



Sudden Hearing Loss Secondary to Cholesteatoma Induced Labyrinthine Fistula

IMAGE

Geng Ju Tuang , Shankar Ramasundram , Abdul Ruhi Ul Azim Liman 

A 30-year-old male complained of sudden onset of right-sided hearing loss and dizziness for two days. There was neither recent loud noise exposure nor head and neck trauma. He also denied other otological symptoms, including otorrhea, otalgia, aural fullness and tinnitus. Upon clinical assessment, horizontal nystagmus was demonstrated through the elicitation of the right external ear canal (EAC) pressure with saccadic movement towards the contralateral side. Rinne's test unveiled better bone conduction on the affected ear, with Weber's test lateralized to the contralateral ear. Endoscopic examination of his right ear showed deep retraction pocket over the attic region filled with keratin flecks (Fig. 1). Pure tone audiometry revealed a severe sensorineural hearing loss in the right ear. Computerized tomography (CT) of the temporal bone showed sclerosis of the right mastoid bone and a hypodense lesion within the mastoid antrum and epitympanum, causing bony defect on the horizontal semi-circular canal (Fig. 2). A subsequent modified radical mastoidectomy established the diagnosis of a cholesteatoma. The cholesteatoma sac was gently removed, revealing the underneath horizontal semi-circular canal endosteum fistula with an intact membranous labyrinthine. The bony defect was sealed with bone pate. Repeated pure tone audiometry at three months postoperative showed improved hearing threshold on the right ear.

Sudden hearing loss is regarded as a devastating symptom with significant impacts on patients. Despite its rare occurrence, such clinical manifestation frequently prompts an emergency visit to the health care providers. A detailed relevant history and clinical assessment are, therefore, paramount for defining potential treatment and prognosis (1). Labyrinthine fistulas represent a well-known complication from cholesteatoma owing to its bone eroding characteristic (2). The clinical manifestation of a labyrinthine fistula encompasses a dynamic spectrum from asymptomatic to severe vertigo and hearing loss. The propagation of inflammatory mediators and toxins from the cholesteatoma into the inner ear fluid through the fistula may further deteriorate hearing (2). Bedside EAC pressure-induced nystagmus or vertigo, also known as the fistula test, may help in detecting the abnormal communication between the middle and inner ear (3). The management of cholesteatoma constitutes a surgical removal with hearing preservation as a secondary intention. Meticulous removal of cholesteatoma matrix preceding grafting of the fistulous site has shown promising postoperative hearing outcomes (2). In conclusion, a heedful clinical evaluation cannot be overemphasized in such cases. Attentive observation of the CT scan aids to provide useful diagnostic information and facilitates the subsequent operative planning.



Figure 1. Otoendoscopy of the right ear showed keratin filled deep retraction pocket of the pars flaccida (blue arrow) and intact pars tensa (white arrow)

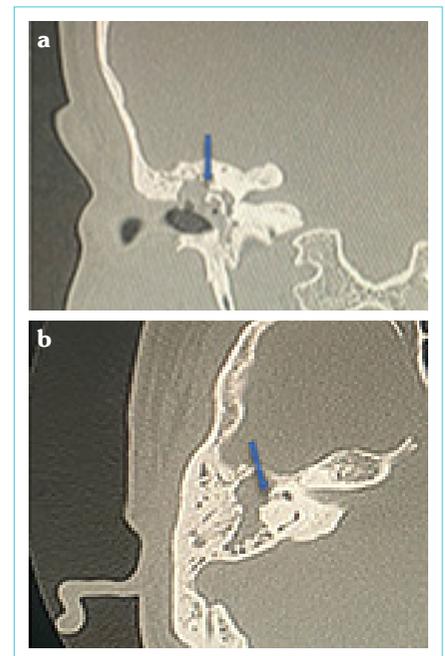


Figure 2. Coronal (panel A) and axial view (panel B) of computerized tomography of the (right) temporal bone demonstrate that a soft tissue fills the mastoid antrum and epitympanum, causing a bony defect on the horizontal semi-circular canal (blue arrow)

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Department of
Otorhinolaryngology, Head
and Neck Surgery,
Hospital Selayang,
Batu Caves, Malaysia

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Correspondence
Geng Ju Tuang,
Hospital Selayang,
Department of
Otorhinolaryngology, Head
and Neck Surgery,
Batu Caves, Malaysia
Phone: +6012- 2298 360
e-mail: erictgj@gmail.com

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