

Case Report

Congenital Cholesteatoma in Adult: Is it Still Possible?

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Abstract:

Congenital cholesteatoma is a mass of squamous epithelium located medial to an intact tympanic membrane without previous history of tympanic membrane perforation, otorrhoea or otological surgery. We described a 24 year old gentleman with a left postauricular discharging fistula for 3 years with recent history of gradual hearing loss, tinnitus and recurrent episodes of positional vertigo. Clinical examination noted left postauricular fistula opening and otoscopy showed a whitish mass medial to a bulging intact tympanic membrane. High-resolution computed tomography of temporal bone was suggestive of cholesteatoma. Left modified radical mastoidectomy was done and he recovered with resolution of symptoms.

Keywords: *Congenital cholesteatoma, fistula, mastoidectomy, computed tomography scan, vertigo*

Introduction :

Congenital cholesteatoma (CC) is an expanding cystic mass of keratinizing squamous epithelium located medial to an intact tympanic membrane in patients without any prior history of tympanic membrane perforation, otorrhoea or otological surgery. However, a previous history of otitis media or effusion does not exclude CC¹. It is most commonly postulated to originate from rests of epithelial cells in middle ear which failed to

involute during intrauterine life. These keratinizing squamous epithelium will progress slowly and ultimately result in a locally invasive cholesteatoma². It is mainly a disease of childhood with a paucity of adult cases reported. We report a case of CC with discharging post auricular fistula as the sole complaint.

Case Report :

A 24 year old gentleman presented with history of discharging left postauricular fistula for the past 2 years. He also complained of gradual hearing loss and tinnitus for the past 1 year with history of positional vertigo lasting minutes for the past month. Otherwise, he denied any history of otorrhoea, ear surgery or local trauma.

Clinical examination showed a left postauricular fistula opening with surrounding erythema and depressed mastoid bone area on palpation. Otoscopy revealed bulging left tympanic membrane with whitish mass seen

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medial to tympanic membrane. However, there was no sagging of posterior external auditory canal seen.

Pure tone audiometry showed conductive hearing loss of 40 dB over left side and a normal hearing contralaterally. Left and right ear had a type C and type A tympanometry respectively.



Fig 1 Otoscopy showing cholesteatoma medial to bulging, intact left tympanic membrane

His high-resolution computed tomography (HRCT) of temporal bone revealed soft tissue density in left middle ear cavity, mainly at mesotympanum extending to hypotympanum with erosion of ossicles and scutum, thus, supporting the diagnosis of cholesteatoma. Dehiscence of lateral mastoid wall with erosion of mastoid air cell septae were seen.

Left modified radical mastoidectomy via postauricular approach was done under general anaesthesia. Unhealthy skin around postauricular fistula was excised. Cholesteatoma sac was seen in middle ear and removed in total. Eroded head of malleus and remnants were removed. Both incus and stapes were unable to be identified. However, oval window was intact. Lateral semicircular canal bony dehiscence was seen with intact endosteum. Facial nerve canal was intact.

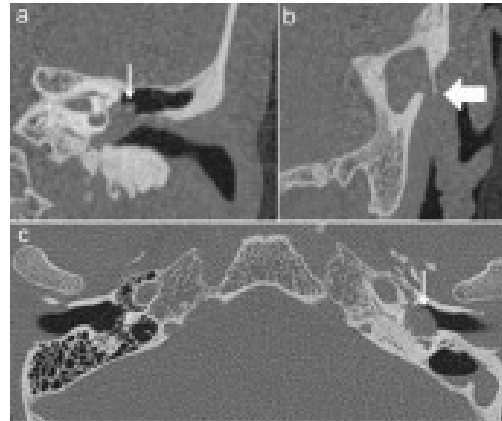


Fig 2 High-resolution computed tomography of left temporal bone in coronal view (a and b) and axial view (c). Thin arrows represent cholesteatoma at mesotympanum extending to hypotympanum with erosion of ossicles and scutum. Thick arrow represents dehiscence of lateral mastoid wall

Noleakage of perilymph seen on Valsalva manoeuvre which was done intraoperatively. Tympanoplasty was done using temporalis fascia graft.

Histopathological examination confirmed the diagnosis of cholesteatoma. Upon follow-up at 4 months postoperatively, he had a full and uneventful recovery with no residual vertigo and no worsening of his hearing.

Discussion:

CC is mostly seen in paediatric age group with a mean age of presentation at 5 years.³ However, there is a scarcity of reports in adult population. Misale et al. reported a series of 6 patients ranging from 18 to 49 year old with a mean of 27.5 year old⁴. Our case illustrates an atypical presentation in an adult.

Due to the slow growing nature of CC, patients with CC can be asymptomatic. Clinical presentation is determined by the site and extend of disease. CC can occur at the petrous apex and in the middle ear cleft.

However, for middle ear cleft CC, it is classically seen at the middle ear, medial to an intact tympanic membrane with the anterosuperior quadrant being the commonest site and posterosuperior quadrant being second most common. Conductive hearing loss is the most common symptom, although unilateral hearing loss is frequently unrecognized in young children. Hearing loss may only be identified as cholesteatoma grew to a large size filling the middle ear cavity or when ossicles are eroded as illustrated in our patient where only remnants of malleus remained^{2,5}.

Priyanka et al. reported a similar case with discharging post auricular fistula as the sole presentation, however, unlike our patient, the cholesteatoma sac was confined to the mastoid region only.⁴ We postulate that the postauricular fistula in our patient is likely a complication from chronic mastoiditis as suggested by the erosion of mastoid air cell septae and dehiscence of lateral mastoid wall. His vertiginous symptom is attributed to the lateral semicircular canal bony dehiscence caused by cholesteatoma. Cholesteatoma sac was able to be removed intraoperatively without further injury to the canal and his symptoms subsequently resolved postoperatively.

Other reported symptoms of CC in adult include facial nerve palsy, loss of taste and meningitis. Fortunately, CC has a lower incidence of intracranial complications compared to acquired cholesteatoma^{4,5,6}.

HRCT of temporal bone is invaluable to assess location and extend of disease. Typical findings seen are well demarcated soft tissue density, blunting of scutum, erosion of ossicles and tympanic tegmen.² Common extradural sites of occurrence are middle ear, mastoid, squamous temporal bone and petrous apex⁷.

Surgery is the mainstay of treatment with the aim of complete eradication of the disease and creation of a safe ear with optimization of hearing as second in priorities. This can be achieved by either canal wall up (CWU) or canal wall down (CWD) mastoidectomy. CWD surgery involves removal of posterior external auditory canal wall to exteriorize middle ear and mastoid cavity, thus, improving visualization intraoperatively. It also facilitates monitoring for recurrence. CWU surgery preserves the external auditory canal wall and confer the benefit of avoiding the need for lifelong ear cleaning and water precautions. However, it is associated with higher rates of residual disease on the virtue of reduced exposure intraoperative and necessitates a second-look procedure^{2,5}. In our case, a CWD mastoidectomy was performed to facilitate total removal of cholesteatoma sac. The choice of surgery should be individualized according to extend of disease and patient's social factors. Nonetheless, long-term follow-up is of paramount importance regardless of technique to monitor for disease recurrence.

Conclusion :

CC is predominantly a childhood disease; however, adults can present with atypical symptoms. It should be a differential in case where white mass is seen medial to an intact tympanic membrane. Early detection of CC is crucial to prevent its complications.

Declarations :

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Conflict of Interest :

The authors declare that they have no conflict of interest.

Ethical Approval :

This case report was conducted in accordance with the Declaration of Helsinki.

Ethics approval was given by National Medical Research Register of Malaysia under research ID 55872.

Consent to Participate :

Informed consent was obtained for all procedures including the publication of all photographs.

Consent to Publish :

Additional informed consent was obtained for the publication of data and photographs.

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