The Buzzing Ear: A Case of Bilateral Superior Semicircular Canal Dehiscence

Case Report

Erica Gima[1], Mohd Normani Zakaria[2], Mohd Khairi Md Daud[1], Nik Adilah Nik Othman[1]

Department of [1]Otorhinolaryngology-Head and Neck Surgery, School of Medical Sciences, [2]Programme of Audiology, School of Health Sciences, Universiti Sains Malaysia, Health Campus, Malaysia.

ABSTRACT

Superior semicircular canal dehiscence syndrome is a condition that mimics various otologic and vestibular problems. Diagnosis can be achieved via a combination of physical examination, audiometry, vestibular-evoked myogenic potential and high resolution computed tomography of temporal bone. We present a case of a 64-year-old lady with pulsatile tinnitus and reduced hearing in the right ear for four years. Her initial audiogram showed a right mild to moderate conductive hearing loss at low frequencies and left normal hearing at low and mid frequencies with moderate sensorineural hearing loss at high frequency, while tympanogram was type A bilaterally. A high-resolution computed tomography of temporal bone was reported as dehiscent of both superior semicircular canals. As she was able to cope with her tinnitus and her symptoms improved with time, we opted for conservative management. We highlight the importance of considering superior semicircular canal dehiscence syndrome as a differential in patients presenting with auditory and vestibular symptoms.

Key Words: Autophony, conductive hearing loss, tinnitus, superior semicircular canal dehiscence, vertigo.

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Corresponding Author: Nik Adilah Nik Othman, MMed, Department of Otorhinolaryngology - Head & Neck Surgery, School of Medical Sciences, Health Campus USM, 16150 Kubang Kerian, Kelantan, Malaysia. **Tel.:** 60199377080, **E-mail**: adilahkk@usm.my

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INTRODUCTION

Superior semicircular canal dehiscence syndrome (SSCDS) occurs when there is a bony defect of the superior semicircular canal, enabling pressure transmission through the dehiscence. This "third window" leads to variable symptomatology which makes diagnosis elusive. Patients usually present with auditory and vestibular symptoms with variable degrees of severity. Patients with severe symptoms may benefit from surgical intervention, but in those with mild symptoms, avoidance of triggering factors and conservative management would suffice.

Case Report:

A 64-year-old Malay lady presented with right tinnitus for four years. It was a pulsatile and low-pitched buzzing sound, which occurred more frequently in the past few months. The tinnitus was worse in a quiet environment and early mornings. There were no identifiable triggers or relieving factors. She had non progressive hearing loss on the right ear for four years with intermittent blocked sensation and autophonia. However, she denied any ear discharge or pain. There was no dizziness or spinning sensation, and the patient did not have any history of head trauma or exposure to loud noise. She had mild and intermittent allergic rhinitis symptoms. Despite the increase in frequency, the tinnitus did not disturb her sleep and daily activities. The patient also denied any difficulties in communication due to her hearing problem.

The otoscopic findings were normal and there was no rising sun sign to indicate the presence of glomus tumor. Upon testing with a 512-Hz tuning fork, Rinne test was positive bilaterally and sound was lateralized to the right ear on Weber test. The cranial nerves were intact. Nasoendoscopy showed no significant findings. The tympanometry was type A bilaterally indicating normal middle ear function, and the pure tone audiometry (PTA) revealed right mild to moderate conductive hearing loss at low frequencies and left normal hearing at low and mid frequencies with moderate sensorineural hearing loss at high frequency (Figure 1). Her computed tomography (CT) angiogram was normal and a high resolution computed tomography (HRCT) of temporal bone showed dehiscence measuring 1mm over the arcuate eminence bilaterally (Figure 2 and 3).

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She was taught distraction methods of masking the tinnitus with external sounds such as music and nature sounds. She was also started on Mecobalamin and Loratadine. Although her tinnitus gradually improved, she started to complain of difficulty understanding speech in noise. A repeated tympanometry showed a type C bilaterally indicating Eustachian tube dysfunction, and her PTA showed right mild to severe mixed hearing loss and left mild to moderate mixed hearing loss (Figure 4). Audiometric Weber was lateralized to her right ear at 250-Hz and 500-Hz, indicating poorer hearing of conductive loss on the right side. Acoustic reflex was absent at all frequencies both ipsilaterally and contralaterally.

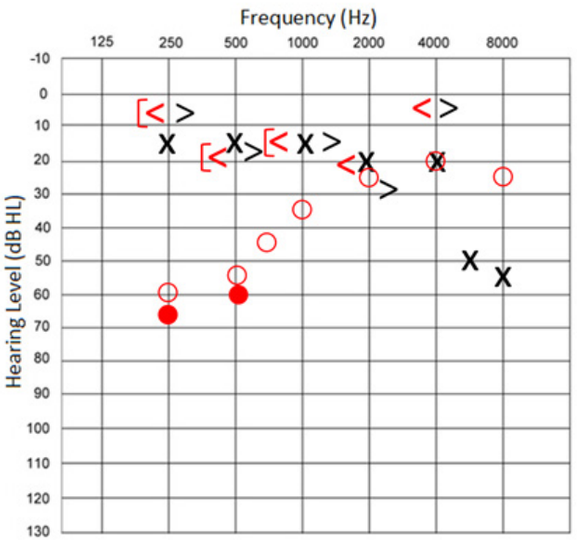

Fig. 1: Pure tone audiogram in March 2020.

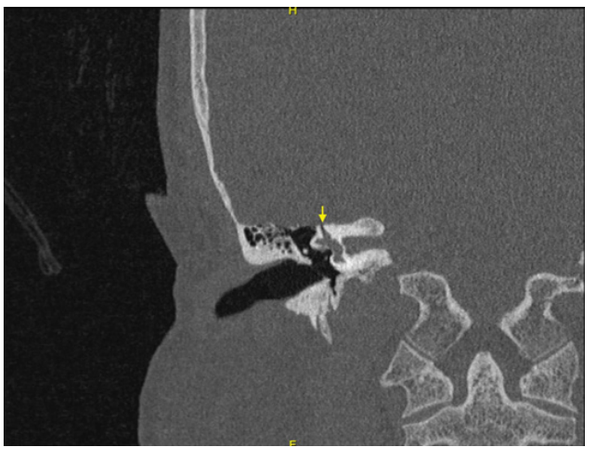

Fig. 2: Coronal view of right temporal bone. Yellow arrow points to area of dehiscence.

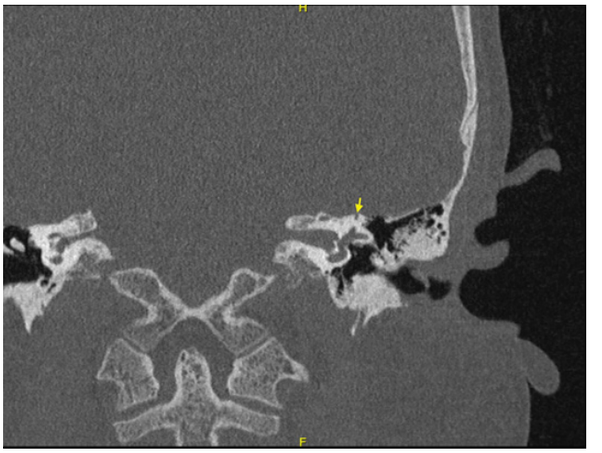

Fig. 3: Coronal view of left temporal bone. Yellow arrow points to area of dehiscence.

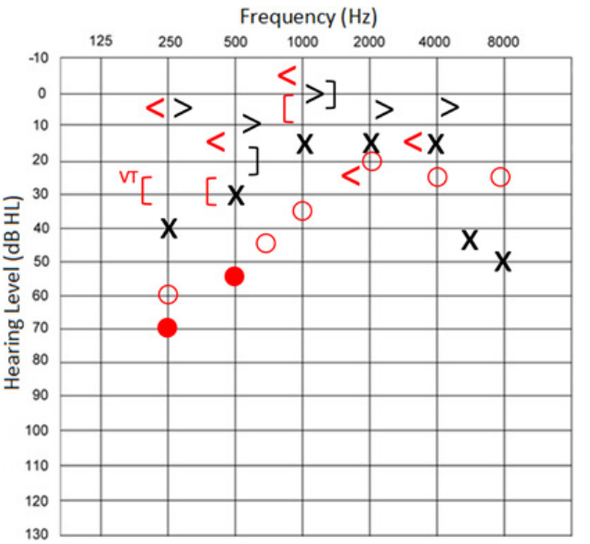

Fig. 4: Pure tone audiogram in April 2021.

DISCUSSION

SSCDS was first described by Minor *et al* in 1998[1]. The pathogenesis can be explained by a "third window" created by the dehiscence at the superior semicircular canal[2]. Consequently, any pressure changes or loud sounds may trigger movement at the dehiscence. Under normal circumstances, mechanical energy transmitted through the oval window is propagated through the scala vestibuli and scala tympani, driving movement of the basilar membrane which generates action potentials. A third window creates an alternate pathway for the inner ear fluid, dispersing some of the acoustic energy away. In PTA, this is reflected by a large air bone gap at the lower frequencies, whereas bone conduction become supranormal.

Patients often suffer from conductive hearing loss with hypersensitivity to bone conducted sounds which may be reported as hearing their eyeballs move or hearing their own footfalls. Other auditory symptoms include pulsatile tinnitus and aural fullness. Patients with vestibular symptoms may report Tullio's phenomenon, described as dizziness induced by loud sounds or display Hennebert's sign, whereby nystagmus is triggered by pressure changes when doing the fistula test or Valsalva maneuver[3]. The severity of symptoms varies, with some presenting with exclusively auditory or vestibular symptoms, or a mix of both. This variability of symptoms has been attributed to the size of dehiscence[4], patency of cochlear aqueduct, the compliance of round window membrane[5] and elasticity of dura[6].

SSCDS is a great mimicker, with symptoms overlapping those of otosclerosis, Meniere's disease and perilymphatic fistula. Our patient's most prominent symptom was pulsatile tinnitus with reduced hearing, and we initially suspected a vascular cause for her symptoms. Bilateral SSCDS was discovered incidentally upon radiographic studies, which did not tally with her unilateral symptom at that time. This could be due to an overestimate by CT imaging, as the bony covering may be thinner than what is detectable by CT. Cadaveric temporal bone studies showed that the prevalence of SSCDS is 0.5%[7], however the percentage increases up to 12%[8] on CT scan with 1-mm thickness slice. To overcome this shortcoming, the slices should be set ideally at 0.65mm or less and the images reformatted to include views parallel and perpendicular to the superior semicircular canal, known as Poschl's and Stenver's view respectively[9]. Studies showed that the bony defect is most commonly found at the arcuate eminence and rarely involves the posteromedial limb[2].

Our patient's tinnitus was improving with treatment, however the patient started to complain of difficulty understanding speech in background noise, which was reflected in the PTA. However, the presence of Carhart's notch and type C tympanogram suggested concomitant middle ear pathology. We did not proceed with vestibular-evoked myogenic potential (VEMP) as the patient's symptoms were mild and VEMP would not change her management.

Surgery for SSCDS is reserved for patients with severely debilitating symptoms. In this patient, conservative management was adequate as her symptoms were limited to tinnitus and reduced hearing. She was able to cope well with her tinnitus and had no problems in communication, except in loud places.

CONCLUSION

The variability of symptoms reported by patients makes the diagnosis of SSCDS challenging. We recommend that SSCDS be included in the differential diagnosis for patients presenting with auditory and vestibular symptoms, bearing in mind that one may occur without the other. This suspicion can then be confirmed by audiometry, VEMP and HRCT. Patients with distressing symptoms will benefit from surgical intervention while those with mild symptoms can be managed conservatively.

CONFLICT OF INTEREST

There are no conflicts of interest.

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