

COMPLETE ENDOSCOPIC EXCISION OF STAGE 4 CONGENITAL CHOLESTEATOMA, A CASE REPORT

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Abstract

KEY POINTS 1. Congenital cholesteatoma (CC) is a relatively uncommon condition, accounting for 2-5% of all cholesteatomas. 2. Endoscopic ear surgery is commonly employed for limited cholesteatomas, usually Potts stage 1 and 2. 3. This is the first reported case in the literature utilized a transcanal endoscopic ear surgery approach for an 11-year-old child with POTSIC 4 Congenital cholesteatoma, ensuring complete cholesteatoma removal and subsequent area reconstruction. 4. Endoscopic approaches offer advantages in terms of visualization and minimizing the need for external incisions and postoperative scarring, CC, improving the quality of life.

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1. Congenital cholesteatoma (CC) is a relatively uncommon condition, accounting for 2-5% of all cholesteatomas.
2. Endoscopic ear surgery is commonly employed for limited cholesteatomas, usually Potts stage 1 and 2.
3. This is the first reported case in the literature utilized a transcanal endoscopic ear surgery approach for an 11-year-old child with POTSIC 4 Congenital cholesteatoma, ensuring complete cholesteatoma removal and subsequent area reconstruction.
4. Endoscopic approaches offer advantages in terms of visualization and minimizing the need for external incisions and postoperative scarring, CC, improving the quality of life.

Introduction

Congenital cholesteatoma(CC) is a relatively uncommon condition that has an incidence of 2-9 per 100000 and consists of 2-5% of all cholesteatomas. The classical description is the presence of a whitish mass medial to the intact tympanic membrane, with no previous history of discharge, perforation, or ear surgery. In 66% of cases it is diagnosed during procedures like grommet insertion and incidentally in the other 34%.(1)

Two types of CC are described in the literature Open type Congenital cholesteatoma (OTCC) and Closed type congenital cholesteatoma (CTCC). Closed CC contains epithelial cyst without keratin exposure whereas OTCC exposure of cholesteatoma matrix occurs which forms part of middle ear mucosa. (2) The closed type can rupture and lead to an Open type CC. The OTCC is technically more challenging. Conventionally POTSIC system is used (3)(Table 1) for staging, another staging system used is by the Japan Otological Society staging system for congenital cholesteatoma. (4) (Table 2). Endoscopic ear surgery is described in the literature for POTSIC 1 and 2 stages, and according to the authors knowledge this is the first reported case

of POTSIC stage 4 congenital cholesteatoma which was operated through a complete transanal endoscopic technique, and the patient is in follow up for past 3 years without any recurrence.

Method

This case report was reported according to CARE guidelines. This report has been approved by the Institutional review board and appropriate consent from the patients parents are taken . Institutional ethics committee clearance has been taken for the same.

Study setting : Tertiary care institute in India.

Case Report : An 11-year-old child was presented to the outpatient department with a history of impaired hearing for one year. No previous history of ear discharge, vertigo, or tinnitus was present. On otoscopic examination, there was a pearly white mass behind the intact tympanic membrane (TM) occupying all 4 quadrants. A provisional diagnosis of congenital cholesteatoma was made which was confirmed by an otoendoscope. Rinne's test was negative on the right side and weber was lateralized to right with a 512 Hz tuning fork. In pure-tone audiometry, the air conduction and bone conduction pure tone average for 250 Hz, 500 Hz, 1000 Hz, and 2000 Hz in the right ear were 32 dB and 15 dB respectively. Hearing levels in left ear were normal.

HRCT Temporal bone showed an intact tympanic membrane with soft tissue density in the middle ear extending to the attic and antrum with blunting of scutum and erosion of ossicles. **(Figure1)**

Surgical Procedure : Written Informed consent was taken from the parents of the child, Trans canal endoscopic ear surgery was performed under general anesthesia. 2.7mm 0-degree and 30-degree endoscopes were used. Head end elevated 15- 30 degrees. Endoscope light intensity was kept at 50% to avoid any thermal injury. Local infiltration was given with 2% xylocaine and 1:100000 adrenaline in the post auricular area and 4 quadrants of the external auditory canal. Tympano-meatal flap elevated from 6'o clock to 12'o clock position. Middle ear entered, the cholesteatoma sac was visualized involving all quadrants of the middle ear, the disease was dissected and its involvements were followed endoscopically. The sac was found to be going posteriorly. The attic, aditus, antrum, and posterior wall of EAC were drilled to follow the disease and the disease was removed completely from the middle ear and mastoid. The sac removed in its entirety and its complete removal is ensured by checking with a 30-degree endoscope. Sinus tympani, facial recess, anterior epitympanic space, and hypotympanum visualized. Incus was eroded and was removed, stapes supra structure was present. After ensuring the complete removal of the disease, hearing reconstruction is done. Type 3 tympanoplasty with ossicular reconstruction and Canal wall/attic defect was reconstructed using cartilage harvested from concha. The tympanomeatal flap was repositioned, and EAC is packed with gel foam and medicated ribbon gauze. **(figure 2) (Video of the procedure in the supplementary information section)**

Results : Complete removal of the cholesteatoma was done with same setting hearing reconstruction by type 3 tympanoplasty and soft wall reconstruction.

Later the pathological diagnosis was confirmed as noted by the keratin debris in the histopathology report. The child is still in follow up for past 3 years without any recurrence.

Discussion

CC most commonly occurs in the Anterior superior quadrant(82%) followed by the postero superior quadrant of the tympanic membrane(49%). In 43% of cases ossicular erosion and in 23% extension to the mastoid cavity is also reported. The increased rate of recurrence also with higher the stage of the disease is reported in previous studies, mostly because of the persistence of disease in hidden areas like sinus tympani and facial recess, anterior epitympanum, etc. (1) Microscope is used for many decades and previous studies had been described the removal of stage 1& 2 POTSIC stages through transcanal endoscopic approach and 3& 4 through the endoscopic assisted approach. (5) As per the authors' knowledge this is the first case report on complete transcanal endoscopic surgery for potsic stage 4 congenital cholesteatoma.

The complete removal of the disease with reconstruction for the maximum possible hearing is always a challenge to the surgeon. Microscopes are the gold standard for past many decades, but the visibility through the microscope is restricted by the narrowest part of the EAC, and a retro auricular incision is needed, also canal wall-up procedures always have a possibility of leaving behind the disease in hidden areas like sinus tympani and facial recess, etc. A previous study by James et al have shown a reduced recidivism rate where endoscopes were used in adjunct with microscopes. (6) Endoscopes are very useful in the pediatric age group with congenital cholesteatoma because of easy visualization of sinus tympani, facial recess, hypotympanum, etc where there is a higher chance of leaving behind the cholesteatoma during microscopic surgery. The pediatric EAC allows the passage of 2.7mm and 3mm endoscopes with the instruments easily.

Complete visualization and removal of congenital cholesteatoma through a trans canal endoscopic approach with hearing reconstruction alleviate the need for a second look surgery, especially when combined with Diffusion-weighted MRI .

An external scar and cavity have a tremendous impact on the quality of life in children. Cavity problems and a postauricular incision may limit children from participating in social activities and water sports also. This also can be sorted out with endoscopes and minimum bone is only removed during the surgery, so the normal ventilation is also maximum preserved.

To conclude in extensive congenital cholesteatomas also we can always try a completely endoscopic approach considering the benefits. In a center where both microscopic and endoscopic facilities are available at any point of difficulty, we can convert to endoscopic-assisted surgery also.

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TABLES

Table 1 – POTSIC Staging of congenital cholesteatoma

Stage	Description
I	Single quadrant disease without ossicular involvement or mastoid extension
II	Disease involving multiple quadrants without ossicular involvement or mastoid extension
III	Ossicular involvement, defined as ossicular erosion or necessity of surgical removal for disease eradication
IV	Disease with any mastoid extension

Table 2- Japan Otological Society staging system for congenital cholesteatoma

Stage	Description
Stage I	Cholesteatoma localized in the tympanic cavity
Stage Ia	Cholesteatoma confined to the anterior half of the tympanic cavity
Stage Ib	Cholesteatoma confined to the posterior half of the tympanic cavity
Stage Ic	Cholesteatoma involving both sides of the tympanic cavity
Stage II	Cholesteatoma involving 2 or more sites Stage
Stage III	Cholesteatoma with infratemporal complications and pathologic conditions
Stage IV	Cholesteatoma with intracranial complications

Figure legends

Figure 1 - HRCT temporal bone axial [a,b,c] & coronal sections [d,e,f] showing soft tissue density medial to tympanic membrane ,involving the middle ear,aditus extending to mastoid antrum.

Figure 2 - Endoscopic image showing [a]- congenital cholesteatoma involving all quadrants of the tympanic membrane [b]- tympanomeatal flap elevated, showing closed type CC [c]-drilling of attic and antrum , only the stapes suprastructure is seen after removal of malleus and incus [d] mastoid cavity after entire cholesteatoma removal [e] ossicular reconstruction with conchal cartilage[f] soft wall reconstruction of posterior canal wall with conchal cartilage [g] reposition of tympano meatal flap [h] healed cavity after 6 months of surgery



