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Case Report

Minor's syndrome: Dehiscence of the superior semicircular canal. A case report ☆

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ABSTRACT

Superior semicircular canal (SSC) dehiscence syndrome, also known as Minor syndrome, is a rare condition characterized by vestibular and cochlear symptoms linked to a defect in the bony roof of the SSC. The prevalence is estimated at 0.5%, with a male predominance. Dehiscence may result from abnormal bone development, becoming symptomatic due to minimal trauma or pressure changes. Clinical presentation varies based on dehiscence size and location, with dizziness and oscillatory movements triggered by pressure changes or loud sounds being common symptoms. Other manifestations include conductive hearing loss due to the formation of a "third window" in the inner ear. Diagnosis typically involves computed tomography, distinguishing SSCDS from otosclerosis. Surgical treatment is reserved for cases of disabling vestibular pathology, often involving sealing the dehiscence through various approaches

Introduction

Superior semicircular canal dehiscence syndrome, first described in 1998 by Lloyd Minor [1], is a rare entity with an estimated prevalence of 0.5%. It designates a set of vestibular and cochlear clinical symptoms associated with the presence of a dehiscence of the superior semicircular canal (SSC); defect in the coverage of the roof of its bony wall. This bony defect creates a "third window" in the inner ear, dispersing the acoustic energy of sound waves, causing abnormal endolymph mobility, and lowering the bone conduction threshold [2]. These pathophysiological consequences manifest themselves clinically as vertigo, autophony, or even hearing loss, which poses a problem of differential diagnosis with otospongiosis [3–5]. CT scans are used to diagnose this syndrome, the exact etiopathogenesis of which remains unknown [1–5].

The principle of surgical treatment of this condition is to plug the dehiscence [6]. It is indicated when clinical symptoms are severe and impair the patient's quality of life [7].

We report a case of a young patient who presented with vertigo and left hearing loss in whom a CT scan of the rocks found a dehisence of the superior semicircular canal, describing the clinical and radiological features of Minor's syndrome.

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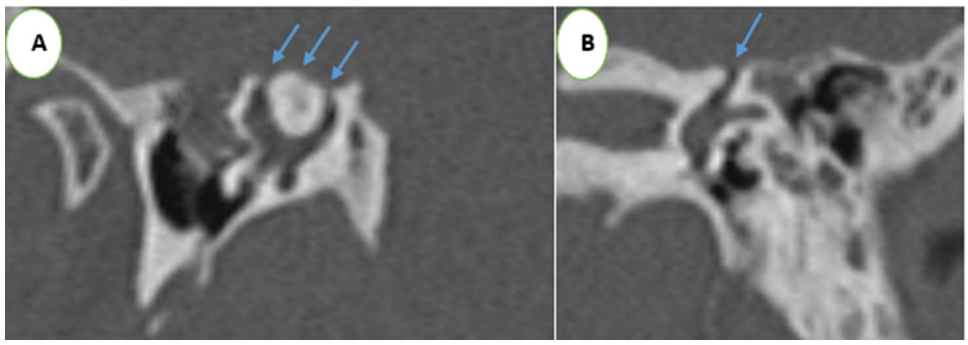

Fig. 1 – CT scan in Pöschl plane (A) and coronal section (B) of the left superior semicircular canal showing a dehiscence in the form of a defect in coverage of the roof of its bony wall *(blue arrow)*.

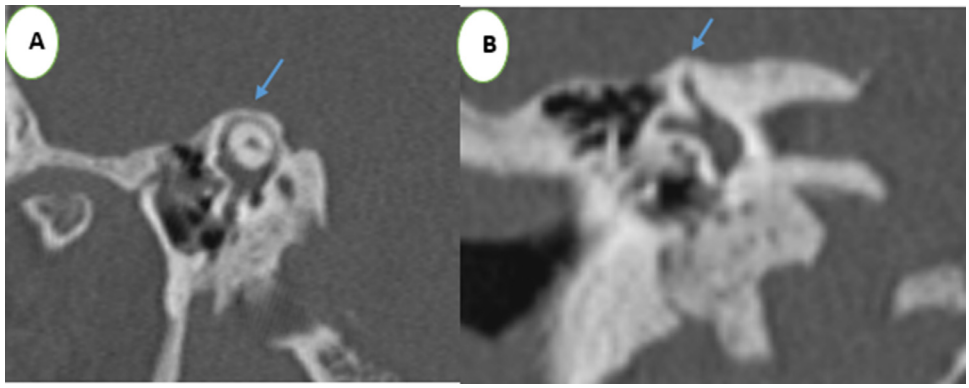

Fig. 2 – CT scan in Pöschl plane (A) and coronal section (B) showing a normal-appearing right semicircular canal in the same patient with respect for the roof of its bone wall *(blue arrow)*.

Case report

A 35-year-old patient had been complaining for a year of vertigo triggered by sudden head movements, coughing, and sneezing. There was no history of head trauma. The patient also complained of a sensation of a "full'' left ear, with a loss of left hearing. On examination, the Valsalva maneuver revealed horizontal rotatory nystagmus, and otoscopic examination revealed no tympanic abnormality. The audiogram revealed a left unilateral conductive hearing loss at frequencies below 1000 Hz and a Carhart notch at 3000 Hz. The stapedial reflex test could not be performed because it triggered vertigo. The CT scan showed a 3 mm dehiscence of the left SSC (Fig. 1). Contralateral side was normal (Fig. 2). The dehiscence was surgically plugged, and the postoperative course was marked by a reduction in vertigo and closure of the air-bone space on audiogram.

Discussion

Superior semicircular canal (SSC) dehiscence syndrome is a rare condition first described in 1998 by Lloyd Minor [1], hence the name Minor syndrome. According to an autoscopic study, its prevalence is estimated at 0.5%, with a clear male predominance [8]. This syndrome designates a set of vestibular and cochlear clinical symptoms associated with the presence of a roof covering defect in the bony wall of the SSC. SSC dehiscence may be secondary to abnormal bone development in the first few weeks of life, becoming symptomatic following a triggering event, either minimal trauma or pressure variation [9].

SSC dehiscence may be unilateral or rarely bilateral. The rate of bilaterality varies from series to series, ranging from 23.5% to 37.5% [10]. Their topography is variable: the posterior or the anterior part of the SSC, or the top of the SSC [6,7]. In our patient, the dehiscence was unilateral and located at the apex of the superior semicircular canal.

Clinical symptomatology varies according to the size and topography of the dehiscence [11,12]. Dizziness and oscillatory movements triggered by pressure changes or loud sounds are the most characteristic symptoms [2,13,14]. Other clinical manifestations of the syndrome include conductive hearing loss, due to the formation of a "third window'' in the inner ear. This causes sound energy to dissipate, raising the threshold for air conduction, while simultaneously lowering the threshold for bone conduction by amplifying the impedance difference between the round and oval window [14].

During the clinical examination, it is important to look for torsional vertical nystagmus triggered by pressure variations, such as the Valsalva maneuver, closed-glot effort, or tympanometry inducing a pressure variation in the external auditory canal. It is also necessary to check for the presence of this

nystagmus after exposure to a sound with an intensity of 100 to 110 decibels and a frequency between 500 and 2000 Hertz. Stapedial reflexes are generally present.

Computed tomography plays a central role in the management of patients with SSCDS. It correlates clinical symptoms with an anatomical lesion by identifying a dehiscence in the roof of the SSC. Otosclerosis represents the main differential diagnosis of this condition [1,3,4], sharing similar clinical features. In both cases, audiograms reveal conductive hearing loss, but imaging allows us to distinguish between them [4,5]. In addition, CT scanning enables us to measure the extent of the exposed area, a criterion for operability, with dehiscences of more than 3 mm considered operable [6], as observed in our case. Postoperatively, CT is useful for detecting possible complications such as fistula or pneumolabyrinth.

MRI is not generally recommended for diagnosis and is often interpreted as normal [13]. Surgical treatment is reserved for cases of disabling vestibular pathology, as in our patient's case. Surgery consists in sealing the dehiscence by performing an obturation from a middle cerebral fossa approach, since this is the only way to confirm dehiscence. Several authors have also proposed a transmastoid approach [15,16].

Conclusion

The presence of conductive or mixed hearing loss with a normal eardrum should raise suspicion of superior semicircular canal dehiscence. Although vertigo triggered by loud sounds or pressure variations is the most characteristic symptom, the diagnosis is confirmed by high-resolution CT scans of the rock with coronal, axial, and axial reconstructions of the superior semicircular canal. Surgical treatment is recommended only in cases of incapacitating vertigo.

Patient consent

Informed consent for publication was obtained from patient.

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