CASE REPORT

Congenital Mastoid Cholesteatoma with Posterior External Auditory Canal Erosion: A Case Report and Literature Review

Danny Kit Chung Wong[1], Aminuddin Saim[2]

Received on: 22 December 2019; *Accepted on:* 26 April 2023; *Published on:* 11 April 2024

Abstract

Background: Congenital cholesteatomas (CCs) are rare epidermal cysts that have been reported to occur most commonly in the middle ear and rarest in the mastoid cavity. They usually present in adulthood and are difficult to detect due to the lack of symptoms. Treatment is usually a canal wall up or down mastoidectomy with good success rates.

Case description: We present a rare case of a mastoid CC with posterior external auditory canal wall erosion treated successfully with a simple canal wall-up mastoidectomy.

Conclusion: Mastoid CCs still remain a rare entity with a relatively uncomplicated surgical approach.

Clinical significance: Clinicians should high index of suspicion and a low threshold for a CT scan in patients with a history of an intermittently discharging ear, a narrowed ear canal, or a posterior canal wall fistula.

Keywords: Case report, Congenital cholesteatoma, Ear discharge, Mastoid, Rare.

Otorhinolaryngology Clinics: An International Journal (2024): 10.5005/jp-journals-10003-1495

Background

Congenital cholesteatoma (CC) is an epidermal cyst that arises usually from the extradural region of the middle ear or mastoid or intradural at the cerebellopontine angle. It has a prevalence of about 2–5% of all cholesteatomas.[1,2] It is most common in the middle ear and rarest in the mastoid region of the temporal bone with an incidence of about 3–4% of all CCs.[3] It can be dormant for years while slowly increasing in size. However, in some cases, it can rapidly progress causing bony destruction and serious complications.[4] We present 34-year-old female who presented with a large CC of the mastoid with posterior external auditory canal (EAC) erosion.

Case Description

A 34-year-old female presented to the ENT Head and Neck clinic with a 5-month history of intermittently discharging the left ear with occasional dizziness. There were no symptoms of retro auricular swelling, neck pain, or hearing loss. There was no history of previous ear discharge, perforation, or otological surgery in that ear. On examination, she had a protruding mass with surrounding keratin that eroded the lateral third of the posterior EAC wall and skin, partially obscuring the view of her tympanic membrane. Following the aural toilette, the tympanic membrane was visualized and found to be grossly normal (Fig. 1).

A pure tone audiogram showed normal hearing bilaterally. A high-resolution computed tomography (CT) scan showed a mass occupying the inferior and superior aspect of the left mastoid cavity with erosion to the posterior canal (Fig. 2A). The mastoid antrum and aditus were normal (Fig. 2B).

Subsequently, she underwent a canal wall-up mastoidectomy. Intraoperatively, there was a large amount of granulation tissue in the mastoid cavity surrounding a cystic mass. This was removed entirely in a piecemeal fashion. On examination, the mastoid cavity appeared to have undergone autocavitation. There was some erosion of the dura and complete destruction of the lateral portion of the posterior EAC wall with communication to the EAC. There was no communication of the mass to the middle ear and the middle ear structures were normal. However, there was a mild retraction of the posterior superior portion of the tympanic membrane.

Post-operative recovery was uneventful with only mild dizziness that persisted for two weeks. Follow-up at three months revealed a dry ear and a normal tympanic membrane with no evidence of recurrence.

Histopathologic examination showed fragments of stratified squamous epithelial lining with numerous keratin flakes and debris consistent with a cholesteatomas.

Discussion

Congenital cholesteatoma's has been reported to originate from the petrous apex, middle ear, mastoid process and external auditory canal.[3] Derlecki and Clemis were the first to describe an isolated

[1]Department of Otorhinolaryngology Head and Neck Surgery, KPJ Healthcare University College, Negeri Sembilan, Malaysia

[2]Department of Otorhinolaryngology Head and Neck Surgery, KPJ Ampang Puteri Specialist Hospital, Selangor, Malaysia

Corresponding Author: Danny Kit Chung Wong, Department of Otorhinolaryngology Head and Neck Surgery, KPJ Healthcare University College, Negeri Sembilan, Malaysia, Phone: +60342895000, e-mail: drdannykcwong@gmail.com

How to cite this article: Wong DKC, Saim A. Congenital Mastoid Cholesteatoma with Posterior External Auditory Canal Erosion: A Case Report and Literature Review. Int J Otorhinolaryngol Clin 2024;16(1):17–20.

Source of support: Nil

Conflict of interest: None

mastoid CC in 1965.[5] They defined CC of the mastoid to have 1) all features of a CC (i.e., normal tympanic membrane, no previous ear surgery, and no ear discharge) and 2) no involvement of middle ear, attic or aditus.[3,5] We performed a literature search and found that there have only been 29 cases of mastoid CCs in 18 case reports, that have met the criteria above.[1–19] Cases where there was an extension of the mastoid CC to the middle ear such as some cases in Warren et al. case series were excluded from our review due to doubts regarding their site of origin.[1]

Pathophysiology

Congenital cholesteatomas are epidermoid cysts that arise as a result of progressive desquamation of the epithelium. It is theorized to be the failure of reabsorption of embryonic epidermoid residue that leads to the development of a cyst behind the tympanic membrane, which in this case is in the mastoid cavity of the temporal bone.[20] Hong et al theorized that the best theory to explain CC in the mastoid tip is the implantation of squamous epithelium within the tympanomastoid suture during the closure of the mastoid fontanelle.[14] It is locally destructive to bone, which could be explained by two predominant mechanisms; firstly, pressure-induced bone resorption and secondly enzymatic dissolution of bone by cytokine-mediated inflammation.[11]

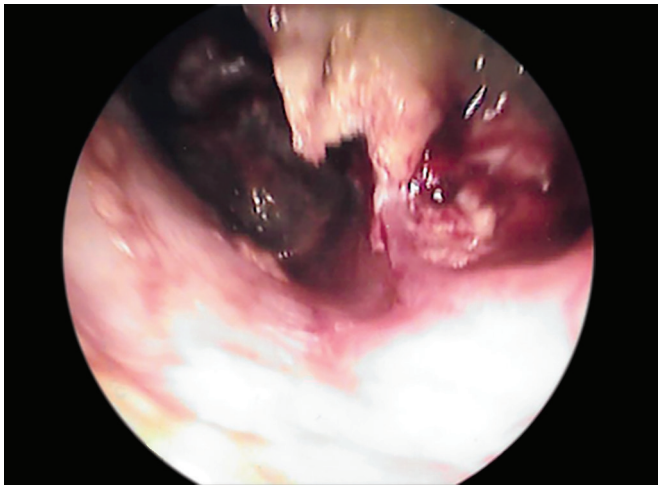


Fig. 1: Otoscopic view of the left EAC showing an eroded posterior canal wall with a mass in the mastoid cavity with keratin debris. The tympanic membrane is seen in the distance and is grossly normal

Clinical Symptoms and Signs

The symptoms of a mastoid CC could be variable, non-specific, and determined by location. They usually present in adulthood due to the lack of symptoms for a long period, unlike their middle ear counterparts which present in childhood.[3] Luntz et al. described the symptoms as neck pain, retro auricular swelling, and persistent disequilibrium.[7,17] However, contrary to Luntz et al., it is quite rare for a mastoid CC to present so typically.[9,10] Majority of the 29 patients in the literature reviewed presented with neck mastoid or ear pain with or without mastoiditis. This was followed by symptoms of ear discharge, hearing loss and ear canal strictures (Table 1).

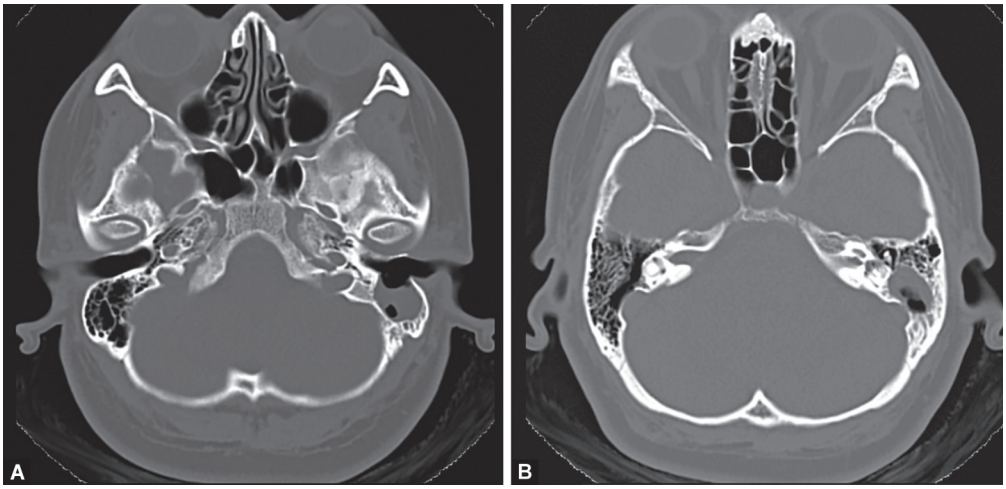
Out of the six cases of reported ear discharge, only three cases had a breach in the skin of the posterior EAC wall in the form of a fistula.[2,14,15] Other cases only noted swelling of the external meatus skin which was probably a signal of an impending fistula.[4,9] Although otorrhea is uncharacteristic of mastoid CCs, it is very possible when posterior EAC wall is eroded. Our case above adds to the few cases where erosion of the posterior canal wall has caused a fistula between the EAC and mastoid cavity.

Diagnosis

Typically, the diagnosis of a CC of the mastoid could be confirmed by a high-resolution CT scan which would demonstrate a non-enhancing, non-specific soft-tissue swelling within the mastoid portion of the temporal bone. The middle ear, aditus, epitympanic recess, and mastoid antrum would be normal.[7] A magnetic resonance imaging (MRI) scan would be helpful to further delineate a mastoid CC as it is hyperintense on T2-weighted images and hypointense on T1-weighted images.[2,17] The most common findings are mastoid cortex erosion, sigmoid plate dehiscence, dural exposure, and external wall destruction.[4] In our case above, the posterior EAC and dura were eroded however the tympanic membrane was intact with no ossicular chain involvement.

Histology

Congenital or acquired cholesteatoma are similar on histo-pathological examination with a bone-eroding skin-lined cavity



Figs 2A and B: (A) Axial CT of the left mastoid cavity filled with the cholesteatoma that has eroded through to the EAC. There is also thinning of the wall of the remainder EAC; (B) Axial CT showing the middle ear cavity and ossicles are normal despite superior extension of the mass

Table 1: Main presentations of previous cases of congenital cholesteatomas of the mastoid

Authors	*Year*	*Main presentation*	*No. of patients*
Derlacki and Clemis[5]	1965	Ear, mastoid or neck pain without mastoiditis	9
El-Kholy and Cherry[6]	1989		
Luntz et al.[7]	1997		
Cüreoglu et al.[8]	2000		
Warren et al.[1] (×2 cases)	2007		
Lee et al.[9]	2007		
Gianuzzi et al.[3]	2011		
Cvorovic et al.[4]	2014		
Rajan et al.[12]	2005	Ear, mastoid or neck pain with mastoiditis	5
Hidaka et al.[10]	2009		
Janardhan et al.[11]	2009		
Cvorovic et al.[4]	2014		
Tabook et al.[13]	2015		
Rashad et al.[15]	1999	Ear discharge	5
Hong et al.[14]	2007		
Cvorovic et al.[4] (×2 cases)	2014		
Chauhan et al.[2]	2014		
	2015		
Mevio et al.[17]	1997	Dizziness	3
Gianuzzi et al.[3]	2002		
Granato et al.[16]	2011		
Warren et al.[1]	2007	Hearing loss	2
Cvorovic et al.[4]	2014		
Warren et al.[1]	2007	Incidental	3
Giannuzzi et al.[3]	2011		
Cvorovic et al.[4]	2014		
Nagato et al.[18]	2012	Ear strictures or bulging posterior canal wall	1

or sac which is filled with concentric layers of the desquamated epithelium.[12] The only difference is that congenital cholesteatomas are meant to be sterile unless contaminated through surgery or erosion through the EAC such as in our case here.

Differential Diagnosis

The differential diagnosis of a mass arising from the mastoid could include cholesterol granulomas, meningiomas, tumors of the endolymphatic sac, anomalous sigmoid sinus, jugular paragangliomas, or other neoplasms.[19]

Treatment

The primary management in all CC whether from the middle ear or mastoid is surgical excision of the tumor. The options range from a simple mastoidectomy to a modified radical mastoidectomy, depending on the extent of the tumor. The decision for a canal wall-up mastoidectomy in our case was to preserve hearing and to avoid a tympanoplasty.

Conclusion

Congenital cholesteatoma of the mastoid is still an uncommon disease that has a relatively uncomplicated treatment approach with good outcomes when detected early. The delay in its diagnosis is likely due to the lack of symptoms and awareness of such a disease. Our case presented is atypical for a mastoid CC due to the otorrhea.

Clinical Significance

Therefore, primary care physicians and otolaryngologist should always maintain a high index of suspicion and a low threshold for a CT scan in patients with a history of an intermittently discharging ear, a narrowed ear canal, or a posterior canal wall fistula.

References

1. Warren FM, Bennett ML, Wiggins RH 3rd, et al. Congenital cholesteatoma of the mastoid temporal bone. Laryngoscope 2007;117(8):1389–1194. DOI: 10.1097/MLG.0b013e3180645d50.
2. Chauhan A, Wadhwa V, Rajan S, et al. Congenital choelasteatoma of the mastoid process: A case report. Otolaryngology Online Journal 2015;21(1):72–78. DOI: 10.4103/0971-7749.152875.
3. Giannuzzi AL, Merkus P, Talbah A, et al. Congenital mastoid cholesteatoma: Case series, definition, surgical key points, and literature review. Ann Otol Rhinol Laryngol 2011;120(11):700–706. DOI: 10.1177/000348941112001102.
4. Cvorovic L, Djeric D, Vlaski L, et al. Congenital cholesteatoma of mastoid origin–a multicenter case series. Vojnosanit Pregl 2014;71(7):619–622. DOI: 10.2298/VSP120130005C.
5. Derlacki EL, Clemis JD. Congenital cholesteatoma of the middle ear and mastoid. Trans Am Otol Soc 1965;53:208–231. PMID: 5834665.
6. El-Kholy AA, Cherry JR. Primary cholesteatoma of the mastoid presenting as cervical abscess. J Laryngol Otol 1989;103(1):92–93. DOI: 10.1017/s0022215100108114.
7. Luntz M, Telischi F, Bowen B, et al. Imaging case study of the month. Congenital cholesteatoma isolated to the mastoid. Ann Otol Rhinol Laryngol 1997;106(7 Pt 1):608–610. DOI: 10.1177/000348949710600715.
8. Cüreoglu A, Osma U, Oktay MF, et al. Congenital cholesteatoma of the mastoid region. J Laryngol Otol 2000;114(10):779–780. DOI: 10.1258/0022215001903906.
9. Lee JH, Hong SJ, Park CH, et al. Congenital cholesteatoma of mastoid origin. J Laryngol Otol 2007;121(11):e20. DOI: 10.1017/S0022215107000825.
10. Hidaka H, Ishida E, Kaku K, et al. Congenital cholesteatoma of mastoid region manifesting as acute mastoiditis: Case report and literature review. J Laryngol Otol 2010;124(7):810–815. DOI: 10.1017/S0022215109992209.
11. Janardhan N, Nara J, Peram I, et al. Congenital cholesteatoma of temporal bone with bezold's abscess: Case report. Indian J Otolaryngol Head Neck Surg 2012;64(1):97–99. DOI: 10.1007/s12070-011-0226-6.
12. Rajan KV, Santhi T. An unusual case of congenital cholesteatoma in the mastoid with epidural abscess and cutaneous fistula. Indian J Otolaryngol Head Neck Surg 2005;57(4):339–341. DOI: 10.1007/BF02907707.
13. Tabook SM, Abdel Tawab HM, Gopal NK. Congenital cholesteatoma localized to the mastoid cavity and presenting as a mastoid abscess. Case Rep Otolaryngol 2015;2015:305494. DOI: 10.1155/2015/305494.
14. Hong SM, Lee JH, Park CH, et al. Congenital cholesteatoma localized to the tip of the mastoid bone: A case report and possible etiology. Korean J Audiol 2014;18(2):85–88. DOI: 10.7874/kja.2014.18.2.85.
15. Rashad U, Hawthorne M, Kumar U, et al. Unusual cases of congenital cholesteatoma of the ear. J Laryngol Otol 1999;113(1):52–54. DOI: 10.1017/s0022215100143130.
16. Granato L, Silva CJ, Yoo HJ. Isolated congenital cholesteatoma of the mastoid process: A case report. Braz J Otorhinolaryngol 2012;78(4):133. DOI: 10.1590/S1808-86942012000400024.
17. Mevio E, Gorini E, Sbrocca M, et al. Congenital cholesteatoma of the mastoid region. Otolaryngol Head Neck Surg 2002;127(4):346–348. DOI: 10.1067/mhn.2002.128604.

18. Nagato T, Otaka R, Wada T, et al. Congenital cholesteatoma isolated to the mastoid presenting as stricture of the external auditory canal. Int J Pediatr Otorhinolaryngol 2012;76(5):754–756. DOI: 10.1016/j.ijporl.2012.02.032.
19. Thakkar KH, Djalilian HR, Mafee MF. Congenital cholesteatoma isolated to the mastoid. Otol Neurotol 2006;27(2):282–283. DOI: 10.1097/00129492-200602000-00023.
20. Davidoss N, Ha J, Banga R, et al. Delayed presentation of a congenital cholesteatoma in a 64-year-old man: Case report and review of the literature. J Neurol Surg Rep 2014;75(1):e113–e116. DOI: 10.1055/s-0034-1376200.