

## Case Report

# Intracochlear Vestibular Schwannoma Presenting with Mixed Hearing Loss

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As for other vestibular schwannomas, intralabyrinthine schwannomas commonly cause a sensorineural hearing loss, contrary to more lateral ear pathology that can cause conductive or mixed hearing loss. This case report features a patient that presented with a mixed and thus partly pseudo-conductive hearing loss due to an intracochlear schwannoma, a finding that is very rare. As a result, the patient was initially misdiagnosed as having otosclerosis and a stapedotomy was performed, without hearing improvement. We discuss the clinical implications of this atypical presentation, which illustrates the importance of performing supplementary audiological testing (e.g., the Gellé test), and the importance of considering vestibular system testing when otosclerosis is suspected. In addition, the importance of imaging and considering differential diagnoses in cases of conductive hearing loss is stressed.

**KEYWORDS:** Vestibulopathy, surgery, hearing rehabilitation, hearing loss, conductive hearing loss, magnetic resonance imaging

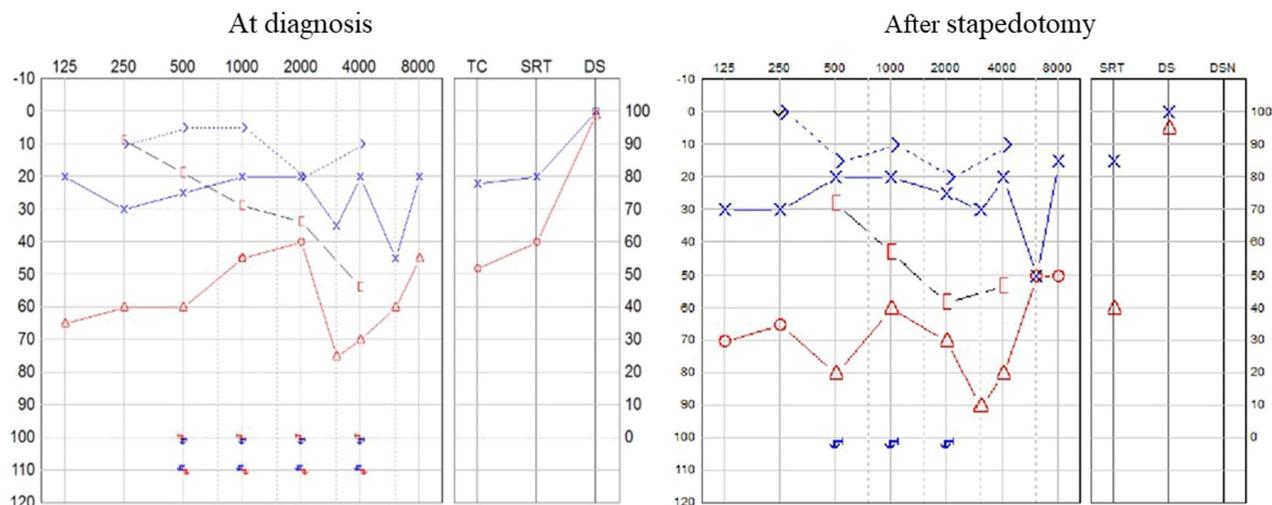
## INTRODUCTION

Vestibular schwannomas (VS) are benign tumors deriving from the Schwann cells of the vestibulocochlear nerve.<sup>1</sup> The incidence is increasing<sup>2</sup> and the most prevalent symptoms are sensorineural hearing loss (SNHL), tinnitus, disequilibrium, and with larger tumors, affection of the neighboring cranial nerves. VS can be divided into 3 subtypes depending on localization: intrameatal schwannomas within the internal auditory canal, extrameatal schwannomas with at least a part of the tumor in the cerebello-pontine angle (CPA), and intralabyrinthine schwannomas located within the cochlea and/or the vestibular system (vestibule and semicircular canals).<sup>3</sup> Purely intrameatal tumors are found most commonly, followed by extrameatal tumors, while intralabyrinthine schwannomas are rare.<sup>4</sup>

The aim of this paper is to report a case of an intracochlear schwannoma presenting with mixed hearing loss, leading erroneously to the diagnosis of otosclerosis and a subsequent stapedotomy, without hearing improvement. The clinical implications of this atypical presentation are discussed.

## CASE PRESENTATION

A 47-year-old male, otherwise healthy, presented to a private otolaryngology clinic with severe tinnitus lasting many years as the main complaint. Furthermore, he had experienced a slowly progressive hearing loss. The otological examination suggested a conductive hearing loss, including a pathological Rinne's test on the right ear and normal on the left ear, along with a Weber test that lateralized to the right ear. Otomicroscopy was normal. Audiometry showed asymmetrical mixed hearing loss on the right ear, and phoneme discrimination within normal range (Figure 1A). The patient was booked for an MRI and referred to the local ENT department. The MRI showed a mass lesion in the basal turn of the right cochlea, strongly suggesting an intracochlear vestibular schwannoma (Figure 2). The patient was subsequently seen by a surgeon in the ENT outclinic at the local hospital. It was concluded that the conductive component of the hearing could be due to otosclerosis, and the patient was booked for a tentative stapedotomy.



**Figure 1.** A (left panel) and B (right panel): Diagnostic pure-tone and phoneme discrimination audiometry showing a light mixed hearing loss on the left side and a severe mixed hearing loss on the right side, mimicking that of otosclerosis (Carharts’ notch apparently present right side). The otoscopy was normal (no Schwartz sign).

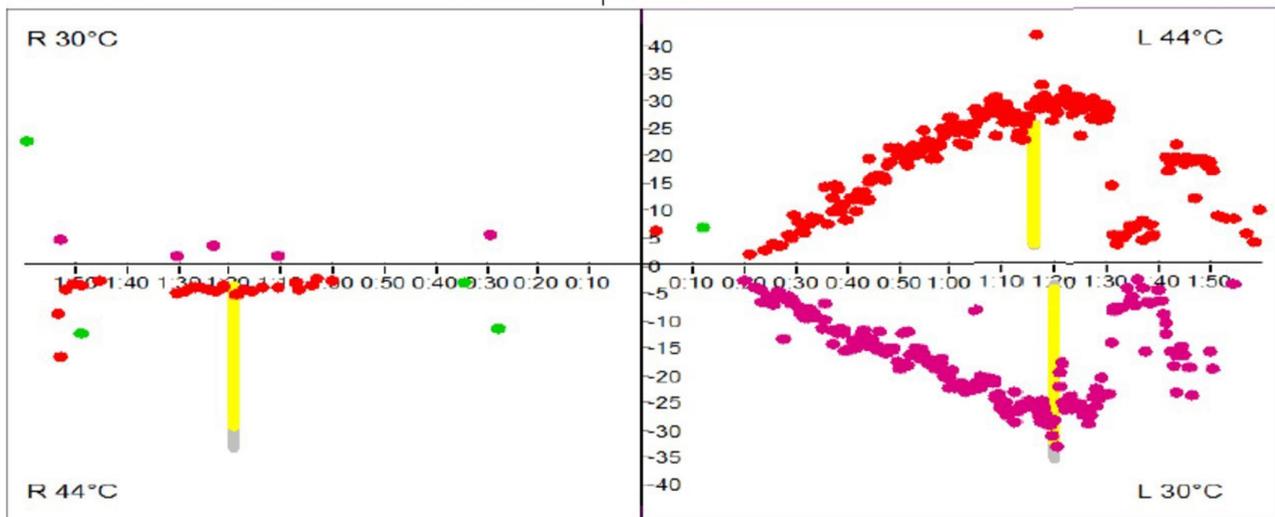
A month later, the patient underwent an uncomplicated stapedotomy, as during explorative tympanotomy, the surgeon estimated the stapes was fixed. At the 3-month follow-up, the patient reported that the hearing loss and tinnitus were unchanged. Audiometry confirmed the lack of improvement (Figure 1B). A senior colleague was then consulted, and based on the MRI, and the patient was referred to our National referral center for VS. At the National Center, vestibular testing with bithermal caloric irrigation demonstrated a unilateral weakness of 86% (canal paresis) ipsilateral to the schwannoma (Figure 3). The hearing loss was effectively managed by conventional

binaural hearing aids, although the patient was informed of the option of surgical tumor removal and cochlear implantation.

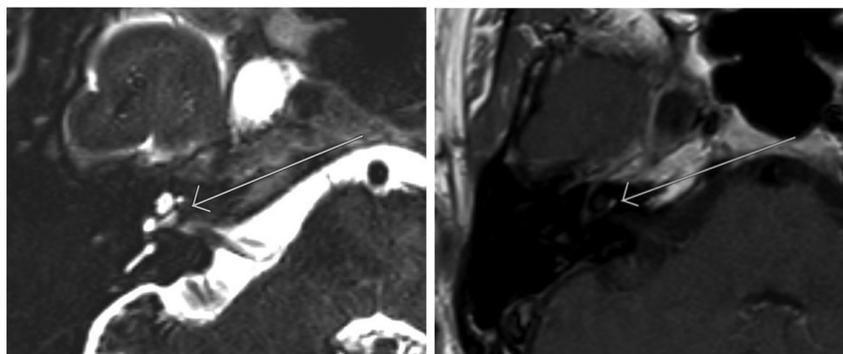
**DISCUSSION**

Here, we here report a very unusual presentation of a VS. The patient presented with a mixed and thus a pseudo-conductive hearing loss and severe tinnitus, leading to the suspicion of otosclerosis. MRI showed a small intracochlear mass in the basal turn, strongly suggesting an intracochlear schwannoma. The pseudo-conductive hearing loss led to a stapedotomy, as the surgeon estimated the

Horiz.avg.	a.SPV[°/s]	Fix.Index	Unilateral Weakness:	86% right
R 44°C:	-4.0	0 %	Right Ear:	3.96°/s
L 44°C:	27.9	22 %	Left Ear:	52.03°/s
L 30°C:	-24.1	27 %	Dirac. Preponderance:	0% right
R 30°C:	0.0	--	LC + RW	28.07°/s
<b>Total SPV:</b>	<b>56.0</b>	<b>SPN:0.0</b>	RC + LW	27.92°/s



**Figure 3.** Caloric test with bithermal irrigation showing canal paresis on the right side (86% unilateral weakness) and normal function in the left ear.



**Figure 2.** T2-weighted MRI with (left) and without contrast (right), disclosing a tumor in the basal turn of the right cochlea (arrow).

stapes was fixed during explorative tympanotomy. Apparently, the schwannoma was not considered as the potential reason for the conductive component of the hearing loss. Neither hearing nor tinnitus improved following stapedotomy, which led to re-assessment and referral to the National VS center. Additional audio-vestibular testing was performed, and the diagnosis settled on an intracochlear schwannoma.

The typical symptoms of VSs are unilateral SNHL (including loss of discrimination), tinnitus, and periodic disequilibrium.<sup>5</sup> Typically, the Rinne test is positive, and the Weber test lateralizes to the contralateral ear. The reported patient's Rinne test was negative, and the Weber test lateralized to the affected ear. The audiometry suggested a mixed, and thus partly pseudo-conductive hearing loss (Figure 1). The discrimination score was within the normal range, and the otomicroscopy was normal (no Schwartze sign). Unfortunately, the ipsilateral thresholds were too high to determine the presence of an acoustic reflex, and no additional tests were performed in order to strengthen the suspicion of otosclerosis (e.g., Gellés test). Yet, the symptoms and findings were erroneously interpreted as most likely caused by otosclerosis. The finding of a fixed stapes during explorative tympanotomy remains obscure, as no hearing improvement occurred upon stapedotomy, leaving otosclerosis as an explanation for the hearing loss highly unlikely.

Conductive hearing loss due to inner ear pathology has been described previously, for example, in regard to third-window lesions of the inner ear, such as superior semicircular canal dehiscence and enlarged vestibular aqueduct.<sup>6</sup> Merchant and Rosowski have posed a plausible reason as to why a third-window lesion may result in a pseudo-conductive hearing loss. Air-conducted sound energy is directed away from the cochlea, whereas bone-conducted energy reaching the cochlea is increased, leading to the finding of a pseudo-conductive hearing loss.<sup>6</sup> The mechanism in the case of an intracochlear mass lesion is plausibly different, as a lesion may hamper the movement of the basilar membrane and thus the traveling wave in a mechanical way, leading to a pseudo-conductive hearing loss. We believe this is the explanation for the configuration of the hearing loss found in the present patient.

Intralabyrinthine schwannomas are reported to be rare. In the present literature, we have only identified 3 cases with a mixed and thus pseudo-conductive hearing loss.<sup>7</sup> The first preoperatively diagnosed intralabyrinthine schwannoma was described by Özlüoğlu and

Herman in 1994.<sup>8</sup> Prior to this discovery, intralabyrinthine schwannomas were an incidental finding, either as a part of the autopsy examination or during surgery.<sup>9-12</sup> The present case report epitomizes that an intralabyrinthine schwannoma should be considered as a rare differential diagnosis in case of mixed hearing loss and normal otomicroscopy. Upon referral of the patient reported, a caloric test was performed, demonstrating canal paresis (Figure 3). This is a common finding in intralabyrinthine schwannomas,<sup>13</sup> meaning that if caloric testing had been performed before the stapedotomy, surgery could probably have been avoided, as the sum of findings (otomicroscopy, MRI, audiometry, and caloric test) would have strongly suggested that the cause of hearing loss was not otosclerosis. Thus, overall, the case illustrates the importance of performing supplementary audiological testing (e.g., Gellés test) and the importance of considering vestibular system testing when otosclerosis is suspected. Also, the importance of performing and scrutinizing findings of imaging, as well as considering alternative diagnoses is evident.

The treatment of VSs typically depends on the size of the tumor, symptoms, patient's age, and comorbidity at the time of diagnosis. Treatment modalities are observation (wait-and-rescan), radiation, or microsurgical excision.<sup>2</sup> Intralabyrinthine VSs have most commonly been treated by observation and repeated MRI. The hearing is most often lost due to the tumor itself, and in case of residual hearing, this would be lost by radiation or surgical opening of the cochlea to remove the tumor. Thus, tumor growth has been the only reasonable indication for active treatment in the past. Cochlear implantation is indicated for single-sided deafness; however, active treatment by surgical removal of the intralabyrinthine tumor and simultaneous or subsequent cochlear implantation is a promising option as surprisingly good results have been reported.<sup>14</sup> Preservation of vestibular function has even been possible after removal of a cochlear tumor.<sup>14</sup>

So far, our patient has found treatment with conventional hearing aids and continued observation by repeated MRI to control for tumor growth satisfactory. Should the hearing deteriorate further or the tumor display growth, the patient will be a candidate for surgical tumor removal and cochlear implantation. Implantation without simultaneous tumor removal is feasible, also leading to surprisingly good results, although this poses problems related to control of tumor growth by MRI.<sup>14</sup>

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**Conflict of Interest:** The authors have no conflict of interest to declare.

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