Ocular vestibular evoked myogenic potentials and intravestibular intralabyrinthine schwannomas

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Abstract

Intravestibular intralabyrinthine schwannomas (ILSs) are uncommon benign tumors that arise from the saccular, utricular, and lateral and superior ampullary nerves. According to the literature, there is an average delay of 8 years between the onset of symptoms and diagnosis. The diagnosis is based on an audiovestibular examination and magnetic resonance imaging (MRI). We describe a case of intravestibular ILS in which we included the ocular vestibular evoked myogenic potentials (oVEMPs) test in the diagnostic workup. The oVEMPs test is a relatively new neurophysiologic diagnostic modality that evaluates the superior vestibular pathway and the ascending contralateral pathway through the vestibulo-ocular reflex. In our case, a 65-year-old man presented with progressive right-sided sensorineural hearing loss, dizziness, and tinnitus and fullness in his right ear. Audiovestibular examination and MRI detected an intravestibular ILS on the right. We found that oVEMPs were absent on the contralateral side, which contributed to the diagnostic process. The detection of oVEMPs can provide detailed information on the functionality of the macula of the utricle and the lateral and superior ampullary nerves, with a precise identification of the affected area. Based on our findings, we discuss the role of oVEMPs in the diagnosis of an intravestibular ILS.

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Introduction

Intralabyrinthine schwannomas (ILSs) are uncommon benign tumors that arise from the perineural Schwann cell sheath of the intralabyrinthine branches of the vestibulocochlear nerve. Little is known about the prevalence of ILSs; a systematic literature review published in 2013 confirmed the rare nature of this lesion, as the authors found no more than 250 reported cases.[1]

ILSs are classified as *intracochlear, intravestibular,* and *intravestibulocochlear*[2]*:*

- Intracochlear ILSs arise from the intracochlear branches of the vestibulocochlear nerve (80.7% of cases).
- Intravestibular ILSs originate in the saccular nerve, the utricular nerve, or the lateral and superior ampullary nerves (13.5%).
- Intravestibulocochlear ILSs involve both areas (5.8%).

ILSs can reach the internal auditory canal (transmodiolar, transmacular, or tympanolabyrinthine growth) and, in rare cases, the middle ear (transotic growth).[3] Consequently, the cochlea, the semicircular canals, the vestibule, or a combination of these structures may be involved.[4]

The presenting symptoms of an ILS usually include a slowly progressive sensorineural hearing loss, fullness, and tinnitus; balance disorders are seen in 16 to 70% of cases.[5-8] In addition to the medical history and clinical examination, the recommended diagnostic steps include a routine audiovestibular examination with pure-tone audiometry (PTA), auditory brainstem response (ABR) testing, infrared videonystagmography, and Fitzgerald-Hallpike caloric stimulation. However, high-resolution magnetic resonance imaging (MRI) with intravenous gadolinium contrast is necessary to confirm the diagnosis of an ILS.[9-11]

The slow, relatively asymptomatic tumor growth in cases of ILS often results in a delayed diagnosis (mean: 8 yr), which may have serious effects on the treatment options available for patients.[12,13]

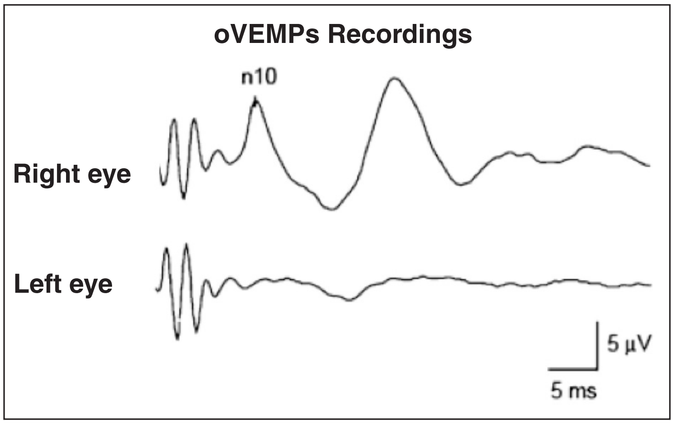

Figure 1. The n10 response of the oVEMPs to bone-conducted vibration is absent in the left eye, which suggests a lesion of the afferent fibers of the contralateral superior vestibular nerve.

Discussion

The more recent series and available reviews confirm the rare nature of ILS. In the English-language literature, only about 250 cases have been described so far.[1] Also, little is known about the patterns of ILS growth.[19]

Presenting symptoms usually include a slowly progressive sensorineural hearing loss, fullness, and tinnitus. In rare cases, the hearing loss is sudden or fluctuant.[20,21] In addition, balance disorders have been reported in 16 to 70% of cases.[5-8] Balance disorders are more common in patients with tumors that involve the vestibule or semicircular canals than in those with schwannomas isolated to the cochlea.[22,23] Ménière-like symptoms have been associated with ILSs in 4 to 43% of cases.[3]

Diagnosis requires an MRI, as initially reported by Doyle and Brackmann[8] in 1994 and now established in current clinical practice and scientific guidelines.[18] MRI is highly sensitive and specific for schwannomas, and it allows radiologists to identify their size, anatomic features, and exact location; its role also extends to postoperative follow-up.[24-28]

The choice of treatment for an ILS depends on the location of the mass and the severity of symptoms.[29,30] In a systematic review of the literature published in 2013, Van Abel et al reported that a wait-and-scan strategy was used in 53% of patients with ILS, and tumor progression was reported in 52% of patients who were studied with serial MRI.[1] Among these patients, 93% had either stable symptoms or a worsening of tinnitus, unsteadiness, or vertigo regardless of management choice, which supports the conservative wait-and-scan approach.[3,9,13] In cases of tumor growth, a complete loss of hearing, unmanageable vertigo or dizziness, or a possible malignancy, treatment options include microsurgical resection, stereotactic radiosurgery, and chemical labyrinthectomy.[31]

The use of oVEMPs testing in the diagnostic workup of a vestibular schwannoma, a condition that is similar to ILS, has been proposed by several authors, especially for assessing the function of the superior and inferior vestibular nerves before and after surgical intervention.[32,33]

Iwasaki et al studied 36 patients affected by vestibular schwannoma and reported that 86% exhibited absent or reduced oVEMPs, 78% had abnormal cVEMPs, and 86% had abnormal responses to caloric vestibular stimulation.[32] These data demonstrate the high degree of sensitivity of oVEMPs as a screening tool to identify affected patients and to evaluate their vestibular nerve function.

In a study published in 2014, Chiarovano et al investigated the clinical utility of VEMPs in 63 patients with a unilateral vestibular schwannoma.[33] They found that cVEMPs were abnormal in 65.1% of patients. In addition, oVEMPs were abnormal in 75.7% of patients using air-conducted sound stimulation and in 56.8% using mastoid bone-conducted vibration. In 16% of patients, VEMPs testing demonstrated a significant clinical value, as it represented the only abnormal test results in some of the enrolled patients. The authors recommended both air-conducted sound stimulation and bone-conducted vibration to screen patients for superior vestibular nerve function. However, in cases of normal results on air-conducted sound stimulation, bone-conducted vibration is not required.

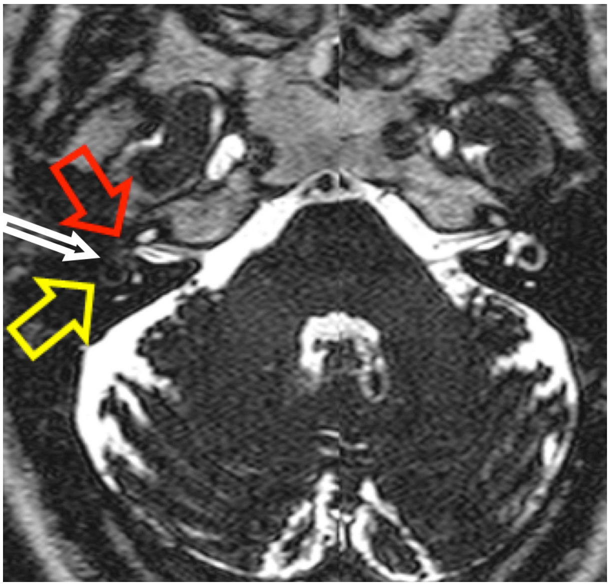

Figure 2. On T2-weighted axial MRI with intravenous gadolinium contrast, the right vestibule (red arrow) and the lateral semicircular canal (yellow arrow) are not clearly visible because of the presence of neoformed tissue and a possible mechanical alteration resulting from an abnormal movement of endo-perilymphatic fluid in the lateral semicircular canal. An area of low intensity is seen in the anterior portion of the lateral semicircular canal (white arrow), which could be interpreted as an initial involvement of the lateral semicircular canal.

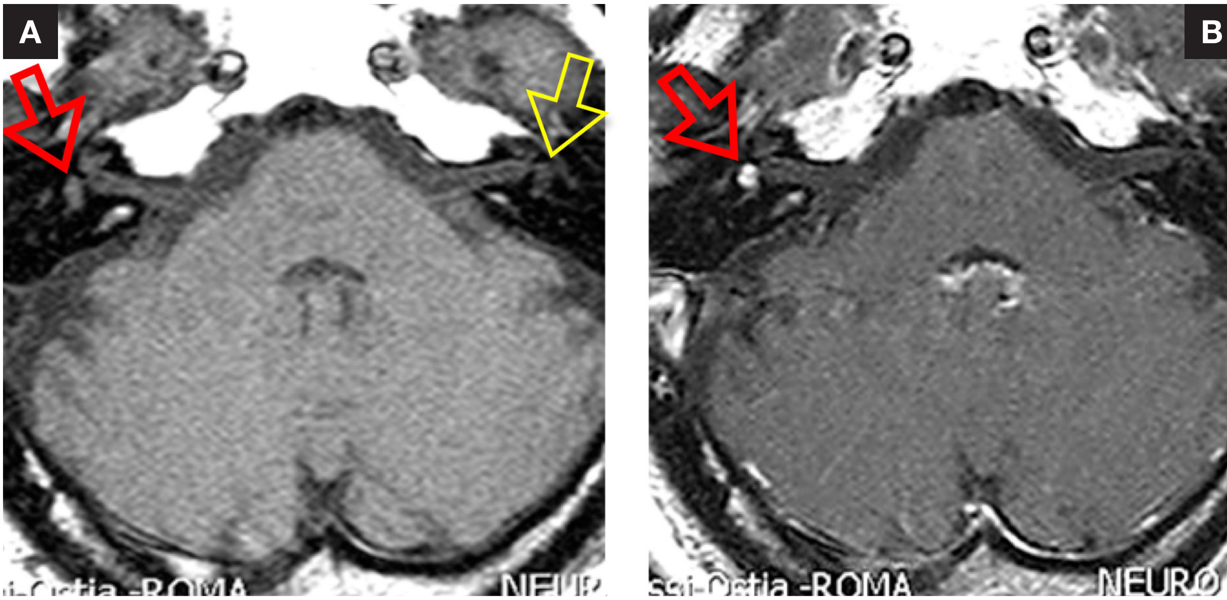

Figure 3. ***A:*** *T1-weighted spin-echo axial MRI without contrast shows the area of altered signal in the right labyrinth. The pathologic tissue (red arrow) was slightly more intense than the endo-perilymphatic fluid on the contralateral side (yellow arrow).* ***B:*** *With contrast, the image shows that the heterogeneously enhancing intravestibular hypervascularized mass is contiguous to the falciform crest with no signs of extension into the internal auditory canal (arrow).*

Recently, He et al used VEMPs and caloric testing to investigate the function of the superior and inferior vestibular nerves in 106 patients with unilateral vestibular schwannoma.[34] Their goal was to preoperatively identify the schwannoma's nerve of origin.

Taylor et al performed cVEMPS and bone-conducted oVEMPS testing in 50 patients with vestibular schwannoma and found that dysfunction of the superior and inferior vestibular nerves evolved in a parallel fashion for most patients with schwannoma.[35] They recommended that VEMPs asymmetry be completed by imaging in patients who present with a nonspecific disequilibrium or vertigo.

The oVEMPs test can provide detailed information on the functionality of the macula of the utricle and the lateral and superior ampullary nerves with precise identification of the affected area. In fact, bone-conducted vibration generates a stimulation similar to linear acceleration in that it selectively activates neurons in the utricular macula. Measuring oVEMPs can therefore be extremely specific in the identification of disorders of these structures, although it cannot replace MRI for diagnosing an ILS.

Another clinical application of oVEMPs testing is to monitor vestibular function before and after surgery. Preoperatively, baseline VEMPs testing provides an indication of residual vestibular function and the likelihood that a patient will need postoperative vestibular rehabilitation. After surgery, VEMPs testing in patients who become unsteady postoperatively can help determine whether the unsteadiness is the result of decompensation or further compromise of vestibular function.

In our patient, the n10 response of the oVEMPs testing to air-conducted sound and bone-conducted vibration was absent on the side contralateral to the lesion. This finding concluded the routine audiovestibular examination, which had already identified an ipsilateral sensorineural hearing loss and areflexia on caloric testing. It also allowed for a precise identification of the affected area, suggesting the diagnosis of an intralabyrinthine intravestibular schwannoma. However, MRI of the brain with gadolinium contrast in the axial planes was necessary to confirm the diagnosis. The use of multiple MRI sequences is necessary to differentiate solid masses and other intralabyrinthine pathologic conditions such as labyrinthine hemorrhage and infective labyrinthitis.

MRI is necessary not only for making the diagnosis of an ILS, but also for subsequent management and follow-up.[24-28] The latest developments in imaging technology allow for a precise regional diagnosis of an ILS, as reported in a study by Yoshida et al published in 2011.[28] They described the case of an ILS that was diagnosed by 3.0 Tesla MRI, which revealed "the size and shape of the tumor via identification of the cochlear nerve and cochlear fluid space."

In our patient, T2-weighted MRI was extremely important not only to show a decrease in signal intensity and the presence of a solid mass in the utricle, but also to highlight signal irregularities. These irregularities were likely caused by mechanical factors resulting from an altered movement of endo-perilymphatic fluids. Also, T2-weighted sequences documented initial involvement of the semicircular canals in the affected side (figure 2). In this case, an area of altered signal in the labyrinth was found on T1-weighted spin-echo sequences without contrast, in which pathologic tissue contrast was slightly more intense than the contrast in endo-perilymphatic fluid on the contralateral side (figure 3, A). When contrast was added, MRI showed a heterogeneously enhancing intravestibular hypervascularized mass (figure 3, B), thus confirming the diagnosis

In conclusion, our case provides evidence of oVEMPs alteration in a patient with an intravestibular ILS. The use of this measurement in the audiovestibular diagnostic process can contribute to precise identification of the involved nerve and should therefore be considered for inclusion in the diagnostic workup of utricular neoplasms.

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