

## Periotic congenital cholesteatoma – A rare case report

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### Abstract

Congenital cholesteatoma is a rare entity and cholesteatoma surrounding the entire otic capsule and involving petrous apex is even rarer. The condition may remain hidden for a long duration destroying the temporal bone and might present as an intracranial space-occupying lesion, loss of hearing, facial nerve palsy without significant otoscopic findings. We report a case of thirty-three years old female who presented with facial palsy and hearing loss for 7 months without any history of ear discharge. Possibility of congenital cholesteatoma was sorted based on clinical and radiological features. Transotic approach was done for clearing the disease. Congenital cholesteatoma should be considered as a differential diagnosis in any patient with unexplained otological signs and symptoms. Radical excision is the mainstay of treatment.

**Keywords:** Congenital cholesteatoma, Facial nerve palsy, Hearing loss, Petrous bone.

### Introduction

Cholesteatoma is a well-demarcated cystic lesion caused by an abnormal growth of keratinizing squamous epithelium in the temporal bone and is “skin in the wrong place”. Congenital cholesteatoma presents a nidus of trapped squamous epithelium which forms a white mass behind an intact eardrum and without any history of previous otologic procedures or otitis media.<sup>1</sup> The annual incidence of congenital cholesteatoma is 0.00012 % in the overall population.<sup>2</sup> Among the cholesteatomas, the overall incidence of congenital cholesteatoma is between 2% and 5% with a male: female ratio of 3:1, the treatment of which is radical excision.<sup>3</sup> We report a rare case of congenital cholesteatoma surrounding the otic capsule. This type of cholesteatoma has scarcely been reported in literature.

### Case Report

Thirty-three years old female who deviation of angle of mouth to right with an inability to close the left eye completely since the past 2 ½ years which responded partially to topical antibiotics presented to Department of Otorhinolaryngology of All India Institute of Medical Sciences, Rishikesh with worsening of these complaints for the past 3 months. She also had occasional left earache with gradually progressive hearing loss for 7 months. There was a history of right ear discharge in childhood.

On examination, left tympanic membrane showed grade I retraction in pars tensa with a whitish patch on the anterior quadrant of pars tensa (Fig. 1) along with left grade IV facial nerve palsy as per House

Brackmann grading (HBG) (Fig. 2). Cerebellar and vestibular signs were negative. Pure tone audiometry revealed conductive hearing loss on both ears (Table 1)

High-Resolution Computed Tomography (HRCT) of Temporal bone showed ill-defined soft-tissue attenuation with lytic destruction of the left petrous bone with destruction of tegmen tympani, the roof of left carotid canal, anterior wall and roof of left internal acoustic meatus and middle ear cavity. (Fig. 3 a & b) Magnetic Resonance Imaging (MRI) of the brain showed a well-defined lobulated lesion of 28x 5x4 mm is involving the petrous part of left temporal bone extending across the petrous apex to reach the internal acoustic meatus showing iso to hypointense on T1 weighted MRI and hyperintense signal on T2 weighted MRI without any post-contrast enhancement. There was no involvement of dura and carotid encasement. (Fig. 4)

Cholesteatoma was removed completely without any residual disease via transotic approach with transection of a necrosed segment of the facial nerve and cable nerve grafting of facial nerve using grater auricular nerve with a complete cavity obliteration using fat from the thigh and cul-de-sac closure of the external auditory canal. (Fig. 5)

Post-operative evaluation showed HBG VI facial palsy with weber lateralisation to right. She was kept on regular follow up.

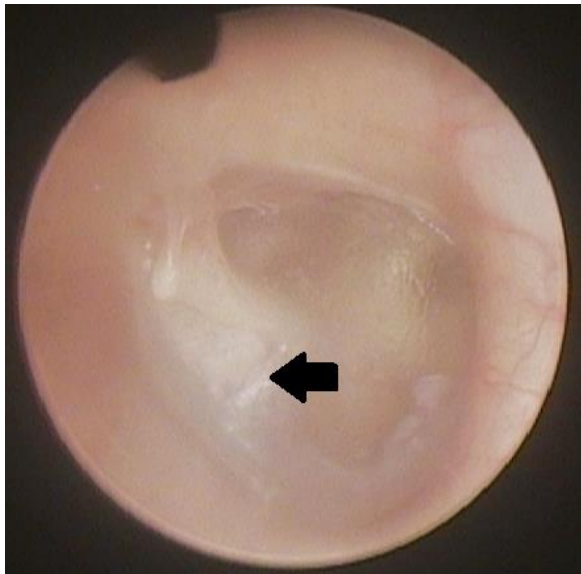


Fig. 1:

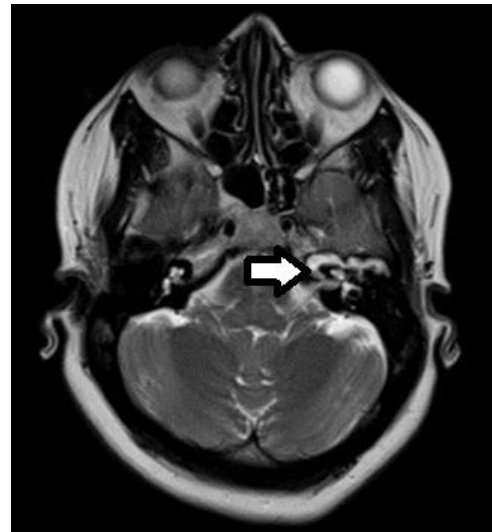


Fig. 4:



Fig. 2:

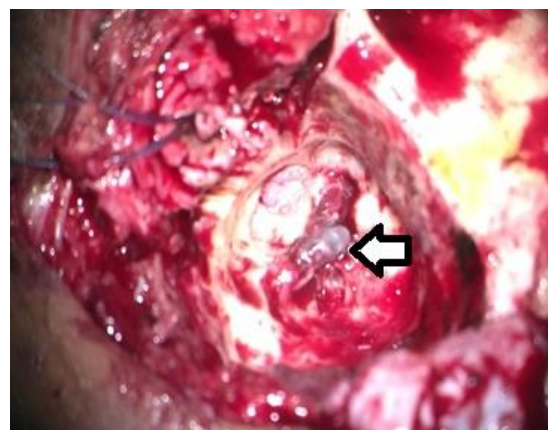


Fig. 5:

**Table 1:** Pure tone audiogram showing Moderate conductive hearing loss

	Right ear	Left ear
Hearing loss	Mild conductive	Moderate conductive
Average Hearing Loss	35dB	50dB
Air - Bone Gap	20dB	35dB

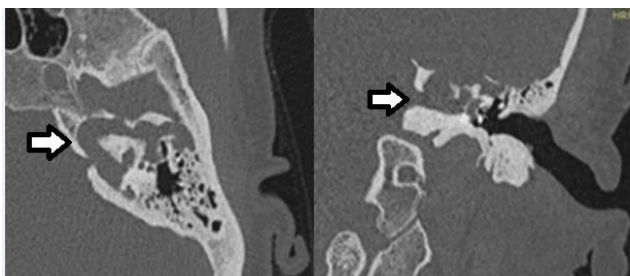


Fig. 3:

**Discussion**

Du Verney, a French anatomist first described a temporal bone tumour probably cholesteatoma in 1683. In 1829, Cruveilhier described the pathologic features of cholesteatoma as pearly tumour (tumeur perl'ee) due to its whitish appearance. Cholesteatoma (chole = cholesterol; steat = fat; oma = tumour), the term was coined by Johannes Muller, a German pathologist, in 1838.<sup>1</sup> There are multiple classification systems for cholesteatoma based on different aspects. Based on pathogenesis, it may be congenital or acquired.<sup>1</sup> Congenital cholesteatoma was first

described by Cawthorne and Griffith. In 1965, Derlacki and Clemis described 6 cases of congenital cholesteatoma and defined congenital cholesteatoma as an embryologic residue of epithelial tissue behind a normal tympanic membrane in the absence of a history of infection or ear surgery which was later used for clinical diagnosis. Levenson, later in 1989, updated the criteria as the presence of uncomplicated acute otitis media does not exclude congenital cholesteatoma.<sup>4</sup> Congenital cholesteatoma is said to be due to inadequate folding of the epidermoid inside the middle ear cleft and accumulation of stratified squamous epithelium through the third and fifth weeks of gestation and slowly grows over time.

The annual incidence of congenital cholesteatoma is 0.00012 % in the overall population.<sup>2</sup> Congenital cholesteatoma may occur in five sites: (a) the external auditory canal (b) the middle ear (c) the mastoid (d) the petrous apex and (e) the cerebellopontine angle. According to Nager congenital cholesteatoma is least common in mastoid process.<sup>4</sup> Congenital cholesteatoma of the temporal bone is rare while the involvement of petrous apex is very rare. A search conducted on 19<sup>th</sup> March 2020 in google scholar with keywords as ‘congenital cholesteatoma’ and ‘India. A total of 4 cases of congenital cholesteatoma of petrous apex were reported in a total of 2 different publications.<sup>2,5</sup>

Congenital cholesteatoma is usually seen as a pearly white mass behind the intact eardrum in the anterosuperior quadrant. Conductive hearing loss is seen in congenital cholesteatoma involving the posterior tympanum. The disease can involve an unusual location in the adults and elderly producing conductive hearing loss and complications.<sup>6</sup>

Even though congenital cholesteatoma has a destructive nature, most of them being asymptomatic and appear to be an innocuous keratin pearl. This makes the diagnosis difficult. They can enlarge and can cause serious complications like ossicular destruction, base of skull involvement and facial paralysis. Symptoms depend on the direction of the extension of the disease. Lesions of posterior mesotympanum have conductive deafness owing to the early erosion of ossicles.

The first symptom of congenital cholesteatoma in petrous bone is the facial nerve deficit followed by unilateral sensorineural hearing loss.<sup>2</sup> To exclude other causes like cerebellopontine angle lesions, CT and MRI scan recommended. Cholesteatoma does not have specific radiological sign making it very difficult to diagnose. In CT cholesteatoma appears iso-dense to cerebrospinal fluid and non-enhancing with contrast

CT and Gadolinium studies. It is hypointense on T1 and hyperintense on T2 weighted MRI. The otologists prefer a transmastoid translabyrinthine approach while neurosurgeons prefer middle cranial fossa and retro mastoid approaches.<sup>7</sup>

Congenital cholesteatoma can cause complications like facial palsy, labyrinthine fistula, meningitis, cranial abscess and even death may occur if diagnosed late. In our case the patient presented with left side HBG IV facial nerve palsy with moderate conductive hearing loss on the left side, with an extension of diseases to the petrous apex causing destruction of tegmen tympani, the roof of left carotid canal, anterior wall and the roof of left internal acoustic meatus.

The basic surgery for removal of petrous apex cholesteatoma is transmastoid translabyrinthine approach with or without transcochlear approach. However, these may be insufficient to remove a deep-seated one in petrous apex adherent to the dura of middle cranial fossa.<sup>2</sup>

In our case, transotic approach was done for clearing the disease. To achieve complete removal, anterior transposition of facial nerve done. Later, necrosed part the nerve was resected and cable nerve grafting done using grater auricular nerve

Cholesteatoma must be treated with complete excision with rehabilitation of ear function either immediately or later. Salvaging or grafting of the facial nerve has the second priority, while next comes hearing. Though hearing could be sacrificed for the sake of complete excision after proper evaluation and patient counseling.<sup>8</sup> Regular follow-up is needed. In case of cul-de-sac closure of the external auditory canal, follow-up can be done using radiological imaging.

## Conclusion

Congenital cholesteatoma can present with varying clinical manifestation and should be suspected in any patient with unexplained otological signs and symptoms. Radical excision is the mainstay of treatment. Management of facial nerve has the second priority followed by hearing. Indefinite follow-up is needed as recurrence may occur.

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## Conflict of interest

None.

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