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Case Report

Intralabyrinthine Schwannoma Mimicking Posterior Canalithiasis

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Intralabyrinthine schwannomas are a rare subgroup of vestibular schwannomas located within the membranous labyrinth of the inner ear and are known for their variable clinical presentations and symptoms. In the present study, we report on a patient with a persistent history of dizziness and positional vertigo, who was misdiagnosed with posterior canalithiasis. As hearing loss was not developed until late in the disease course, the patient was not properly diagnosed until magnetic resonance imaging revealed an intralabyrinthine schwannoma, which was not discovered on earlier imaging. In addition to the unusual clinical presentation, we describe the audio-vestibular profile of our patient. We suggest that a thorough vestibular evaluation, including caloric testing and a careful examination of the inner ear on imaging, is warranted in cases of treatment of refractory vertigo, even in patients where a diagnosis seems certain.

KEYWORDS: Acoustic neuroma, BPPV, canalithiasis, hearing loss, intralabyrinthine schwannoma, TRV chair

INTRODUCTION

Vestibular schwannomas (VS) are benign tumors arising from the Schwann cells of the eighth cranial nerve. The common symptoms of VS are unilateral sensorineural hearing loss (SNHL), tinnitus, and mild disequilibrium. Vestibular schwannomas are found in the internal auditory canal (IAC) sometimes extending extrameatally into the cerebello-pontine angle (CPA), and the diagnosis is established using magnetic resonance imaging (MRI) of the posterior fossa and IAC.[1] A rare subgroup of VS is intralabyrinthine schwannomas (ILSs) that emerge from the most lateral part of the vestibulocochlear nerve within the membranous labyrinth.[2] As per the Kennedy classification, ILSs can be further subdivided according to the location of the tumor within the labyrinth.[3]

Clinically, ILSs may masquerade as Ménière's disease (MD).[2,4] Some ILS patients describe episodic spontaneous vertigo and fluctuating hearing loss resembling the symptomatology of MD.[4] Indeed, ILSs have been identified as the cause of audio-vestibular symptoms in some patients previously diagnosed with MD.[5,6]

In this paper, we describe an atypical case of a patient diagnosed with an ILS mimicking posterior semicircular canal canalithiasis (P-CAN). In addition to the clinical presentation, we describe the vestibular profile of our ILS patient and discuss the difficulty often encountered in diagnosing ILS and the unusual case of canalithiasis as a differential diagnosis of VS/ILS.

CASE PRESENTATION

A 31-year-old female patient consulted her private otolaryngologist with complaints of dizziness triggered by loud sounds and fullness of the right ear. She was referred to her local ENT department on suspicion of a right-sided perilymphatic fistula. Subsequent fistula test turned out negative, alongside a negative Dix–Hallpike test, and both MRI and computer tomography findings were seemingly normal. Audiological examination showed a bilateral positive Rinne's test and lateralization to the left ear on Weber's test; however, her audiometry displayed normal hearing. In addition, the video head impulse test (VHIT) evaluating the vestibulo-ocular reflex was also performed and showed no vestibular deficit.

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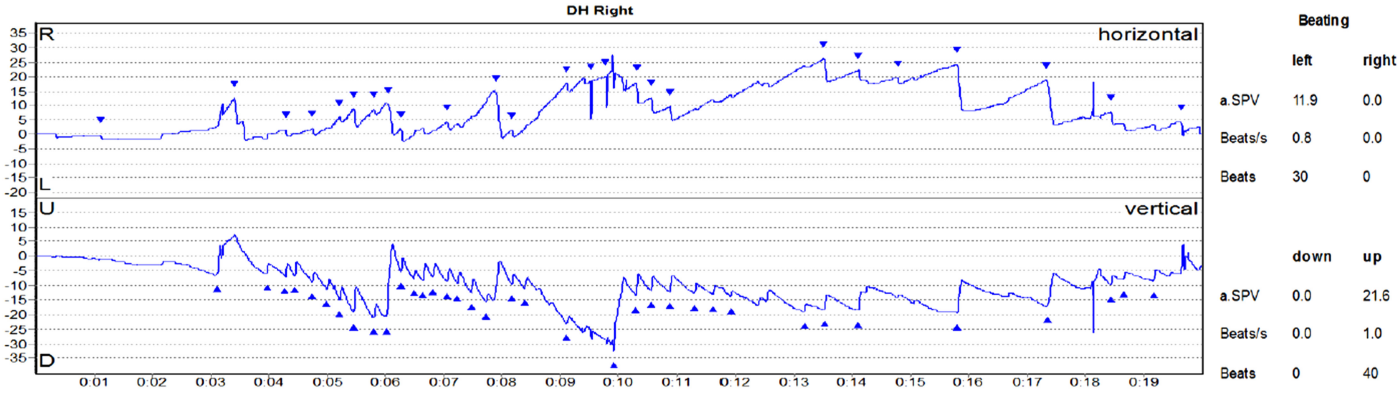

Figure 1. Videonystagmography recording during right Dix–Hallpike revealed a fatigable, geotropic up-beat nystagmus with little latency, compatible with right posterior semicircular canal canalithiasis.

Over the next 4 years, her symptoms worsened as she developed positional dizziness when resting on her right side, and she was diagnosed with benign paroxysmal positional vertigo (BPPV). The patient received numerous treatments in a TRV chair for BPPV, but no lasting relief of her symptoms was observed.

Eventually, she was referred to our national referral center, now also suffering from recurrent attacks of severe rotatory dizziness along with complaints of right-sided hearing loss and tinnitus. During videonystagmography, the patient exhibited positional nystagmus when placed in the Dix–Hallpike position (Figure 1), which along with the clinical presentation of positional vertigo suggested P-CAN. A new audiometry was performed at our department, which confirmed right-sided hearing loss (Figure 2).

Her asymmetrical hearing loss coupled with treatment refractory BPPV necessitated a repeat MRI, which revealed a 3 mm mass lesion in the vestibule of her right inner ear (Figure 3), providing a final diagnosis of ILS of the intravestibular subtype. Subsequent caloric irrigation showed 100% right canal paresis (Figure 4a), and a new VHIT displayed a pathological vestibulo-ocular reflex gain and presence of covert saccades on testing of the right lateral semicircular canal (LSCC) (Figure 4b).

The patient was counseled about the possibility of surgery for tumor resection, but she opted for the wait-and-scan approach and the hearing loss was effectively managed using a conventional hearing aid while she received vestibular rehabilitation therapy to combat dizziness. Informed consent to publish was obtained from the patient.

DISCUSSION

In this case report, we present an abnormal case of ILS mimicking otolith disease represented by P-CAN. The ILS patient had a 4-year history of slowly progressing vestibular symptoms and did not develop hearing loss until late in the disease course, leading to a

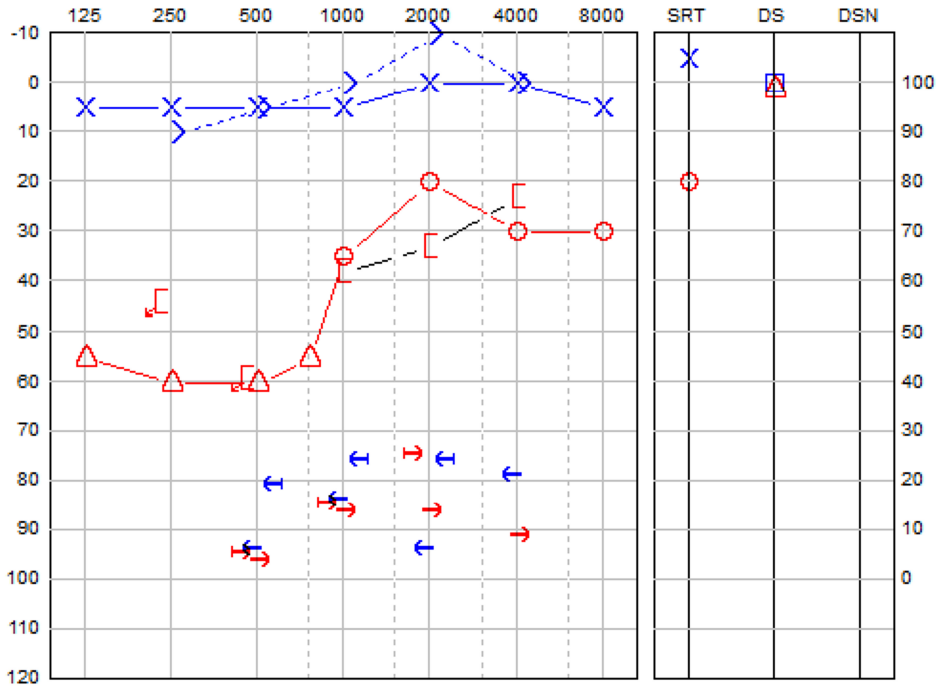

Figure 2. Pure-tone audiometry and word discrimination score testing performed late in the disease course in our center showed a right-sided sensorineural hearing loss, mostly pronounced in the low frequencies.

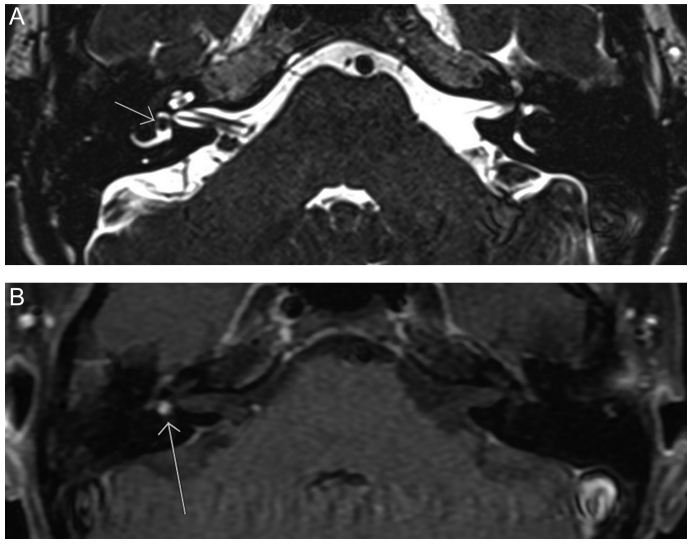

Figure 3. A, B. Magnetic resonance imaging of our patient showing a T2-weighted SPACE sequence without contrast (A) and a contrast-enhanced T1-weighted sequence (B), both revealing a 3 mm lesion in the vestibule of the right inner ear.

new MRI and the correct diagnosis. The diagnostic delay between the first clinical sign and her diagnosis is explained in part because her schwannoma was undetected on her first MRI and in part because her clinical presentation strongly suggested P-CAN. The case illustrates how ILS can be a difficult diagnosis to make, which is in concordance with the findings of Dubernard et al,[7] who in their multicenter study of 110 ILS cases identified an average 6-year delay between the first clinical sign and final diagnosis of ILS. Likewise, in a systematic review including 72 studies and a total of 234 ILS cases, Van Abel et al[8] found an average delay of 7 years between symptom onset and diagnosis.

Allowing for several differential diagnoses, the heterogeneous clinical presentation of ILS complicates tumor identification as the cause of audio-vestibular symptoms. Positional vertigo in ILS patients has been observed before,[2] but the presenting case is unique, as BPPV was diagnosed in a repositioning chair using videonystagmography goggles, regarded as the gold standard within BPPV diagnostics,[9] allowing the recording of a reproducible canalithiasis-like nystagmography pattern in the Dix–Hallpike maneuver (Figure 1).

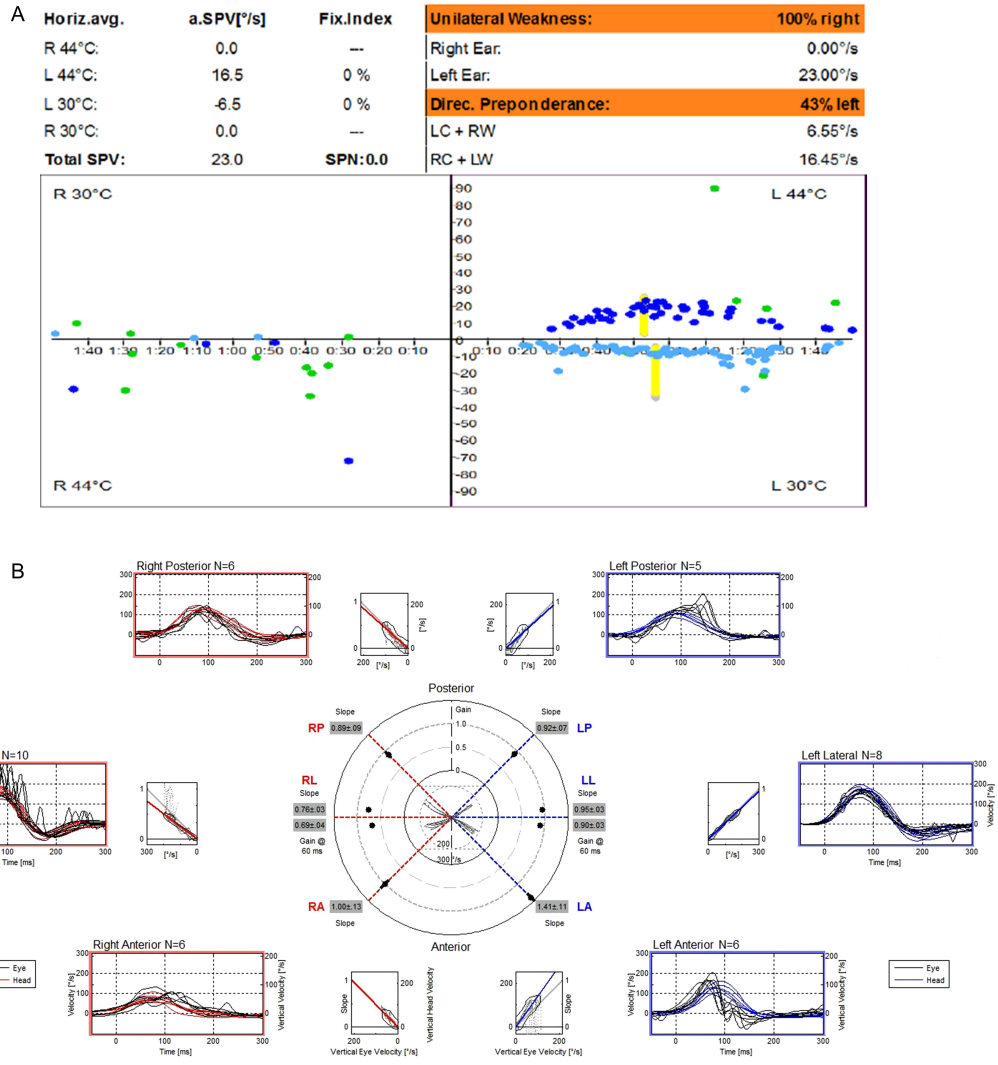

Figure 4. A, B. Caloric irrigation during videonystagmography (A) showed a right-sided slow-phase velocity of 0 degrees/second for both warm and cold water, indicating a 100% right canal paresis, and video head impulse test (B) of the vestibulo-ocular reflex of all semicircular canals showed a 0.69 gain and presence of covert saccades on evaluation of the right lateral semicircular canal.

Regarding hearing, ILS has also been reported to present as a mixed hearing loss rather than a pure sensorineural hearing loss and as both sudden, progressive, and fluctuating.[3,8] Intralabyrinthine schwannomas may also mimic otosclerosis when presenting with a pseudo-conductive hearing loss.[10]

The explanation for this lack of specificity in symptoms of ILS may be attributed to the exact location of the tumor within the labyrinth. It would be reasonable to assume that a localized mass effect on the different sensory end-organs of the inner ear would produce a variable clinical picture, including positional vertigo mimicking BPPV.[2] In their review, Van Abel et al[8] found a strong association between vertigo and tumors localized in the vestibular part of the labyrinth, suggesting at least a partial location effect of the tumor mass.

It is therefore important that the otologist keeps in mind the varied ways in which ILS may present itself depending on the affected sensory end-organs and consider the possibility of an ILS in cases of intractable, positional, or Ménière's like vertigo, especially when coinciding with unilateral hearing loss.

As evident on the MRI, the intravestibular lesion in our case was found in close proximity to the utriculus and the LSCC on the right ear. Thus, a localized mass effect acting upon the right LSCC could explain the pathological gain and presence of covert saccades on VHIT of the LSCC, as well as the 100% canal paresis of the LSCC on caloric testing. The P-CAN-like vertigo experienced by our patient could be hypothesized to be the cause of a herniation effect of the schwannoma triggered in the supine Dix–Hallpike position. In addition to direct nerve compression of the schwannoma, endolymphatic hydrops and metabolic changes of the endolymph caused by the tumor may also account for the pathophysiology of hearing loss and imbalance experienced by our and other ILS patients.[2,8,11] If caloric testing of our patient had been performed earlier in the disease course, it is likely that the diagnosis of posterior canalithiasis would have been discarded, permitting an earlier diagnosis of ILS.

Magnetic resonance imaging is the gold standard for diagnosing vestibular schwannomas, including ILS. However, because of their rarity and small sizes, intralabyrinthine lesions may elude detection on MRI, making underreporting likely.[12] Upon suspicion of a retrocochlear disorder, special attention to imaging should not only be given to the posterior fossa and IAC but also to the labyrinth, and contrast MRI should be favored. Furthermore, ILS alongside VS should be suspected in cases of refractory vertigo, as discussed above. Our case stresses the importance of careful examination of the inner ear on contrast MRI as well as a thorough vestibular assessment, including caloric testing, of a patient with long-lasting vestibular complaints.

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

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Declaration of Interests: The authors have no conflicts of interest to declare.

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REFERENCES

1. Dang L, Tu NC, Chan EY. Current imaging tools for vestibular schwannoma. *Curr Opin Otolaryngol Head Neck Surg*. 2020;28(5):302-307. [CrossRef]
2. Slattery EL, Babu SC, Chole RA, Zappia JJ. Intralabyrinthine schwannomas mimic cochleovestibular disease: symptoms from tumor mass effect in the labyrinth. *Otol Neurotol*. 2015;36(1):167-171. [CrossRef]
3. Kennedy RJ, Shelton C, Salzman KL, Davidson HC, Harnsberger HR. Intralabyrinthine schwannomas: diagnosis, management, and a new classification system. *Otol Neurotol*. 2004;25(2):160-167. [CrossRef]
4. Lee SU, Bae YJ, Kim HJ, et al. Intralabyrinthine schwannoma: distinct features for differential diagnosis. *Front Neurol*. 2019;10:750. [CrossRef]
5. Choudhury B, Carlson ML, Jethanamest D. Intralabyrinthine schwannomas: disease presentation, tumor management, and hearing rehabilitation. *J Neurol Surg B Skull Base*. 2019;80(2):196-202. [CrossRef]
6. Neff BA, Willcox TO Jr, Sataloff RT. Intralabyrinthine schwannomas. *Otol Neurotol*. 2003;24(2):299-307. [CrossRef]
7. Dubernard X, Somers T, Veros K, et al. Clinical presentation of intralabyrinthine schwannomas: a multicenter study of 110 cases. *Otol Neurotol*. 2014;35(9):1641-1649. [CrossRef]
8. Van Abel KM, Carlson ML, Link MJ, et al. Primary inner ear schwannomas: a case series and systematic review of the literature. *Laryngoscope*. 2013;123(8):1957-1966. [CrossRef]
9. West N, Hansen S, Møller MN, Bloch SL, Klokker M. Repositioning chairs in benign paroxysmal positional vertigo: implications and clinical outcome. *Eur Arch Otorhinolaryngol*. 2016;273(3):573-580. [CrossRef]
10. Reda JD, West N, Cayé-Thomasen P. Intracochlear vestibular schwannoma presenting with mixed hearing loss. *J Int Adv Otol*. 2021;17(3):265-268. [CrossRef]
11. Elias TGA, Perez Neto A, Zica ATS, Antunes ML, Penido NO. Different clinical presentation of intralabyrinthine schwannomas - a systematic review. *Braz J Otorhinolaryngol*. 2019;85(1):111-120. [CrossRef]
12. Salzman KL, Childs AM, Davidson HC, Kennedy RJ, Shelton C, Harnsberger HR. Intralabyrinthine schwannomas: imaging diagnosis and classification. *AJNR Am J Neuroradiol*. 2012;33(1):104-109. [CrossRef]