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Isolated horizontal canal hypofunction differentiating a canalith jam from an acute peripheral vestibular loss

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ABSTRACT

Objectives: To describe a unique case of acute vertigo presenting with spontaneous horizontal nystagmus (SHN) and a clinical picture consistent with right acute peripheral vestibular loss (APVL) in which an isolated hypofunction of a horizontal semicircular canal (HSC) permitted to detect a spontaneous canalith jam and treat the patient accordingly.
Methods: Case report and literature review.
Results: A 74-year old woman presented with acute vertigo, left-beating SHN and a clinical picture consistent with right APVL. Nevertheless, vestibular evoked myogenic potentials were normal with symmetrical amplitudes and the video head impulse test (vHIT) revealed an isolated hypofunction of the right HSC. After repeated head shakings, the supine roll test evoked bilaterally a positioning paroxysmal geotropic horizontal nystagmus suggesting benign paroxysmal positional vertigo involving the non-ampullated arm of the right HSC. vHIT and caloric testing confirmed restitution of HSC function after repositioning maneuvers.
Conclusions: In case of acute vertigo with SHN, a complete functional assessment of vestibular receptors and afferents should always be given in order to avoid misdiagnosis. Canalith jam should be considered in case of spontaneous nystagmus and isolated canal hypofunction.

1. Introduction

The differential diagnosis in case of acute vertigo with spontaneous horizontal nystagmus (SHN) can be challenging. It can result from lesions or dysfunctions affecting both central and peripheral vestibular pathways. An accurate bedside examination following the HINTS protocol has demonstrated to detect small lesions of the posterior fossa with higher accuracy than brain magnetic resonance imaging (MRI) within the first 48 h [1]. In fact, in case of gaze-evoked direction-changing nystagmus, skew deviation of the eyes and normal head impulse test (HIT), acute vertigo should be considered of central origin. Conversely, corrective saccades after passive horizontal impulses of the patient's head toward the opposite side of the fast-beating component of the SHN unequivocally indicate an acute peripheral vestibular loss (APVL) [1]. APVL could be due to a sudden functional loss of the vestibular nerve divisions (generally attributed to a viral reactivation) [2] or is supposed to follow an ischemic damage involving labyrinthine end-organs (likely a selective ischemia of a terminal branch of the internal auditory artery) [3]. Nowadays, thanks to the introduction of modern and fast tools for vestibular testing, information about the functional status of peripheral vestibular receptors and afferents can be easily provided. While cervical- and ocular-vestibular evoked myogenic potentials (cVEMPs and oVEMPs, respectively) test otolith organs and their afferents [4], semicircular canals function can be reliably evaluated with the video HIT (vHIT) in all canal planes [5]. The interpretation of data obtained combining the results of these tests provides a more accurate assessment of vestibular function, enabling the identification of specific patterns of lesion affecting inner ear sensors or vestibular nerve divisions [6]. In case of an unusual combination of instrumental findings in an acute setting, an alternative diagnosis to APVL should be investigated.

We present a unique case of acute vertigo with SHN in which an

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isolated hypofunction of a horizontal semicircular canal (HSC) among instrumental findings permitted to hypothesize a spontaneous canalith jam as possible underlying mechanism and treat the patient accordingly.

2. Case report

A 74-year old woman presented to the outpatient unit reporting an abrupt onset of acute vertigo with nausea and vomiting since one day prior to presentation. She denied auditory symptoms or previous complaints of vestibular symptoms. Vestibular examination with the patient upright revealed a left-beating SHN with a slow phase velocity (SPV) of 7°/s on video-oculography (VOG). The nystagmus showed peripheral features as it increased during left gaze, decreased during the gaze in the opposite direction and was suppressed by visual fixation. Intensity and direction of SHN remained unchanged during the head pitch test and the supine roll test. A positive clinical head impulse test (HIT) on the horizontal plane with refixation saccades was clearly detected by rapidly turning the patient's head to the right. Alternate cover test excluded signs of skew deviation, neither saccadic nor smooth pursuit abnormalities could be found at the bedside ocular motor testing. Neurological examination was otherwise unremarkable. The patient was diagnosed with a right APVL and submitted to an extensive otoneurologic evaluation to determine the amount of vestibular end-organs and afferents involved. Pure tone audiometry showed a mild and symmetrical high-tone sensorineural hearing loss consistent with the patient's age. Bithermal caloric test (BCT) at temperatures of 30 °C and 44 °C revealed a severe right unilateral weakness (Fig. 1A). Nevertheless, both cVEMPs and oVEMPs to air-conducted stimuli (500 Hz-tone bursts) showed symmetrical and normal amplitudes (cVEMPs: 56 μV on the right and 68 μV on the left, oVEMPs: 9 μV on the right and 7 μV on the left) and the vHIT revealed an isolated reduction of the vestibulo-ocular reflex (VOR) gain in the right HSC (Fig. 1B). This unusual selective deficit among instrumental finding leads us to hypothesize an alternative diagnosis to APVL. An isolated hypofunction of the right HSC due to a canalith jam obstructing the lumen of the membranous duct was postulated. Indeed, after repeated head shakings, the supine roll test elicited bilaterally a positioning paroxysmal geotropic horizontal nystagmus. Nystagmus was more severe during right head turning (SPV = 35°/s) than the left (SPV = 10°/s) suggesting a benign paroxysmal positional vertigo (BPPV) involving the non-ampullated arm of the right HSC. The patient was then submitted to canalith repositioning maneuvers (CRM) for HSC-BPPV [7] and was instructed to lay with the head on the healthy side for two days. When the patient returned to our clinic, her symptoms had subsided. Neither SHN nor positional nystagmus were observed on VOG, verifying the therapeutic success of CRM. BCT and vHIT demonstrated a complete functional recovery of the HSC VOR for both low- and high-frequency domains, respectively (Fig. 2). Brain MRI did not reveal any lesions in the cerebellum or brainstem.

3. Discussion

BPPV is considered the most common peripheral vestibular disorder. It is responsible of short-lasting vertigo spells triggered by changes in head position. Otoliths detachment from utricular macula perturbing semicircular canals micromechanics has been accepted as the underlying pathophysiological mechanism. Clinical features of BPPV depend on the semicircular canals involved and the location of debris within the membranous duct. Otoconia can be free to float causing a paroxysmal positioning nystagmus (canalolithiasis) or settle on the cupula resulting in a persistent positional nystagmus (cupulolithiasis). HSC can be involved in 10–20% of all BPPV cases. Particles can fall in the non-ampullated arm (geotropic variant) or in the ampullated arm of the HSC (apogeotropic variant). Several CRM have been proposed to treat each type of BPPV with a high success rate [7].

Canalith jam represents a rare condition, occurring when an otoliths clot plugs a narrow portion of the membranous duct causing a blockage

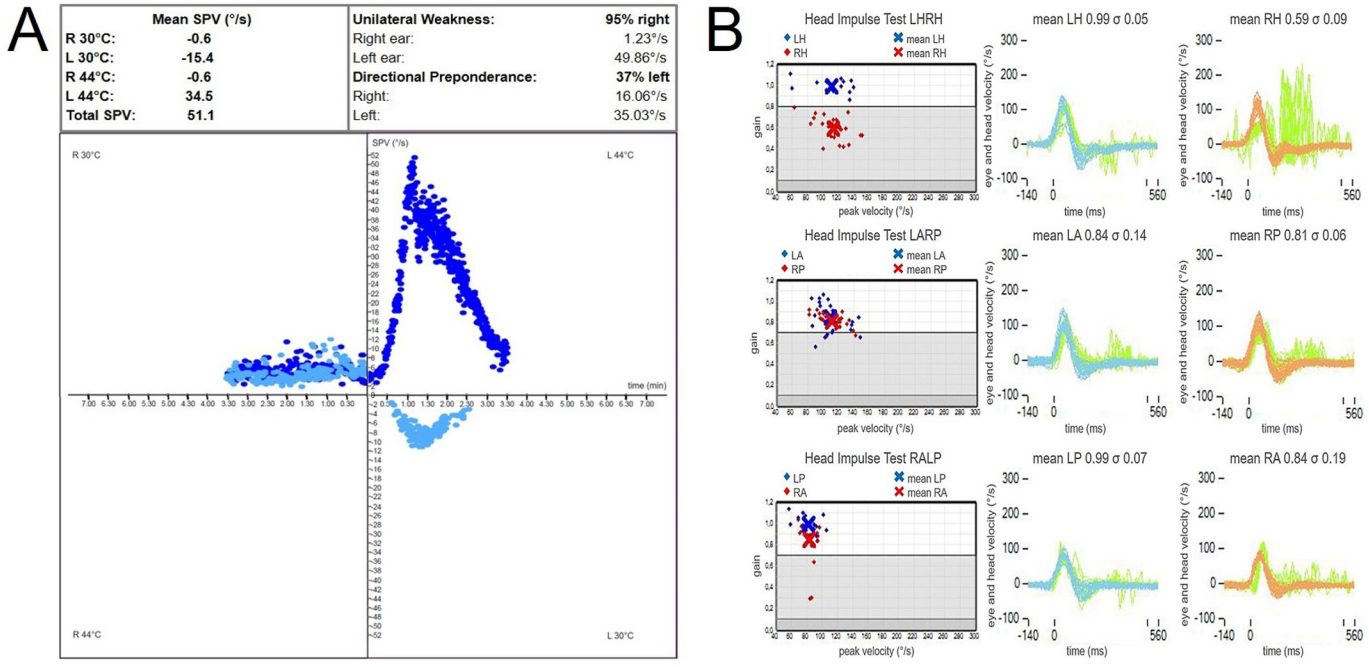

Fig. 1. Instrumental evaluation of semicircular canals function at presentation. (A) Standard bithermal caloric testing (30 and 44 °C ear irrigations), evaluated using a VNG system (model VN415/VO425 Firewire, Interacoustics A/S, Denmark). Jongkees formula demonstrated a severe right canal paresis (unilateral weakness 95%). L: left. R: right. SPV: slow phase velocity. (B) Video Head Impulse Test performed using an ICS Video-oculographic system (GN Otometrics A/S, Denmark) as reported by Halmagyi et al. [5]. Mean value of the vestibulo-ocular reflex (VOR) gain (eye velocity/head velocity) and corresponding standard deviation (σ) is reported for each canal. A selective deficient VOR gain for the right horizontal canal (0.59) with corrective saccades could be clearly observed. LH: left horizontal, RH: right horizontal, LA: left anterior, RP: right posterior, LP: left posterior, RA: right anterior.

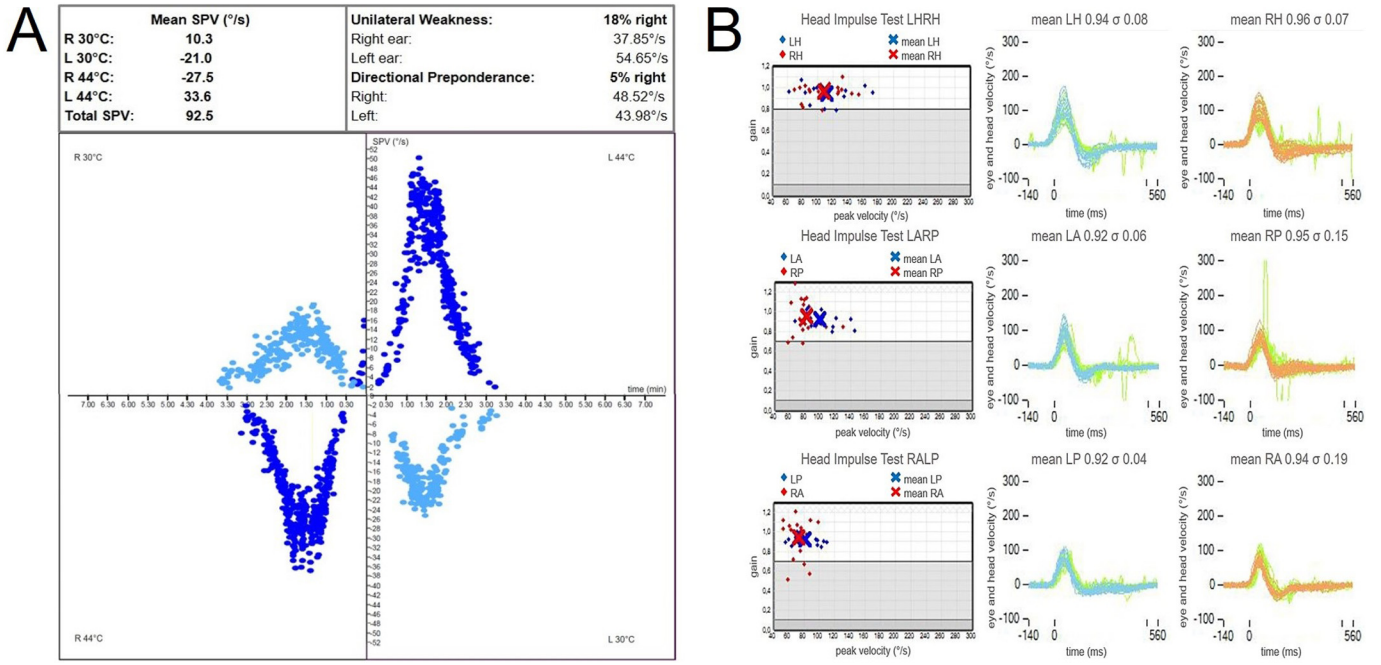

Fig. 2. Instrumental assessment of semicircular canals after canalith repositioning maneuvers. (A) Caloric testing documented a complete recovery of the right side paresis with a value of unilateral weakness within normal range (18%). (B) Even the video Head Impulse Test supported a functional recovery of the right horizontal canal with a vestibulo-ocular reflex gain of 0.96.

of the endolymphatic flow. In this condition, a continuous alteration of hydrostatic pressure between the otoliths clump and the cupula occurs, leading to a persistent deflection of the cupula itself. It results in a spontaneous nystagmus regardless of head position [8]. Cases of canalith jam of the HSC have already been described in literature, occurring spontaneously [9,10] or following CRM [11–13]. Vigorous head shakings or skull vibrations have demonstrated to release otoconia from the clot and mobilize them in the canal lumen [8–13].

Spontaneous nystagmus can also be observed in patients with HSC-BPPV. Nevertheless, in these cases it develops because the natural canal inclination of 30° relative to the horizontal plane in the upright position results in a gravity-induced slow falling of free-floating debris or in a persistent deflection of the overloaded cupula [14]. Due to its gravity dependency, SHN in HSC-BPPV usually changes direction with the head pitch test and the supine roll test; therefore, it should be more properly called “pseudo-spontaneous nystagmus” [15]. Conversely, the patient of our report showed a persistent direction-fixed nystagmus insensitive to gravity. A continuous bending of the cupula due to a spontaneous canalith jam, rather than an acute suppression of activity of right vestibular afferents as in case of APVL, more likely accounts for this clinical feature. Several observations support this theory.

First, presenting left-beating SHN that did not change with positioning test could be likely explained by the persistent utriculofugal displacement of the cupula due to the negative pressure between the otolith clot and the cupula itself (Fig. 3A), as already proposed in literature [11]. Subsequent direction-changing paroxysmal geotropic horizontal nystagmus elicited by the supine roll test after repeated head shakings indicates that the debris have been mobilized in the non-ampullated arm of the right HSC (Fig. 3B). Furthermore, in case of APVL with SHN, a global vestibular loss (due to a complete damage of vestibular nerve branches) or a partial vestibular hypofunction sparing saccular and posterior canal activity (as in case of selective lesions involving the superior nerve division or ischemia of the territories supplied by the anterior vestibular artery) would be rather expected than an isolated HSC hypofunction as reported here [2,3,6]. This peculiar instrumental pattern of lesion in an acute setting has so far never been described. A similar picture, even though without an instrumental evaluation of macular receptors, has been already reported in four cases with slight, selective HSC damage fully recovered over time [16]. In our patient, the transient HSC hypofunction, restored after CRM, would be less likely to occur if vestibular receptors had been damaged. It should be more properly considered a high- and low-frequency “pseudo-hyporeflexia”, consistently with previous findings after selective canal plugging on humans and animals using high-acceleration head impulses [17] and caloric testing [18]. Recovery of canal function after CRM has been already reported in patients with canalith jam of the HSC [9–12]. Nevertheless, vertical canals function has never been instrumentally tested in these cases. Our report is unique in literature since a spontaneous canalith jam affecting the right HSC could have been hypothesized and treated on the basis of an isolated HSC hypofunction detected evaluating all vestibular sensors and afferents. Though presenting signs where consistent with an APVL, no plausible combination of end-organs lesion could have been responsible for instrumental data and following findings.

4. Conclusions

To the best of our knowledge, an isolated HSC hypofunction detected evaluating all vestibular receptors in an acute setting has never been described before. This case report is unique since the unusual pattern of lesion represented the key for differentiating between a canalith jam and an APVL. An extensive evaluation of all vestibular sensors and afferents should always be given in case of acute vertigo with SHN to avoid misdiagnosis. In fact, if an accurate vestibular assessment had been neglected, the patient would have been improperly diagnosed with APVL and treated accordingly basing on the results of the clinical examination. A canalith jam should always be considered in case of spontaneous nystagmus and isolated canal hypofunction on vestibular assessment.

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None.

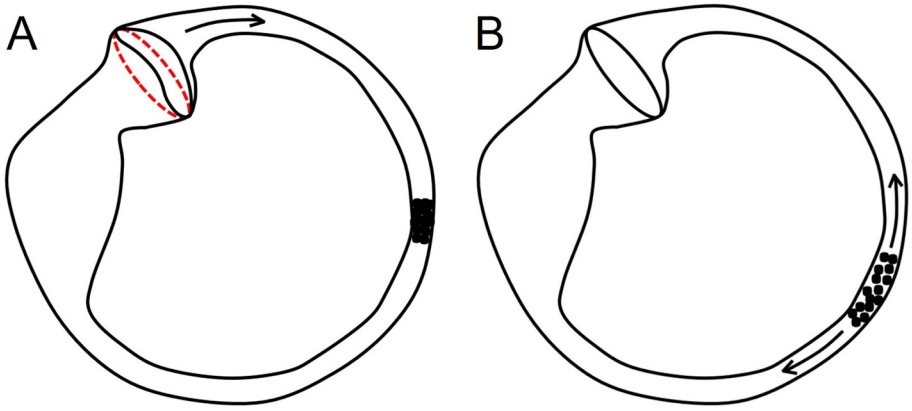

Fig. 3. Proposed mechanism for presenting left-beating spontaneous horizontal nystagmus (SHN) with right horizontal semicircular canal (HSC) hypofunction and for direction-changing positioning paroxysmal nystagmus at the supine roll test after head shakings. (A) An otoliths clot entrapped within the membranous duct could likely exert a negative pressure (arrow) on the cupula preventing dynamic responses of the right HSC by blocking endolymphatic flow. The cupula is shown deflected utriculofugally and the red dashed line indicates its original position. This condition resulted in a left-beating SHN insensitive to gravity. (B) After repeated head shakings, debris were mobilized. Otoconia are shown free to float toward the ampulla or in the opposite direction (arrows) within the membranous duct, resulting bilaterally in a positioning paroxysmal horizontal nystagmus at the supine roll test. Since direction-changing nystagmus was geotropic and more severe during right head turning, otoconia likely fell in the non-ampullated arm of the right HSC. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

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