

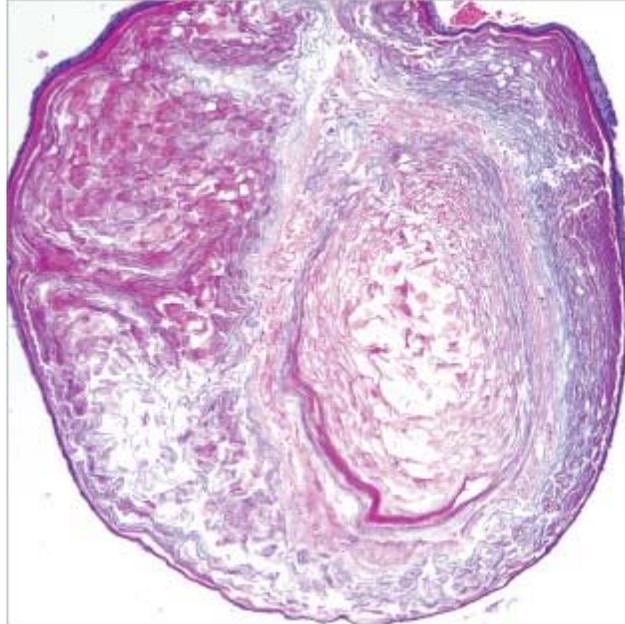
Cholesteatoma

by Gabriel Caponetti, MD, Lester D. R. Thompson, MD, and Liron Pantanowitz, MD

Cholesteatoma is a lesion formed from keratinizing stratified squamous epithelium. It may present intradurally (an epidermoid) or extradurally. Extradural lesions most commonly involve the middle ear cleft; involvement of the mastoid or external auditory canal is less common. The term cholesteatoma is actually a misnomer as these masses rarely contain cholesterol. Although they are not true neoplasms either, clinically they can mimic malignant neoplasms because of their propensity to destroy surrounding tissue and recur after excision.

Cholesteatomas can be congenital (primary) or acquired. Congenital lesions are uncommon; they are seen in young children, they occur in the anterosuperior part of the middle ear, they measure 3 mm or more in diameter, and they usually present as a pearly-white, closed keratinous cyst behind an intact tympanic membrane (figure 1). They likely arise from an epidermoid cell rest. Congenital malformations (i.e., cleft palate) are often accompanied by congenital cholesteatomas; in such cases, the cholesteatoma is more likely to be bilateral.

Figure 1. This congenital cholesteatoma appears as a closed (intact), squamous-epithelium-lined cyst that is filled with keratinous material.



Acquired cholesteatomas are much more common, and they typically occur in older children and young adults. Acquired lesions are usually associated with chronic otitis media, they are primarily unilateral (90% of cases), and they present as a white-gray or yellow irregular and open (noncystic) mass lesion behind a perforated tympanic membrane, often in the upper and posterior part of the middle ear. They may be accompanied by otorrhea, conductive hearing loss, otalgia, and possibly signs indicative of advanced destruction (e.g., facial nerve palsy, vertigo, meningitis, or brain abscess). It is unclear whether they are acquired from squamous metaplasia of the middle ear mucosa, invagination (retraction) of the tympanic membrane, invasion of squamous epithelium into the middle ear, or as a result of trauma.

By histology, a combination of keratinous material and stratified squamous epithelium is required to make a pathologic diagnosis of cholesteatoma (figure 2). The presence of squamous epithelium in the middle ear (which is normally lined with glandular epithelium) is abnormal. The abundant anucleate (dead) keratin squames account for the flaky, pearly-white otoscopic (gross) appearance of cholesteatomas. Unlike the epidermis of the skin, the squamous epithelium does not contain adnexal structures or rete ridges. There may be adjacent inflamed granulation or fibrous tissue, as well as a giant-cell reaction to keratin material (figure 3). Recent studies have shown that immunohistochemical staining for cytokeratin 16 (a marker for hyperproliferative keratinocytes) and Ki-67 (a marker of proliferative activity) shows strong expression in cholesteatomas. Extra numbers of chromosome 7 detected by fluorescence in situ hybridization (FISH) also appears to correlate with proliferative activity. Extradural cholesteatomas must be differentiated from squamous cell carcinoma (which has malignant squamous epithelium) and cholesterol granuloma (which contains cholesterol clefts).

Figure 2. On histology, the cholesteatoma shows bland, keratinizing, stratified squamous epithelium and anucleate squames. Note the lack of rete.

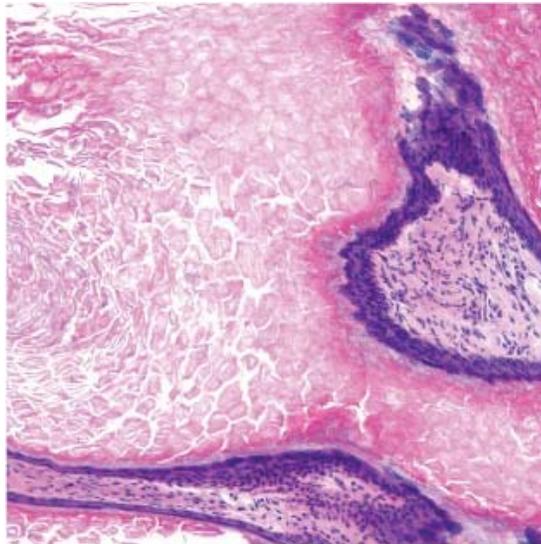
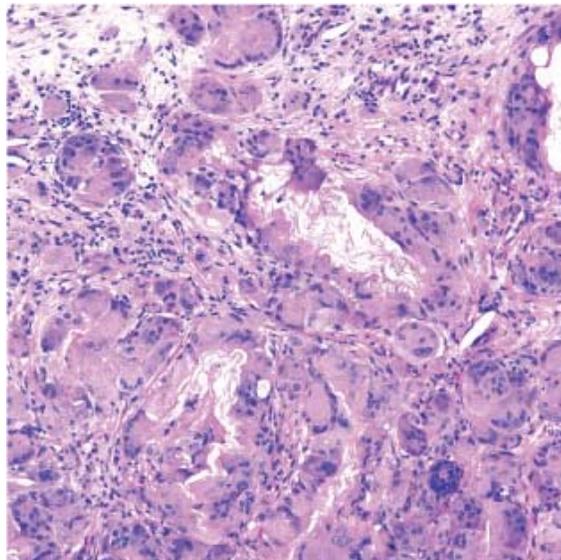


Figure 3. In this cholesteatoma, a granulomatous giant-cell reaction is seen in response to keratin material.



Complete excision or exteriorization is the goal of treatment. However, cholesteatomas may recur, especially if they are incompletely excised and if the patient is younger than 20 years and has extensive disease with marked ossicular erosion and associated polypoid mucosal inflammatory disease.

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Suggested reading

1. Adair C. Non-neoplastic lesions of the ear and temporal bone. In: Thompson LDR, ed. Head and Neck Pathology. Philadelphia: Elsevier Health Sciences; 2006:371-96.
2. Semaan MT, Megerian CA. The pathophysiology of cholesteatoma. *Otolaryngol Clin North Am* 2006; 39 (6): 1143-59.