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Superior Semicircular Canal Dehiscence Symptoms Unmasked by Ossicular Chain Reconstruction

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Keywords

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Superior semicircular canal dehiscence (SSCD) was first described in 1998 among patients experiencing an unusual combination of auditory and vestibular symptoms.¹ Common symptoms include autophony, aural fullness, and dizziness/vertigo.² Patients often exhibit sound- and pressure-induced eye movements on examination and low-frequency conductive hearing loss on audiometric testing.^{1,2} The pathophysiology of SSCD lies in the thinning or absence of the bone between the middle cranial fossa and the superior semicircular canal, thus creating a “third window” into the inner ear.³ Otosclerosis, which can generate oval window immobility, can coexist with SSCD and suppress SSCD symptoms, and accordingly stapes surgery can “unmask” the third window.⁴ In the current case study, we describe, for the first time, the unmasking of a third window following an ossicular chain reconstruction, resulting in symptomatic SSCD.

Case Report

A 32-year-old man was referred to our institution for left-sided, slowly progressive, and near-maximal conductive hearing loss and presumptive otosclerosis. Notably, the patient reported that due to his hearing loss, he would habitually auto-insufflate his left ear to achieve a momentary improvement in hearing and, in doing so, experience mild dizziness for several seconds. The patient had present acoustic reflexes, and accordingly, a computed tomography scan was performed, demonstrating left-sided SSCD (**Figure 1A**), a possible calcified anterior malleolar ligament (**Figure 1B**), and the absence of an otosclerotic focus in the fissula ante fenestram.

The identification of the SSCD prior to treatment allowed for appropriate patient counseling regarding the possibility of the contribution of one or both pathologies to

his conductive hearing loss. In addition, it was explained that repair of an ossicular fixation may introduce additional SSCD symptoms. Given the severe impact of the hearing loss on his life, the patient desired all possible interventions to improve his hearing. A left-sided transcanal middle ear exploration was performed, which revealed a rigidly fixed malleus and incus. The incus and malleus head were removed and replaced with an incus interposition prosthesis, successfully narrowing the air-bone gap (**Figure 2**).

Three weeks following surgery, the patient experienced dizziness with exertion, as well as new SSCD symptoms, including autophony and increased awareness of internal body sounds—namely, the appreciation of his own footsteps in his left ear. Nevertheless, the patient was exceptionally pleased with his improved hearing and, at 5 months after the procedure, was not sufficiently bothered by his SSCD symptoms to pursue any surgical intervention.

University of California–Irvine Institutional Review Board exemption was obtained for this case report.

Discussion

Masking of a third window can theoretically occur when middle ear pathologies are present, such as otosclerosis and ossicular chain fixation. These pathologies can result in hypomobility or immobility at the level of the oval window and mitigate third-window effects by maintaining a 2-window system. Third-window physiology and symptoms may emerge if the middle ear pathology is repaired and oval window mobility is restored.

The unmasking of SSCD has been described in the context of poststapedotomy vertigo and persistent auditory symptoms.⁴ In the current and novel case study, we report

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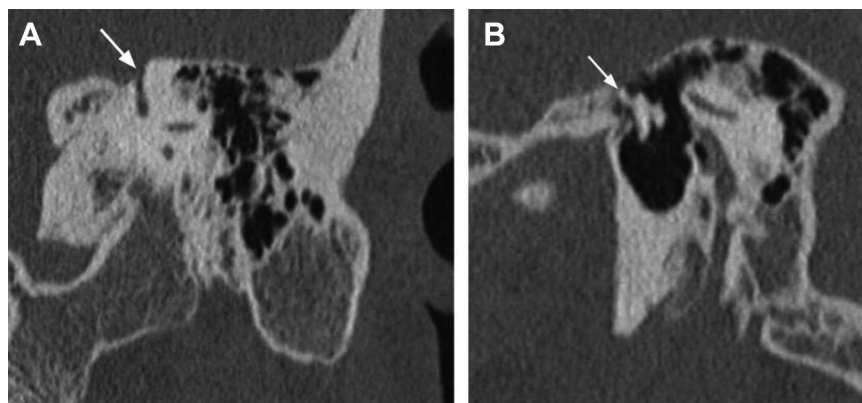


Figure 1. Computed tomography: A, coronal image demonstrating a large superior semicircular canal dehiscence (arrow); B, sagittal image showing calcification of the anterior malleal ligament (arrow).

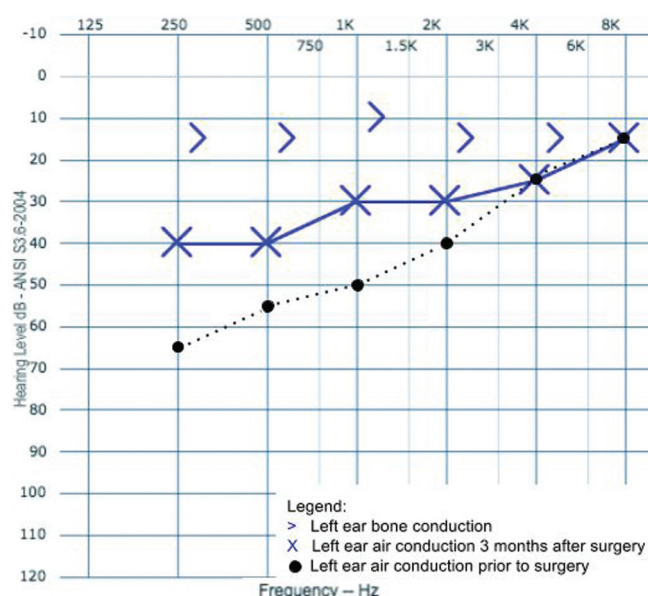


Figure 2. Patient audiogram showing pre- and postoperative hearing.

that the restoration of mobility to the oval window by means of an ossicular chain reconstruction for ossicular fixation can also unmask the third-window phenomenon and reveal SSCD symptoms.

It is paramount to consider evaluation for SSCD for any patient with conductive hearing loss when considering surgical treatment for otosclerosis. On rare occasion, middle and inner ear pathologies can coexist, and a low threshold to obtain imaging should be held. Vestibular evoked myogenic potential (VEMP) testing can also be performed to assist in the diagnosis of SSCD.² In this case, VEMP was considered but not obtained due to the clear diagnosis of SSCD on computed tomography imaging and the radiographic evidence of a middle ear pathology, which would undermine the results of VEMP testing. Although restoration of mobility to the oval window in the setting of comorbid SSCD does not always introduce third-window symptoms,⁵

surgeons should counsel all patients with otosclerosis and ossicular chain fixation with asymptomatic SSCD regarding the possible risk of surgically unmasking a third window.

Author Contributions

Omid Moshtaghi, analyzed clinical findings, drafting the article and final approval of the version to be published; **Hossein Mahboubi**, analyzed clinical findings, drafting the article and final approval of the version to be published; **Hamid R. Djalilian**, analyzed clinical findings, drafting the article and final approval of the version to be published; **Harrison W. Lin**, treating physician, analyzed clinical findings, drafting the article and final approval of the version to be published.

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