Case Report

Pediatric Middle Ear Congenital Cholesteatoma: A Case Report

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Congenital cholesteatoma (CC) is a rarely seen benign tumor of the temporal bone. There are five general sites of extradural occurrence: the middle ear, external auditory meatus, mastoid, squamous portion and the petrous apex of the temporal bone. CC grows slowly and presents no symptoms at the early stage. Delayed and mis-diagnosis are common with this condition.

Case report

A 10-year-old boy presented with a 3-month history of hearing loss on right side. There was no history of otorrhea, facial palsy, previous otological procedures or trauma. Otoscopy revealed a bulging posterosuperior quadrant in the otherwise intact right tympanic membrane (Fig. 1). Pure tone audiometry showed an average threshold of 51 dB for 500, 1000, 2000 and 4000Hz, with a 40 dB air-bone gap, suggesting a moderate conductive hearing loss (Fig. 4). CT scan of the temporal bone showed an isolated soft tissue density lesion in the middle ear (Fig. 2).

Following the 1986 Levenson diagnostic criteria for CC (i.e. intact tympanic membrane, no history of otorrhea or middle ear surgery), a working diagnosis of middle ear CC was made.

Middle ear exploration was performed under general anesthesia, via a postaural incision. The tympanic cavity was filled with cholesteatoma with extension into the facial recess and sinus tympani. The facial canal was intact, The malleus and incus were partially eroded, the suprastructures of the stapes were missing. The footplate itself was intact with good mobility. Residual malleus, incus and crus of stapes were removed. There was no cholesteatoma in the mastoid and posterior tympanic cavity, although local mucosa appeared slightly swollen (Fig 3 a b c). Following complete removal of cholesteatoma, the ossicular chain was reconstructed in a type III tympanoplasty fashion using...
a piece of tragus cartilage carved to fit between the residual malleus and footplate, which was covered using a piece of temporalis fascia graft. Histological examination of the specimen confirmed the diagnosis of middle ear CC. At 4-month follow-up, audiometric tests showed improved hearing (Fig 5).

Discussion

The middle ear is involved in 80% of CC cases, which can present at any age from infancy to adulthood. CC is believed to occur secondary to failure of normal involution of the embryonic epidermoid tissue. This disease develops not only slowly but also without symptoms, and is difficult to discover in the early stage. As the lesion grows in size, symptoms will develop, including conductive hearing loss, facial palsy and vertigo as a result of facial canal and semicircular canal damage. Mixed hearing loss can also occur.

The temporal bone pathology research by Mc Gill suggests that there are two types of middle ear CC. In the more common type, the lesion may be visualized in the anterosuperior quadrant of tympanic membrane, implying its location in the mesotympanum and hypotympanum. This type of CC does not affect the ossicular chain, and can present before 4-years of age. The other type is rare, in which the cholesteatoma lesion tends to appear in the posterosuperior quadrant of the tympanic membrane. Involvement of the ossicular chain, especially the incostapedial joint, is usually the rule in this type. Removal of such lesion often requires sacrificing the malleus and incus. This type can present in individuals older than 5-years of age.

The present case belongs to the second type,
where the cholesteatoma was mainly in the pos-
terosuperior portion of the tympanic cavity, including the facial recess and sinus tympani. The pa-
tient had mild hearing loss, which was mainly con-
ductive. CT scan located the lesion in the middle ear, with erosion of the ossicular chain and no inner ear involvement.

Treatment of CC entails surgical excision, once the diagnosis is confirmed. Children with extensive disease may require a second-stage procedure to reconstruct the ossicular chain. In the present case autologous cartilage was successfully used to reconstruct the ossicular chain with significant hearing improvement. The technique is simple and inexpensive, and can easily be adopted in the clinic. Its long-term results in treating middle ear CC need to be further assessed.

References