissue

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Pars flaccida acquired cholesteatoma. (A) Axial CT shows a cholesteatoma (arrow) in Prussak's space with erosion of the lateral margin of the malleus & incus. (B) Coronal CT reveals the cholesteatoma originates from the pars flaccida portion of the TM, truncates the scutum (arrow) as it enters Prussak's space.

- If there is tegmen tympani erosion, coronal MR is recommended to exclude intracranial extension
- T1 C+ coronal: Dural enhancement at bony defect

Imaging Recommendations

- High-resolution temporal bone CT, without contrast, is test of choice
- Prussak's space mass, attic, scutum are best seen on coronal CT
- Enhanced MR, including coronal plane, is used when cephalocele, middle cranial fossa infection or intracranial cholesteatoma extension is an issue

Differential Diagnosis: Middle Ear Mass

Pars Tensa Cholesteatoma

- Less common than pars flaccida type
- Results from posterosuperior TM perforation
- Sinus tympani & facial recess involvement are classic
- Ossicles pushed laterally

Congenital Cholesteatoma of Middle Ear

- Only 2% of cholesteatomas are congenital
- Focal mass behind intact TM
- Ossicular erosion may occur

Cholesterol Granuloma of Middle Ear

- TM intact
- Ossicular & bony erosions may be similar to cholesteatoma
- Hyperintense on T1 C- images

Glomus Tympanicum Paraganglioma

- Focal mass on cochlear promontory without ossicular or bone erosions
- Intact TM with pulsatile vascular retrotympanic mass

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Pathology

General

- General Path Comments
 - Development of cholesteatoma begins with negative middle ear pressure leading to a TM retraction pocket
 - Moisture and desquamation of epithelial cells result in invaginated pouch filling with epidermal debris
 - Exfoliated squamous epithelium grows into cholesteatoma
 - Second theory: TM perforation leads to squamous epithelium from TM surface extending into middle ear, resulting in cholesteatoma
 - Bottom line: TM perforation or retraction results in middle ear accumulation of stratified epithelial squamous cells
- Etiology-Pathogenesis
 - TM perforation or retraction results in middle ear accumulation of stratified epithelial squamous cells
- Epidemiology
 - Most common middle ear/mastoid lesion

Gross Pathologic, Surgical Features

- "Pearly tumor," composed of soft, waxy, white material

Microscopic Features

- Identical to epidermoid cyst
- Stratified squamous epithelium, with progressive exfoliation of keratinous material; contents rich in cholesterol crystals

Clinical Issues

Presentation

- Principal presenting symptom: Chronic otorrhea + conductive hearing loss
- Other symptoms: Recurrent or chronic middle ear infections, vertigo if the lateral semicircular canal is dehiscid
- Otologic examination: Retraction pocket, perforation or obvious pearly white tumor at pars flaccida, located at supero-anterior quadrant of TM

Natural History

- Progressive increase in size of cholesteatoma, with destruction of surrounding structures, including ossicular chain, otic capsule and semicircular canals, tegmen tympani, and transverse sinus invasion
- Facial nerve involvement, venous sinus thrombosis, and intracranial extension are late complications

Treatment

- Surgical goal is complete eradication of cholesteatoma & infection
- Hearing preservation or restoration are secondary goals
- Surgery includes mastoidectomy & formation of a common cavity between mastoid antrum & external auditory canal + TM & ossicle reconstruction

Prognosis

- Recurrence rate = 6-8%
- Small cholesteatoma: Excellent for total eradication and normal hearing
- Large cholesteatoma: Residual conductive hearing loss is possible

Selected References

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3. Swartz JD: Cholesteatomas of the middle ear: Diagnosis, etiology and complications. Radiol Clin North Am 22:15-35, 1984