Case Report

Multiple dehiscences of bony labyrinthine capsule. A rare case report and review of the literature

Deiscenze multiple della capsula ossea del labirinto. Descrizione di un caso raro e revisione della letteratura

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SUMMARY

Multiple semicircular canal dehiscences are clinical entities characterised by vestibular and cochlear symptoms induced by enhanced sensitivity of labyrinthine receptors due to a multiple bone defect of the otic capsule. The case is presented of a 38-year-old male with bilateral posterior semicircular canal dehiscence associated with unilateral (right) superior semicircular canal dehiscence. The man suffered from vestibular (recurrent Tullio Phenomenon or sound-induced vertigo) and cochlear symptoms (persistent aural fullness associated with mixed hearing loss and disabling tinnitus).

KEY WORDS: Vertigo • Superior and posterior semicircular canal dehiscence • Tullio Phenomenon • CT scans

RIASSUNTO

Le deiscenze multiple dei canali semicircolari sono entità cliniche recenti, caratterizzate da un corteo clinico e sintomatologico della sfera vestibolare e di quella cocleare. I sintomi in special modo sono provocati dall'aumentata sensibilità dei recettori labirintici dovuta proprio alla molteplicità dei difetti ossei della capsula labirintica. Si presenta il caso di un uomo di 38 anni con deiscenza bilaterale del canale semicircolare posteriore associata ad una deiscenza del canale semicircolare superiore destro. Il soggetto lamentava sintomi tanto della sfera vestibolare (fenomeno di Tullio ricorrente altrimenti definito come la vertigine indotta dall'esposizione al suono) che della sfera cocleare (fullness auricolare persistente con associata ipoacusia di tipo misto ed acufene disabilitante).

PAROLE CHIAVE: *Vertigine • Deiscenza del canale semicircolare superiore e posteriore • Fenomeno di Tullio • TC ad alta risoluzione*

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Introduction

In 1998, Lloyd B. Minor et al. [1] described, for the first time, a new clinical entity characterised by the onset of unspecific vestibular and cochlear symptoms induced by an enhanced sensitivity of labyrinthine receptors due to a bony defect usually located in the external wall of the superior semicircular canal.

The exact incidence of posterior semicircular canal dehiscence (PSCD) is unknown.

In a radiographic survey of 112 temporal bones, known to have a high jugular bulb, this was detected in 4 patients [2]. In contrast, the incidence of superior semicircular canal dehiscence (SSCD) in a temporal bone survey was 0.7% [3]. In this report, a case of association between two alterations of the otic capsule is presented supporting the notion that there is probably a developmental abnormality underlying the SSCD and PSCD syndromes.

Case report

A 38-year-old male was referred to our tertiary referral neurotological centre with vestibular (recurrent Tullio Phenomenon or sound-induced vertigo) and cochlear symptoms (persistent aural fullness) with mixed hearing loss (Fig. 1) associated with disabling pulsatile tinnitus.

A detailed case history revealed oscillopsia related to intense exertions and "Tullio Phenomenon" evoked by loud sounds. The patient did not remember closed cranial trauma. The patient also referred to and reported pulsatile tinnitus which was particularly irritating and disabling. Otoscopy findings were normal.

All the symptoms mentioned by the patient were then evaluated with a standardized set of tests including those during "Bed-side examination" with a video oculography (VOG) recorder. The pattern of horizontal, torsional and vertical eye movements was evaluated and recorded with three-dimensional infra-red video oculography (50

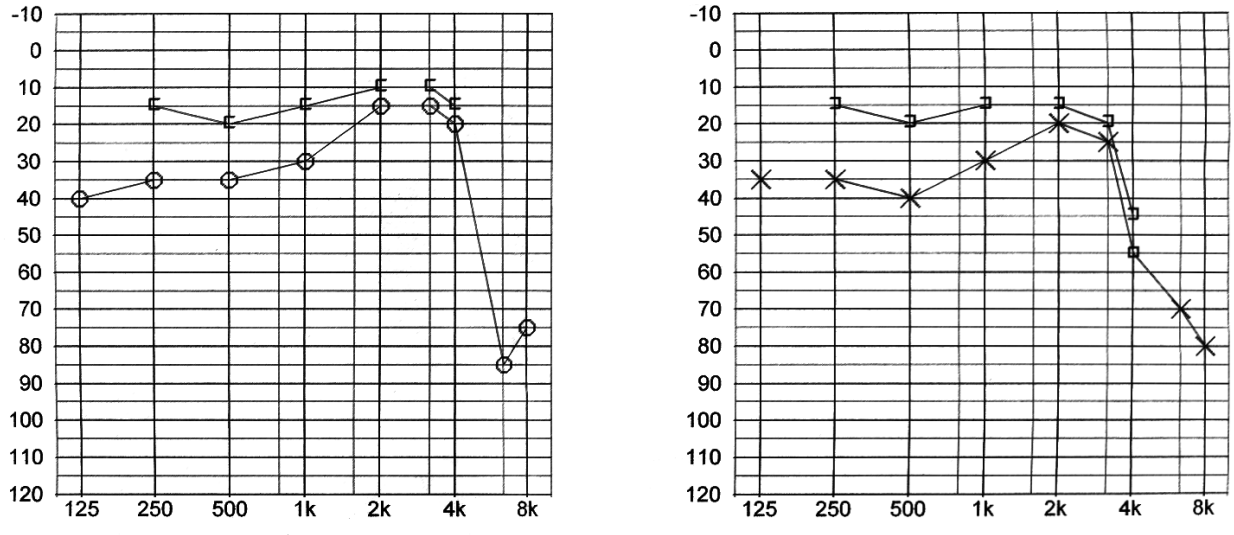

Fig. 1. Audiogram showing bilateral mixed hearing loss (conductive component of low-middle frequencies).

Hz sampling; Torsio VNG Ulmer; Synapsys, Marseille, France): spontaneous nystagmus, head shaking nystagmus, research of the position and positioning nystagmus (Dix-Hallpike manoeuvre and head roll manoeuvre), observation under VOG of the nystagmus after Valsalva manoeuvre, observation after hyperventilation test and after exposure to 110 dB sound stimulation, at a frequency of 3 kHz, for the detection of the so-called "Tullio Phenomenon" and, finally, after mastoid vibration at 100 Hz.

The patient was then submitted to a specific set of instrumental audio-vestibular tests: caloric tests showed a normal pattern (methodology of stimulation according to Fitzgerald-Hallpike), Fz bone conducted ocular and cervical vestibular evoked myogenic potentials (VEMPs) performed with a short tone burst of 500 Hz lasting 7 milliseconds with a repetition frequency of 4 Hz for 50 times (Fig. 2A, B) showing enhanced responses in terms of amplitude. Stapedial reflexes and auditory brainstem response were present.

For this reason, the patient was first referred to a tertiary radiology centre for magnetic resonance imaging (MRI) of the posterior cranial fossa using paramagnetic contrast enhancement. This radiological evaluation revealed a normal and symmetrical eighth cranial nerve. At this point, and on account of the ocular and cervical VEMPs results, it was necessary to perform high resolution computed tomography (CT) that revealed bilateral dehiscence of the posterior semicircular canal (Fig. 3c, d) and right SSCD with marked thinning of the bone covering the left superior semicircular canal dehiscence. This last imaging result was fundamental in explaining the vestibular symptoms (Fig. 3 a, b).

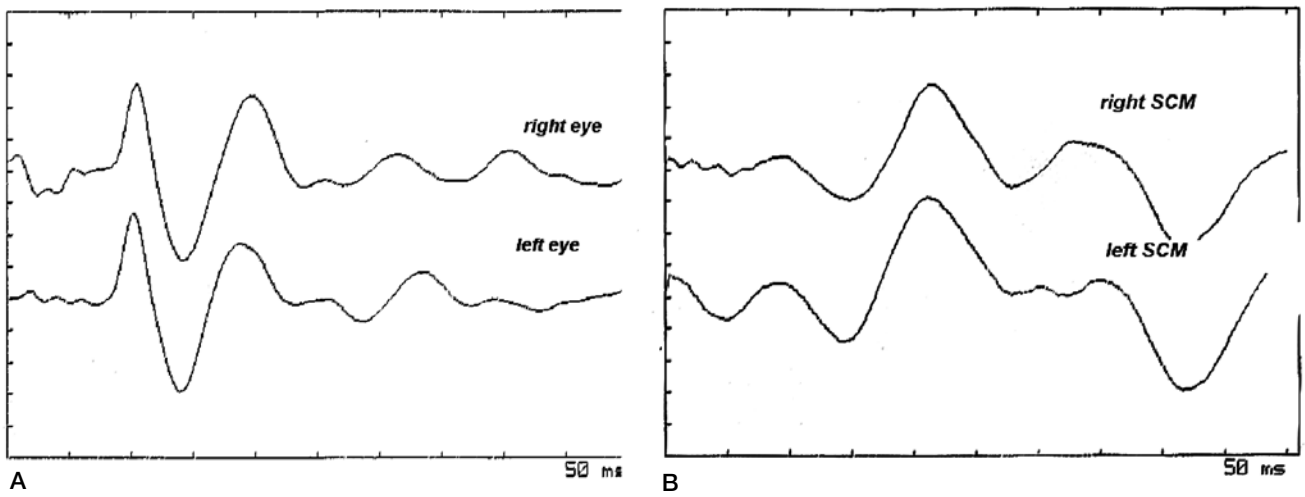

Fig. 2. Recordings of bone conducted vibratory Fz ocular VEMPs (A) and cervical VEMPs (B) from patient, showing bilaterally enhanced amplitudes of n-10 in oVEMPs and p-13/n-19 complex in cVEMPs.

Discussion

In 1998, Minor et al. [1] described, for the first time, a new clinical entity characterized by the onset of non-specific vestibular and cochlear symptoms induced by hypersensitivity of the labyrinthine receptors due to a bony defect usually located in the external wall of the superior semicircular canal.

Diagnosis can be based on characteristic symptoms, as the latter may include dizziness, Tullio Phenomenon, positional vertigo, pulsatile tinnitus, conductive and/or sensorineural hearing loss, etc.; the possibility of canal dehiscences must be considered in the presence of certain signs (Hennebert sign, nystagmus induced by Valsalva manoeuvre, etc.) and may be confirmed by the results of some instrumental investigations (essentially, cervical vestibular evoked myogenic potentials (c-VEMPs) with threshold analysis) [4].

However, the final confirmation requires cross-sectional imaging by means of high resolution computed tomography (HRCT) able to follow, and tracing, the circumference of the superior and posterior semicircular canal.

The contrast between bone and soft tissue obtained by a bone dedicated window provides excellent definition of bone structure. However, all projections help the clinician and radiologist to identify dehiscences (Fig. 3). CT scans also provide diagnostic information not only in genetic but, especially, as in the present case, in lifestyle counselling.

HRCT has proven to have a sensitivity of 100% and a specificity of 99% for the diagnosis of canal dehiscences [4]. The bilateral nature of the defect supports the hypothesis of a developmental abnormality, even if of a mild degree, as already suggested for SSCD [5].

The aetiology of SSCD is debated [5]. The independent development of the individual semicircular canals in relation to the cochlea and vestibule is a complex embryologic process rather than merely an arrest in development [6]. This syndrome may also result from an abnormal vascular anatomy and may present late in life without further evidence of precipitation [3 7 8]. SSCD caused a significant reduction in sound-induced round-window velocity at low frequencies, small but significant increases in sound-

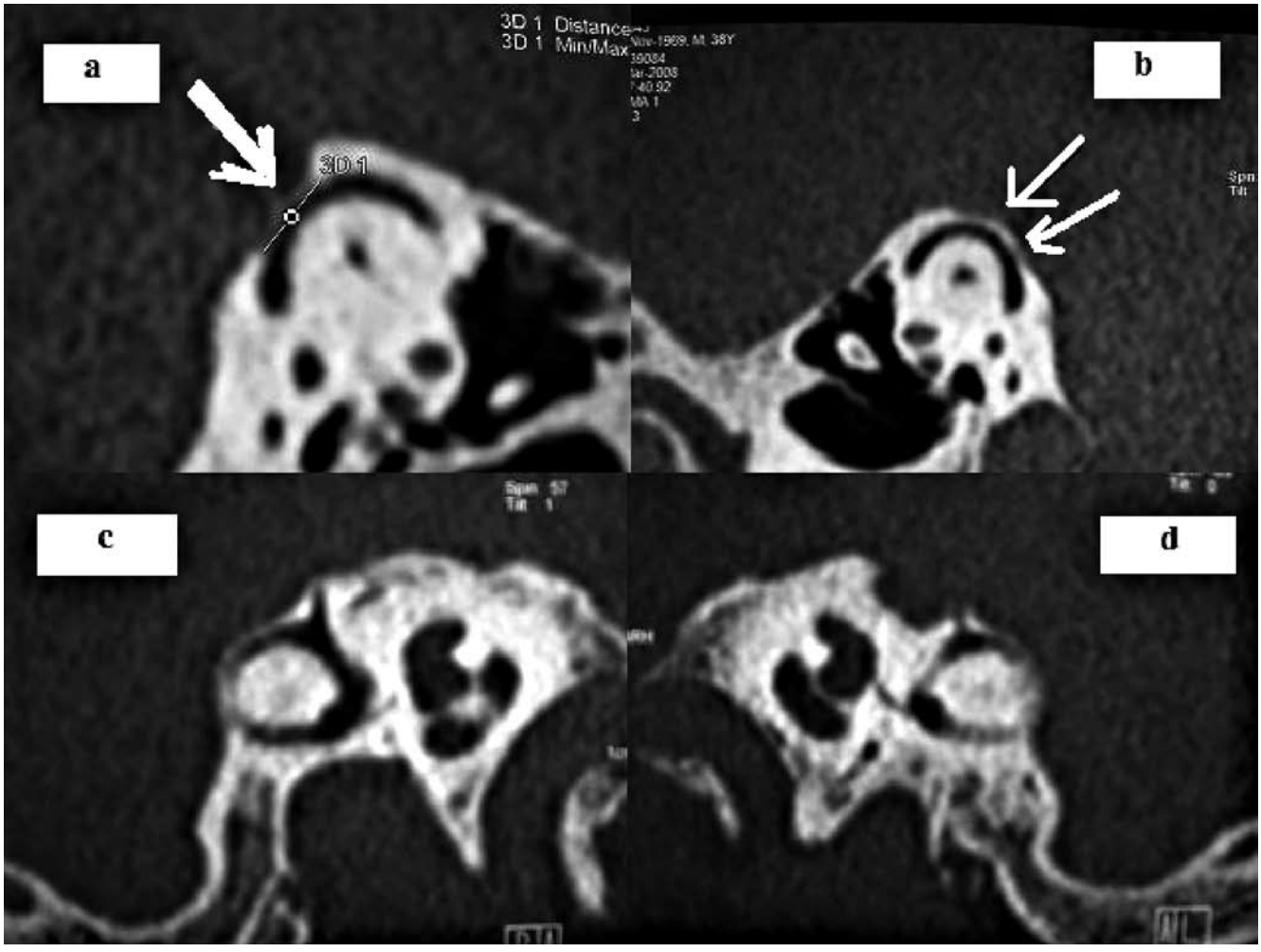

Fig. 3 a-d. CT scan, sagittal projections showed bilateral dehiscence of posterior semicircular canal in right ear (3c) and left ear (3d), and unilateral right dehiscence of the superior semicircular canal (3a white arrow). White arrows in 3b instead show the bone covering the left superior semicircular canal.

induced stapes and umbo rates, and a measurable fluid velocity inside the dehiscence. Findings from the cadaveric temporal-bone preparation were consistent with the third-window hypothesis.

The present case, with its association of two lesions [10], with no previous history of trauma, confirms the hypothesis that congenital and/or genetic factors are involved in some cases. The present case also alerts clinicians about associations of auditory and vestibular symptoms.

In the absence of reports of such cases [11 12], it was deemed important to obtain CT scans in order to reach a diagnosis and a differential diagnosis (for example with PSCD in patients presenting with vertigo, sensorineuronal hearing loss) [13].

The management of superior semicircular canal dehiscence syndrome involves conservative and surgical approaches. Surgical options for patients with disabling vestibular symptoms include a middle fossa craniotomy [14] or a viable alternative to this technique, trans-mastoid superior semicircular canal occlusion [14]. Primary middle fossa or trans-mastoid superior semicircular canal occlusion repair are not associated with sensorineural hearing loss and, in some cases, can lead to a return to normal of the conductive hearing loss [14].

In conclusion, this case also demonstrates the need, for the clinician, to be highly suspicious of the associations between auditory and vestibular symptoms.

The bilateral nature of the defect would appear to suggest a developmental abnormality or a more complex embryologic process [6], albeit mild, as already suggested for SSCD [5].

References

1 Minor LB, Solomon D, Zinreich JS, et al. *Sound- and/or pressure-induced vertigo due to bone dehiscence of the superior semicircular canal*. Arch Otolaryngol Head Neck Surg 1998;124:249-58.

2 Wadin K, Thomander L, Wilbrand H. *Effects of a high jugular fossa and jugular bulb diverticulum on the inner ear. A clinical and radiologic investigation*. Acta Radiol Diagn (Stockh) 1986;27:629-36.

3 Carey JP, Minor LB, Nager T. *Dehiscence or thinning of bone overlying the superior semicircular canal in a temporal bone survey*. Arch Otolaryngol Head Neck Surg 2000;126:37-47.

4 Hirvonen TP, Weg N, Zinreich SJ, et al. *High-resolution CT findings suggest a developmental abnormality underlying superior canal dehiscence syndrome*. Acta Otolaryngol 2003;123:477-81.

5 Minor LB. *Clinical manifestation of superior semicircular canal dehiscence*. Laryngoscope 2005;115:1717-27.

6 Yu KK, Mukherji S, Carrasco V, et al. *Molecular genetic advances in semicircular canal abnormalities and sensorineural hearing loss: a report of 16 cases*. Otolaryngol Head Neck Surg 2003;129:637-46.

7 Puwanarajah P, Pretorius P, Bottrill I. *Superior semicircular canal dehiscence syndrome: a new aetiology*. J Laryngol Otol 2008;122:741-4.

8 Brantberg K, Greitz D, Pansell T. *Subarcuate venous malformation causing audio-vestibular symptoms similar to those in superior canal dehiscence syndrome.* Otol Neurotol 2004;25:993-7.

9 Chien W, Ravicz ME, Rosowski JJ, et al. *Measurements of human middle- and inner-ear mechanics with dehiscence of the superior semicircular canal*. Otol Neurotol 2007;28:250-7.

10 Mahendran S, Sunkaraneni VS, Baguley DM, et al. *Superior semicircular canal dehiscence with a large tegmental defect*. J Laryngol Otol 2007;121:189-91.

11 Rajenderkumar D, Farrell KL, Alles RM, et al. *Multiple dehiscence of semicircular canals*. J Laryngol Otol 2007;121:80-2.

12 Manzari L, Modugno GC. *Bilateral dehiscence of both superior and posterior semicircular canals*. Otol Neurotol 2009;30:423-5.

13 Krombach GA, DiMartino E, Schmitz-Rode T, et al. *Posterior semicircular canal dehiscence: a morphologic cause of vertigo similar to superior semicircular canal dehiscence*. Eur Radiol 2003;13:1444-50.

14 Limb CJ, Carey JP, Srireddy S, et al. *Auditory function in patients with surgically treated superior semicircular canal dehiscence*. Otol Neurotol 2006;27:969-80.

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