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Superior semicircular canal dehiscence in the only hearing ear with no symptom: a case presentation

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ABSTRACT

We herein present a case of Superior Semicircular Canal Dehiscence (SSCD) syndrome in the only hearing ear. The audiogram of the patient showed mild mixed-type hearing loss in the left ear and total hearing loss in the right ear. Although SSCD syndrome has many symptoms, the reported patient had no complaints as a sign of Semicircular Canal Dehiscence (SCD) in the left ear in daily life. There has been a suspicion for SCD occurred during audiological evaluation. The superior SCD was identified in High-Resolution Computed Tomography (HRCT) scanning. The patient, for whom a conservative approach with a hearing aid was recommended, did not attend follow-up visits.

Keywords: semicircular canal dehiscence, hearing loss, air-bone gap, third window, autophony

ÖZ

İşiten tek kulakta asemptomatik süperior semisirküler kanal dehissansı: olgu sunumu

Bu raporda, işiten tek kulakta Superior Semisirküler Kanal Dehissansı (SSKD) sendromu olgusu sunulmaktadır. Hastanın odyogramında sol kulakta hafif derecede mikst tip, sağ kulakta total işitme kaybı görülmüştür. Superior semisirküler kanal dehissansı sendromunun birçok semptomu olmasına rağmen, bildirilen hastanın günlük yaşamda sol kulağında Semisirküler Kanal Dehissans (SKD) belirtisi olarak herhangi bir şikâyeti olmamıştır. Odyolojik değerlendirme sırasında SKD şüphesi oluşmuştur. Süperior SKD tanısı Yüksek Çözünürlüklü Bilgisayarlı Tomografi (YÇBT) ile konulmuştur. İşitme cihazı ile koruyucu yaklaşım önerilen hasta, takip ziyaretlerine gelmemiştir.

Anahtar kelimeler: semisirküler kanal dehissansı, işitme kaybı, hava kemik aralığı, üçüncü pencere etkisi, otofoni

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INTRODUCTION

Superior Semicircular Canal Dehiscence (SSCD) is a defective opening on the bony covering of the superior canals of the inner ear (Minor, Solomon, Zinreich, & Zee, 1998). The etiology of Semicircular Canal Dehiscence SCD is not understood, however, congenital and acquired types may exist. It's possible that SCD will result in the failure of prenatal and postnatal development of the temporal bone. Some of the potential causes of acquired SCD include intracranial hypertension, neoplasms, vascular malformations, chronic osteomyelitis, fibrous dysplasia, and head trauma with fractures to the temporal bone (Eberhard, Chari, Nakajima, Klokker, Cayé-Thomasen, & Lee, 2021). Superior semicircular canal dehiscence has many symptoms seen as abnormal auditory or vestibular signs. The common symptoms of SSCD are hearing loss, autophony, hyperacusis, vertigo, and ossilopsia (Minor, 2005; Minor et al., 1998). The audiogram frequently shows air-bone gap, especially in low-frequency regions and an intact acoustic reflex response (Merchant & Rosowski, 2008; Merchant, Rosowski, & McKenna, 2007). The third window hypothesis can help to understand air-bone gap in low-frequency regions which is also seen in Conductive Hearing Loss (CHL). Vertigo can be brought on by the third mobile window because it causes the auditory stimuli that are present at the oval window to spread out across the vestibular labyrinth (Merchant et al., 2007). An air-bone gap develops as a result of the SCD's introduction of a third window into the inner ear, which also results in a changed inner ear volume velocity. Vestibular symptoms are quite common in SCD, and they can be brought on by loud sounds (Tullio Phenomenon), by changes in the pressure of the external canal that is conveyed to the middle ear (Hennebert sign), or by performing the Valsalva maneuver (Minor et al., 1998).

Diagnostic criteria for SSCD include clinical findings that are unique to SSCD, high-resolution temporal bone computed tomography that demonstrates dehiscence, and supportive evidence such as pure tone audiometry and Vestibular-Evoked

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Myogenic Potentials (VEMPs) (Eberhard et al., 2021; Ward, Carey, & Minor, 2017). Established clinical testing that indicates supranormal bone conduction thresholds, low-frequency air-bone gap with present acoustic reflexes, low threshold cervical VEMP, and elevated ocular VEMP amplitudes are beneficial in directing therapeutic options for symptomatic patients who have radiologic SSCD. Even while conservative treatment is an acceptable option for the majority of patients with SSCD, there is a possibility that certain cases will suffer difficulty with communication and activities of daily life (Remenschneider, Owoc, Kozin, McKenna, Lee, & Jung, 2015). Surgical repair of dehiscence is the only treatment that has been shown to be beneficial for patients with disabling symptoms. The influence of the third window is to be minimized through surgical intervention. Current therapeutic options for dehiscence plugging include the middle cranial fossa technique and the transmastoid route (Banakis Hartl & Cass, 2018; Schwartz, Almosnino, Noonan, Banakis Hartl, Zeitler, Saunders, & Cass, 2019). It may be challenging to diagnose SSCD in certain patients because the condition may not manifest any symptoms at all. If SSCD is not diagnosed in time, many of these patients will need to have surgery on their middle ears, which may not be beneficial or may even be harmful.

CASE REPORT

We are going to describe the medical history of a 50-year-old male patient whose only symptom was right ear hearing loss since childhood. He sometimes feels fullness in his left ear when having a cold. He had no complaints of hyperacusis, autophony, vertigo, and tinnitus.

On physical examination his external ear canals and eardrums were normal. 512 and 1024 cps Rinne test was positive on the left side, while no response was achieved on the right side. The Weber fork exam was lateralized to the left ear. When asked to perform the Valsalva maneuver, he had no complaints of dizziness or ossilopsia. Eustachian tube function was normal on otoscopic evaluation of the tympanic membrane on both sides.

An audiometric assessment showed mild mixed-type hearing loss in the left ear and total hearing loss in the right ear (Figure 1). The right acoustic reflexes were absent while the acoustic reflexes were seen on high-intensity levels on the left ear (Table 1). The Eustachian tube was patent on the left ear and not patent on the right ear. The Speech Reception Threshold (SRT) in the left ear was consistent with Pure Tone Average (Gupta, Eavey, Wang, Curhan, & Curhan, 2019) level (Table 2). The patient had felt discomfort and dizziness while measuring the right ear's speech awareness/reception levels due to contralateral masking although it was on the appropriate level. The patient's feeling of dizzy with the sound stimulus indicated that Tulio Phenomenon was positive in the right ear, and it was decided to perform the Fistula Test based on this phenomenon seen during the speech test. There was no sign of nystagmus and vertigo (Hennebert Sign) in both ears, so the Fistula test was negative bilaterally. Due to the lack of equipment in the clinic, VEMP, which was suggested to strengthen the clinical diagnosis, could not be applied.

Table 1. Immitancemetry Measurements

	Right Ear	Left Ear
Middle Ear Pressure (daPa)	-120 dapa	-170 dapa
Compliance	0,8	0,3
Volume (ml)	1,2	1,4
Ipsilateral AR	(-)	(+)
Contralateral AR	(-)	(+)

AR: Acoustic Reflex

Table 2. Pure Tone Measurements and Speech Tests

	Right Ear	Left Ear
Pure Tone Average	NA	28 dB
Speech Reception Threshold (dB)	NA (M) (SAT)	30 dB
Most Comfortable Level	NA (M)	70 dB
Speech Discrimination (%)	%	% 96
Uncomfortable Level (dB)	**100+**	**100+**

M: Masked, NA: Not Achieved, SAT: Speech Awareness Threshold
*Right Bone SRT= 20 dB, Left Bone SAT= NA

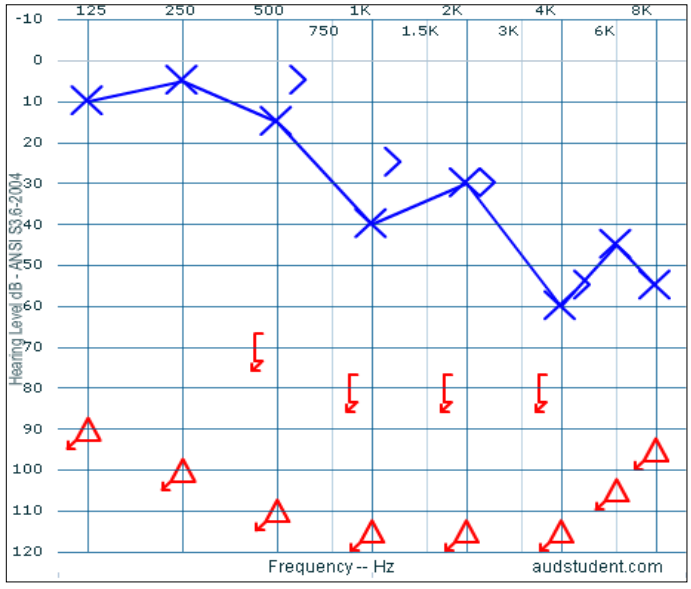

Figure 1. Pure Tone Audiogram: demonstrating mild mixed type hearing loss in left ear and total hearing loss in right ear.

We performed High-Resolution Computed Tomography (HRCT) and Magnetic Resonance Imaging (MRI) for the patient. High-resolution computed tomography indicated SSCD in the left ear (Figure 2) and MRI showed vestibulocochlear nerve aplasia in the right ear. As a rehabilitation approach, it was recommended to try a conventional air conduction hearing aid in the left ear or to use the contralateral routing of the signal (McMinn, Wiens, & Crossen, 1988) hearing aid system as another option. A follow-up visit was asked based on a conservative approach.

Since the patient had nerve aplasia in the right ear, it was thought that amplification would not be beneficial in this ear. However,

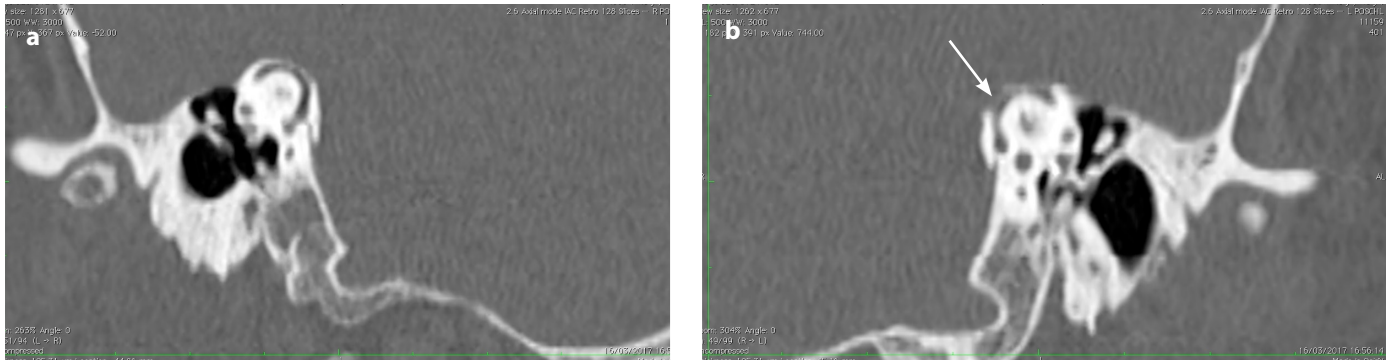

Figure 2. High-resolution computed tomography images of right (a) and left ear (b). Arrow points to dehiscence.

it is possible that the disadvantages of single-sided hearing loss could be reduced by passing the auditory information from the right ear to the left ear. It was determined that there was some degree of hearing loss in the left ear, therefore it would be appropriate to amplify the sound here as well. It was thought that the signals sent from the transmitter in the right ear to the hearing aid in the left ear by using bilateral cross-hearing aid would increase hearing performance. The patient, who was diagnosed with SSCD despite being asymptomatic, was given recommendations consistent with these considerations, but the patient did not return to the clinic for follow-ups. Therefore, we were unable to form an opinion regarding the patient's progression.

DISCUSSION

Since SSCD was first described in 1998, there have been many SSCD cases presented with vestibular and auditory signs. The patient we had presented here, had no vestibular or auditory symptoms in daily life. During our clinical evaluation, intolerance of masking stimulus in the left ear raised suspicion for SSCD and we diagnosed SSCD in the only hearing ear via HRCT.

In the audiometric evaluation of SSCD patients, bone conduction thresholds would be less than 0 dB normal hearing level and the audiogram frequently shows air-bone gap, especially in low-frequency regions like in CHL (Merchant et al., 2008; Minor, 2005). The pathophysiology of CHL can be explained by the third window hypothesis. The dehiscence on the canal would be accepted as a third window opening into the inner ear. This window causes the impedance between scala tympani and scala vestibuli to be increased and hence bone conduction thresholds improve. On the other hand, air conduction thresholds worsen due to changing way of acoustic energy reaches to the cochlea. These mechanisms creates air-bone gap and hearing loss occurs (Merchant et al., 2007).

The patient's audiogram showed air-bone gap which was 10 dB at 0.5 kHz, 15 dB at 1.0 kHz, and 5 dB at 4.00 kHz but none of the assessed bone conduction hearing levels was less than 0 dB. The patient had no complaints of autophony and hypersensitivity to bone-conducted sounds due to the third window effect.

Superior semicircular canal dehiscence is thought to be mimicking otosclerosis**.** In differential diagnosis, the Acoustic Reflexes (AR) are absent in otosclerosis whereas they are seen in SSCD. In addition to intact AR, the cervical Vestibular Evoked Myogenic Potential (VEMP) thresholds are decreased in SCD patients (Noij & Rauch, 2020). Otosclerosis is not associated with decreased VEMP responses, vertigo, and computed tomography findings of SCD (Minor et al., 1998). In this case, no signs of otosclerosis were present. Vestibular evoked myogenic potential could not be applied to this case for differential diagnosis since there was no test device in the clinic. This can be considered a limitation of this report.

The initial symptoms creating suspicion for SCD are generally sound or pressure-induced vertigo and ossilopsia. It is known as Tulio Phenomenon when loud sounds cause eye movements and it is known Hennebert Sign when changing in the pressure of the external ear causes eye movements (Minor, 2005). Frenzel lenses or infrared video goggles may help to examine the eye movements in response to loud sound and increased external ear pressure or Valsalva maneuvers, in this way it would be possible to predict the related defective semicircular canal (Minor et al., 1998).

Although rarely seen, the posterior and lateral semicircular canals would have defective openings and may show similar (Belden, Weg, Minor, & Zinreich, 2003; Williamson, Vrabec, Coker, & Sandlin, 2003) dehiscence. For example, high-riding jugular bulbs and fibrous dysplasia may be found together with Posterior Semicircular Canal Dehiscence (PSCD). The presence of chronic otitis media with cholesteatoma may cause Lateral Semicircular Canal Dehiscence (LSCD) (Spasic et al., 2015). When cholesteatoma or infection like syphilis destroys the bony labyrinthine, a perilymphathic fistula would exist consequently. Although cholesteatoma and perilymphathic fistula have different causes, their clinical findings may be like SSCDs. For differential diagnosis of perilymphathic fistula, Fistula Test and electrocochleography may help but the verification of diagnosis would be done at the time of surgery (Belden et al., 2003).

In suspicion of SSCD, high-resolution temporal bone computed tomography scans are importantly needed in terms of their specificity and positive predictive value in the diagnosis of SSCD (Belden et al., 2003; Williamson et al., 2003). Although there was not a strong clinical sign for SCD in our case, HRCT is performed, and it indicated dehiscence in the superior semicircular canal. In addition to HRCT, MRI is also performed, and vestibulocochlear nerve aplasia is found in the right ear. We can say it was important to diagnose SSCD in the only hearing ear although the patient was asymptomatic. The patient's everyday activities had not been significantly impacted; thus, it was decided that a conservative therapy strategy would be best. Due to this, amplification was advised along with certain recommendations to prevent the symptoms of dehiscence from becoming worse. The patient was informed about the risks of developing symptoms by trauma or intracranial pressure increase and to take precautions for this possibility. The patient's lack of participation in the follow ups made it impossible to report on the process' progress in this report. This could be considered a limitation; however, the case's informative feature is the discovery of dehiscence in an asymptomatic condition.

CONCLUSION

We report a case of superior semicircular canal dehiscence in the only hearing ear. Although the patient was asymptomatic, suspicion of SCD occurred during performing speech test in the bad ear in audiological evaluation. We recommend being careful to catch the patient's reaction to any sound stimulus as it can provide a clue for SCD.

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