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Abstract

Introduction: Superior semicircular canal dehiscence syndrome (SSCDS) is a rare medical condition of the inner ear that was initially described in 1998 by Minor and al. It is defined as a structural bony defect of the roof of the superior semi-circular canal and is responsible for the creation of a third window, which alters the dynamics of the inner ear.

Objective : The purpose of this study is to describe the clinical and paraclinical findings of symptomatic SSCDS.

Patients and methods : A case series of 3 patients with SSCDS, collected at the ENT department of Mongi Slim hospital over a period of 2 years .

Case presentation :

Patient 1 : A 63-year-old female with a history of diabetes, hypothyroidism, sarcoidosis consulted for left tinnitus and hypoacusis with rotatory vertigo evolving for 1 year. Physical examination was normal. Audiometry showed a low-frequency conductive hearing loss at 40dB on the left side. Tympanometry showed on the left ear a type B curve. Stapedial reflexes were present. A high density temporal bone CT Scan showed a left dehiscence of superior semicircular canal.

Patient 2 : A 49-year-old female with no medical history consulted for right tinnitus and hypoacusis with rotatory vertigo evolving for 2 years. Physical examination was unremarkable. Audiometry showed a left conductive hearing loss at 30dB in the low frequencies. Tympanometry showed on the left a dome-shaped curve. Stapedial reflexes were present. Temporal bone CT Scan was requested which confirmed the presence of a bilateral dehiscence of superior semicircular canal.

Patient 3 : A 66 year-old man with no history of otological disease, presented with months of progressively hearing loss in the right ear associated with tinnitus and rotatory vertigo. The patient claimed that the vertigo was usually triggered by loud noises and lasted for few minutes. The physical examination revealed a normal aspect of the tympanic membrane, the Weber test was lateralized to the right ear. The audiogram showed in the right ear a mixed hearing loss of 60dB with a Gap of 20dB. The impedancemetry showed a type A tympanometry and a present acoustic stapedial reflex. A high-resolution CT scan of the temporal bones objectified a dehiscence of the right superior semicircular canal.

All 3 patients received medical treatment based on low salt diet and a vestibular suppressant (Betahistine) that relieved most of the symptoms.

Conclusion: SSCDS is a recently described medical condition giving rise to vestibular and auditory symptoms. This case series showed that subjective, clinical and paraclinical presentation of SSCDS can widely vary from a patient to another, making the diagnosis more challenging.

Results

	Patient 1	Patient 2	Patient 3
Age	63 yo	49 yo	66 yo
Gender	female	female	male
Medical history	Diabetes, sarcoidosis, hypothyroidism	-	-
Symptoms	Left tinnitus and hypoacusis, rotatory vertigo Evolving for a year	right tinnitus and hypoacusis with rotatory vertigo evolving for 2 years	Right hypoacusis and tinnitus and rotatory vertigoevolving for months
Physical examination	Normal	Normal	Normal
Audiometry	low-frequency conductive hearing loss at 40dB on the left ear	left conductive hearing loss at 30dB in the low frequencies	mixed hearing loss of 60dB with a Gap of 20dB (Figure 1)
Stapedial reflex	present	present	present
VNG + VHIT	Bilateral vestibulopathy	-	-
Temporal bone CT SCAN	left superior semicircular canal dehiscence(SSCD) (Figure 2)	Bilateral SSCD (Figure 3)	Right SSCD (Figure 4)

- All 3 patients received a medical treatment based on low salt diet and a vestibular suppressant (Betahistine) that relieved most of the symptoms.

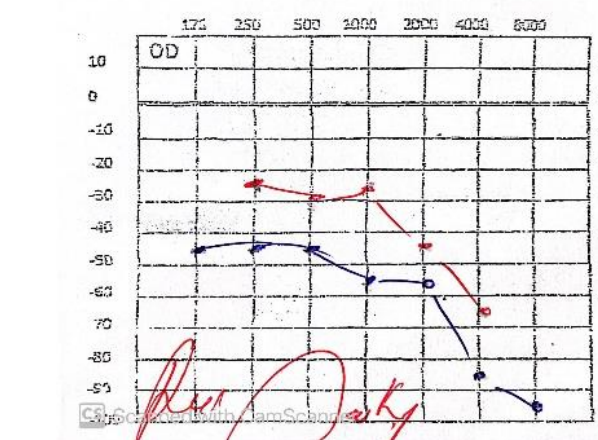


Fig.1 audiogram showing a mixed hearing loss of 60dB with a Gap of 20dB and a dip to 90dB at 4000Hz (patient3)



Fig.2 CT Scan of the temporal bone showing dehiscence of the apex of the left superior semicircular canal (patient1)



Fig.3 Coronal CT Scan of the temporal bone showing dehiscence of the right superior semicircular canal (patient 2)

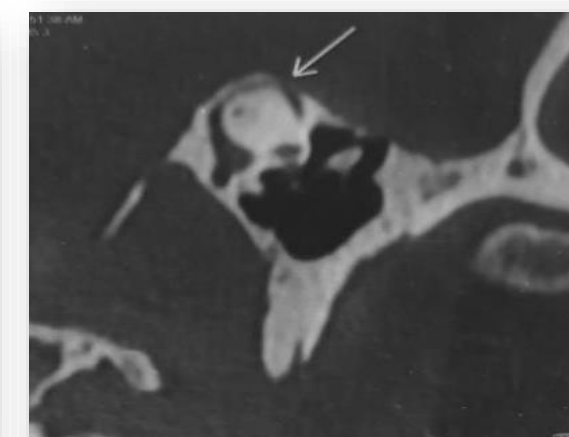


Fig.4 Temporal bone CT Scan showing a dehiscence of the right superior semicircular canal (patient3)

Objectives

- The purpose of this study is to describe the clinical and paraclinical findings of symptomatic SSCDS

Conclusion

- SSCDS is a recently described medical condition giving rise to vestibular and auditory symptoms. In our case series, the hearing loss was the main reason encouraging patients to consult in addition to the vertigo and tinnitus.
- Patients with intact acoustic reflex responses and an air-bone gap on audiometry should undergo further investigation in order to detect superior canal dehiscence.
- This case series showed that subjective, clinical and paraclinical presentation of SSCDS can widely vary from a patient to another, making the diagnosis more challenging.

Materials and methods

- We present a case series of 3 patients with SSCDS, collected at the ENT department of Mongi Slim hospital over a period of 2 years .

References

- 1.Minor LB. Clinical Manifestations of Superior Semicircular Canal Dehiscence: The Laryngoscope. oct 2005;115(10):1717-27.
- 2.Palma Diaz M, Cisneros Lesser J, Vega Alarcón A. Superior Semicircular Canal Dehiscence Syndrome – Diagnosis and Surgical Management. Int Arch Otorhinolaryngol. avr 2017;21(02):195-8.
- 3.Kontorinis G, Lenarz T. Superior semicircular canal dehiscence: a narrative review. J Laryngol Otol. avr 2022;136(4):284-92.