International Journal of Research in Medical Sciences
Solanki KJ et al. Int J Res Med Sci. 2026 Mar;14(3):1162-1166
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012
DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20260638

Case Series

Congenital cholesteatoma of middle ear: clinical features and surgical outcomes

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Received: 28 December 2025
Revised: 02 February 2026
Accepted: 12 February 2026

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ABSTRACT

Congenital cholesteatoma of the middle ear is an uncommon entity, often detected incidentally in children. The present case series evaluates the clinical presentation and surgical outcomes of patients with congenital cholesteatoma. Ten patients diagnosed with congenital cholesteatoma underwent surgical eradication of the disease. Postoperative cavity status and hearing outcomes were assessed at a 3-month follow-up using pure tone audiometry, and air–bone gaps were calculated. The mean preoperative air–bone gap was 54.50±14.62 dB, which improved to 35.50±11.41 dB postoperatively, demonstrating a mean hearing gain of 19.00 ± 5.68 dB. Histopathological examination confirmed the diagnosis of cholesteatoma in all cases. No recurrence was observed during the follow-up period. Early diagnosis based on micro-otoscopy, supported by computed tomography of the temporal bone, and complete surgical excision provide satisfactory anatomical and audiological outcomes in congenital cholesteatoma.

Keywords: Cholesteatoma, Hearing, Reconstructive surgical procedures, Recurrence

INTRODUCTION

The first reported case of congenital cholesteatoma of the petrous portion of the temporal bone was published by Jefferson and Smalley in 1938.[1] It was not until 1953 that house described middle ear cholesteatoma behind an intact tympanic membrane.[2] Congenital cholesteatoma (CC) of the middle ear was first described by House in 1953.[3]

Later, Derlacki and Clemis established the clinical criteria for the diagnosis. These include a white mass behind an intact tympanic membrane, a normal pars tensa and flaccida and no history of otorrhea, perforation or a previous otologic procedure.[4] Levenson et al revised the criteria by adding that previous episodes of otitis media or effusion should not exclude a diagnosis of CC.[5]CC accounts for 1-5% of all cholesteatomas and is more common in boys by a ratio of 3:1.[7] The mean age of diagnosis is four to five years.[7-9] The origin of the disease is unclear.[6,10,11] The most accepted hypothesis is that CC is a remnant of embryonic epithelial tissue.[12] The anterior superior quadrant of the middle ear is the most frequent site for CC.[7] CC usually grows slowly and is asymptomatic until the presence of secondary ossicular erosion, local infection, rupture of the tympanic membrane and otorrhea. Thus, in advanced stages of the disease differentiating between acquired and CC may be impossible. Mastoid air cell (MAC) pneumatization varies between individuals and is usually symmetric in both ears.[14,15]

MAC are known not to be well developed in individuals with chronic otitis media with and without cholesteatoma.[16–19] MAC were described as more pneumatized in children with CC than in children with middle ear inflammatory diseases.

Table 1: Clinical criteria for congenital cholesteatoma.

Clinical criteria for congenital cholesteatoma	
1	A whitish abnormality in the tympanic cavity visible through the tympanic membrane
2	Normal appearance of the pars flaccida and pars tensa of the tympanic membrane
3	No ear discharge, no tympanic perforation, no ear trauma
4	Negative history of ear surgery
5	A history of uncomplicated acute otitis media does not warrant the exclusion of congenital cholesteatoma

Table 2: Staging systems as proposed by Potsic et al and Nelson et al for congenital cholesteatoma based upon disease findings and recurrence rates.[5,6]

Author	Stage	Description
Nelson et al	I	Confined to middle ear with no ossicular involvement
	II	Involves ossicular chain, posterior mesotympanum and/or superior quadrant of the attic
	III	Middle ear and mastoid obliteration with ossicular erosion
Potsic et al	I-II	Involves one or more middle ear quadrants without ossicular invasion or mastoid extension
	III	Ossicular erosion but no mastoid extension
	IV	Mastoid infiltration

Potsic distinguished between 4 stages of congenital middle ear cholesteatoma: cholesteatoma confined to the anterosuperior quadrant of the tympanic membrane, cholesteatoma of the mesotympanum and a few quadrants of the tympanic membrane without ossicular involvement, cholesteatoma of the mesotympanum and a few quadrants of the tympanic membrane with ossicular involvement but with no mastoid involvement, mastoid involvement.

CASE SERIES

A total of 10 patients were included in the study, all of them had CC.

A detailed ENT and systemic examination were done and informed consent was taken from all the patients planned for surgery. On admission a thorough ENT history was taken, the patients not had any episodes of otitis media or middle ear trauma and not had history of any ear surgery. The patients did not report any ear pain, tinnitus, a sense of fullness in the ear, dizziness or headaches.

The patient's history was negative for chronic diseases. No family history of cholesteatoma was found either. Otoscopy revealed a normal, wide and inflammation free external auditory canal in all patients, no foreign body was found there. However, a whitish spherical mass was found in the tympanic cavity behind an intact tympanic membrane. Preoperative and postoperative hearing results were measured at 500, 1,000, 2,000, and the air-bone gap was calculated. Preoperative CT scan of temporal bone done in all patients.

Surgery was performed under general anesthesia via a postauricular approach. A tympano-meatal flap was elevated and the mesotympanum was exposed. The status of the ossicular chain and the extent of the cholesteatoma in the middle ear were both evaluated.

The lateral epitympanic wall was removed with drill burrs to make a wide opening into the epitympanum and thus visualize the extent of the cholesteatoma. If necessary, the malleus head and/or incus were also removed. Canal wall down mastoidectomy or modified radical mastoidectomy was performed and cholesteatoma was removed as completely as possible.

We reviewed the medical records for intraoperative findings, pre-and postoperative hearing levels, postoperative complications and any problems arising during follow-up of 3 months. A total of 10 patients having CC were included in the study with a postoperative follow up of 3 months.

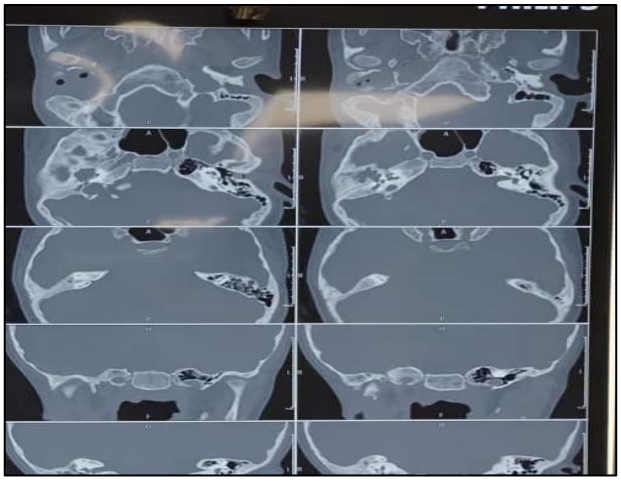

Figure 1. HRCT temporal bone showing right-sided otomastoiditis with congenital cholesteatoma and hypoplastic ossicles.

Among all patients only one patient having a history of facial weakness. Micro-otoscopy revealed a normal, intact, translucent tympanic membrane which had no perforation, whitish abnormality, signs of thickening or inflammatory lesions of the myringosclerosis type. In the hospitalized patient's congenital cholesteatoma limited to the tympanic cavity of the middle ear was diagnosed. The mean age of the patients was 7 years and the mean hospitalization time was 5 days.

All patients having main complaint of decreased hearing and earache which was on and off in onset, no history of ear discharge and negative history of ear surgery. All CCs adhered tightly to the tympanic membrane. Normal sound reception was observed on whisper and tuning fork hearing tests in a quiet room with no external noise. In our study, the mean preoperative and postoperative air-bone gaps of all the patients (n=10) were 54.50+14.62db and 35.50+11.41db respectively, suggesting significant improvement of postoperative hearing with an improvement of around 19.00+5.68db in the postoperative period. The histopathologic assessment confirmed the diagnosis of cholesteatoma. Post-operatively the patient recovered well and retained full function of the facial nerve.

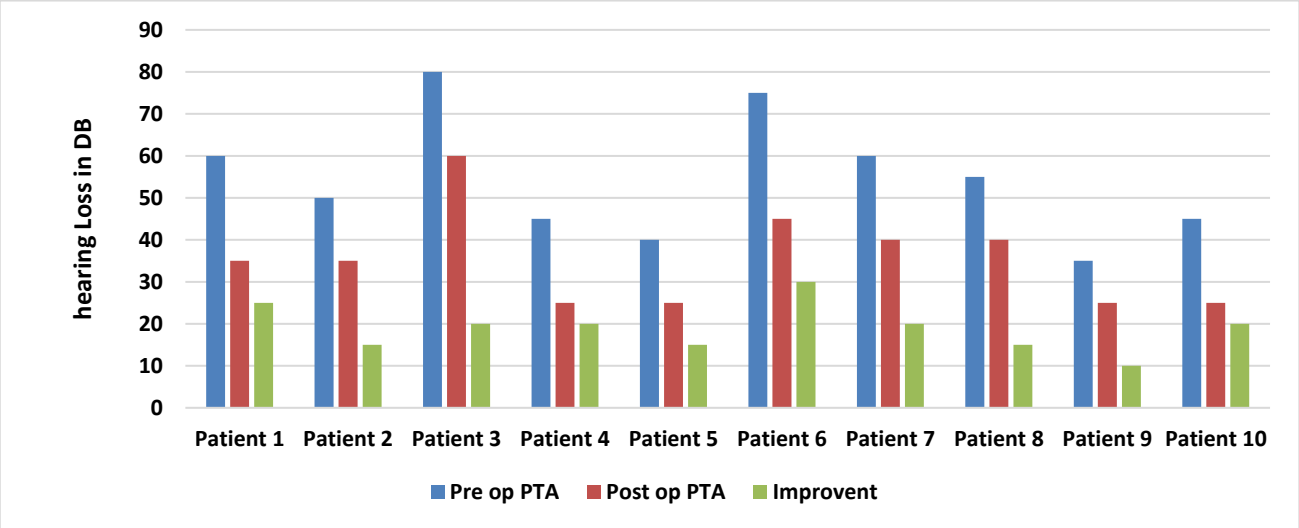

Figure 2: Describe hearing outcome after surgery. The mean preoperative and postoperative air-bone gaps of all the patients (n=10) were 54.50+14.62db and 35.50+11.41db respectively, suggesting significant improvement of postoperative hearing with an improvement of around 19.00+5.68db in the postoperative period.

There was no recurrence of cholesteatoma noted in any of the patients in the follow up period of 3 months.

DISCUSSION

CC of the middle ear remains a rare but clinically significant entity, primarily affecting the pediatric population. The principal objectives of surgical management are complete eradication of disease, prevention of recurrence, and preservation or improvement of hearing. In the present study, all patients underwent surgical excision with satisfactory anatomical outcomes and a statistically and clinically significant improvement in hearing thresholds. In this study, the mean preoperative air–bone gap (ABG) was 54.50±14.62 dB, which improved to 35.50±11.41 dB postoperatively, resulting in a mean hearing gain of 19.00±5.68 dB. This improvement reflects successful disease clearance and effective ossicular reconstruction or preservation wherever feasible.

These results are comparable to those reported by Potsic et al, who observed postoperative ABG closure to within 20–30 dB in a significant proportion of children undergoing surgery for CC, particularly in early-stage disease.[6] Similarly, McGill et al demonstrated meaningful hearing improvement following complete excision, emphasizing that postoperative hearing outcomes are closely related to the extent of ossicular involvement at presentation.[9] Levenson et al reported that patients with disease limited to the mesotympanum (Potsic stages I and II) achieved better postoperative hearing outcomes than those with ossicular erosion or mastoid extension.[8] The relatively higher preoperative ABG observed in our series suggests that many patients presented at a more advanced stage, which may explain why postoperative ABG values did not normalize completely despite significant improvement.

No recurrence was observed in any patient during the 3-month follow-up period in the present study. Previous studies have reported recurrence rates ranging from 5% to 30%, depending on disease stage, surgical technique, and length of follow-up. Potsic et al demonstrated that recurrence is significantly higher in stage III and IV disease and emphasized the importance of long-term surveillance.[6] Similarly, Semaan and Megerian noted that microscopic residual disease may remain dormant for several years before becoming clinically evident,

underscoring the need for prolonged follow-up with imaging when indicated.

Table 3: Differences between congenital cholesteatoma of middle ear and that of mastoid process.[24]

	Middle ear	Mastoid
Presentation	Hearing loss	Temporal area and neck pain, dizziness
Age at diagnosis	Childhood	Adulthood
Findings of otoscopy	Cholesteatoma behind eardrum	No abnormalities
Findings of imaging	Restricted to middle ear	Restricted to mastoid process

The purpose of this study is to assess the anatomical and audiological outcome of mastoid surgery in congenital cholesteatoma cases. Despite favorable results, the present study has certain limitations. The small sample size and relatively short follow-up period may underestimate the true recurrence rate. Previous long-term studies, such as that by Potsic et al, have shown that recurrence may occur several years after surgery. Therefore, extended follow-up is necessary to confirm long-term disease control and hearing stability. CC has been reported to originate in the petrous apex, the middle ear, the mastoid process and the external auditory canal. The middle ear and the petrous apex are well established as sites of origin for CC.[20] In contrast, CC of the external auditory canal is usually caused by congenital aural stenosis and the true existence of primary CC in this site is uncertain.[21] The mastoid process is undoubtedly the least-reported site for the onset of CC. Overall, the findings of this study are comparable with previously published literature, demonstrating that early diagnosis, appropriate staging, meticulous surgical technique, and adequate postoperative surveillance are crucial for achieving optimal outcomes in CC.[23]

CONCLUSION

CCs often extend beyond the typically described anterosuperior location of the middle ear. Ossicular destruction and mastoid infiltration occur more frequently than previously thought. Preoperative assessment and radiographic imaging should focus on the extent of disease with respect to ossicular and mastoid involvement. Infiltrations of these structures dictate the risk of recurrence and surgical approach to CCs. Early-stage CC of the middle ear in children is usually diagnosed incidentally. The most common location is the anterosuperior part of the tympanic cavity. Investigation and diagnosis of CC is based on micro-otoscopy; computed tomography of the temporal bones is also a useful procedure in the process. The diagnosis is made by imaging; MRI is currently superior, but a CT scan may better show details of the eroded bone and exposed structures. Surgery is the only treatment method, and careful manipulation of the exposed delicate structures is required in order to avoid complications.

ACKNOWLEDGEMENTS

The authors thank their colleagues for academic support and insightful contributions. Appreciation is extended to the departmental staff for assistance with procedural aspects of the study. Sincere gratitude is expressed to the research participants for their trust and cooperation. The authors also acknowledge the institute for providing the necessary infrastructure and research opportunities that enabled completion of this study.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Solanki KJ, Nakum NN, Mehta MV. Congenital cholesteatoma of middle ear: clinical features and surgical outcomes. Int J Res Med Sci 2026;14:1162-6.