Posterior semicircular canal dehiscence: value of VEMP and multidetector CT


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Abstract. Posterior semicircular canal dehiscence: value of VEMP and multidetector CT. Objective: To illustrate that posterior semicircular canal dehiscence can present similarly to superior semicircular canal dehiscence.

Case study: The symptomatology initially presented as probable Menière’s disease evolving into a mixed conductive hearing loss with a Carhart notch-type perceptive component suggestive of otosclerosis-type stapes fixation. A small hole stapedotomy resulted in a dead ear and a horizontal semicircular canal hypofunction. Recurrent incapacitating vertigo attacks developed. Vestibular evoked myogenic potential (VEMP) testing demonstrated intact vestibulocollic reflexes. Additional evaluation with high resolution multidetector computed tomography (MDCT) of the temporal bone showed a dehiscence of the left posterior semicircular canal.

Conclusions: Besides superior semicircular canal dehiscence, posterior semicircular canal dehiscence has to be included in the differential diagnosis of atypical Menière’s disease and/or low tone conductive hearing loss. The value of performing MDCT before otosclerosis-type surgery is stressed. VEMP might contribute to establishing the differential diagnosis.

Introduction

Superior semicircular canal dehiscence has recently been added to the differential diagnosis of Menière-like symptoms and low frequency sensorineural or conductive hearing loss.1 Review of CT scans of patients with these symptoms have shown that the diagnosis of superior semicircular canal dehiscence is not extremely uncommon. Up to 83% of patients with superior semicircular canal dehiscence presented with symptoms of vertigo and 10% of them with hearing loss and tinnitus.2 Although several reports showed a favourable outcome of a fossa media approach with plugging or resurfacing of the dehiscence of the superior semicircular canal, all focused on the high incidence of “missed diagnosis”. This resulted in long term treatment for Menière’s disease or in otosclerosis surgery, which led to complete hearing loss in certain cases.

Recently, the occurrence of posterior semicircular canal dehiscence was reported.2 Apart from perilymph leakage, which is an important component in establishing a differential diagnosis in patients with Menière-like symptoms, the authors stress the inclusion of semicircular canal dehiscences in the differential diagnosis of these patients. The consequences of semicircular canal dehiscences are illustrated by means of a case study of posterior semicircular canal dehiscence.

Case report

The 40-year-old female patient started with a period of unsteadiness and vertigo, initiated by a drop-type attack. After an initial evaluation, the diagnosis of probable Menière’s disease on the left was proposed. Conservative medical therapy followed. The vertigo sensations became insignificant but the patient reported that she experienced progressive hearing loss and tinnitus on the left. Audiometry showed a mainly conductive hearing loss, with a pronounced Carhart notch-type perceptive component around 2 kHz on that side (Figure 1). The tympanometry was normal and acoustic reflexes were absent. There was no history of otitis or ear pain. Based on these findings, an uneventful surgical intervention was performed using a classical endaural approach with small hole fenestration of the footplate and teflon Causse prosthesis interposition between the incus and the stapedotomy.

In the early postoperative period total hearing loss occurred, followed by recurrent attacks of violent incapacitating vertigo. At this
stage, the patient was first seen in our department. Upon neuro-otological evaluation, audiometry showed a dead ear on the left. The right ear had a normal hearing threshold. The caloric test showed a left horizontal semicircular canal paresis (50%) and a spontaneous nystagmus to the right together with a down beating nystagmus. The gain of the vestibular ocular reflex (VOR), tested by rotating the patient sinusoidally around an earth vertical axis (maximum angular velocity = $50^\circ/s$; frequency = 0.05 Hz), was within normal limits.\(^3\) The saccadic and optokinetic eye tracking was normal. Vestibular evoked myogenic potentials (VEMPs), which were performed by means of unilateral stimulation using 500 Hz tone bursts (rise/fall time = 2 ms, plateau time = 2 ms; methodology: demonstrated a clear response in both ears.\(^4\) The sound threshold was 90 dB nHL in the left and 85 dB nHL in the right ear (Figure 2).

High resolution multidetector computed tomography (MDCT) scans of the temporal bone showed a dehiscence of the left posterior semicircular canal (Figures 3, 4).

Drug therapy did not yield an improvement of the rotatory vertigo attacks, which had resulted from the combination of the stapedotomy and the dehiscence of the

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**Figure 1**

Audiogram of the patient before otosclerosis surgery (o: air-conduction threshold right ear; x: air-conduction threshold left ear; ]: bone conduction threshold left ear).

**Figure 2**

VEMP with determination of the tone burst threshold (500 Hz; rise/fall time = 2 ms, plateau time = 2 ms) for left (a) and right (b) side.
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Posterior semicircular canal. A selective retrosigmoid vestibular neurectomy was performed on the left side and followed by a vestibular rehabilitation programme. During the six-months period after the surgery, the patient experienced no rotatory vertigo attacks.

Discussion

This case illustrates that a similar clinical history can occur with posterior as well as superior semicircular canal dehiscence. Initially, symptomatology was suggestive of Menière’s disease. In a second phase, a mixed low and mid-tone conductive and high-tone sensorineural hearing loss, normal tympanometry and absent acoustic reflexes were seen, mimicking otosclerosis.

Mikulec et al. reported absent acoustic reflexes in 1 out of 4 ears with posterior SCC dehiscence that had not undergone stapedectomy.

The MDCT scans of the temporal bone showed a posterior canal dehiscence on the left. Cross-sectional imaging, such as CT and magnetic resonance imaging (MRI), is performed to confirm the clinical diagnosis and to localize the exact portion (anterior limb, apex, or posterior limb) of

Figure 3

a. Posterior semicircular dehiscence. Axial 0.5 mm reconstructed CT images. b. Orthogonal reconstructed 0.5 mm images in the coronal plane. A small defect of the bone overlying the posterior semicircular canal can be observed (white arrows). Normal appearance of the superior semicircular canal.

Figure 4

Added value of additional oblique reconstructions in semicircular canal dehiscence. a. Orthogonal axial reference image shows the plane of reconstruction through the roof of the left posterior semicircular canal. b. Single-oblique sagittal reconstruction shows dehiscence of the left posterior semicircular canal (white arrow).
the semicircular canal that is dehiscent. In general, a bony defect can be present at both limbs of the superior semicircular canal and/or at the posterior limb of the posterior semicircular canal. Bony defects over the lateral semicircular canal, other than those caused by erosion from cholesteatoma, have not been described. In the early case reports, direct axial and coronal images were used to confirm the diagnosis of superior semicircular canal dehiscence. However, many anatomic structures of the middle and inner ear, such as the semicircular canals, are not optimally depicted when using the standard axial and coronal planes. Multidetector CT allows the isotropic voxels to be reconstructed in any plane, thus permitting optimal depiction of clinically relevant temporal bone anatomy, and this without additional radiation or scanning time.

In the evaluation of the semicircular canals, orthogonal axial and coronal images usually suffice for the assessment of the bony integrity of the canals (Figure 3). However, in individuals in whom the covering of bone is thin, problems with volume averaging effects could cause confusion regarding the integrity of the bone. Oblique sagittal reconstructions parallel to (Stenvers plane) and perpendicular to (Pöschl plane) the superior and posterior semicircular canal can help eliminate problems with volume averaging (Figure 4).

As in many superior semicircular canal dehiscence cases reported, this case also underwent an otosclerosis operation. Similar to some reported cases and to some situations known to occur with a gusher, in which there is a direct connection between the internal auditory canal and the scala tympani of the basal turning of the cochlea which may lead to a deaf ear, a total hearing loss occurred after the surgery. The persistent vertigo spells necessitated a selective retrosigmoid vestibular neurectomy. The physiopathology of the existence of Ménière-type symptoms and the conductive (or mixed) type audiogram in these patients is well explained physiopathologically and is supported by experimental work by Mikulec et al. The dehiscence is depicted as a third mobile window in the inner ear (in addition to the oval and round window), which results in an elevation of thresholds for air-conducted sounds and reduction of thresholds for bone-conducted sounds. Dissipation of acoustic energy through the dehiscence away from the cochlea could create an elevation of air-conducted hearing thresholds. Acoustic impedance can lead to the loss being more pronounced in the low frequencies. The effect of a dehiscence on bone-conducted thresholds can be understood based on the compressional mechanism of bone conduction. A semicircular canal dehiscence serves to increase the inequality of impedance between the round and oval windows, thereby increasing the cochlea’s response to compressional bone conduction and can thus explain the negative bone-conduction thresholds observed in some patients. Vestibular complaints in patients with dehiscence of bone overlying the superior semicircular canal occur mostly after stimulation by sound or pressure stimuli, although this was not reported explicitly by this patient. These vertigo sensations can be explained by the greater compliance of the vestibular system due to the lack of bone over the semicircular canal. Stimuli which result in an inward pressure at the round or oval window result in an outward bulging of the membranous canal near the dehiscence and an ampullofugal deflection of the cupula. These mechanisms of cupula stimulation can also be responsible for the vertigo attacks in patients with posterior semicircular canal dehiscence. The audiogram before the surgery showed mainly a conductive hearing loss. A hearing loss with a significant air-bone gap due to ossicular fixation, as with otosclerosis for instance, would lead to absent VEMP’s. Performing a VEMP test might lead to etiologies other than otosclerosis in this condition. This had not been carried out before the otosclerosis surgery.

Nine months after the surgery, the caloric test showed a diminished left horizontal semicircular canal function. The VEMP thresholds were in the normal range and were similar (90 dB nHL in the left ear and 85 dB nHL in the right ear). The VEMP examination in this case shows the functionality of the sacculus in spite of a total deaf ear on the left, and explains its vestibular origin. This study is the first reported case of the performance of a VEMP test in posterior semicircular canal dehiscence. In most reports of superior semicircular canal dehiscence, the VEMP response occurs at a lower threshold than in normal subjects, which can be explained by the
increased transmission of sound energy to the saccule due to the increased compliance of the endolymphatic system. In this case, VEMP responses were present, although not at a lower threshold. The authors hypothesise that in the case of a semicircular canal dehiscence the VEMP threshold is determined by the compliance of the endolymphatic system and this can vary from case to case. This has to be investigated by performing VEMP measurements in a significant number of patients with posterior or superior canal dehiscence.

**Conclusion**

The authors conclude that, besides superior semicircular canal dehiscence, posterior semicircular canal dehiscence has also to be taken into account in the differential diagnosis of Menière’s disease and/or low tone conductive hearing loss.

In order not to overlook the diagnosis of superior/posterior semicircular canal dehiscence, the authors emphasize the need to evaluate the VEMP response and to perform a high resolution MDCT scan in cases with vertigo or unsteadiness and an ‘otosclerosis-like’ audiogram.

**References**


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