Case Report

Bilateral Superior Semi-Circular Canal Dehiscence Presenting as Conductive Hearing Loss with Vestibular Symptoms: A Client Centre Case Report

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Highlights

- The SSCD is often diagnosed by audiometric and balance tests
- Definitive diagnosis of SSCD is made by CT scan along with clinical symptoms
- Increased amplitude and latency in cVEMP test are the main symptoms of SSCD

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ABSTRACT

Background: Superior semicircular canal dehiscence (SSCD) is a rare bone defect in the petrous part of the temporal bone. In this syndrome, a third window is created between the middle cranial fossa and the bony labyrinth of the inner ear which can lead to stimulation of the vestibular system by sound. Patients usually have symptoms such as vertigo, imbalance, autophony, pulsatile tinnitus, and aural fullness. The clinical symptoms of this disease vary from person to person.

The Case: This study reports a case of a rare bilateral SSCD in a 39-year-old woman with imbalance and autophony problems. The audiological findings showed a bilateral mild conductive hearing loss at low frequencies with symptoms of vertigo and torsional nystagmus. The SSCD was diagnosed by computed tomography. Then, the patient underwent bone repair surgery. Auditory and balance tests were performed again after the surgery to evaluate the extent of recovery.

Conclusion: The SSCD can be diagnosed by using the mentioned test battery. By choosing the appropriate treatment methods and following up of the outcomes, most of the problems of patients with SSCD can be solved.

Keywords: Superior semicircular canal dehiscence; bilateral; conductive hearing loss

Introduction

Superior Semicircular Canal Dehiscence (SSCD) is the formation of a pathological new window between the bony labyrinth of the inner ear and the middle cranial fossa [1, 2]. The most common bony defects in the superior semicircular canal occur on the arcuate eminence of the canal, and rarely in the posterior and lateral canals [3]. In previous studies on temporal bones using high-resolution Computed Tomography (CT), SSCD was observed in 9.6% of patients, 8% of cases were within the superior canal, 1.2% within the posterior canal, and 0.4% within the lateral canal [4-6]. Although the symptoms vary from one patient to another in general, SSCD is detected with high resolution CT scan, audiometry test, and Vestibular Evoked Myogenic Potential (VEMP) test [7-9]. The SSCD was first described by Minor et al. [10]. In their reports, 8 patients were examined with symptoms such as vertigo induced by sound and activities that cause a temporary intracranial pressure (e.g., Valsalva maneuver, cough). These activities can also lead to torsional nystagmus caused by stimulation of the superior semicircular canal [10]. The clinical findings of sound-induced vertigo and the pressure-induced torsional nystagmus associated with SSCD have been termed "Minor's syndrome" [10-12]. Ear fullness, a decrease in bone conduction thresholds, nausea, hyperacusis, and pulsatile tinnitus are other symptoms of SSCD, the amount and type of which are different in patients [10]. Tullio's phenomenon is the presence of disequilibrium and oscillopsia in response to acoustic stimuli at low frequency [13]. Hennebert's sign is defined as eye motion caused by pressure changes on the external auditory canal [14].

Patients with SSCD show a variety of visual, auditory, and balance problems in their fourth and fifth decades of life [4, 12, 15]. Considering that these patients are typically more sensitive to low-intensity stimuli in the VEMP test, this test is often used in the early stages of SSCD [16, 17]. Although audiometric tests are quite effective in diagnosing SSCD, high-resolution temporal CT scan is capable of confirming this disease; however, without audio and vestibular findings, CT scan may have high number of false positive detections. In general, SSCD and specially the bilateral SSCD is very rare. This study reports a case of bilateral SSCD with vestibular and auditory symptoms.

Case presentation

The patient was a 39-year-old woman with severe autophony for approximately 4 years. She frequently claimed that she could hear her footsteps inside her head during walking, which was quite unpleasant to her. Her primary occupation was farming. Due to her living and working conditions, she frequently complained of symptoms such as ear fullness and hearing impairment. She also had acute vertigo when lifting heavy objects. Her symptoms had become more severe in the last 3 months, forced her to visit a specialist. She had not visited a doctor before. In her first visit to an otolaryngologist, audiometry and balance tests and CT scan were prescribed for her. According to the assessments, the patient and her family members had no history of any particular diseases, such as bone abnormalities, metabolic or vascular diseases, or ear infections.

Pure tone audiometry revealed a mild conductive hearing loss in both ears at low frequencies, where the bone conduction threshold was –15 dB (Figure 1). The speech discrimination score in both ears was within the normal range. The otoscopy results were completely normal and the tympanometry test showed a type A (normal) result for both sides and the acoustic middle ear reflexes in both ears were present. The acoustic middle ear reflexes elicited at an intensity level of 85 dB, and no symptoms of imbalance was visible at this level (Tullio's phenomenon) (Figure 2). The auditory and balance symptoms were the same in both ears, although when the patient turned her head to the left, the symptoms and vertigo relatively increased.

For the VEMP test, a two-channel auditory evoked potential device was used. In this method, the non-inverted electrode is placed on the midpoint of the sternocleidomastoid muscle. The response parameters include threshold, latency, and amplitude of P1 and N1, one-way stimulations at intensities of 70, 85, and 90 dB, and the polarity of rarefaction [18]. The results of cervical VEMP (cVEMP) test (as a balance test) were abnormal in both ears. The cVEMP thresholds in the left ear were lower and their amplitude was higher than in the right ear (Figure 3). The Tullio's phenomenon was observed bilaterally at the frequency of 500 Hz and the intensity of 105 dB, where the Valsalva maneuver caused symptoms such as vertigo (Hennebert's sign) and imbalance. Torsional nystagmus was present in both sides; it was not lateralized to one side, since the lesion was bilateral. The results of the high-resolution CT scan showed bilateral SSCD.

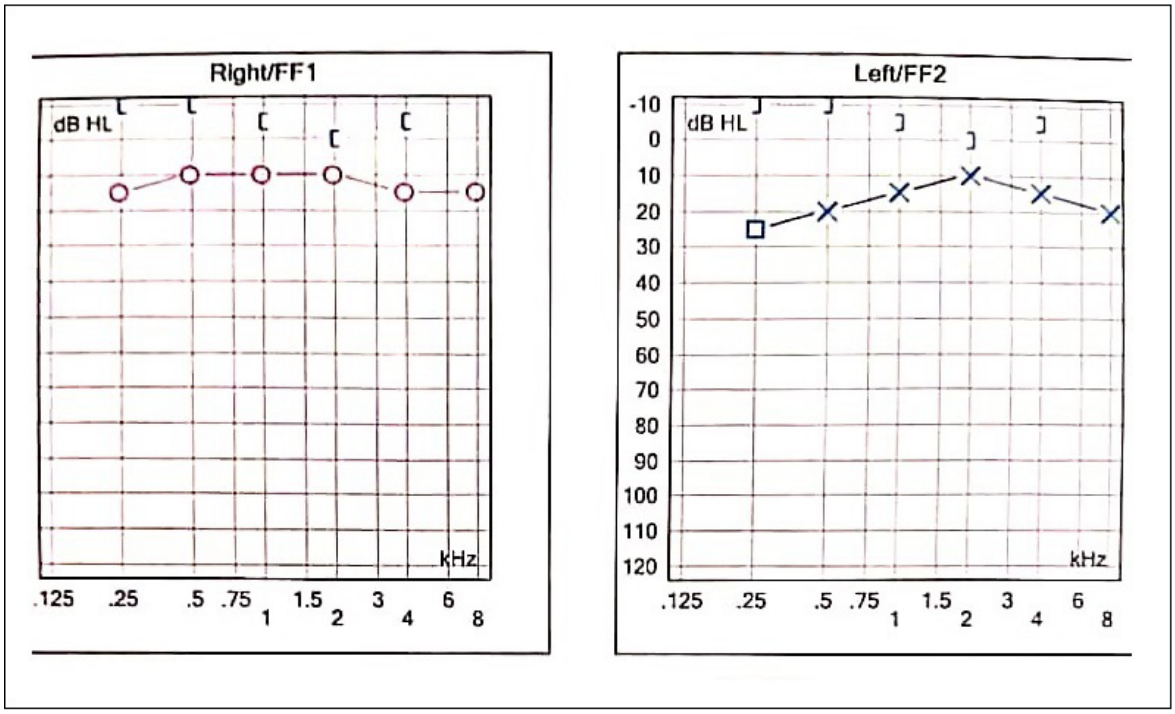


Figure 1. The results of initial audiometry test before the operation (bilateral conductive hearing loss in low frequencies). FF; free field

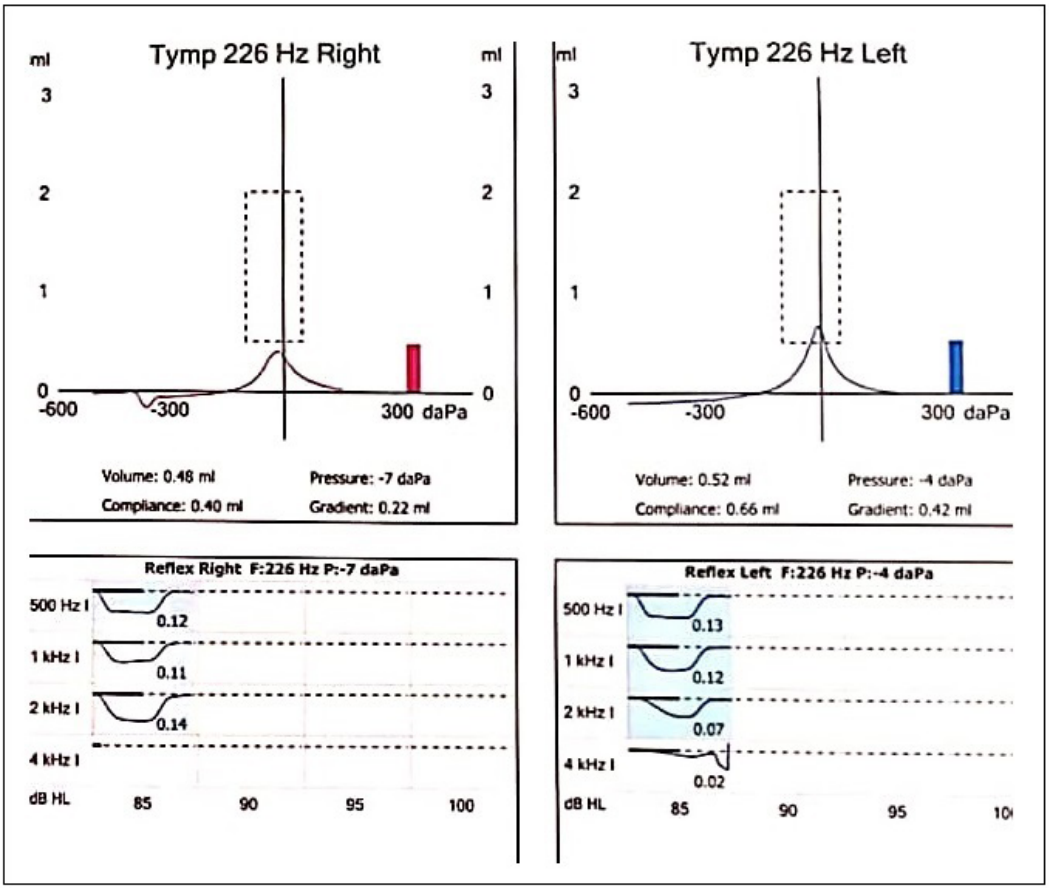
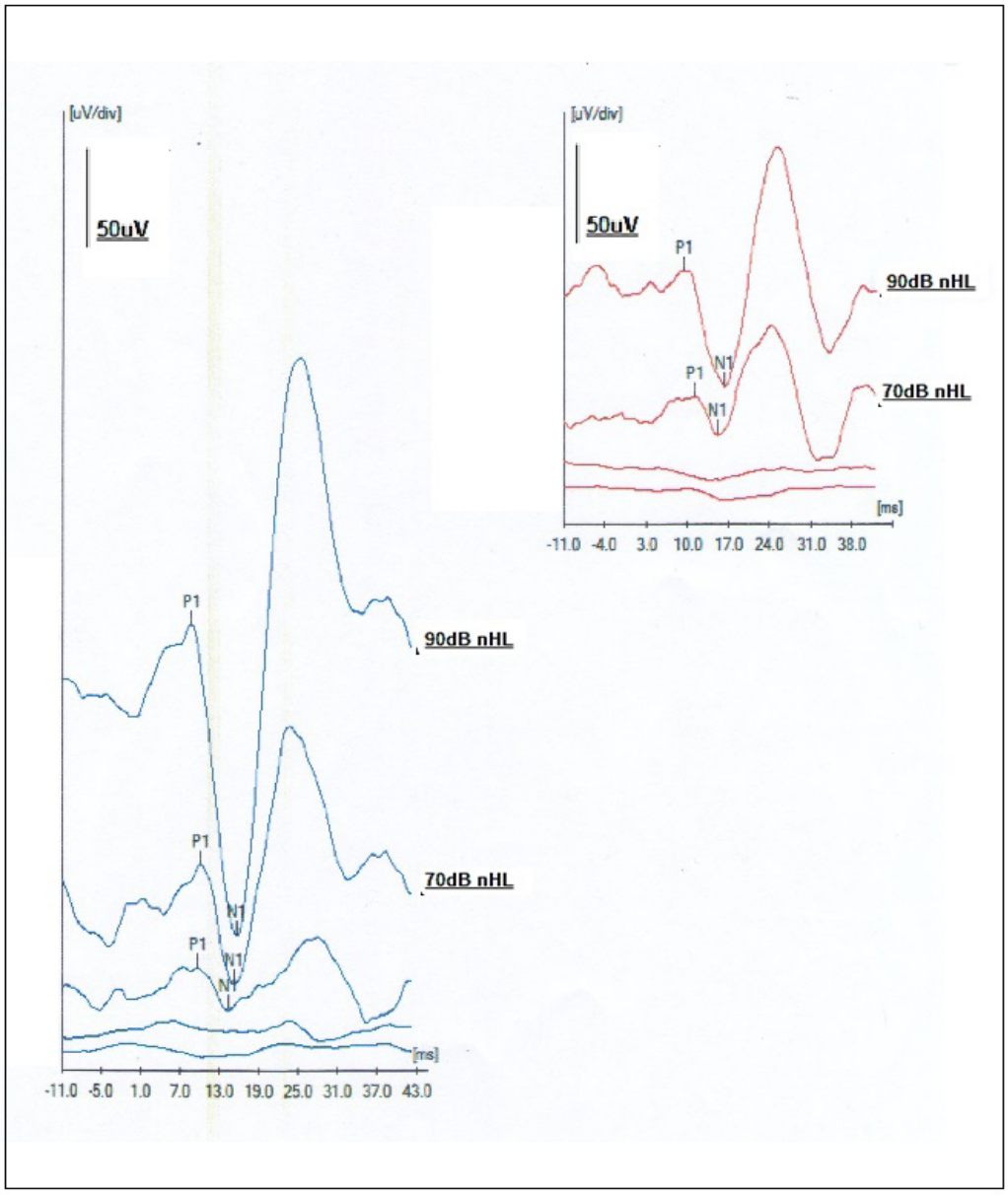


Figure 2. The results of tympanometry test and middle ear reflexes in both ears. (An type in tympanometry test, and normal reflex thresholds in all frequencies)



Latencies (ms)

Label Index	SmartTag	P1	N1
A1	L VEMP	8.67	15.54
A2	L VEMP	10.12	15.12
B1	R VEMP	9.40	16.37
B2	R VEMP	11.17	15.12

Interlatencies (ms)

Label Index	SmartTag	P1-N1
A1	L VEMP	-6.87
A2	L VEMP	-5.00
B1	R VEMP	-6.97
B2	R VEMP	-3.96

Interamplitudes (uV)

Label Index	SmartTag	P1-N1
A1	L VEMP	184.490
A2	L VEMP	70.840
B1	R VEMP	75.260
B2	R VEMP	25.090

Figure 3. The results of initial cervical vestibular evoked myogenic potential test in both ears before the surgery (x axis: time (ms), y axis: amplitude (μv))

Based on symptoms and test results, first the left ear underwent bone repair surgery followed by the repair surgery in the right ear. The patient was in a supine position with her head inclined approximately 35 degrees to the left during the operation. The surgical procedure was craniotomy using the Keyhole infratemporal approach. Before the surgery, a CT scan was performed by employing the stereotactic head frame and the images were reconstructed to coordinate and identify the nerve pathways. In addition, a number of components including cartilage and fascia were used to fill in the pathology holes. The patient returned to the clinic two months after the left ear surgery to repeat the hearing and balance diagnostic tests. The audiometry results were completely

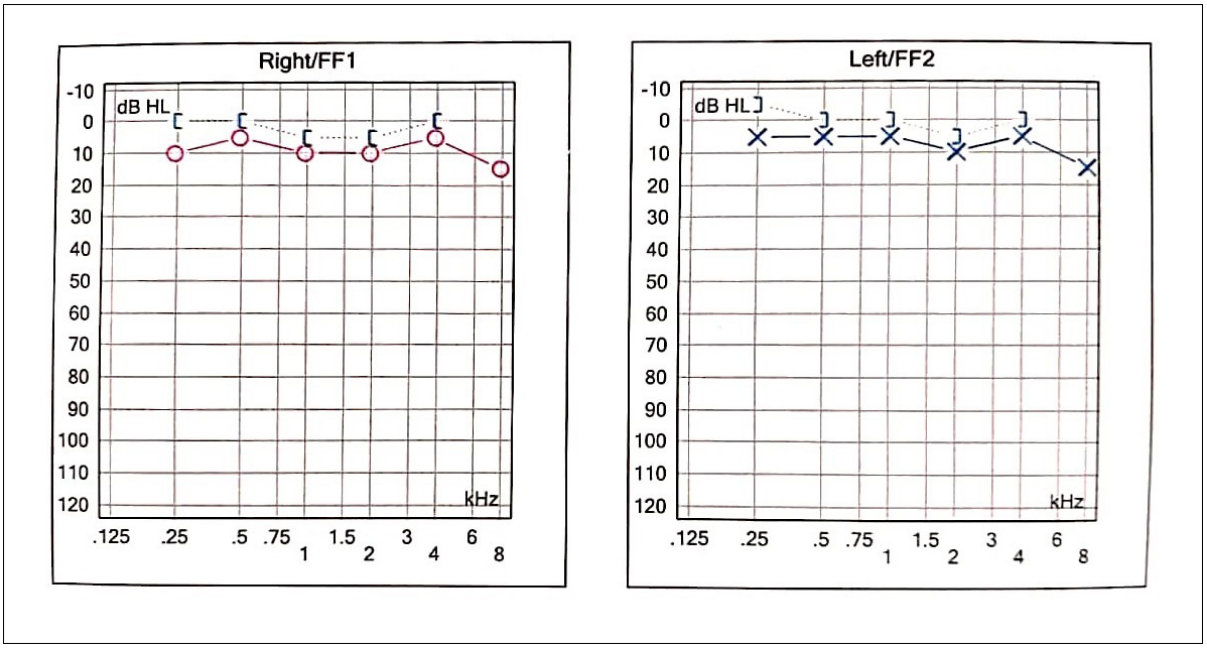
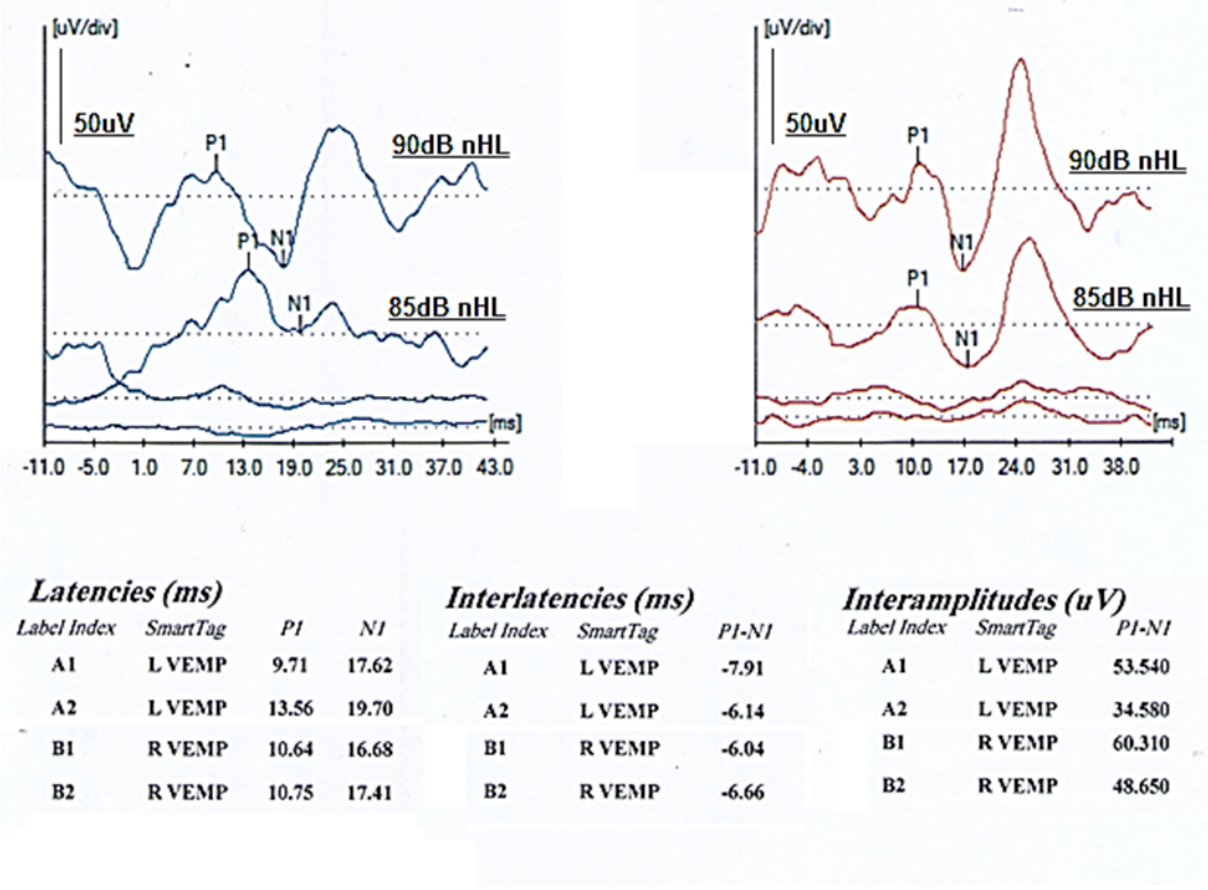


Figure 4. The results of audiometry test after surgery (elimination of the gap between air conduction and bone conduction). FF; free field



Latencies (ms)

Label Index	*SmartTag*	*P1*	*N1*
A1	L VEMP	9.71	17.62
A2	L VEMP	13.56	19.70
B1	R VEMP	10.64	16.68
B2	R VEMP	10.75	17.41

Interlatencies (ms)

Label Index	*SmartTag*	*P1-N1*
A1	L VEMP	-7.91
A2	L VEMP	-6.14
B1	R VEMP	-6.04
B2	R VEMP	-6.66

Interamplitudes (uV)

Label Index	*SmartTag*	*P1-N1*
A1	L VEMP	53.540
A2	L VEMP	34.580
B1	R VEMP	60.310
B2	R VEMP	48.650

Figure 5. The results of cervical vestibular evoked myogenic potential test after surgery (normal amplitude and latency in both ears) (x axis: time (ms), y axis: amplitude (μv))

normal and the gaps between air conduction and bone conduction were completely closed. The VEMP test showed that the symptoms mostly disappeared and the cervical VEMP threshold and amplitude in the left ear returned to normal levels. Two months after the right ear surgery, the audiometry results were within the normal range and the gaps were eliminated. The VEMP test also showed that the patient's responses were within the normal range, and the cervical VEMP thresholds and amplitudes returned to the normal levels (Figures 4, 5). Despite these improvements, the patient still complained of poor balance; therefore, vestibular tests such as caloric test and videonystagmography were conducted. All the test results were normal, but the results of the video head impulse test were abnormal for both ears, which indicated weakness in the superior semicircular canals. She was advised for vestibular rehabilitation, but due to her living conditions and financial difficulties, she rejected the recommendations.

Discussion

In patients with SSCD syndrome, both conductive and sensorineural hearing loss are observed, although conductive hearing loss is more likely due to the presence of a third window in the superior semicircular canal which causes acoustic energy to leak from through the third window. Many assumptions have been proposed for the acquired formation of SSCD, including trauma, increased intracranial pressure, or osteopenia [16, 17]. In children, the existence of SSCD can be related to congenital issues that lead to the thinning of the temporal bone and the subsequent head trauma. In our study, the patient had no history of trauma and it was less likely that bone defect was acquired. In such cases, there may be congenital abnormalities or mild bone problems that lead to the emergence and aggravation of symptoms over time. To our knowledge, there is scant reports of the bilateral SSCD. In most of the previous reports, the diagnostic and treatment aspects have been discussed from the medical point of view, but the present case report study, focused on the symptoms that caused the patient's chief complaints, in addition to the medical aspects. Before bone repair surgery, the patient had vertigo and relatively severe imbalance as well as many problems in performing her daily activities. After the surgical intervention, all the auditory and balance problems disappeared; there was only a slight impairment in her balance. According to the patient, her physical conditions significantly improved in comparison with the preoperative conditions.

Conclusion

The superior semicircular canal dehiscence can be diagnosed by using the vestibular evoked myogenic potential and audiometry test with CT scan. By choosing the appropriate treatment methods and following up of the outcomes, most of the problems of patients with superior semicircular canal dehiscence can be solved.

Ethical Considerations

Compliance with ethical guidelines

The ethical clearance was obtained from the research Ethics Committee of Nianin Neurorehabilitation Research Complex (Code: UN. NNRC.2023.7).

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Authors' contributions

MRA: Study design, interpretation of the results, and drafting the manuscript; HB: Study design, acquisition of data, and drafting the manuscript; SK: Acquisition of data and drafting the manuscript; AK: Study design, interpretation of the results and drafting the manuscript. All authors approved the final version of the manuscript.

Conflict of interest

The author does not have any financial or other interests relating to the study.

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