

Bilateral Superior Semicircular Canal Dehiscence and Tullio Phenomenon

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A 44-year-old female presented to a neurology clinic with changes to her hearing and balance for the past 3 years. She endorsed hearing her own voice, heartbeat, and footsteps. Notably, loud noises initiated rotatory nystagmus and vertigo. The echolalia was present in the left ear. Patient denied any subjective hearing loss but described longstanding nonpulsatile tinnitus that had not changed over the past 3 years.

On examination, tympanic membranes appeared normal bilaterally. Full cranial nerve examination, including extraocular movements, was normal. Pneumatic otoscopy and palpation of the tragus to perform a fistula test demonstrated a subtle reproducible rotatory nystagmus synchronous with pulsations. This also elicited visual symptoms that the patient had been experiencing previously. Audiogram demonstrated a left 20-dB conductive

hearing at 2000 Hz and a symmetric 25-dB loss in the high frequencies. Her acoustic reflexes were all intact, in the setting of a conductive hearing loss, making it suspicious for a possible “third-window” type pathology.

Computed tomography (CT) scan of the temporal bone was obtained demonstrating intact tympanic membranes and ossicles bilaterally. The facial nerve followed a normal course and the internal auditory canal was also normal. However, dehiscence of the superior semicircular canal was noted bilaterally (Figures 1 and 2).

DISCUSSION

Superior semicircular canal dehiscence (SSCD) is a radiological diagnosis and is associated with Tullio phenomenon, which refers to

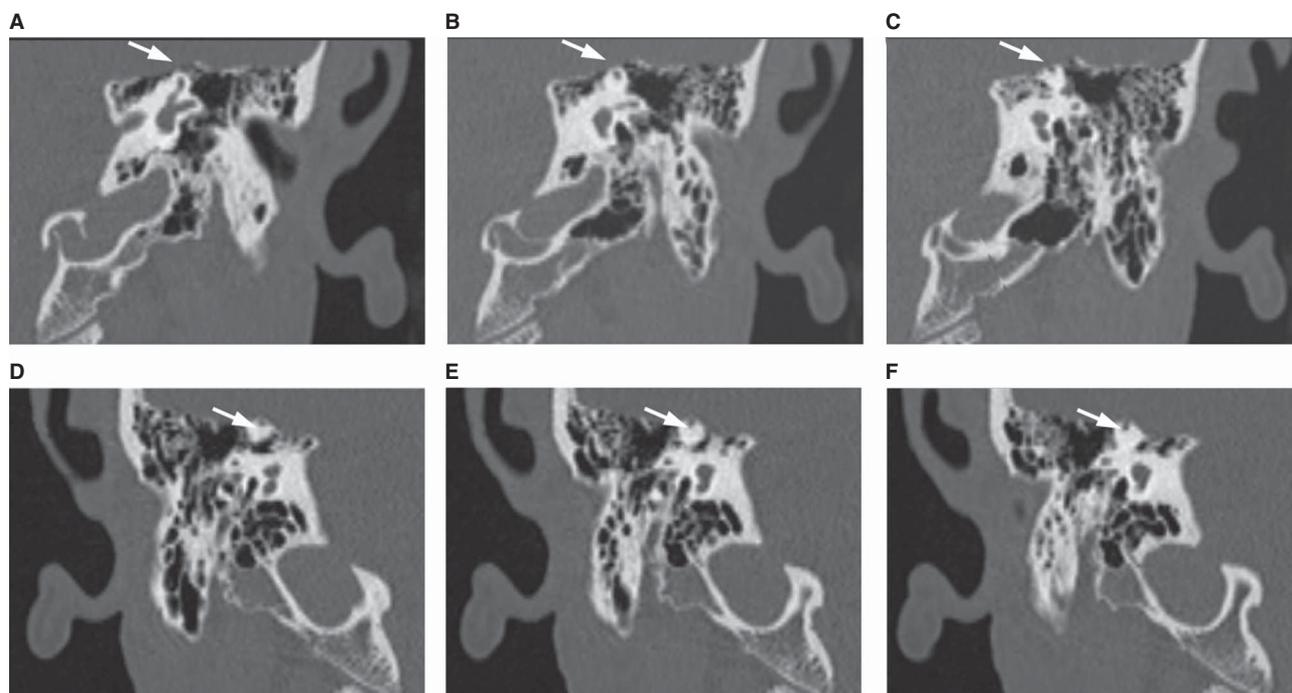


Figure 1: Coronal reformats of the left (A-C) and right temporal bone (D, E) of unenhanced computed tomography demonstrating dehiscence of the superior semicircular canal (white arrow).

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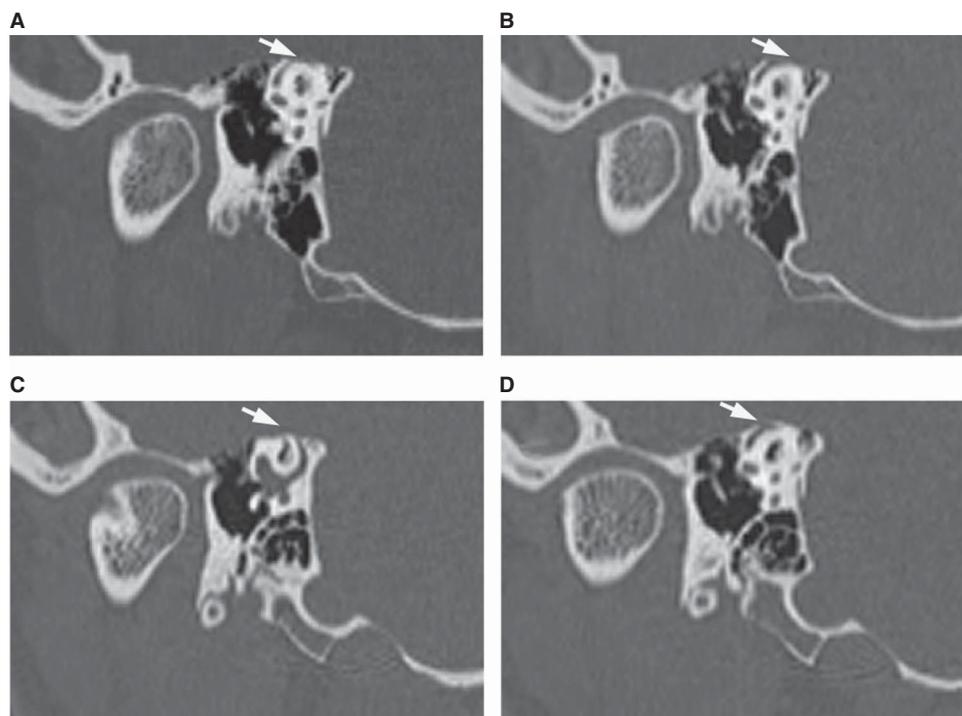


Figure 2: Poschl view of the left (A, B) and right temporal bone (C, D) of unenhanced computed tomography, demonstrating dehiscence of the superior semicircular canal. The size of the maximal dehiscence on the Poschl view was 3.9 mm on the left and 1.6 mm on the right superior semicircular canal.

sound-induced hearing loss, vertigo, nystagmus, and oscillopsia.¹ In one study, the prevalence of SSCD was 0.7%, and 1.3% had a bony layer of less than 0.1 mm covering the canal in normal subjects.² Though pathogenesis is uncertain, prior studies have implicated possible genetic, embryologic, metabolic and traumatic etiologies.³⁻⁵

In SSCD, an additional conduction from bone to fluid through the dehiscence is present, acting as a “third window” of conduction. The transmitted oscillations from high-intensity acoustic stimuli and increased pressure in the canal cause direct stimulation of the cochlear hair cells. This mechanism results in tinnitus and autophony, including vertigo, nystagmus, and oscillopsia.^{1,5}

Diagnosis is made initially on clinical grounds and vestibular evoked myogenic potentials causing vestibular response to stimuli of lower intensity than expected. The diagnosis is confirmed with CT scan of the temporal bone.⁶ A 1-mm CT scan of the temporal bone has a sensitivity and specificity of 100% and 77%, respectively, for SSCD.⁷ In addition, there seems to be no association between the size of temporal bone dehiscence and symptoms.⁷ However in our case, the dehiscence was larger on the left ear (3.9 mm on left vs 1.6 mm on right), which correlated with the clinical findings of echolalia on the left ear.

Additional reformations in the planes of Stenver (perpendicular to canal) and Poschl (parallel to the canal) do not seem to change the diagnostic interpretation for SSCD.⁸ In addition, cone beam CT has emerged as a low-dose technique in visualizing high-contrast structures in the head and neck. However, there is insufficient evidence at present to conclude that cone beam CT is superior to the conventional CT.⁹

Surgical repair of the canal defect is offered to individuals with debilitating symptoms.¹⁰ Surgical techniques include canal

plugging and resurfacing of the canal. Improvement in 94% of the patients who underwent surgery has been previously demonstrated; however, most patients with SSCD opt for conservative management.¹¹

DISCLOSURES

Jonghun John Lee, Pavlo Ohorodnyk, Manas Sharma, and Sachin Kishore Pandey do not have anything to disclose.

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