OXFORD

Oxford Medical Case Reports, 2022, **7**, 249–250
https://doi.org/10.1093/omcr/omab125
Clinical Image

Asymptomatic multiple semicircular canal dehiscence: a rare entity

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A 55-year-old male patient presented to our department with chief complaint of left-sided hearing loss. The onset of the hearing loss was reportedly within the last 2 years. He denied vertigo, tinnitus and autophony. The audiologic case history was unremarkable. Otoscopy revealed normal-appearing eardrums bilaterally. Vestibular examination, including Valsalva maneuver, was normal. Pure tone audiometry revealed middle sensorineural hearing loss in the right ear and moderate mixed hearing loss in the left ear. A temporal bone CT-scan revealed multiple dehiscence of bony labyrinthine capsule: right superior, right posterior (Fig. 1A), left superior and left posterior (Fig. 1B). Cervical vestibular evoked myogenic potentials (VEMPs) were later performed and showed normal thresholds and amplitudes on both sides.

Dehiscence of the semicircular canal is a relatively rare entity. It is defined as the absence of bone overlying the semicircular canal facing toward the dura of the middle cranial fossa. It was first described by Minor back in 1998 [1]. It mainly affects the superior semicircular canal (SSCD), and the posterior canal less frequently [2].

Clinically, this pathology presents as a third window syndrome with sound induced vertigo (Tullio's phenomenon), hearing loss, pulsatile tinnitus and autophony. Physical examination occasionally finds a characteristic torsional nystagmus. This nystagmus can be induced by sound or pressure change (Hennebert's sign; [3]). High resolution computerized tomography (CT) usually confirms the diagnosis by showing the dehiscent canal [3]. VEMPs are also useful to detect this disease with high sensitivity and specificity. They typically show high responses and low thresholds [3]. Our patient was asymptomatic from the vestibular viewpoint, which is really surprising given the CT images.

The Barany society has recently proposed diagnostic criteria for SSCD syndrome [4]. Bilateral dehiscence of the two superior semicircular canals has been described on the literature with similar features; however, multiple canal involvement remains exceptional [5]. The bilateral nature of the defects would appear to suggest a developmental abnormality [6]. Management of SSCD involves conservative and surgical approaches. Surgical treatment is generally considered for patients with disabling vestibular symptoms [3].

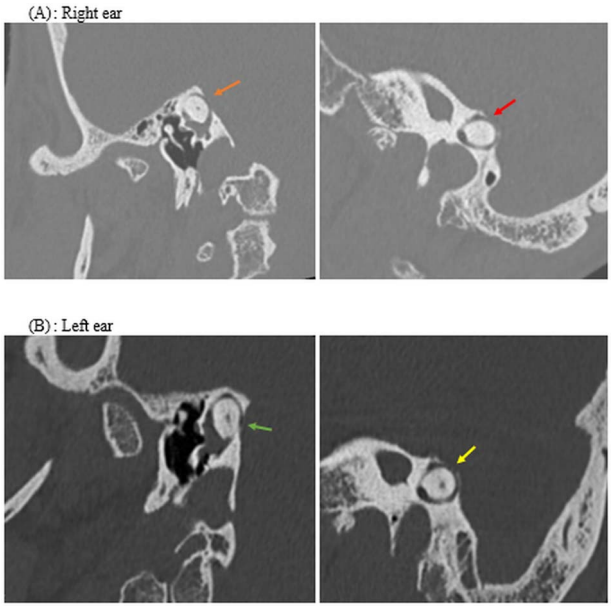

Figure 1. Composite CT scan picture, sagittal projections. (**A**): Right ear: Dehiscent superior canal (red arrow) and posterior canal (orange arrow). (**B**): Left ear: Similar aspect, dehiscent superior canal (green arrow) and posterior canal (yellow arrow).

CONFLICT OF INTEREST STATEMENT

There is no conflict of interest.

FUNDING

There was no funding for this publication.

Received: May 3, 2021. **Revised:** July 14, 2021. **Accepted:** November 5, 2021

ETHICAL APPROVAL

Ethical approval has been exempted by our institution.

CONSENT

Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient.

GUARANTOR

Walid Bijou is the guarantor for this publication.

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