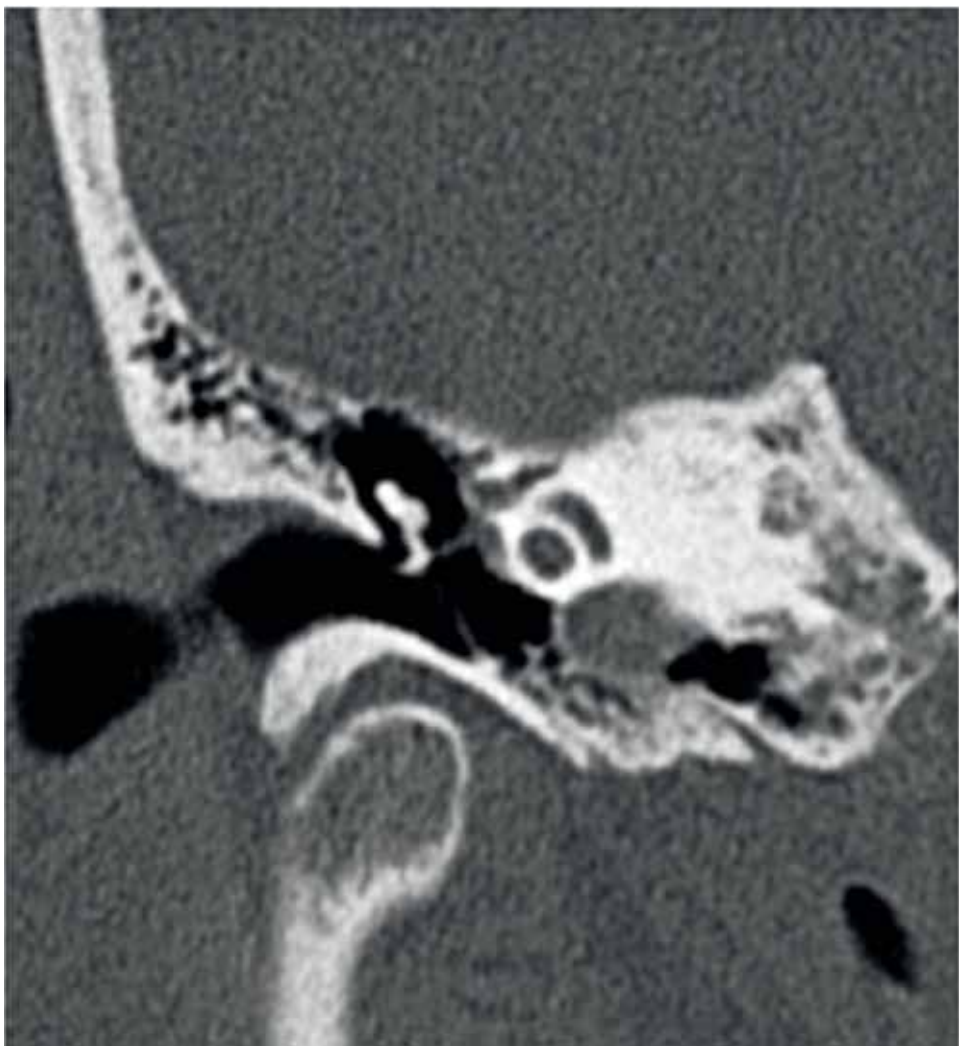

Figure 1: Normal coronal CT scan section of a right ear, illustrating intact external, middle and inner ear structures.

Figure 2: Coronal CT scan of a cholesteatoma in a right ear. Note the opacification in the middle ear, eroding into the inner ear. This patient presented with vertigo and a discharging ear.

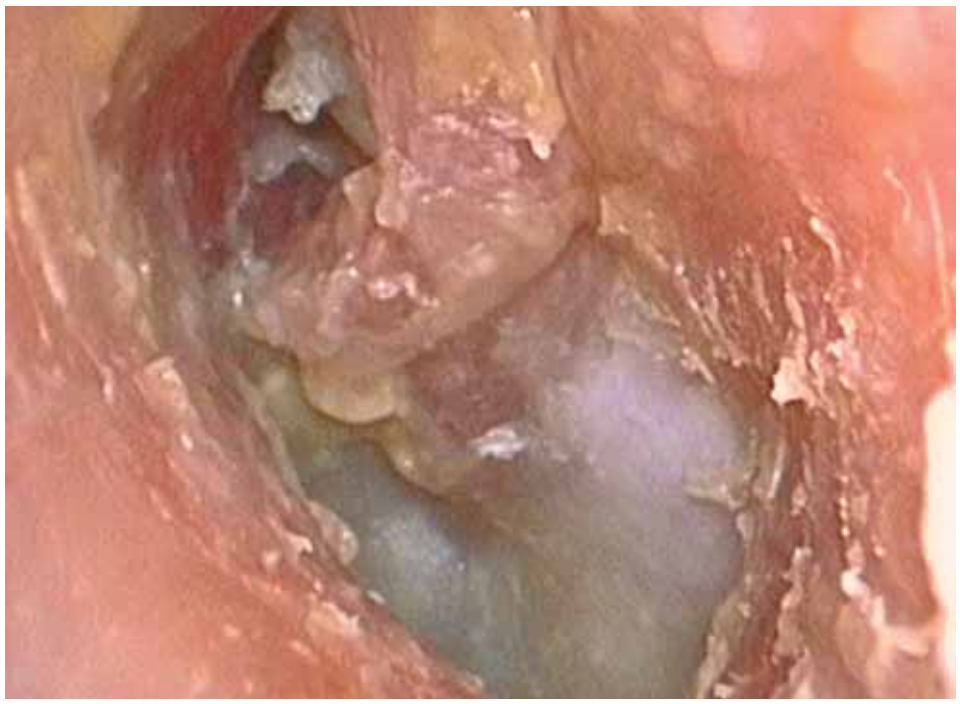

Figure 3: Otoscopic view of a left ear with cholesteatoma — with an attic perforation and a squamous debris collection.

Update on cholesteatoma

The goal in treating this epidermal inclusion cyst is to remove the disease and create a safe, dry ear.

DR PHIL SALE AND
DR NIRMAL PATEL

Definition

CHOLESTEATOMA is an epidermal inclusion cyst within the middle ear, temporal or tympanic bone with a propensity for expansion, recurrent infection and destruction of surrounding structures.

Histopathologically, cholesteatoma is made up of a keratinising, squamous epithelium and keratinous debris with a margin of granulation tissue. The condition has an incidence of approximately 1 in 10,000 in adults and 1 in 30,000 in children.

Most cases of cholesteatoma are acquired, with most of these being the 'primary acquired' type. This type of cholesteatoma is thought to evolve when negative pressure in the middle ear space, from long-term eustachian tube dysfunction, causes development of a collapsed eardrum known as a "retraction pocket" in the tympanic membrane.

Formation of the retraction pocket is associated with alteration of the usual pattern of epithelial migration

Formation of the retraction pocket is associated with alteration of the usual pattern of epithelial migration, with accumulation, rather than clearance of the keratin debris.

Accumulation of debris within the retraction pocket leads to gradual expansion, made more rapid periodically because of recurrent infection.

Less common causes of acquired cholesteatoma include marginal tympanic membrane perforations, iatrogenic causes from sur-

cont'd next page

from previous page
gical manipulation (during grommet insertion or tympanic membrane grafting) and squamous metaplasia in the middle ear mucosa due to chronic inflammation.

Diagnosis

A diagnosis of cholesteatoma is made based on history-taking, microscopic examination of the ear, and imaging (see figures 1 and 2).

Many patients will remain asymptomatic for a significant period of time, with some having incidentally discovered cholesteatomas.

Others will be symptomatic due to repeated infections presenting as either otorrhoea (with or without pain) or destruction of adjacent structures manifesting as a conductive hearing loss, vertigo or facial nerve palsy.

Examination findings are often variable, though the most common form of cholesteatoma will appear as a defect in the superior portion of the tympanic membrane, with keratin debris in its centre (see figure 3).

Associated infection and pre-existing abnormalities of the tympanic membrane and middle ear contents in patients with chronic otitis media introduce further variability in the microscopic appearance.

Congenital cholesteatomas typically appear as a dull white mass behind the intact tympanic membrane, most often in the anterosuperior quadrant.

Regardless of aetiology, cholesteatoma is a problem because of its relentless expansion.

Tympanic membrane anatomy

The pars flaccida region of the tympanic membrane is that region above the short process of the malleus (figure 4).

The clinician must have a

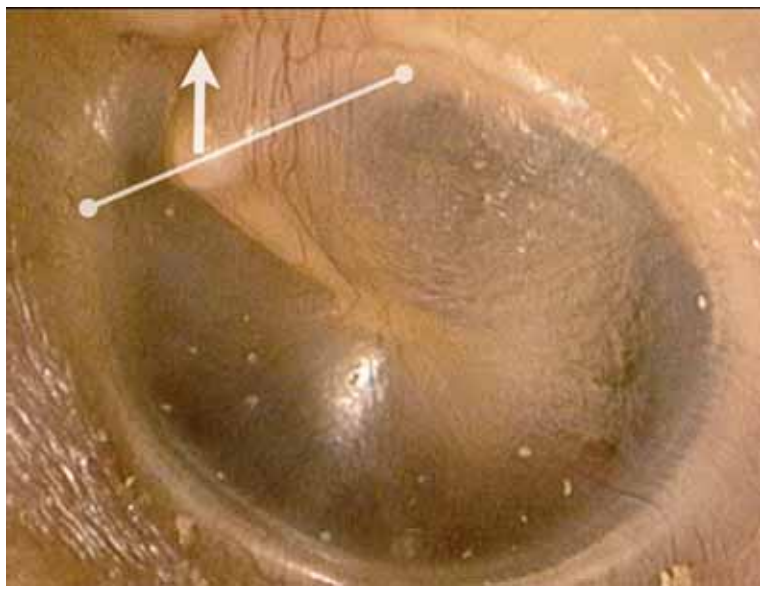

Figure 4: Otoscopic view of a normal left ear. The oblique line illustrates the demarcation between pars flaccida (superior) and pars tensa (below). The solid white arrow begins on the short process of the malleus and extends up to show a normal non-diseased attic region.

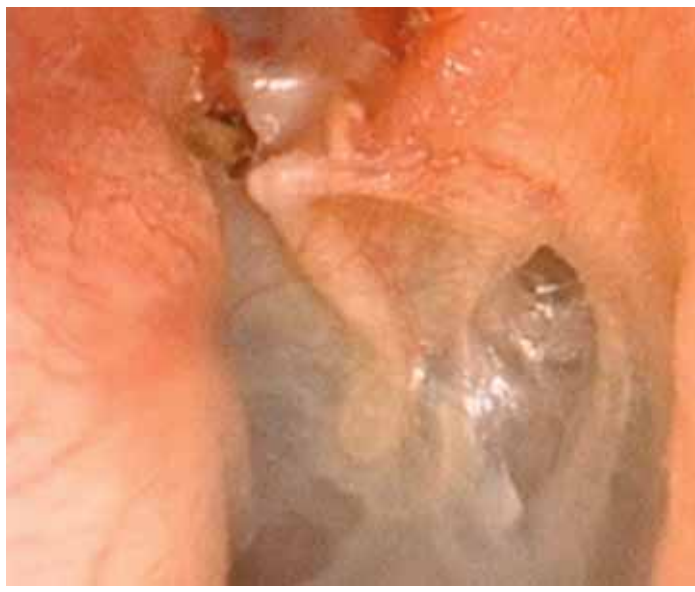

Figure 5: Otoscopic view of a left attic retraction pocket. An attic retraction pocket is visible above the short process of the malleus. This situation may be a prelude to cholesteatoma formation.

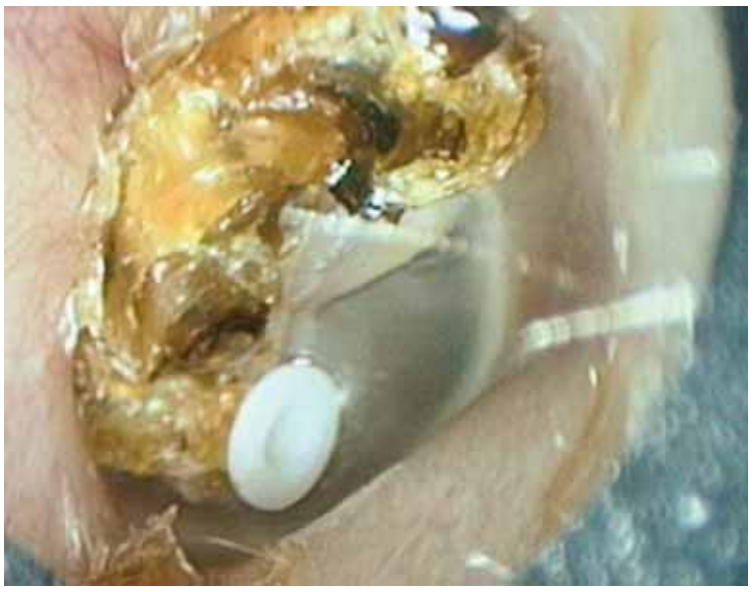

Figure 6: Otoscopic view of a right ear with extruding grommet and attic debris. With intermittent discharge and pain, this patient was eventually noted to have a cholesteatoma underneath the attic debris

clear view of the region above this area to identify most cholesteatomas. Typically this area is called the attic region and is clean and smooth with a few blood vessels noted.

If this region is not smooth and free of wax and or debris, a cholesteatoma may be suspected (see figures 5 and 6), especially if the patient has symptoms.

Red flags in diagnosis

- Meningitis-type symptoms — headache, photophobia, stiff neck.
- Cranial nerve deficits.
- Sensorineural hearing loss
- Deep ear pain, particularly in immunocompromised patients.

Differential diagnosis of tympanic membrane perforations

Safe perforations

will not progress to significant morbidity.

- Dry central perforations — away from the attic or the annulus margins (see figure 7).

Unsafe perforations

may progress to significant morbidity.

- Attic perforations.
- Marginal perforations — near the annulus of the ear drum (see figure 8).

Management

Most cholesteatomas will require surgical management, but this is generally not urgent unless intracranial complications have occurred.

In small cholesteatomas, frequent toileting of the cholesteatoma may delay the requirement for surgical management or help avoid

Congenital cholesteatoma

CHOLESTEATOMA is defined as congenital in the presence of an intact tympanic membrane, with no history of ear discharge, surgery or perforation of the tympanic membrane.

These criteria mean that even a paediatric otologist can expect fewer than 1 in 20 cholesteatomas they diagnose to be congenital.

Congenital cholesteatoma is commonly thought to be caused by the retention of epithelial cells in the middle ear during fetal development. Known as embryonic cell rest theory, this explains why the cholesteatoma is present at birth, with gradual expansion until the cholesteatoma is diagnosed because of the development of symptoms or as an incidental finding.

Regardless of aetiology, cholesteatoma is a problem because of its relentless expansion. Bone is no barrier to cholesteatoma expansion, with the inflammatory infiltrate at the advancing margin producing proteolytic enzymes as well as attracting and stimulating osteoclasts.

Without surgical management, this can eventually lead to destruction of vital structures such as the otic capsule, facial nerve, invasion of dural venous sinuses and the middle and posterior cranial fossae causing intracranial symptoms.

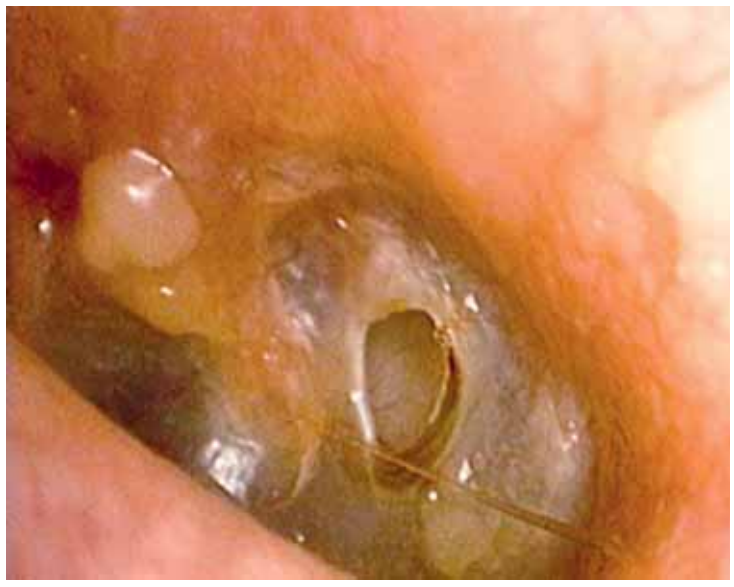

Figure 7: Otoscopic view of a left ear with a dry central perforation. This is a safe ear.

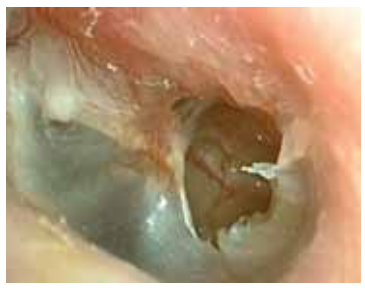

Figure 8: Otoscopic view of a left ear with a marginal unsafe perforation. If left untreated this ear may develop into cholesteatoma.

Whichever technique is chosen, the main purpose is remove the disease and create a safe, dry ear.

from previous page

surgery in patients unfit for anaesthesia.

When managed surgically, the exact approach will vary according to several factors, including the extent of disease, patient anatomy and surgeon's experience with various techniques.

Broadly speaking, two techniques are offered: either conservative (intact canal wall mastoidectomy/atticotomy) or radical (modified radical mastoidectomy/radical mastoidectomy).

Whichever technique is chosen, the main purpose is remove the disease and create a safe, dry ear. A secondary benefit may be restoration of hearing.

Modern surgical techniques seek to minimise alteration of middle ear anatomy wherever possible in less extensive disease. More recently, the use of intraoperative endoscopy has meant that the extent of disease can be further evaluated using a transcanal/endaural approach (through the ear canal), which can sometimes reduce or avoid a post auricular scar.

Surgical management of cholesteatoma is often staged in an attempt to reconstruct hearing 9-12 months later if there is no recurrence of disease. A downside to this is the need for a second surgery.

Newer research suggests that MRI — in particular non-echoplanar diffusion-weighted MRI — can be utilised to image ears prior to second-look surgery, with good sensitivity for residual or recurrent cholesteatoma in postoperative ears.

As well as predicting the presence and location of cholesteatomas in second-look surgery, MRI can be utilised in the surveillance of ears that would not otherwise be readily examinable, such as ears in which the ossicular chain has already been reconstructed, or the mastoid cavity has been deliberately obliterated.

When to refer

- Attic or marginal perforation (especially if patient is symptomatic).
- Central perforation that is symptomatic (discharge or hearing loss).
- Painful, discharging ear where the attic cannot be clearly visualised.

Dr Sale is a Cochlear Implant Research Fellow at the Kolling Deafness Research Centre at the University of Sydney. Dr Patel is associate professor of surgery at Macquarie University, clinical senior lecturer at the University of Sydney, and director of the Kolling Deafness Research Centre.

Acknowledgements

The authors thank Dr John Kelly for providing figures 3, 5, 6 and 7.

NBM1057-22.02.11-AU. 9086-AD