Discussion

Cholesteatomas are non-neoplastic, keratinizing squamous epithelial cysts that can develop in various locations, including the middle ear, mastoid, and eustachian tube. Congenital cholesteatoma is a rare condition that can involve the eustachian tube, presenting with chronic otorrhea and tympanic membrane perforation.

The most common presentation is an asymptomatic, white mass limited to the mesotympanum just adjacent to the eustachian tube. The progression of middle ear cholesteatoma usually takes the pathway of least resistance. We would expect the mesotympanum to be full of cholesteatoma if the lesion had initiated in the middle ear such as in cholesteatoma secondary to tympanic membrane perforation, prior to eustachian tube involvement.

The incidence of all cholesteatomas in children is 3 in 100,000 and the incidence of congenital cholesteatoma (CC) has been reported to be 0.12 in 100,000, but the actual incidence may be higher since the lesion can present at any age.

Ear fluid pH should be investigated. An alternative would be eustachian tube occlusion with pressure equalization tube placement to improve the outcome.

The most common location of CC is in the anterior superior quadrant of the middle ear, which would suggest a chronic process of disease, including remodeling of the eustachian tube during pre-partum development. The patient's symptoms started as early as one month after birth, which would not give sufficient time for development of such a massive cholesteatoma inside the eustachian tube with such a significant change in eustachian tube size.

Surgery with complete removal of squamous epithelium is the main treatment. With such extensive disease, a second look at 12 months after surgery should be considered to rule out any recurrence of her ear infection.

Although a small residual cholesteatoma was expected to be present, the postoperative course was uneventful with resolution of symptoms. The underlying pathology of the otorrhea was not clear. Differential diagnoses included: recurrent otitis media, chronic otitis media secondary to neutropenia, histiocytosis of the temporal bone, eustachian tube obstruction, and cholestochal cholesteatoma at other sites. Surgical management with complete removal of squamous epithelium is the main treatment. With such extensive disease, a second look at 12 months after surgery should be considered to rule out any recurrence of her ear infection.

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References