Acquired Cholesteatoma - a pictorial review.

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Learning Objectives

• To review the anatomy, classification and imaging of acquired cholesteatoma

Background

Background

Cholesteatoma may be congenital or acquired and can be radiologically and histologically indistinguishable. Acquired cholesteatomas occur exclusively within the middle ear and are related to recurrent middle ear disease/eustachian tube dysfunction. Several complications can occur which may result in hearing loss, vertigo, headache and intracranial complications. Because of the serious potential complications, surgical treatment is necessary. High resolution temporal bone CT and MRI aid in evaluation, surgical planning and postoperative assessment.

Definition

Cholesteatomas are a non neoplastic expansile growth which consist of keratinising squamous epithelium. They can develop anywhere in the pneumatised portion of the temporal bone but are most common in the middle ear cavity. They consist of a cystic keratin centre with a surrounding matrix of keratinising squamous epithelium and an outer perimatrix of granulation tissue. The perimatrix granulation tissue is responsible for bony destruction through the release of proteolytic enzymes.

Classification

Cholesteatomas can be classified as congenital or acquired. Congenital cholesteatomas occur in children without a history of otitis media with an intact tympanic membrane. Acquired cholesteatomas can be categorised as primary or secondary. Special cholesteatoma variants include External Auditory Canal Cholesteatomas and Mural Cholesteatomas.

Primary Acquired Cholesteatoma

Primary acquired cholesteatomas constitute approximately 80% of middle ear cholesteatomas¹.

The aetiology is related to eustachian tube dysfunction/chronic otitis media where there is retraction of the tympanic membrane forming a retraction pocket, most commonly in
the upper one third of the tympanic membrane (pars flaccida). This serves as a potential site of accumulation of keratin which may lead to cholesteatoma formation. Retraction pockets progressing to cholesteatomas also commonly form in the postero-superior pars tensa of the tympanic membrane. Erosion of the long process of the incus is a common association with this type.

**Secondary Acquired Cholesteatoma**

Secondary acquired cholesteatomas account for approximately 18% of middle ear cholesteatomas¹. There are several theories of pathogenesis but they develop by growing through a defect in the tympanic membrane.

**Middle Ear Anatomy**

An understanding of the middle ear’s complex anatomy is essential for adequately assessing cholesteatoma and the associated disease complications. The middle ear is roughly divided into three parts based on the relationship to the superior and inferior limits of the external auditory canal (EAC). Above the superior limit is the epitympanum or attic. At the level of the EAC is the mesotympanum. The hypotympanum is below the mesotympanum. Several important structures are located in each of the three parts, as outlined in table 1.

<table>
<thead>
<tr>
<th>Epitympanum (Attic)</th>
<th>Mesotympanum</th>
<th>Hypotympanum</th>
</tr>
</thead>
<tbody>
<tr>
<td>Head of Malleus</td>
<td>Handle of Malleus</td>
<td>Bony canal for the jugular bulb (posterior)</td>
</tr>
<tr>
<td>Body and short process of Incus</td>
<td>Long Process of Incus</td>
<td>Bony canal for the carotid canal (anterior)</td>
</tr>
<tr>
<td>Prussak’s space</td>
<td>Stapes</td>
<td></td>
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<tr>
<td>Communication with the mastoid (posterior)</td>
<td>Oval and Round windows</td>
<td></td>
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<tr>
<td>Communication with the supratubal recess (anterior)</td>
<td>Cochlear Promontory</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Sinus Tympani</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Facial nerve tympanic segment within facial nerve canal</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Eustachian tube orifice anteriorly</td>
<td></td>
</tr>
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</table>
Table 1. Important Structures related to Divisions of the Middle Ear

*Prussak’s Space*

This is located medial to pars flaccida (upper one third of the tympanic membrane) and lateral to the head and neck of the malleus. It is bounded by malleal ligaments anteriorly and posteriorly. The resultant folding of the retracting tympanic membrane results in a predictable location for the formation of attic cholesteatomas³.

Fig. 1 on page 4 demonstrates relevant anatomy.

Images for this section:
Imaging Findings OR Procedure Details

High Resolution Computerised Tomography (HRCT)

High resolution temporal bone CT is the primary imaging investigation for cholesteatoma. CT alone is generally adequate in defining key surgically relevant landmarks such as:

- Degree of mastoid pneumatization
- Extent of the primary lesion
- Extent of bone erosion
- Identifying specific surgical hazards e.g. sinus tympani extension, lateral semicircular canal and tegmen tympani dehiscence.

Unfortunately, there are a number of important clinical situations where CT lacks specificity. Examples include small non-erosive lesions, atypical lesions, co-existent inflammatory middle ear disease and in the postoperative setting.

At our institution, helical scanning is performed on a Siemens Sensation 16 slice CT scanner and a Toshiba Aquilion One 128 slice CT scanner with a slice thickness of 0.6mm. Axial and coronal 0.6mm bone reconstructions are standard.

Pars Flaccida / Attic Cholesteatoma.

Cholesteatomas arising in the pars flaccida region are usually primary acquired lesions but secondary acquired cholesteatomas can also develop at this location. (see Fig. 2 on page 9)

Typical imaging features include:

- Retracted tympanic membrane
- Soft tissue mass in Prussak's space
- Soft tissue lateral to the ossicles
- Erosion of the scutum
- Posterior extension into the mastoid antrum

Pars Tensa / Mesotympanic Cholesteatoma

Pars tensa cholesteatomas usually occur in the setting of a perforated tympanic membrane, generally secondary to chronic otitis media. They are often referred to as sinus cholesteatomas as the lesions extend toward the sinus tympani and the facial recess of the mesotympanum. (see Fig. 3 on page 10)
Typical imaging features include:

- Soft tissue mass medial to the ossicles
- Lateral displacement of the ossicles
- Ossicular erosion, particularly the long process of the Incus
- Opacification of the sinus tympani

**Advanced Lesions**

Extensive cholesteatomas are at times difficult to categorise but assessment of the overall extent of the lesion and associated complications is valuable for surgical planning. (see Fig. 4 on page 11)

**External Auditory Canal Cholesteatoma (EACC).**

External auditory canal cholesteatomas are rare lesions and constitute 1-1.5% of cholesteatomas. The aetiology probably relates to poor keratin migration in the external auditory canal but may relate to previous trauma or recurrent otitis externa. The CT features of EACC are not particularly specific but they include:

- External auditory canal soft tissue mass
- Erosion of adjacent bone
- Intralosional bone fragments#

(see example in Fig. 5 on page 12)

EACC may extend deeply into the middle ear or mastoid air cells. The major differential diagnoses are keratitis obturans (circumferential expansion of the deep ear canal with tenacious squamous epithelium) and malignant otitis externa (pseudomonas skull base osteomyelitis or squamous cell carcinoma).

**Mural Cholesteatoma**

Mural cholesteatoma is a rare variant where a rind of cholesteatoma remains in the middle ear after it has invaded through the bony wall of the external auditory canal. There is drainage of material through the external auditory canal, giving the appearance of an "automastoidectomy"#. (see Fig. 6 on page 12)

**Complications**

The major complications are either as a result of direct involvement of important adjacent structures or secondary infection# (outlined in Table 2). Fig. 7 on page 13, Fig. 8 on page 14 and Fig. 9 on page 15 demonstrate common complications.
<table>
<thead>
<tr>
<th>Complication</th>
<th>Clinical Outcome</th>
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</thead>
<tbody>
<tr>
<td>Ossicular/cochlear erosion</td>
<td>Conductive or sensorineural hearing loss</td>
</tr>
<tr>
<td>Labyrinthine Fistula</td>
<td>Vertigo</td>
</tr>
<tr>
<td>Facial nerve involvement</td>
<td>Facial nerve paralysis, especially if the Geniculate ganglion is injured. Abnormal taste sensation if Chorda tympani nerve is injured.</td>
</tr>
<tr>
<td>Secondary infection of Cholesteatoma</td>
<td>Mastoiditis, Meningitis/Cerebritis, Temporal lobe abscess</td>
</tr>
<tr>
<td>Secondary Internal Jugular vein or Sigmoid sinus thrombosis/thrombophlebitis</td>
<td>Headache, intracranial infection, raised intracranial pressure.</td>
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</tbody>
</table>

**Table 2. Complications of Cholesteatoma**

**Magnetic Resonance Imaging (MRI).**

MRI has the advantage of high tissue specificity and contrast resolution in cholesteatoma imaging. There are several clinical circumstances that MRI may be useful for and this includes:

- Confirmation of cholesteatoma when there is clinical or CT diagnostic uncertainty.
- Detection of residual or recurrent disease following surgery.

At our institution, we routinely perform cholesteatoma studies on a Philips Achiever 1.5T MRI scanner with standard sequences being axial turbo spin echo (TSE) T1 and T2 sequences, axial T2 3D DRIVE, thin slice coronal Balanced Turbo Field Echo (BTFE) and coronal Multishot TSE Diffusion Weighted Imaging (DWI).

**Conventional Techniques**

This includes routine T1 and T2 - weighted spin echo sequences. Cholesteatomas have variable signal intensity on these sequences and can be difficult to delineate from adjacent inflammatory change. Thin slice sequences can aid localisation.

**Delayed Post Contrast Imaging.**

Cholesteatomas are avascular and therefore do not enhance following contrast administration. Conversely, granulation tissue is vascularised, albeit poorly and therefore shows some enhancement on delayed T1 post contrast sequences. Ayache et
al# prospectively studied this technique and demonstrated reasonable detection of cholesteatomas with a sensitivity of 90% and specificity of 100%. The sensitivity is reduced in the detection of small lesions (e.g. ≤3mm). The other major disadvantage of this technique is the necessity to acquire images 30 - 45 minutes following contrast administration. Subsequently, diffusion weighted imaging techniques have proved to be as sensitive, if not more so#.

**Diffusion Weighted Imaging (DWI)**

Diffusion weighted imaging of cholesteatomas is highly specific due to the high keratin content of the lesion. Routine echo planar DWI has its limitations due to susceptibility artefacts at the air-bone interfaces of the skull base. New techniques have been developed over the last decade utilising non-echo planar DWI which are less susceptible to these artefacts and have the advantage of thinner imaging sections and improved spatial resolution.

The novel techniques developed include singleshot and multishot Turbo Spin Echo (TSE) DWI, half-Fourier acquisition single-shot turbo spin-echo (HASTE) DWI, BLADE DWI and periodically rotated overlapping parallel lines with enhanced reconstruction (PROPELLER) DWI. Sensitivities between 90 - 100% have been achieved with these techniques for lesions as small as 2mm#. (see Fig. 10 on page 16 and Fig. 11 on page 17).

The diagnostic criteria for cholesteatoma is hyperintensity relative to brain parenchyma on the b = 0 sec/mm2 which is persistent on the high b value (800-1000 sec/mm2) images.

**Residual and Recurrent disease**

High resolution CT in the postoperative setting is useful to confirm that there is no residual abnormal middle ear soft tissue. However, in several clinical situations, CT is not specific enough to distinguish between residual/recurrent disease and post surgical scar tissue. MRI has the advantage of tissue specificity, allowing discrimination between these entities. Fig. 11 on page 17. demonstrates a small recurrent cholesteatoma.

There are two major surgical approaches employed in the treatment of cholesteatoma². In "Canal Wall Down" mastoidectomy surgery, the posterior wall of the external auditory canal is removed with subsequent formation of a common mastoid/external auditory canal cavity. This is usually easy to evaluate otoscopically except when there is significant postoperative granulation tissue/fibrosis. A cholesteatoma may be difficult to detect in this setting and MRI may serve as a useful problem solving tool.
In the more conservative "Canal Wall Up" mastoidectomy surgery, the bony external auditory canal is preserved. Otoscopic blind spots are comparatively more common and there is also a higher rate of residual disease. Fig. 12 on page 18 demonstrates intraoperative views of a recurrent cholesteatoma with preservation of the bony external auditory canal.

Cholesteatoma recurrence generally occurs within the first 2 postoperative years, with over half occurring within a year of surgery\textsuperscript{19}. MRI increasingly has a role in reducing the requirement for second or third look surgery, particularly in clinical situations where otoscopic visualisation is difficult (as encountered with canal wall up surgery or opaque tympanic membrane reconstruction). In an early small prospective study by Dubrulle et al\textsuperscript{11}, the positive predictive value for MRI DWI detection of recurrent cholesteatoma was 93% and the negative predictive value was 100%. Subsequent studies using non-echo planar DWI techniques have demonstrated similar very promising results\textsuperscript{12,13}.

**Limitations of MRI in Residual/Recurrent disease.**

Residual disease can be very small and MRI is not as sensitive at detecting lesions #2-3mm#. Other false negative studies have been attributed to motion artifact and in mural cholesteatomas which expel their central keratin content. Falsely positive studies may be encountered in the setting of acute middle ear/mastoid abscess formation, cholesterol granuloma or immediate postoperative haemorrhage.

**Images for this section:**
Figure 2. Pars Flaccida / Attic Cholesteatoma. HRCT Coronal MPR images.
A - Attic cholesteatoma (arrow). B - Scutum erosion (arrow). C - Ossicular erosion (arrow).

Fig. 2: Pars Flaccida Cholesteatoma
Fig. 3: Pars Tensa Cholesteatoma

Figure 3. Pars Tensa Cholesteatoma. HRCT Coronal MPR images.
The cholesteatoma (arrowed in images A-C) is located medial to the ossicular chain. Prussak’s space (arrow) is demonstrated in image D and is noted to be clear. No ossicular or other bone destruction.

Fig. 4. Advanced Left Cholesteatoma. HRCT Coronal MPR images
A - Erosion of the ossicles (arrow).
B - Erosion of the scutum (arrow) and extension into the mesotympanum.
C - Extension posteriorly into the mastoid air cells (arrow). Semicircular canals and facial nerve canal remain intact.
Fig. 4: Advanced Left Cholesteatoma.

Fig. 5: External Auditory Canal Cholesteatoma
Fig. 6: Mural Cholesteatoma

Figure 6. Mural Cholesteatoma.
HRCT Axial (A - C) and Coronal MPR (D)
A - Residual cholesteatoma in autamastoidectomy cavity (arrow). B - Absence of ossicles.
C - Communication with the external auditory canal following erosion of the posterior wall of the external auditory canal (arrow). D - Coronal MPR further demonstrates erosion of the scutum/mastoid air cells (arrow).
Fig. 7: Complicated Attic Cholesteatoma

**HRCT Coronal MPR images.**

A - Scutum erosion  
B - Labyrinthine fistula (arrow) and facial nerve canal dehiscence (*dashed arrow*)  
C - Tegmen tympani dehiscence

Ossicular erosion was also present but is not demonstrated on these images.
Figure 8. Complicated Attic Cholesteatoma. HRCT Coronal MPR (A - C) and Axial (D).
A - Ossicular erosion. B - Superior (arrow) and lateral (dashed arrow) semicircular canal labyrinthine fistula, facial nerve canal tympanic segment dehiscence (arrowhead). C - Facial nerve mastoid segment erosion and Tegmen tympani/mastoideum dehiscence (arrowheads). D - Jugular Bulb (arrow), Sigmoid plate dehiscence (arrowhead) and External Auditory Canal dehiscence (dashed arrow).

Fig. 8: Complicated Attic Cholesteatoma
Figure 9. Extensive Right Cholesteatoma Sac.
Intraoperative photographs (A - B) Courtesy of Dr Melanie Collins, Otorhinolaryngologist, North Shore Hospital.
Axial HRCT image (C)
A - Cholesteatoma sac exposed (outlined) after the attic wall and cortex have been drilled away. The cholesteatoma extends from the attic into the mastoid air cells.
B - Cholesteatoma excised revealing the eroded tegmen tympani and lateral semicircular canal (arrows).
C - CT correlate demonstrating lateral semicircular canal dehiscence (arrow) and mastoid extension (dashed arrow).

Fig. 9: Extensive Cholesteatoma - Intraoperative Correlation
**Fig. 10:** Recurrent Left Cholesteatoma on MRI

*Figure 10. Recurrent Left Cholesteatoma. MRI Coronal BTFE (A), Multishot TSE DWI (B), Axial T1 (C), Axial T2 (D). Previous modified radical left mastoidectomy for cholesteatoma was performed 3 years prior. **A to D** - Recurrent cholesteatoma is demonstrated (arrows). **B** - Note is made of restricted diffusion and central inspissated secretions (which are not restricted on DWI).*
Fig. 11: Small Recurrent Cholesteatoma on MRI

Figure 11. Recurrent Left Cholesteatoma.
MRI Axial T1 (A), Axial T2 (B), Coronal BTFE (C) and Coronal Non EPI Multishot TSE DWI (D) images.
This patient had previously undergone atticootomy and tympanoplasty for cholesteatoma. Follow up clinical assessment was suspicious for recurrent cholesteatoma which was confirmed with MRI and second look surgery.
A - B Cholesteatoma "pearl" T1 isointense, T2 hyperintense (arrows). C - Cholesteatoma located inferior to the lateral semicircular canal (arrow). D - Corresponding restricted diffusion (arrow).
Figure 12. Recurrent Right Cholesteatoma. *Intraoperative photographs courtesy of Mr NJ Holland, Otorhinolaryngologist, North Shore Hospital.*

Combined approach tympanoplasty with intact external auditory canal and exposed mastoid air cells.

A - Cholesteatoma located adjacent to the lateral semicircular canal (*Lateral SCC*)

B - Close up of cholesteatoma "pearl" measuring 3mm.

**Fig. 12:** Intraoperative Photographs of a Recurrent Cholesteatoma "Pearl"
Conclusion

High resolution temporal bone CT remains the primary imaging investigation for acquired cholesteatoma and aids in the classification and localisation of disease extent. Both CT and MRI are useful for investigating complications, with MRI being particularly sensitive for detecting residual/recurrent disease in the postoperative setting.

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References


