

# Congenital Mastoid Cholesteatoma Presenting as a Mass Obstructing External Auditory Canal

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## SUMMARY

Cholesteatoma is one of the common disorders encountered by the otorhinolaryngologist. However, there are few cases with an atypical clinical presentation and computed tomography scan findings which make cholesteatoma difficult to diagnose. We report a rare case of congenital mastoid cholesteatoma that presented as a mass obstructing the external auditory canal. The disease was successfully treated with a mastoidectomy and histopathological examination confirmed the diagnosis of cholesteatoma. Clinicians should consider congenital mastoid cholesteatoma in the differential diagnosis of patients with a mass in the external auditory canal. The diagnosis is likely to be made based on surgical and histopathological findings.

## KEY WORDS:

*Cholesteatoma; congenital; mastoid; external auditory canal*

## INTRODUCTION

Cholesteatoma is still a common disease seen in otology clinic nowadays. It implies retention of keratinous debris in middle ear cleft. The disease is potentially dangerous as it may lead to serious complications intracranially and extracranially. It is due to the mass effect and destructive properties to the adjacent bony wall structures. Therefore prompt diagnosis and treatment is necessary to avoid any consequences. Cholesteatoma is categorized either as congenital or acquired. Congenital cholesteatoma is a rare entity and accounts only 2% to 4% of cases presenting to paediatric otologists.<sup>1</sup> However, it is difficult to diagnose due to its atypical presentation. We report a rare case of congenital mastoid cholesteatoma that presenting as a mass obstructing the external auditory canal.

## CASE REPORT

A 21-year-old Malay lady with no known medical illness presented with progressive reduced hearing over left ear for one year. Otherwise, there was no history of earache, ear discharge, vertigo or tinnitus. There were no associated nasal symptoms. She denied any history of trauma or previous history of ear operation. On examination, she looked comfortable. The facial nerve was intact. Otoscopic examination showed mass occupying whole left external auditory canal which was soft in consistency upon probing and obscuring the left tympanic membrane (Fig. 1). The

probe was not able to be moved surrounding the mass. The right external auditory canal and tympanic membrane were normal. Nasal examination revealed no significant findings. Tuning fork test showed positive Rinne's test bilaterally and Weber's test was lateralized to the left. Pure tone audiogram showed normal hearing of right ear and moderate conductive hearing loss of left ear.

High resolution computed tomography (CT) scan revealed a large lobulated cystic lesion measuring 1.3 cm x 1.5 cm x 1.6 cm seen within the left mastoid cavity extending to middle ear and external auditory canal causing expansion of external auditory canal (Fig. 2). There was no calcification or soft tissue component within the lesion. There was erosion of mastoid air cell seen. The scutum and tympanic membrane were not visualized. Otherwise, the ossicles, semicircular canals and cochlea were intact. Initial diagnosis of a left mastoid cyst was made.

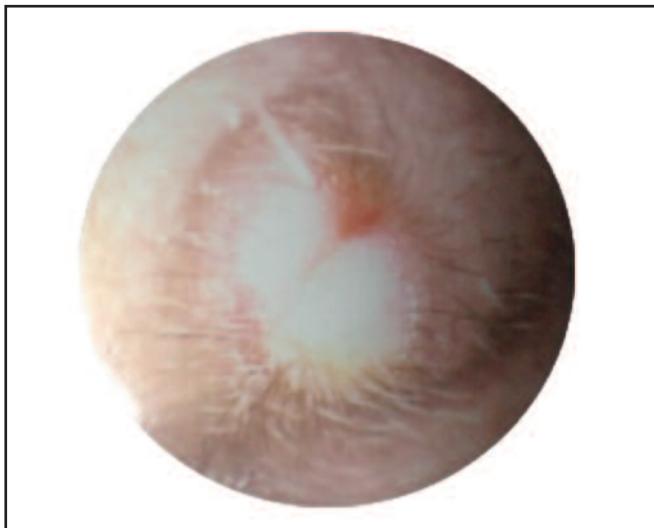
She underwent left mastoid exploration, tympanoplasty type III and meatoplasty. Intraoperatively, there was cystic mass containing whitish material seen at left mastoid cavity extending to external auditory canal and middle ear. The cystic mass pushed the ossicles medially. The tympanic membrane was plastered to the promontory. Posterior wall of external auditory canal eroded and formed autocavitation. The malleus was intact but there was erosion of the incus. The mastoid cyst was sent for histopathological examination and demonstrated numerous keratin flakes which consistent with cholesteatoma. Postoperatively, the left external auditory canal and mastoid cavity were dry and no discharge or debris seen.

## DISCUSSION

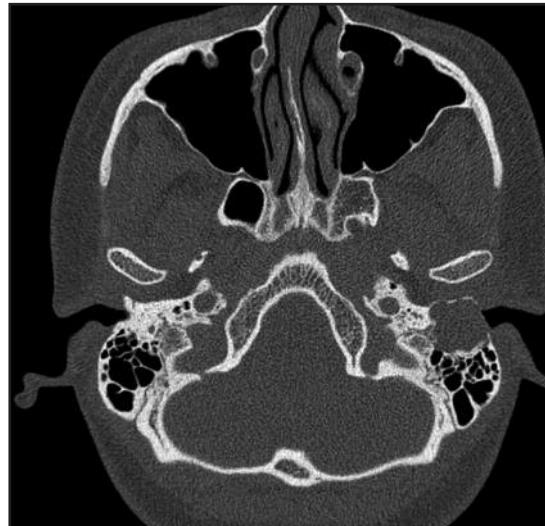
The pathogenesis of the cholesteatoma is still debatable. Congenital cholesteatoma generally appears during childhood. The most acceptable theories of congenital cholesteatoma is embryonic epithelial cell rest and epithelial migration theory. The rest of the cells which is known as embryonic formation is normally appear by 10 weeks of gestation and is not seen by 33 weeks of gestation. However in congenital cholesteatoma the embryonic formation persists and it was observed in half of the cases.<sup>2</sup> On the other hand, the epithelial migration theory suggests that the embryonic ectodermal layer of external auditory canal migrates into middle ear. The acquired cholesteatoma is postulated due to Eustachian tube dysfunction which result in

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**Fig. 1:** Otoscopic examination showed soft mass occupying whole left external auditory canal.



**Fig. 2:** Axial view CT scan showed lobulated cystic lesion within left mastoid cavity.

poor ventilation of epitympanic space and finally draw the pars flaccida inwards. As a result, it forms retraction pocket and disrupts normal epithelial migration of tympanic membrane and ultimately, promotes accumulation of keratin debris. Another theory of acquired cholesteatoma includes implantation of squamous epithelium to middle ear cleft due to previous ear surgery, trauma or infection. The most probable explanation for the pathogenesis of cholesteatoma in our patient is congenital type as the tympanic membrane was intact and there was no evidence of a retraction pocket, chronic ear infection or ear surgery.

Clinical presentations of congenital cholesteatoma are more variable and non specific as compared to acquired cholesteatoma. Congenital cholesteatoma usually presents as a whitish pearl mass medial to an intact tympanic membrane. It is commonly starts at the anterosuperior quadrant, and then progressively erodes the ossicular chain and surrounding bony wall and rarely does it involve the mastoid. According to Nelson *et al.*, 49% of the patients had a history of acute otitis media, serous otitis media, or both at presentation.<sup>3</sup> Disruption of ossicular chain by the disease results in conductive hearing loss. Unlike acquired cholesteatoma, congenital cholesteatoma does not typically present with otorrhoea, tympanic membrane perforation or previous history or trauma and ear surgery. The mass in external auditory canal may be due to ear canal stenosis, otitis externa or osteoma.

Atypical findings in otoscopy which showed presence of soft mass in external auditory canal together with CT scan findings of mastoid cyst made the diagnosis of cholesteatoma seems to be unlikely for this patient initially. The presence of mass in the external auditory canal was explained by intraoperative findings whereby the mastoid mass extends to the external auditory canal due to erosion of its posterior wall. The histopathological examination confirmed the

presence of keratin flakes which was consistent with cholesteatoma. Congenital mastoid cholesteatoma is uncommon and is infrequently mentioned in the literature but with different manifestations.<sup>4</sup> In view of its atypical presentation, congenital mastoid cholesteatoma is difficult to diagnose. The patient might not have any otological symptoms and the radiological imaging is helpful in establishing the diagnosis. As for primary mastoid cyst, the incidence is also rare and it is usually due to infection or trauma.<sup>5</sup>

Almost all congenital cholesteatoma warrant surgical intervention. The most common procedures being performed is modified radical mastoidectomy which include canal wall up or canal wall down approaches. In our case the posterior wall of external auditory canal was already eroded by the disease resulting in autocavitation of mastoid cavity and external auditory canal.

In conclusion, clinicians should consider congenital mastoid cholesteatoma in the differential diagnosis of patients with a mass in the external auditory canal. The diagnosis is likely to be made based on surgical and histopathological findings.

## REFERENCES

1. Richter GT, Lee KH. Contemporary assessment and management of congenital cholesteatoma. *Curr Opin Otolaryngol Head Neck Surg* 2009; 17: 339-45.
2. Koltai PJ, Nelson M, Castellon RJ, *et al.* The natural history of congenital cholesteatoma. *Arch Otolaryngol Head Neck Surg* 2002; 128: 804-9.
3. Nelson M, Roger G, Koltai PJ, *et al.* Congenital cholesteatoma: classification, management, and outcome. *Arch Otolaryngol Head Neck Surg* 2002; 128: 810-4.
4. Hidaka H, Ishida E, Kaku K, Nishikawa H, Kobayashi T. Congenital cholesteatoma of mastoid region manifesting as acute mastoiditis: case report and literature review. *J Laryngol Otol* 2010; 124: 810-5.
5. Tan CY, Chong S, Shaw CKL. Primary mastoid cyst. *J Laryngol Otol* 2013; 127: 48-50.