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Endoscopic management of congenital cholesteatoma

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Congenital cholesteatomas are epithelial cell rests found within the middle ear or mastoid that are present behind an intact tympanic membrane. Typically found early in life, these expansile skin cysts can result in conductive hearing loss by impeding or eroding the ossicular chain. If left unaddressed, congenital cholesteatoma can result in erosion of the bony labyrinth, tegmen, or facial nerve resulting in significant patient morbidity. Surgical removal of congenital cholesteatoma is recommended, preferably when the lesions are small. When considering a surgical approach for the removal of congenital cholesteatoma, surgeons should consider the size and location of the lesion as well as the extent of expected reconstruction. Transcanal endoscopic ear surgery can successfully be used to manage congenital cholesteatoma limited to the tympanic cavity. Patient features that favor an endoscopic approach include a circumscribed, unruptured congenital cholesteatoma, intact external auditory canal wall, and absence of secondary complications such as labyrinthine fistula, middle fossa extension, or facial nerve paresis. We describe our endoscopic operative approach to remove congenital cholesteatoma and also highlight effective methods of tympanic reconstruction.

Introduction

Aural congenital cholesteatomas are keratinizing skin cysts found within the middle ear or mastoid in children without a history of middle ear disease. Thought to arise from rests of epithelial cells, congenital cholesteatomas typically occur within the anterior superior quadrant of the tympanic space. Derlacki and Clemis[1] first clinically defined congenital cholesteatomas as pearly white masses medial to an intact tympanic membrane. Their criteria included the findings of a normal pars tensa and pars flaccida, no history of otorrhea or tympanic membrane perforation, or prior otologic surgery. The incidence of congenital cholesteatoma varies by report, but in general it is thought to be quite rare at ~3% of all cholesteatoma cases.[2]

Some ambiguity exists regarding the diagnosis of congential cholesteatoma, as often the patient will not present until the cyst has grown large enough to result in local inflammation and rupture of the tympanic membrane, with resultant otorrhea. Absence of prior otologic disease, health of the contralateral ear, lack of a defined retraction pocket, and imaging characteristics may suggest congenital origin. In some cases, it may be impossible to be sure whether a cholesteatoma is acquired or congenital.

The management of congenital middle ear cholesteatoma is distinctly different from that of acquired cholesteatoma given that affected patients frequently have normal Eustachian tube function. As such, the tympanic membrane is often uninvolved and can be preserved, without the need for ventilation or cartilage reinforcement. When complete

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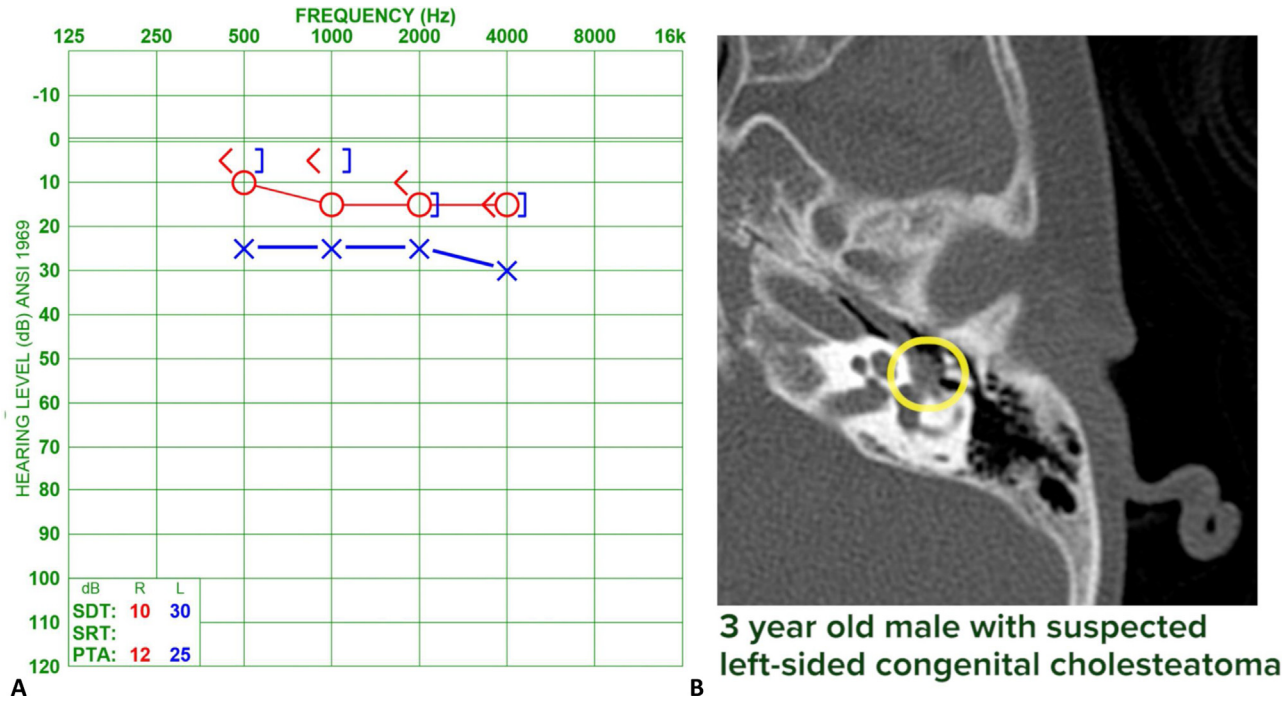

Figure 1 Preoperative evaluation of a patient with left congenital cholesteatoma. (A) 25 dB conductive hearing loss on threshold audiogram. (B) High-resolution CT scan demonstrates an opacification in the anterior superior quadrant, typical for congenital cholesteatoma. CT, computed tomography. (Color version of figure is available online.)

removal of the cholesteatoma is accomplished, rapid reconstruction of the ossicular chain is possible.

In this article, we will outline the workup of patients with congenital cholesteatoma, then discuss patient selection for transcanal endoscopic ear surgery. We will delineate the necessary steps for safe and effective endoscopic removal of congenital cholesteatomas and then describe the methods of tympanic reconstruction.

Patient evaluation

The evaluation of patients with congenital cholesteatoma begins with clinical history and physical examination. Although patients in the past were often not diagnosed until secondary complications such as hearing loss, vertigo, or facial weakness developed,[3] today most congenital cholesteatomas are diagnosed incidentally on routine otoscopy by pediatricians.[4] A white mass behind and intact tympanic membrane without other coincident middle ear disease is the most frequent finding.[4] Symptomatic patients typically present with unilateral hearing loss that is conductive in nature.

Threshold audiometry should be performed in children who are capable of participation. If there is a question about the reliability of threshold audiometry, evoked potentials can be used to obtain accurate air conduction thresholds (Figure 1A).

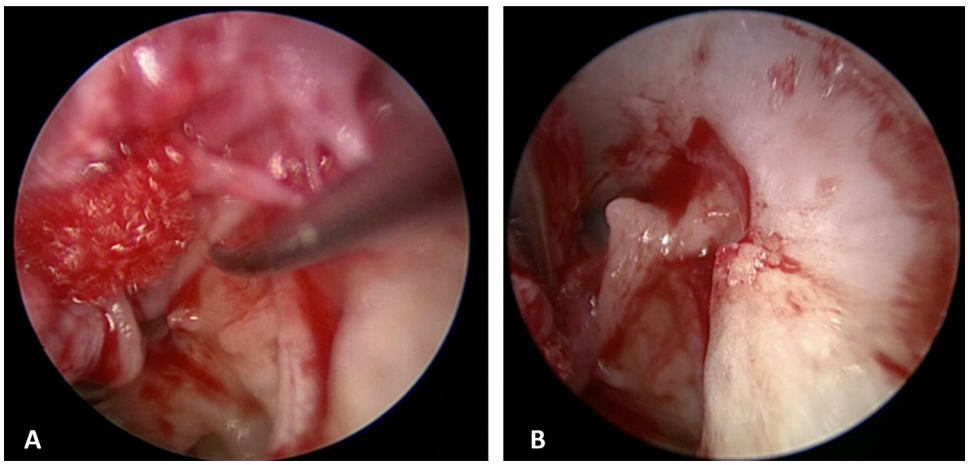

Figure 2 Inferiorly based tympanomeatal flap elevation, left ear. (A) Dissection along the manubrium of the malleus allows inferior folding of the tympanic membrane for visualization of the anterior quadrant of the tympanic cavity. (B) Lateral process of the malleus and anterior tympanic space visualized. (Color version of figure is available online.)