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CASE REPORT

Primary Cholesteatoma within the Stenotic Ear Canal: Report of two Cases, one with Extra-Temporal Complications

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Congenital aural stenosis can be complicated with primary cholesteatomas which may be overlooked until the occurrences of resultant complications or incidentally diagnosed during or before corrective surgeries.

In this report, samples of both "complicated" and "occult" types of cholesteatomas associated with aural stenosis are presented with the aim of illustrating their clinical course and shedding light on their possible pathogenesis.

We herewith report 2 patients with congenital aural stenosis who developed cholesteatoma related to the external ear canal (EEC) stenosis. One of the cases, had previously undergone craniotomy for intracranial complication of the disease, and also had postauricular fistula, whereas in the second patient it was silent and confined within the relatively enlarged stenotic ear canal in front of a rudimentary tympanic membrane.

In the first case cholesteatoma was removed with mastoidectomy in the first phase and then atresioplasty was carried out with tympanoplasty. In both cases, a novel two-stage technique was used for EEC reconstruction with success.

Congenital aural stenosis predisposes cholesteatoma most likely due to entrapping of the squamous epithelium within the stenotic EEC or tympanum, and these cases were more prone to the extra-tympanic complications of cholesteatoma than those with the normal EEC.

In conclusion, otologists should hold a high grade of suspicion of cholesteatoma in a patient with stenotic EEC, and their presence should be ruled out prior to any definitive corrective surgery.

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Congenital external earcanal (EEC) stenosis is classified as a sub-group of congenital aural atresia. Schuknecht divided aural atresia into 4 types: meatal atresia (Type-A), partial atresia with narrow tortuous EEC (Type-B), total atresia (Type-C) and with hypo-pneumatic mastoids (Type-D), Type B of which can also be defined as aural stenosis.[1] Similarly, Cremers et al classified EEC malformations as Type I: mild, Type II: medium and Type III: severe atresia. The Type I was described as a small canal usually ending up with a rudimentary tympanic membrane corresponds with EEC stenosis.[2] In both reports, EEC stenoses were associated with high risk of primary cholesteatoma occurrence. According to the Levenson's criteria of congenital cholesteatoma; cases of primary cholesteatoma with canal atresia/stenosis as well as intramembranous cholesteatomas and giant temporal bone cholesteatomas are considered as subgroups different from the main group of congenital cholestetomas.[3]

Case Reports

Case 1

A 20 year old male was referred to our clinic with right otorrheae and bilateral hearing difficulty that was more severe on the right side. He had had an emergency surgery for the drainage of otogenic subdural abscess in the posterior cranial fossa some 5 years ago. Large craniotomy defect and incision scar on the right tem-

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poro-parietal region was detected. However, neither an incision scar was present nor a mastoidectomy cavity was noted on CT scan as to indicate a previous mastoid surgery. He had bilateral stenotic meatae at the diameter of around 3 mm. He had right postauricular cutaneous fistula in the center of an area of enduration and purulent otorrhea from the ear canal on the same side. His auriculae were also slightly deformed (Figure 1). Computerized tomography (CT) of the temporal region showed a large radiolucent area in the mastoid cortex as well as an image of a mass in the tympanum strongly suggestive of cholesteatoma (Figure 2). His pure tone hearing threshold was measured (the avarage of 0.5, 1 and 2 kHz thresholds) as air conduction (AC)/ bone conduction (BC): 67/13 dBHL.

He was operated under general anesthesia primarily for the eradication of the concomitant chronic otitis media with cholesteatoma and secondarily for the repair of atresia. A spindle shaped postauricular skin incision was made. The cutaneous fistula and its subcutaneous tract were excised with the surrounding granulation tissue. Mastoid cortex was found defective and thinned as expected, under which a giant cholesteatoma was found extending from the sinus plate to the mastoid tip; filling rudimentary aditus ad antrum and posterior mesotympanum. Mastoid tegmen was found partly eroded and replaced by fibrous tissue. Initially, no ossicular structure was identified but a mobile stapes footplate. Fallopian canal was also eroded in the tympanic part by the cholesteatoma. There was rudimentary pouch in place of EEC, 2 cm deep and 4 mm wide at the distal end, where it was deficient and infected by the cholesteatoma located in its close proximity. Cholesteatoma was removed totally by intact canal wall procedure with posterior tympanotomy. Antrum was used as landmark and hugging the tegmen, EEC was drilled anteriorly leaving a thin bridge so that the tympanum was entered. A remnant of a rudimentary malleus was identified behind the atresia plate. The canal was widened as to reach the diameter of 1.5- 2 cm. A piece of bone harvested from the mastoid cortex was sculpted to the size and placed between the remains of the malleus and stapes footplate in order to create a columellar effect and to support the tympanic membrane graft. A piece of autologous temporal muscle fascia graft was prepared as tympanic membrane and peripherally tacked into the bony sulcus circumferentially carved at the ideal level of the annulus. Atresioplasty was completed by a wide meatoplasty, during which a large portion of conchal cartilage and infected part of the EEC skin stump were excised. Bony canal wall was covered by split thickness skin graft obtained from the leg.

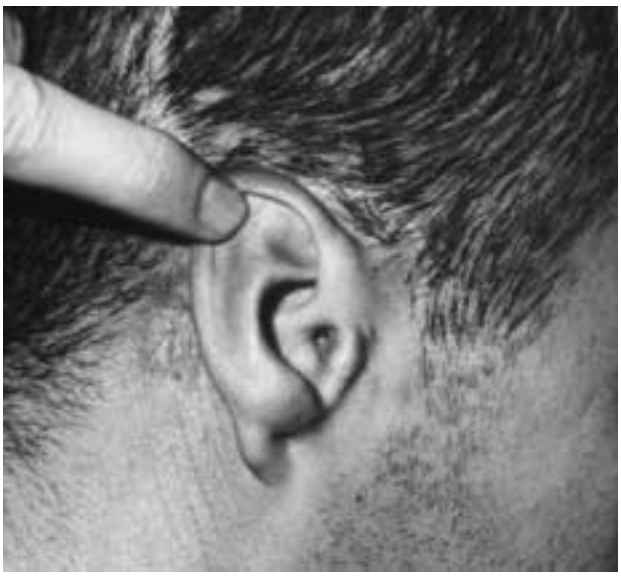

Figure 1. Pre-operative view of the stenotic ear canal. Note temporo-parietal incision scar and postoperative fistula (Case 1).

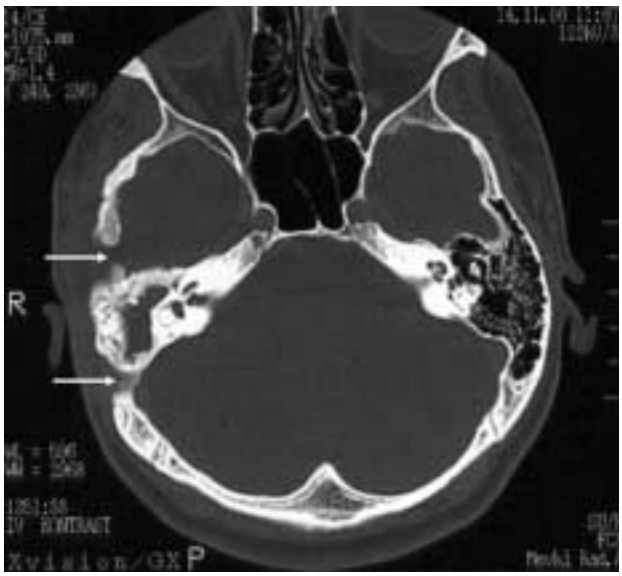

Figure 2. Tympano-mastoid cholesteatoma is seen with the cranial defect (arrows) (Case 1)

Three months after the surgery, the newly created ear canal was found considerably narrowed and a revision surgery by a novel two-stage technique was planned and carried out. At the first stage, excess fibrous tissue was removed from the meatus subcutaneously and the ear canal was cleared off from the scar tissue. The bony EEC was widened and its surface was smoothed. Lateralized tympanic membrane graft was re-positioned, and fixed reconstructed ossicular graft was mobilized. No recurrence of cholesteatoma was detected. Finally, the re-constructed canal was lined with the synthetic skin substitute (Epigard®, Medisave Medical Products, Wiesbaden, Germany) cut to the size. Second stage of the operation was carried out two weeks later. Synthetic skin substitute (SSS) was detached and removed carefully and underlying bleeding canal wall is covered with a tube of split-thickness skin graft harvested from the leg. During the first 4 months, the newly created ear canal kept slightly re-narrowing, however during the following months and years its patency was maintained. (Figure 3) At the first postoperative year follow-up, AC/BC thresholds were assessed as 53/12 dBHL, which meant a 13 dB HL gain in air-bone conduction gap (ABG).

Case 2

A 26 year old man was admitted in 2007 with the right ear canal stenosis and hypoacusis on the same side. The size of the ear canal was about 3 mm wide and his pinna was slightly deformed. Preoperative audiometric assessment (0.5, 1 and 2 kHz average) showed a pure tone threshold of AC/BC: 62/12 dB HL. Preoperative CT scan showed the stenotic ear canal filled with a hypodense material that seemed to enlarge the canal in antero-posterior direction (Figure 4). Atresioplasty was carried out using the two stage technique described above, initially drilling the ear canal as to leave the surfacing to the second phase. The material filling the EEC was a mass of cholesteatoma located in front of a rudimentary tympanic membrane that was macerated but intact (Figure 5). It seemed to be originated from the antero-inferior tympanic recess, extending towards the mastoid by partly eroding the posterior canal wall. Cholesteatoma was cleared, ear canal was widened, tympanic membrane was reconstructed using a thin slice of auricular cartilage and temporalis muscle fascia autograft. Ossicular chain was intact and mobile, yet malleus was deformed. Reconstructed ear canal was covered with SSS to be replaced by split-thickness skin graft at the second stage in two-week time.

His postoperative pure tone hearing threshold was measured as AC/BC: 27/4 dB HL 6 months postoperatively, and the gain in the ABG was 23 dBHL.

Discussion

Cholesteatoma formation in the atretic ears seems to occur in two different patterns. In EEC stenosis, that is

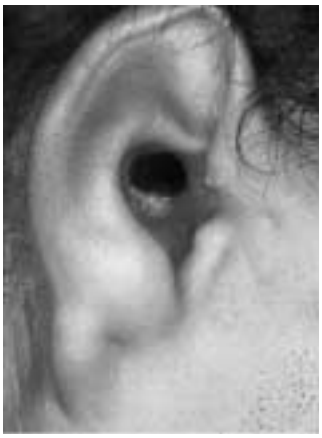

Figure 3. 4th postoperative month view of the constructed EEC meatus. Note auricular deformity (Case 1).

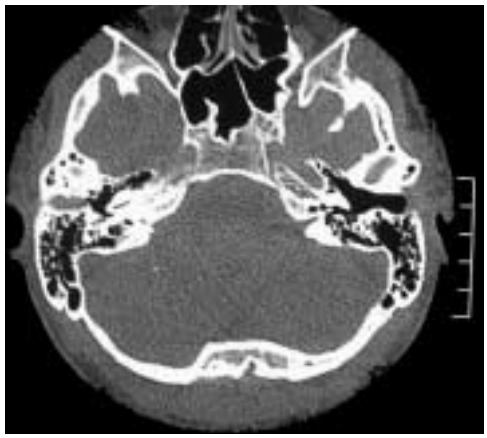

Figure 4. Cholesteatoma in the ear canal. Note rudimentary tympanic membrane (Case 2).

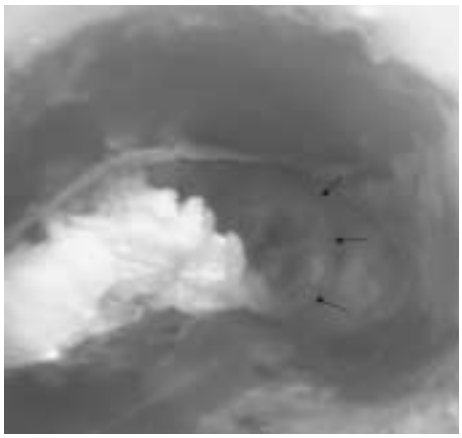

Figure 5. Cholesteatoma in the ear canal. Note rudimentary tympanic membrane (Arrows) (Case 2).

defined as an ear canal with the diameter of 4 mm or smaller, was attributed to the invagination of a small pouch lined with ectoderm within the canal itself into which cholesteatoma grew. The second mode is the development of the cholesteatoma medial to the atretic plate or inside the tympanic cavity. The occurrence of this type is explained by the trapping of the epidermoid elements during the embryological arrest of the EEC formation as to form a nidus for the cholesteatoma behind the atresia plate.[4] This theory is known as "embryological rest" theory and was credited as one of the possible mechanisms of congenital cholestetoma development in general.[5]

Zalzal reported a case of a 2 year old boy with congenital aural atresia complicated by acute mastoiditis and lateral sinus thrombosis due to concurrent cholesteatoma.[6] Nishimura et al reported another cholesteatoma case with aural atresia, arising behind the atresia plate that also caused mastoditis and subperiostal abcess.[7] Gill reported only one case of his 95 congenital aural atresia patients having mastoiditis and cholesteatoma.[8] Cole and Jahrsdorfer reported the largest series of cholesteatoma and aural stenosis association. They found 48% of their EEC stenosis patients (with the ear canal smaller than 4 mm diameter) developing cholesteatoma and none of them were under the age of three. More strikingly, they found cholesteatomous developments in 91 % (10 out of 11) patients with ear canals at the diameters of 2 mm or less. They included squamous trappings along with real cholesteatomas in their series, as mostly the patients at the age of 20 or older demonstrated bony erosions due to the direct effect of cholesteatoma.[9] We also had a case with acquired meatal stenosis with keratinous debris trapped behind the stenotic aural segment and this patient was not considered as having cholesteatoma as there was no discernable matrix unlike the above presented cases. Schuknecht reported 3 cases of occult cholesteatoma of his 11 cases (27%) with partial canal atresia or canal stenosis, in one of whom cholesteatoma was in the middle ear. 1 Caughley et al incidentally found an occult cholesteatoma within the middle ear in a child with complete EEC atresia.[10]

Our patients in conjunction with the reported similar cases in literature confirm that cholesteatoma is more commonly associated with EEC stenosis than atresia and they seem to be developing through more than one etiopathogenetic mechanism. Hence, the locations of cholestomatous developments in both of our cases suggested invagination of epithelial pouch as they were in close contact with the ear canal skin stump. This co-morbidity is most likely due to the existence of a space in stenotic ear canals where epidermal trappings can occur as to form a cholesteatoma nidus that is more freely develop than the completely atretic ear canals.[11] When cholesteatoma is located behind atretic or stenotic canal segment, the risk of extra- temporal complications, such as mastoiditis, cutaneous fistula and intracranial extensions are greater due to the hindrance of growth and drainage towards the non-existent or severely narrowed EEC.

The novel two stage technique we developed for the repair of EEC atresia and stenosis is based on the support of the graft by the naturally growing and smooth local soft tissue, stimulated by the synthetic skin substitute, which constitutes a more hospitable graft bad in comparison to the bare cortical bone. We have observed that the results are favorable, as it did not result in re-stenosis and infection. Our functional result, on the other hand, was modest for the first patient as his sound conduction system was severely affected by both congenital deformity itself and chronic otitis media with cholesteatoma. However, the audiometric gain achieved in the second patient was rated as satisfactory.

Stenosis of the EEC, therefore, should alert the otologist of the possibility of cholesteatoma occurrence, indicates a high resolution CT scanning and when found, constitute an absolute indication of reconstructive surgery and cholesteatoma removal in order to prevent potential complications.[12] However, for the uncomplicated pediatric cases, Schucknecht suggested watchful waiting until an age when the child becomes more cooperative during the postoperative follow-ups.[1]

Our observation is that the above described two stage technique with posterior (mastoid) approach is suitable for cases with EEC stenosis and cholesteatoma as it reduces the chance of re-stenosis and allows a second look at which residual cholesteatoma can be cleared.

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