



Congenital Cholesteatoma in an Adolescent - A Rare, Delayed Presentation in a 16 Year Old Boy.

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Abstract

Background: Congenital cholesteatoma is a rare lesion arising behind an intact tympanic membrane. We report a 16-year-old boy with progressive unilateral hearing loss. Otoscopy revealed a pearly white middle-ear mass, and imaging confirmed congenital cholesteatoma. Surgical excision was performed successfully. This case highlights the rare adolescent presentation of congenital cholesteatoma.

Keywords: Congenital Cholesteatoma, Adolescent, Conductive Hearing Loss, Middle Ear, Mastoidectomy.



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INTRODUCTION

Congenital cholesteatoma is a rare lesion accounting for 1–5% of all cholesteatomas.[1] It is characterized by keratinizing squamous epithelium within the middle ear behind an intact tympanic membrane in the absence of previous otorrhea, tympanic membrane perforation, or ear surgery. Most cases are diagnosed in early childhood with mean age being 5.6 +/- 2.8 years.[2] Adolescent presentation is uncommon and may indicate delayed diagnosis or slow disease progression. We report a rare case of congenital cholesteatoma in a 16-year-old boy presenting with long-standing unilateral hearing loss.

CASE REPORT

A 16-year-old boy presented to the Department of Otorhinolaryngology at Dhanalakshmi Srinivasan Medical College at Perambalur with complaints of progressive hearing loss in the left ear for 10 years. There was no history of ear discharge, otalgia, tinnitus, vertigo, facial weakness, trauma, previous ear surgery, or recurrent episodes of otitis media.

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General physical examination was unremarkable. Otoscopic examination of the left ear (Fig. 1) revealed an intact tympanic membrane with a pearly white mass involving all 4 quadrants of the middle ear. The external auditory canal was normal. Examination of the right ear (Fig. 2) revealed Grade II retraction of the tympanic membrane.



Figure 1: Left ear revealed an intact tympanic membrane with a pearly white mass involving all 4 quadrants of the middle ear. The external auditory canal was normal



Figure 2: Right ear revealed an intact tympanic membrane with Grade II retraction

Tuning Fork Test	Right	Left
Rinne's Test	Positive	Negative
Weber's Test	Lateralised to Left Ear	
Absolute Bone Conduction Test	Not Reduced	Not Reduced

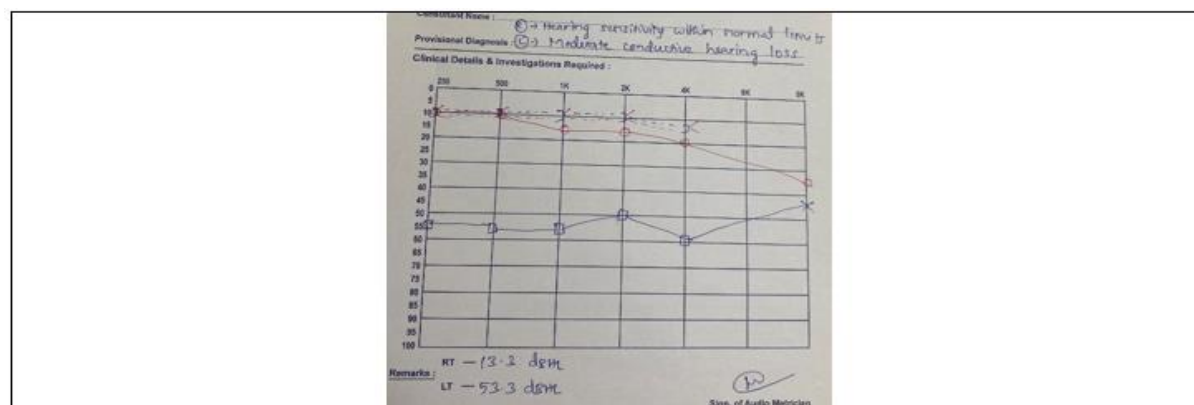


Figure 3: Pure-tone audiometry demonstrated moderate conductive hearing loss in the left ear with an air-bone gap of approximately 40 dB. Right ear hearing was within normal limits

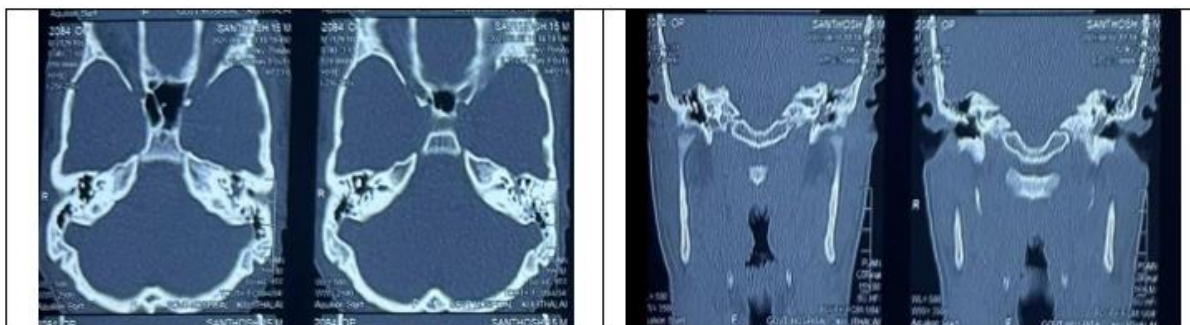


Figure 4 & 5: Computed tomography (CT) of the temporal bone revealed a abnormal soft tissue density of size 10.0 x 8.8 mm seen in middle ear, it is seen encasing the stapes and handle of malleolus. Mastoid air cells were well pneumatized, and no evidence of chronic otitis media was noted

Based on the clinical and radiological findings, a diagnosis of congenital cholesteatoma was made. The lesion was classified as Stage III according to the Potts classification due to ossicular involvement.

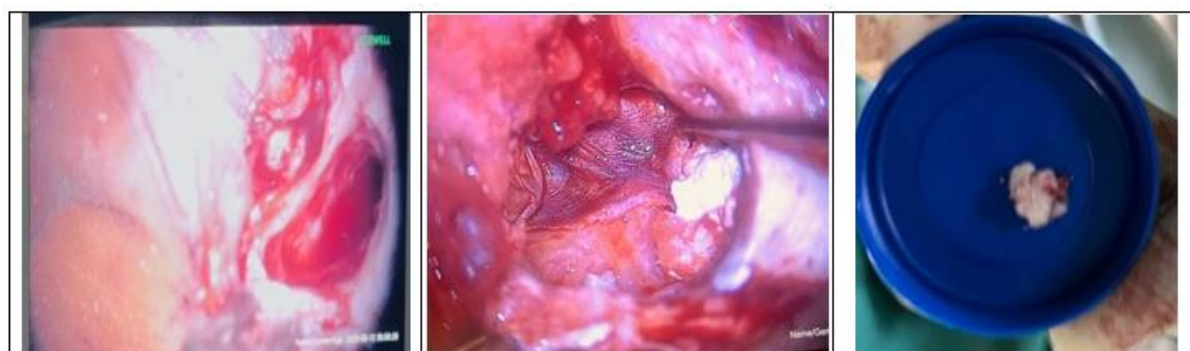


Figure 6, 7 & 8: The patient underwent modified radical mastoidectomy with type III tympanoplasty under general anesthesia. Intraoperatively, a pearly white encapsulated cholesteatoma sac was identified within the epitympanum and mesotympanum. The lesion was completely excised. Erosion of the incus was noted

Histopathological examination confirmed the diagnosis of cholesteatoma, showing a cyst lined by keratinizing stratified squamous epithelium containing laminated keratin debris.

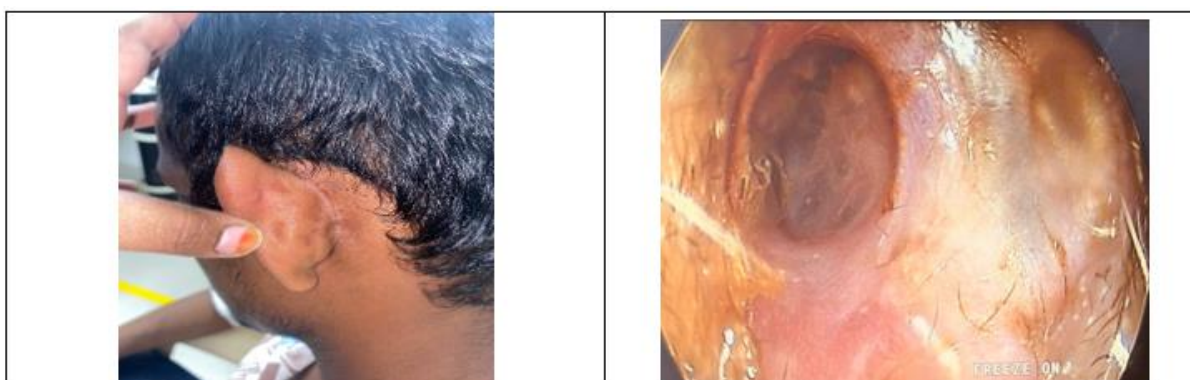


Figure 9 & 10: The postoperative period was uneventful. Follow-up at 3 months demonstrated a well-healed ear, no evidence of residual disease, and significant improvement in hearing thresholds

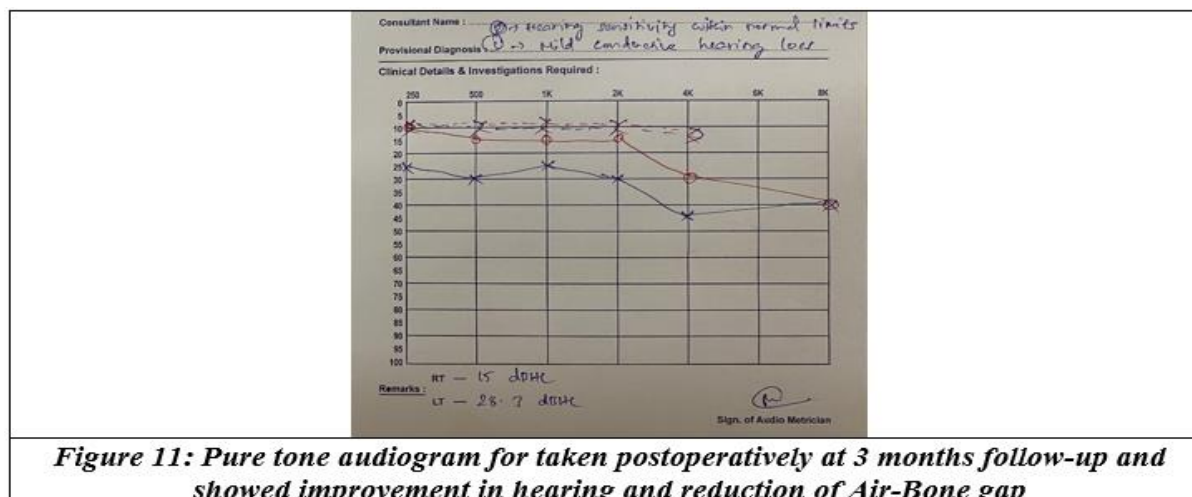


Figure 11: Pure tone audiogram for taken postoperatively at 3 months follow-up and showed improvement in hearing and reduction of Air-Bone gap

DISCUSSION

Congenital cholesteatoma is believed to arise from embryonic epithelial cell rests that persist within the middle ear cavity.[3] Several theories have been proposed regarding its origin, including the epidermoid formation theory, amniotic fluid contamination theory, and epithelial metaplasia theory. Levenson et al. defined congenital cholesteatoma as a white mass medial to an intact tympanic membrane in the absence of previous otorrhea, perforation, or ear surgery.[4] Most cases are diagnosed before the age of ten years. Presentation in adolescence is unusual and may indicate slow-growing disease or delayed diagnosis. Progressive unilateral conductive hearing impairment is the symptom most frequently reported at presentation. Otoscopic examination typically reveals a white, pearly lesion behind an intact tympanic membrane, most frequently in the anterosuperior quadrant.

The Potsic staging system[5] is widely used:

- Stage I: Single quadrant involvement
- Stage II: Multiple quadrants without ossicular or mastoid involvement
- Stage III: Ossicular involvement
- Stage IV: Mastoid extension

Surgical excision remains the definitive treatment.[6] The choice of procedure depends on disease extent and surgeon preference. Early diagnosis and complete removal are essential to prevent recurrence and preserve hearing.[7,8]

This case is noteworthy because of the unusually late presentation at 16 years of age despite the congenital origin of the lesion. Awareness of this possibility can facilitate timely diagnosis and management.

CONCLUSION

Congenital cholesteatoma should be considered in adolescents presenting with unilateral conductive hearing loss and an intact tympanic membrane. Careful otoscopic examination and appropriate imaging are crucial for diagnosis. Surgical excision provides excellent outcomes when performed before the development of extensive complications.

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