

Congenital Cholesteatoma Associated with Blue Eardrum

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Abstract. The classic presentation of congenital cholesteatoma is a pearl behind the anterior-superior quadrant of an intact tympanic membrane. Idiopathic hemotympanum is characterized by a dark blue eardrum, the most prominent feature of which is the presence of cholesterol granulomas. Blue eardrum is associated with eustachian tube dysfunction. Despite the well-established relationship between eustachian tube dysfunction and the development of pediatric cholesteatoma, little has been written concerning the appropriate timing of tympanostomy tube placement. To date, there are no reports of congenital cholesteatoma associated with blue eardrum. A recent case of advanced congenital cholesteatoma (stage IV) associated with blue eardrum was treated using preoperative tympanostomy tube insertion. Tympanostomy tubes were helpful in preventing recurrence of the cholesteatoma after surgery. The case is presented along with a review of the literature.

Congenital cholesteatoma involving the mastoid also typically involves all of the ossicles, is inconsistently controlled with canal wall-up tympanomastoidectomy and is characterized by poor prognosis with respect to treatment of conductive hearing loss. There is a strong association between stage and residual disease. Idiopathic hemotympanum is a term used to designate a chronic condition characterized by dark blue eardrum. The most characteristic feature in blue eardrum is the presence of cholesterol granuloma which was first reported by Ranger (1). Cholesterol granuloma may occur anywhere in the air

cell system of the temporal bone. It commonly occurs in cases of chronic otitis media with effusion (OME) and eustachian tube blockage.

Otoscopically, middle ear congenital cholesteatoma appears as a whitish globular mass lying medial to an intact tympanic membrane. Typically, there is no history of inflammatory otitis media; however, there may be a history of OME related to blockage of the eustachian tube by the cholesteatoma. The mastoid bones of patients with congenital cholesteatoma are usually well aerated compared with those in patients with a history of recurrent or chronic middle ear suppuration (2-4). To date, congenital cholesteatoma associated with middle ear effusion has only rarely been reported.

Poistic *et al.* (5) suggested four stages for the classification of middle ear congenital cholesteatomas: stage I, disease confined to a single quadrant; stage II, cholesteatoma in multiple quadrants, but without ossicular involvement or mastoid extension; stage III, ossicular involvement without mastoid extension; and stage IV, mastoid disease. There is a strong association between stage and residual disease, ranging from a 13% risk in stage I to a 67% risk in stage IV. Significant risk factors for recurrent cholesteatoma in pediatric patients include male gender, pars flaccida type of cholesteatoma and association of otitis media with effusion either in the affected side or in the contralateral side (6). Therefore, the dark blue effusion induced by cholesterol granuloma formation may be one of the risk factors associated with the recurrence of pediatric cholesteatoma. However, little has been written concerning the appropriate timing of tympanostomy tube placement, despite the well-established relationship between eustachian tube dysfunction and pediatric cholesteatoma. A case of advanced congenital cholesteatoma (stage IV) associated with blue eardrum came to our attention. The patient was managed with preoperative tympanostomy tube insertion, which was helpful in preventing recurrence of the cholesteatoma after surgery. The details of this case are hereby reported in addition to a review of the literature.

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Case Report

A 14-year-old boy presented with left-sided hearing loss and middle ear fullness. Otoendoscopy of the left ear revealed a typical blue eardrum with whitish mass in a posterior-inferior location (Figure 1). The patient had no history of otological surgery, tympanic membrane perforation or otorrhea. Mean pure tone threshold was 40 dB and tympanogram was B type. A ventilation tube was inserted, which resulted in evacuation of dark brown effusion fluid (Figure 2). No recurrence of blue eardrum was noted during the subsequent follow-up period of two months, but the white mass remained behind the tympanic membrane. A temporal bone CT was taken two months after ventilation tube insertion; it showed soft tissue density in the attic and mastoid antrum (Figure 3). Intact canal wall tympanomastoidectomy was subsequently performed with the patient under general anesthesia. After elevation of the tympanomeatal flap, a white keratin mass was found at the posterior tympanum. The cholesteatoma did not obstruct the eustachian tube. The mastoid cavity was drilled and the important anatomic landmarks were identified. The sigmoid sinus and the mastoid tegmen were skeletonized. Upon drilling of the mastoid cortex, a cholesterol granuloma was observed to be penetrating rather closely to the surface of the cortex. This was completely drilled out and exenterated from the rest of the mastoid cell central tract. The cholesteatoma sac was easily visible upon entry into the antrum. This cholesteatoma sac was debulked and mobilized anteriorly, and a drill was used to open the surgical field further into the aditus ad antrum and to continue anteriorly into the epitympanum. The posterior canal wall was skeletonized down toward the facial ridge posteriorly. Once the mastoid cavity was exenterated of all diseased cells and the cholesteatoma had been mobilized toward the middle ear, the dissection was continued from the middle ear. Using a 45-degree Rosen needle and suction, the cholesteatoma was mobilized and removed from the middle ear. It was particularly adherent in the tympanic sinus and a whirlybird was used to help remove it. At this point, a 30-degree endoscope (4 mm diameter) was used. The cholesteatoma sac, which was found to extend into the hypotympanic cells, was carefully removed from the round window niche. The stapes footplate was intact. To ensure complete removal of the cholesteatoma from the tympanic sinus, a facial recess approach was used. The long process of the incus was eroded by the cholesteatoma, so the incus and the head of the malleus were removed. After thorough irrigation, a silastic sheet was placed in the middle ear space, a rosebud dressing was placed and the soft tissue and skin were re-approximated.

Nine months after the operation, the ventilation tube had extruded spontaneously and no recurrence of blue eardrum was observed (Figure 4). A year and a half after the

operation a temporal bone CT revealed no recurrence of cholesteatoma (Figure 5). The patient is scheduled to have a second stage ossiculoplasty, two years' postoperatively.

Discussion

The pathogenesis of blue eardrum is controversial as it is still undetermined whether it is a variant form of OME or is an independent clinical entity. Histological examination shows that blue eardrum is the sequela of hemorrhage in the form of brownish secretions, cholesterol granulomas, foreign body giant cells and macrophages. A number of possible causes have been suggested, and the etiopathogenesis has been discussed mainly in relation to OME. However, some authors (7, 8) believe that OME and blue eardrum might be different phases of the same disease process. Congenital cholesteatoma cannot be considered a consequence of cholesterol granuloma. On the contrary, acquired cholesteatoma produces a lack of aeration in the mastoid air cells with subsequent hemorrhage and this gives rise to the formation of cholesterol granulomas (9). The most common sites of cholesterol granuloma formation are the epitympanum, round window, mastoid and tympanic sinus. They are found slightly more frequently in the tympanic cavity than in the mastoid (10). Arachidonic acid metabolites play a role in bone resorption of cholesteatoma (11) and the level of arachidonic acid metabolites such as 12-hydroxyeicosatetraenoic acid has been found to be higher in cholesterol granulomas compared to the granulation tissue seen in chronic otitis media (12).

Congenital cholesteatoma involves the mastoid and may be associated with ossicular erosion and moderate conductive hearing loss. It is best approached via a canal wall-up tympanomastoidectomy. However, the management of blue eardrum with cholesteatoma has been rarely reported. In the present case, the patient was initially treated using ventilation tube insertion. Interval tympanomastoidectomy was then performed for congenital cholesteatoma. Blue eardrum and congenital cholesteatoma can be treated simultaneously. Most surgeons find it easier to tease a cholesteatoma from the recesses of the mesotympanum and the surfaces of the ossicles in a dry, pneumatized middle ear. A bloodless field during definitive cholesteatoma surgery makes it less likely that portions of a pediatric cholesteatoma will be left behind and thus less likely that residual disease will appear.

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Figure 1. Otoendoscopic examination showing a dark blue color associated with a whitish mass at the postero-inferior quadrant of the left side.

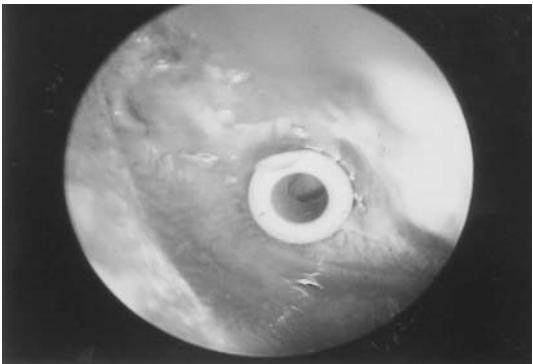


Figure 2. Otoendoscopic examination showing an inserted ventilation tube without recurrence of a dark blue-colored eardrum.

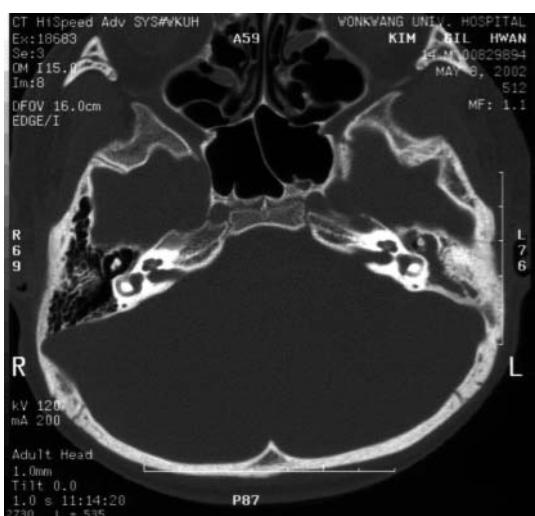


Figure 3. Temporal bone CT (axial view) showing haziness in the epitympanum and mastoid cavity.

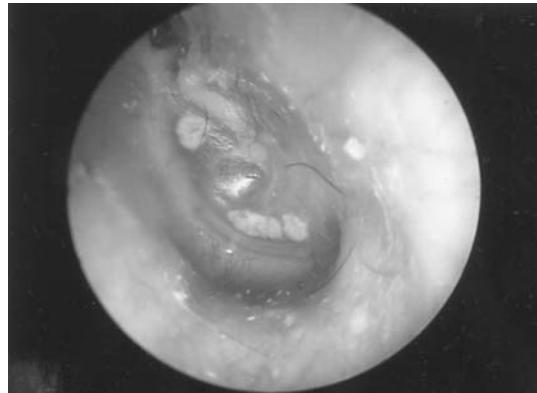


Figure 4. Otoendoscopic examination showing an intact eardrum without recurrence of blue eardrum or congenital cholesteatoma. Ventilation tube-induced tympanosclerosis was noted.

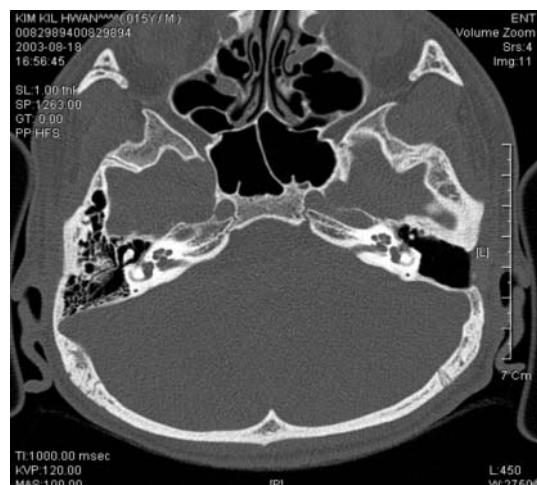


Figure 5. Temporal bone CT at a year and a half after operation, showing a well-ventilated tympanomastoid cavity without recurrence of cholesteatoma.

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