Do signs of natural plugging of superior semicircular canal dehiscence exist?
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Abstract
Our experience with 102 patients having superior semicircular canal dehiscence confirm that the clinical manifestations of the disease are very diverse; we also identified 3 patients who showed Meniere-like symptoms. Clinical examination during an acute vertigo attack of a patient with Meniere disease for several years and whom we subsequently diagnosed as having large superior semicircular canal dehiscence on the affected side allowed us to hypothesize that a natural plugging of the superior semicircular canal by the overhanging dura mater could be responsible for the recurrence of symptoms. Clinical and instrumental data were very similar to those recorded in 7 of 9 patients immediately after surgical plugging. The aim of the study was to understand which semiological and instrumental elements could be clinically useful, first in distinguishing Meniere disease from superior semicircular canal dehiscence and, secondly, in understanding if signs of natural plugging are present.
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1. Introduction
Dehiscence of the bone overlying the superior semicircular canal (SSCD) is considered a possible structural cause of sound-induced vertigo (Tullio phenomenon) and pressure-induced vertigo and/or hyperacusis to bone-conducted sound [1].

Electrophysiologic data by vestibular evoked myogenic potentials (VEMPs) and temporal bone high-resolution computed tomography (CT) (HRCT), carried out using 0.5-mm contiguous reconstructions of the oblique plane of the superior semicircular canal (SSC), should be used to confirm the presence of a “mobile third window,” such as SSCD [1-3].

Clinical manifestations of SSCD are so diverse and patient specific that its typical features can be totally absent. Patients with a debilitating symptoms may benefit from surgical plugging of the SSCD through the middle fossa or using the transmastoid approach (TM) [2].

Ever since SSCD was first characterized, only a few authors have described the fact that cochleovestibular symptoms can mimic that of the Meniere disease [4]; a few cases of SSCD associated with Meniere disease have been reported by Pyykkö and Poe [4]. It is interesting that, in his study, magnetic resonance imaging was used to demonstrate endolymphatic hydrops both before and after surgery [4].

Clinical observation during an acute vertigo attack of a patient with Meniere disease for several years and who we subsequently diagnosed as having large SSCD on the affected side allowed us to hypothesize, based on several clinical and semiological elements, that recurrent Meniere-like symptoms could be justified by natural plugging of the SSC endolymphatic compartment by the overhanging dura mater. Our hypothesis was borne out by the strict similarity between the symptoms of our case report and that of a small group of patients observed immediately after SSC surgical plugging.

2. Patients and methods
One hundred two patients were identified as having SSCD at the ENT Unit of the Specialistic Surgical and...
Nine patients with incapacitating SSCD symptoms underwent surgical plugging by the senior author (GCM) during the period from October 2005 to December 2010 (Table 1).

Of 102 SSCD patients, many of them reported an acute vertigo attack (33%, benign paroxysmal positional vertigo excluded), but only 3 of them described Meniere-like symptoms. We will now describe shortly the only 1 case that we had the opportunity of examining in the course of an acute vertigo attack.

### 3. Case report

The patient, a 63-year-old woman, presented to our tertiary otology center with a long history of audiological examinations, which hypothesized left Meniere disease. Cochleovestibular examinations revealed a mild mixed hearing loss (air-bone gap at 250 and 500 Hz) in the left ear, which had slightly worsened with respect to her earlier audiometries, right beating vibration-induced nystagmus, and normal bilateral cervical VEMPs thresholds (left 110 dB sound pressure level [SPL], right 115 dB SPL). Contrast-enhanced brain magnetic resonance imaging study revealed no enhancing lesions.

One year later, she again presented at our hospital complaining of continuous sense of vertigo and nausea, which had begun suddenly 2 days after an international flight. Auditory symptoms, such as left tinnitus and fullness, were also present. The anamnesis disclosed interesting elements, such as considerable intolerance for low sounds and vibrations, and great distress when encountering bumps in the road (slight oscillopsia). Pure-tone audiometry revealed a mild mixed hearing loss (slight air-bone gap at 250 and 500 Hz) in the left ear and a slight sensorineural hearing loss in the right ear. Imittance audiometry showed normal bilateral tympanic membrane mobility and normal bilateral acoustic reflexes.

Video nystagmoscopy revealed evident left beating spontaneous nystagmus (fast phase, observer’s view) enhanced by bilateral mastoid vibration, which added a torsional clockwise component to it. Cervical and periocular air-conducted VEMP testing showed a lowered left (100 dB SPL) vs right threshold (132 dB SPL).

At this stage, a temporal bone HRCT performed on a multislice light speed GE Medical Systems (Buc, France) scanner revealed left SSCD and very thin right bone layer covering the SSC (Fig. 1).

Over the first 10 days, vestibular bedside examination was carried out daily. The left spontaneous nystagmus gradually decreased and disappeared in approximately a month, whereas the torsional components of the vibration-induced nystagmus continued for a longer time (approximately 4 months). Nearly 8 months later, vestibular examination recorded a total absence of spontaneous nystagmus, but afterwards, the patient returned, complaining of a slight vertigo episode without cochlear symptoms: we recorded right beating spontaneous nystagmus, unchanged in the course of a head pitch test, mastoid vibration, and head shaking. The TM SSC plugging was proposed but not accepted.

Actually, 20 months after the first observation, the patient is free from vestibular symptoms and dizziness. She has only complained of left hearing loss.

### 4. Discussion

Endolymphatic hydrops as a consequence of membranous labyrinth SSC compression by a prolapsing dura in large SSCD was hypothesized by Minor et al [3] about 10 years ago.

Other authors have recently also correlated SSCD to the possibility of endolymphatic hydrops in 4 patients, without, however, expressing themselves as to the physiopathology of this condition [4].

None of these authors has, however, reported the bedside examinations with video nystagmoscopy in an acute vertigo attack in patients having Meniere-like symptoms.

The patient aforementioned had been considered to be affected by left Meniere disease by other otorhinolaryngologists.
During her vertigo crisis, we recorded left spontaneous nystagmus (which persisted for several days) and left mixed low frequencies hearing loss. This condition could be consistent with an attack of Meniere disease in which brief ipsilesional spontaneous nystagmus can be present at the beginning. In this patient, however, the ipsilesional spontaneous nystagmus had lasted for more than a few hours but even as long as 1 month. In addition, the mastoid vibration had generated torsional clockwise/vertical up-beating nystagmus components. These important functional data suggested the hypothesis of left SSCD, which was subsequently confirmed by VEMP and CT testing.

Approximately 8 months later, we recorded an inversion of the nystagmus direction (soft right spontaneous nystagmus) and the well-being of our patient.

Similarly, in our small group of patients with SSCD treated with surgical plugging (carried out using different techniques), immediately after surgery, intense vestibular symptoms were accompanied by strong spontaneous nystagmus directed toward the side operated on, which appeared that it underwent a change in direction a few days later (Table 1).

Up to now, numerous publications have shown the vestibular function assessment of SSCD patients, both before and after SSC surgical plugging. Studies of the vestibuloocular reflex have demonstrated that plugging produces a selective reduction in the functioning of the canal operated on while preserving relatively normal functioning in the other canals [5]. Cervical and ocular VEMP tests have demonstrated that surgical plugging produces a normalization of potential thresholds [2]. The vestibular semiology was not analyzed, even during first postoperative period. The only description of horizontal ipsilesional spontaneous nystagmus resulting from TM surgical SSC plugging, carried out to resolve intractable SSC benign paroxysmal positional vertigo (BPPV), was by Brantberg and Bergenius [6], who hypothesized an irritated state of the left lateral semicircular canal; his patient also showed ipsilesional fullness during vertigo and a reduced energy response. However, we do not know if the author considered the hypothesis of SSCD in his case report.

The natural SSC plugging in our patient was hypothesized, above all, thanks to a nystagmus pattern during the vertigo attack, more precisely ipsilesional nystagmus.

On the contrary, other semiological elements, such as the torsional component in vibration-induced nystagmus and electrophysiologic data resulting from VEMP testing, suggested an active “third window effect.” Indeed, we hypothesized that the natural plugging could be “temporary” or realized in only 1 of the SSC arms; the membranous canal can be closed, but, at the same time, the bony canal can be opened. Vestibuloocular reflex analysis with high-speed camera could be soon useful in confirming eventual selective SSC damage.

Although the evidence accumulated during this study is inferential, it does suggest sufficiently that the natural plugging can be a dynamic condition predisposed to evoke recurrent cochlear and vestibular symptoms, such as Meniere disease.

Continued longitudinal follow-up of a large quantity of similar patients may provide additional insights. We recommend following the situation daily using video nystagmoscopy for all patients having ipsilesional nystagmus during their vertigo attack to eventually evaluate its directional change and/or to evoke possible vertical/torsional vibration-induced nystagmus.

We conclude by proposing the inclusion of a the temporal bone HRCT study, completed by reformatted images aligned SSC plane, in the diagnostic protocol of all patients affected by Meniere disease or Meniere-like symptoms.
References


