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Superior canal dehiscence with tegmen defect revealed by otoscopy: Video clip demonstration of pulsatile tympanic membrane

Andrea Castellucci [a], Cristina Brandolini [b], Gianluca Piras [c], Ignacio Javier Fernandez [b], Davide Giordano [a], Carmine Pernice [a], Giovanni Carlo Modugno [b], Antonio Pirodda [b], Gian Gaetano Ferri [b,*]

[a] *ENT Unit, Department of Surgery, Arcispedale Santa Maria Nuova, Center for Clinical and Basic Research (IRCCS), Viale Risorgimento 80, 42123 Reggio Emilia, Italy*
[b] *ENT & Audiology Unit, S.Orsola-Malpighi University Hospital, Via Massarenti 9, 40138 Bologna, Italy*
[c] *Department of Otology and Skull Base Surgery, Gruppo Otologico, Via Antonio Emmanueli 42, 29121 Piacenza, Italy*

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ABSTRACT

Superior canal dehiscence is a pathologic condition of the otic capsule acting as aberrant window of the inner ear. It results in reduction of inner ear impedance and in abnormal exposure of the labyrinthine neuroepithelium to the action of the surrounding structures. The sum of these phenomena leads to the onset of typical cochleo-vestibular symptoms and signs. Among them, pulsatile tinnitus has been attributed to a direct transmission of intracranial vascular activities to labyrinthine fluids. We present the first video-otoscopic documentation of spontaneous pulse-synchronous movements of the tympanic membrane in two patients with superior canal dehiscence. Pulsating eardrum may represent an additional sign of third-mobile window lesion.

1. Introduction

Superior canal dehiscence syndrome (SCDS) is one of the latest clinical entities introduced in neuro-otology [1]. It is a relatively rare condition occurring in patients with a congenital or acquired bony defect overlying the superior semicircular canal (SSC). It is often associated to a dehiscence or thinning of the contralateral SSC roof [2,3], and/or to other concomitant middle fossa floor defects, likely suggesting a common morphologic abnormality of the skull base [4]. It can result in a variable combination of peculiar symptoms such as vertigo induced by pressure and/or loud sounds, hyperacusis to bodily sounds, autophony and pulsatile tinnitus [5]. While the latter has been attributed to a direct transmission of the intracranial pulse-related pressure changes to the inner ear fluids through the bony defect, most symptoms recognize the so-called ''third-widow mechanism'' as underlying physiopathological substrate [5]. In fact, similar to other morphological abnormalities of the otic capsule acting as aberrant inner ear windows (such as enlarged vestibular aqueduct and posterior canal dehiscence), a bony defect over the SSC reduces the impedance of the vestibular partition of the inner ear. This condition induces an abnormal vestibular activation to acoustic stimuli, thereby shunting part of the air-conducted acoustic energy away from the cochlear compartment [6]. Typical signs of third-window

* Corresponding author at: Department of Experimental, Diagnostic and Specialty Medicine (DIMES), S.Orsola-Malpighi University Hospital, Via Massarenti 9, 40138 Bologna, Italy.
E-mail address: giangaetano.ferri@unibo.it (G.G. Ferri).

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lesions include sound/pressure-elicited torsional nystagmus, mild low-frequency air-bone gap (ABG) with preserved acoustic reflexes, cervical vestibular evoked myogenic potentials (VEMPs) with increased amplitude and lowered threshold [5]. Since imaging tends to overestimate the incidence of SSC dehiscence, an appropriate diagnosis of SCDS should be formulated not only by means of high resolution CT (HRCT) scans of the temporal bones showing a bony defect over the SSC, but also by abnormal electrophysiological data, thus confirming an increased inner ear admittance on the lesion side [7]. Recently, pulse-synchronous waves on impedance audiometry have been described as an additional rare sign of third-window lesion [8]. We present, for the first time, to the best of our knowledge, video-recorded pulsating activities of the tympanic membrane (TM) in two patients with SCD associated with an extremely thinned/dehiscent tegmen tympani.

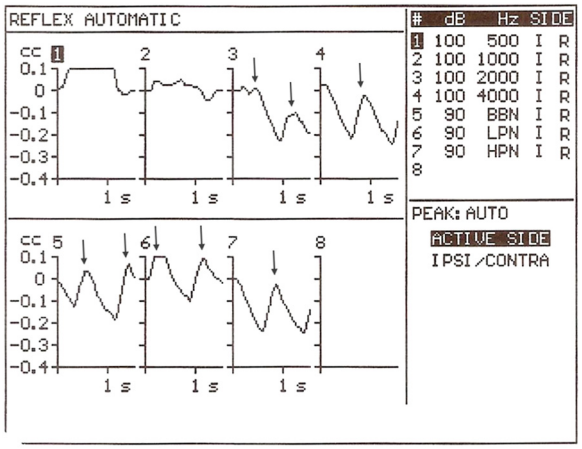

Fig. 1. Pulse-synchronous waves recorded during ipsilateral acoustic reflex testing on the right ear of patient 1.

2. Case reports

Patient 1. A 62-year-old woman affected by a right intracanalicular vestibular schwannoma (VS) presented to our attention for the gradual onset of right pulsatile tinnitus worsening during physical efforts. She also complained of chronic imbalance and recurrent benign paroxysmal positional vertigo (BPPV) attacks involving the right posterior semicircular canal. She has been undergoing annual brain Magnetic Resonance Imaging (MRI) which showed a not-growing VS without other inner ear abnormalities. She had neither history of head injury nor ear surgery and was submitted to an extensive otoneurological examination. Standard otoscopy was uneventful, while a video-otoscopy performed with a Karl Storz 0° endoscope recorded pulse-synchronous movements of the right eardrum (Video 1). Pulsations stopped when the patient was supine and while she was performing a Valsalva maneuver with closed glottis. No retrotympanic masses or TM movements in response to forced breathing were detected. Audiometry revealed a right, mixed hearing loss with a 30 dB ABG for the mild-low frequencies (0.25–1 kHz) and a left down-sloping age-related sensorineural hearing loss. Tympanometry produced type A-shaped curves and normal acoustic reflexes were evoked on both sides. The patient experienced subjective dizziness during right impedance testing. Right ipsilateral acoustic reflex recording detected a pulsating wave synchronous with her heartbeat (Fig. 1). Neither spontaneous/positional nystagmus nor Valsalva-induced eye movements could be seen by video-oculography. Bi-mastoid vibratory test elicited a transitory vertical up-beating nystagmus. Right canal hypo-reflectivity (Canal Paresis (CP) of 29%) was detected by standard bithermal caloric test (30 and 44 °C ear irrigations with a VN415/VO425 FireWire VNG system, Interacoustics, Denmark). Normal high-frequency VOR gains were found by testing each semicircular canal function with the video Head Impulse Test (vHIT) using an ICS video-oculographic system (GN Otometrics, Denmark). Air-conducted (AC) cervical-VEMPs, recorded using an Epic Plus evoked potentials system (Labat, Mestre, Italy) with a 2-channels averaging capacity, showed enlarged potentials with lowered threshold (85 dB SPL) in the right side. Abnormally enhanced VEMPs to bone-conducted (BC) stimuli were also detected under the left eye. Temporal bone HRCT scans detected a wide bony dehiscence of the right SSC, an extremely thin bone overlying the left canal and bilateral tegmen defects (Fig. 2).

Supplementary Video 1 related to this article can be found, in the online version, at http://dx.doi.org/10.1016/j.anl.2016.11.013.

Patient 2. A 68-year-old woman visited our outpatient clinic with a right-sided hearing loss, autophony and pulsatile tinnitus. The patient referred her symptoms beginning immediately after suffering a head trauma occurred 3 months before. She also started to experience recurrent short-lasting vertigo attacks while quickly shifting from the supine to the sitting position or when performing sudden vertical head tilts during everyday activities. She reported intolerance toward loud sounds and dizziness while straining. Both brain CT and MRI scans were normal. Thus she was submitted to a detailed otoneurologic investigation. Otoscopy showed myringosclerosis in both atrophic eardrums. The right TM moved in time with her pulse rate but not with respiration, as it is shown in the animation (Video 2). Unlike patient 1, no variations of TM activity were induced by body-position changes or Valsalva maneuvers. No pathologic mass behind the eardrum could be seen. Pure-tone audiometry evidenced a right-sided mixed hearing loss with a mean mild-low frequency (0.25–1 kHz) ABG of 26.7 dB and a left sensorineural dip at 8 kHz. A 226 Hz Weber tuning fork test was lateralized to the right side. Tympanometry produced normal compliance curves and the measurement of right baseline impedance detected pulsating waveforms (Fig. 3). A transitory downbeating nystagmus with a counterclockwise torsional component (with respect to the observer's point of view) could be noted while performing a Valsalva maneuver with pinched nostrils. Bi-mastoid vibratory test induced analogous transitory eye movements. Standard caloric ear irrigations revealed a slight right canal hypo-reflectivity with a border-line value of CP (21%), whereas the vHIT detected normal high-frequency VOR gain for each semicircular canal. As for patient 1, right AC cervical-VEMPs were abnormally enlarged and with lowered threshold (90 dB SPL) compared

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