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Case Report

Bilateral Vestibular Atelectasis as a Fluctuating Bilateral Vestibulopathy: A Case Report Care

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The vestibular atelectasis (VA) is a very rarely reported clinical and radiological condition characterized by the collapse of the labyrinthine membrane. Clinical features include symptoms which mimic endolymphatic hydrops; bilateral forms are rarely reported in the literature. Tullio's sign was reported in a few cases. In this paper, we report an original case of bilateral fluctuating VA associated with a rare posterior semicircular canal (SSC) dehiscence variant. A 54-year-old patient presented with evolving and fluctuating bilateral vestibulopathy associated with a pressure-induced nystagmus. Vestibular assessment revealed a bilateral vestibulopathy affecting low and mid velocities, with fluctuating high-velocity impairment. Exploration of the otolithic system showed preserved saccular activity and an absence of utriculo-ocular response. Radiological evaluation demonstrated bilateral VA and a dehiscence of the right posterior SSC. We provide clinical elements to better understand this entity, and propose a new hypothesis for the presence of a previously reported Tullio sign in VA.

KEYWORDS: Vestibular atelectasis, valsalva, bilateral vestibulopathy, otic capsule dehiscence

INTRODUCTION

The vestibular atelectasis (VA) is a very rarely reported condition, characterized by the collapse of the labyrinthine membrane, therefore being from an anatomopathological point of view the opposite of endolymphatic hydrops.[1] This collapse seems to primarily affect the "pars superior" of the vestibular end organ, including the utricular organ and/or the semicircular ampullae. By contrast, endolymphatic hydrops mostly appears to primarily affect the "pars inferior," including the cochlea and the saccule. Clinical features mimic those of endolymphatic hydrops, although there are very rare or no cochlear symptoms. Some bilateral forms were reported in the literature.[2,3] Tullio's phenomenon has been sparsely reported in a few cases, in which a *superior* semicircular canal (SSC) dehiscence was also diagnosed.[2-5] In this paper, we report the first case of bilateral fluctuating VA associated with a rare variant of otic capsule dehiscence (*posterior* SSC dehiscence).

Case Presentation

Written and verbal informed consent was obtained from the patient.

A 54-year-old patient was referred to our audiology and otoneurology department in Lyon for recurrent vertigo lasting approximately 24 hours, accompanied by significant neurovegetative symptoms and persistent unsteadiness between the episodes. No auditory symptoms were associated with the attacks (e.g., tinnitus, ear fullness, and/or hearing loss). He denied any history of headaches, including migraines. He described the same complaints 10 years earlier with recurrent rotatory vertigo with no auditory symptoms. No positive diagnosis was made at that time but most of his symptoms were alleviated by vestibular/physical therapy.

Vestibular examination demonstrated a vibration-induced nystagmus (VIN) when stimulating the vertex and left mastoid process with a 100 Hz-vestibular vibrator (Synapsys, France). The VIN had a horizontal component beating to the right side at 3.7°/s and a

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vertical component beating downwards at 3.2°/s. No spontaneous or gaze-evoked nystagmus was detected. Videonystagmography (Synapsys, France) revealed severe bilateral vestibular hyporeflexia (<4°/s) with the caloric tests. The Video-Head Impulse Test (v-HIT) (Synapsys, France) showed vestibulo-ocular reflex (VOR) gain impairment in the 3 SSC on the left side. The right posterior SSC was slightly impaired. Ocular vestibular-evoked myogenic potentials were absent, but cervical vestibular-evoked myogenic potentials (VEMPs) were preserved bilaterally. Pure tone audiometry revealed normal and symmetrical hearing thresholds.

So, we first proposed an "anti-hydrops" treatment with betahistine 24 mg twice a day and requested a cerebral magnetic resonance imaging (MRI) with delayed fluid-attenuated inversion recovery (FLAIR) sequences at 4 hours after gadolinium injection. A few weeks later, the patient self-recorded and shared with the medical team (as instructed previously) a few video clips which were taken during 2 episodes of rotatory vertigo crisis. A pure downbeating nystagmus was observed. He was also able to identify the "nose blowing effort" as a trigger factor for brief episodes of dizziness. Therefore, pressure sensitivity maneuvers were carried out further in our department. The pinched nostrils Valsalva maneuver revealed a symptomatic upbeating nystagmus (with a slight horizontal component), which reversed upon pressure release. No nystagmus was induced by the closed glottis Valsalva maneuver. The Tullio test (with pure tones at intensities ranging from 85 to 110 dB HL in each ear at 1 kHz) was negative.

A high-resolution computed tomography (HRCT) scan of the temporal bones was performed and revealed a limited dehiscence of the right posterior SSC (Figure 1). Fluid-attenuated inversion recovery magnetic resonance imaging demonstrated bilateral VA, much clearer on the utricle and the ampullae of the 3 SSC on the left side (Figure 2).

Clinical follow-up was conducted over several months. We observed a progressive deterioration of the VOR gain on both sides. However, VOR gain improved for all SCC 8 months after the first consultation (Figure 3). At that point, the patient reported no other new vertigo episodes for weeks and also indicated being less sensitive to "nose blowing."

DISCUSSION

The concept of VA was first introduced by Merchant and Schuknecht in 1988.[1] They demonstrated the existence of a collapse in the "pars superior" of the membranous labyrinth. The collapse of the walls of the vestibular labyrinth could disrupt the functioning of the vestibular sensory organs through the displacement or compression of the cupulae and otolithic membranes. Two primary presentation patterns in VA were described: in the paroxysmal form, patients experience sudden episodes of vertigo without accompanying auditory or neurological symptoms. In contrast, the insidious form displays a more gradual onset. In both scenarios, the initial occurrence is followed by persistent instability or sporadic episodes of vertigo.

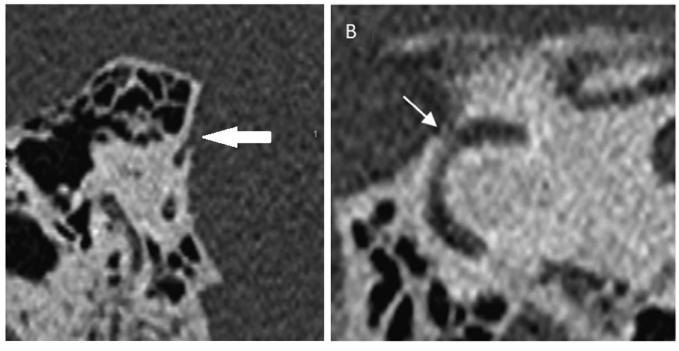

Figure 1. A. High-resolution computed tomography imaging of the temporal bones showing a dehiscence of the right posterior semicircular canal in a sagittal plane (A). Multiplanar reconstruction plane of the right posterior semicircular canal (B).

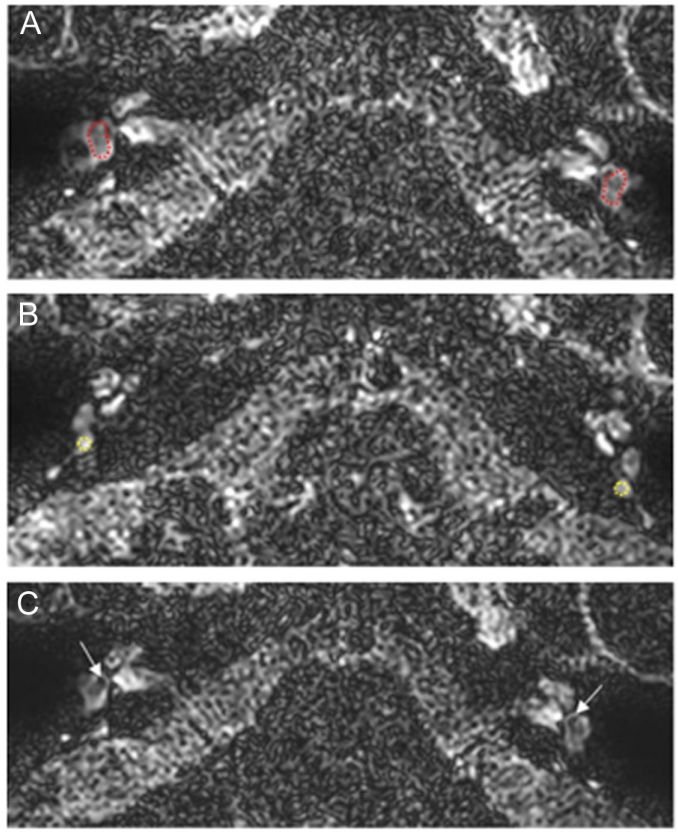

Figure 2. (A) Delayed FLAIR MRI 4 hours post-injection gadolinium in axial plane, showing a bilateral VA with bilateral utricular atelectasis (dotted red circle, A). (B) Bilateral atelectasis of the posterior semicircular canal ampullae (dotted yellow circle, B). (C) Bilateral atelectasis sparing the saccule bilaterally (white arrow, C).

Since this pioneering study, a few cases of VA were published in the literature, contributing to the support of this new clinical entity.[2-4] In a recent study, Eliezer et al[5] presented a case series involving patients with unilateral vestibular loss with a unilateral collapse of the pars superior on delayed post-contrast MRI.

These findings were supported by a second clinical study focusing on clinical and electrophysiological characteristics of patients with unilateral VA confirmed by a FLAIR delayed post-contrast MRI;[6] in a series of 22 patients, there were electrophysiological arguments for utricular dysfunction (90% of cases) and saccular preservation (77% of cases). All patients showed canal function impairment at low velocities (with partial or complete paresis in caloric tests) as well as at high velocities (involving 1, 2, or all SSC).

In this study, the clinical presentation is consistent with the cases of VA previously reported. The patient complained of recurrent sudden

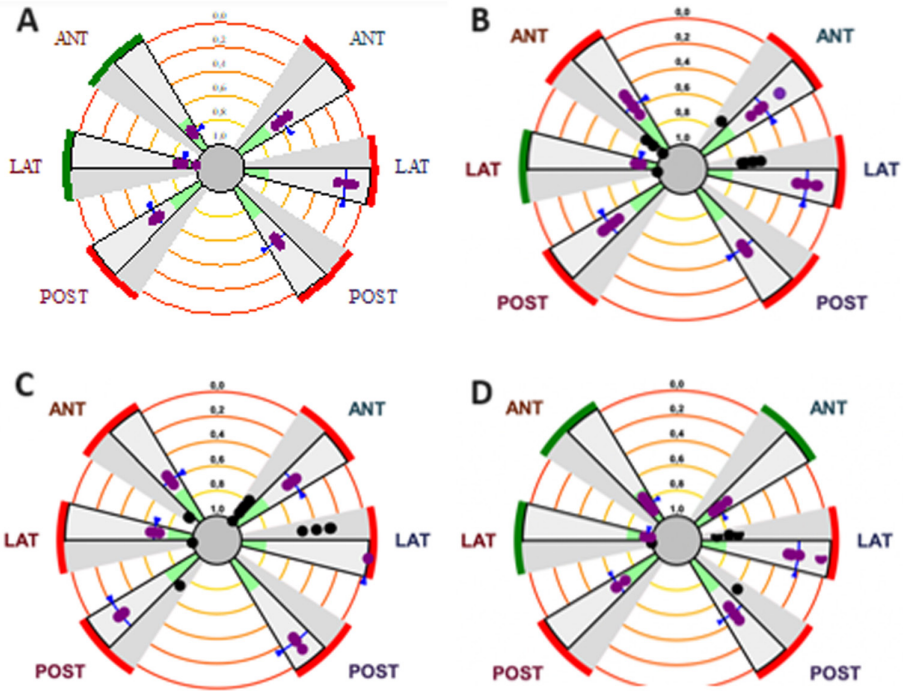

Figure 3. Video-head impulse test gain evolution (8 months follow up). A minimum of 5 acquisitions was performed for each canal.

episodes of vertigo with no auditory symptoms, followed by a persistent unsteadiness ameliorated by vestibular rehabilitation. The notable aspect of this case resides in the sequential onset, with bilaterality of atelectasis occurring years later, and in the fluctuating nature of the vestibulopathy, involving all the SSCs. Fluctuating vestibulopathy is usually associated with pressure-related pathologies of the inner ear, in particular, the endolymphatic hydrops, which is the histopathological hallmark of Menière's disease. In this condition, the disturbance of the endolymph compartment consists of the distension of the endolymphatic spaces involving the "inferior pars" of the membranous labyrinth, including the cochlea and the saccule, with a potential progression towards the entire vestibular organ. Contrary to endolymphatic hydrops, VA consists of the collapse of endolymphatic spaces, which primarily affects the "superior pars," involving the utricule and/or the SSC system. The cases of VA previously reported suggested that v-HIT may be deficient. This would clinically distinguish it from endolymphatic hydrops where v-HIT mostly remains unaffected.[7]

A concomitant, clinically intriguing element in our case is the simultaneous ipsilateral presence of a symptomatic posterior SSC dehiscence. The features of the nystagmus induced by the pinched nostrils Valsalva maneuver are well explained by the dehiscence: increased pressure in the middle ear might cause an "inward" displacement of the stapes, generating a pressure wave in the endolymphatic compartment towards the area of lower impedance. As this induced endolymphatic flow is ampullofugal, the second law of Ewald states that it leads to an excitatory response in the hair cells of the ampullary canal, resulting in an upward vertical nystagmus, theoretically accompanied by a clockwise or counterclockwise component.

Another case of VA with a Tullio phenomenon was published recently. The authors did not attribute the Tullio phenomenon (as this is assumed to be an important sign of the third window syndrome)[8] to a superior SSC dehiscence, since the HRCT and VEMPs did not support this diagnosis.[9] Wenzel et al[4] related sound or pressure-induced nystagmus in 4 VA cases with no radiological sign of superior SSC dehiscence. They hypothesized that the collapse of the membranous labyrinth may bring the stapes closer to the vestibule and therefore facilitate the stimulation of the vestibular organ by auditory or pressure stimulation.

In the present case, Valsalva's maneuver induced a specific nystagmus whose characteristics correspond to a symptomatic posterior SSC dehiscence. These findings raise the question of whether atelectasis is a true contributor to pressure-induced vertigo as described in the cases reported in the literature. Even if the previously mentioned cases did not confirm a symptomatic superior SSC dehiscence, it is important to remember that there are multiple locations of otic capsule dehiscence—including posterior SSC—which still remain underdiagnosed due to limited awareness.[10] Further, Ward et al[11] introduced the notion of near-dehiscence as an extreme thinning of the bone layer that may act similarly to a "true" symptomatic dehiscence.

Finally, although there is growing evidence in the literature supporting this new clinical entity, the underlying mechanism for VA remains unclear. A plausible pathophysiological explanation is based on the dysfunction of Bast's valve, previously mentioned for endolymphatic hydrops.[12] Alternative genetic hypotheses have been proposed for the physiopathological models of endolymphatic hydrops, which can be extended to atelectasis. This theory involves membrane proteins such as the cotransporter NKCC1 found in dark cells, or aquaporin ionic channels.[13,14]

This article provides new clinical elements, helping to better understand this entity which is still far too poorly described and understood. It also suggests a possible new explanation for the association

of a Tullio phenomenon, already reported previously in a few cases of VA, since the link between these pathologies is not clear.

Availability of Data and Materials: The data presented in this paper are available on request from the corresponding author. The data are not publicly available due to ethical, legal, and privacy issues.

Informed Consent: Verbal and written informed consent was obtained from the patient who agreed to take part in the study.

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REFERENCES

1. Merchant SN, Schuknecht HF. Vestibular atelectasis. *Ann Otol Rhinol Laryngol*. 1988;97(6 Pt 1):565-576. [CrossRef]
2. Finn S, Dietzek M, Karvouniari P, et al. Bilateral vestibulopathy with positive Tullio phenomenon. *Laryngoscope*. 2018;128(5):1223-1225. [CrossRef]
3. Roy S, Kumarasamy G, Salim R. Bilateral vestibular atelectasis with Tullio's phenomenon. *Indian J Otolaryngol Head Neck Surg*. 2019;71(suppl 2):1599-1602. [CrossRef]
4. Wenzel A, Ward BK, Schubert MC, et al. Patients with vestibular loss, tullio phenomenon, and pressure-induced nystagmus: vestibular atelectasis? *Otol Neurotol*. 2014;35(5):866-872. [CrossRef]
5. Eliezer M, Attyé A, Guichard JP, et al. Vestibular atelectasis: myth or reality? *Laryngoscope*. 2019;129(7):1689-1695. [CrossRef]
6. Marc M, Hautefort C, Guichard JP, et al. Clinical characteristics in unilateral vestibular atelectasis. *J Neurol*. 2021;268(2):689-700. [CrossRef]
7. Hannigan IP, Welgampola MS, Watson SRD. Dissociation of caloric and head impulse tests: a marker of Meniere's disease. *J Neurol*. 2021;268(2):431-439. [CrossRef]
8. Minor LB, Solomon D, Zinreich JS, Zee DS. Sound- and/or pressure-induced vertigo due to bone dehiscence of the superior semicircular canal. *Arch Otolaryngol Head Neck Surg*. 1998;124(3):249-258. [CrossRef]
9. Maslovara S, Butkovic-Soldo S, Pajic-Matic I, Sestak A. Vestibular atelectasis: decoding pressure and sound-induced nystagmus with bilateral vestibulopathy. *Laryngoscope*. 2019;129(7):1685-1688. [CrossRef]
10. Reynard P, Idriss S, Ltaief-Boudrigua A, et al. Proposal for a unitary anatomo-clinical and radiological classification of third mobile window abnormalities. *Front Neurol*. 2021;12:792545. [CrossRef]
11. Ward BK, Wenzel A, Ritzl EK, et al. Near-dehiscence: clinical findings in patients with thin bone over the superior semicircular canal. *Otol Neurotol*. 2013;34(8):1421-1428. [CrossRef]
12. Hofman R, Segenhout JM, Buytaert JA, Dirckx JJ, Wit HP. Morphology and function of Bast's valve: additional insight in its functioning using 3D-reconstruction. *Eur Arch Otorhinolaryngol*. 2008;265(2):153-157. [CrossRef]
13. Hibino H, Kurachi Y. Molecular and physiological bases of the K+ circulation in the mammalian inner ear. *Physiology (Bethesda)*. 2006;21:336-345. [CrossRef]
14. Teggi R, Carpini SD, Zagato L. Endolymphatic hydrops and ionic transporters: genetic and biohumoral aspects. *J Neurol*. 2019;266(suppl 1):47-51. [CrossRef]