Hearing Loss Owing to Intralabyrinthine Schwannoma Responsive to Intratympanic Steroid Treatment

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Vestibular schwannoma is a benign tumour that originates from the distal parts of the vestibular or cochlear nerve. The vestibule is an unusual location for acoustic neuromas. The primary symptoms of such tumours are unilateral hearing loss, tinnitus, and vertigo. In this article, we report a patient with vestibular schwannoma who presented with the typical complaints and audiologic findings of Meniere disease. On magnetic resonance imaging (MRI), the patient was found to have a 5 × 4 mm mass that was slightly hyperintense on T1-weighted MRI and showed contrast enhancement (Figure 1, Figure 2, and Figure 3). The associated hearing loss responded to intratympanic steroid therapy.

Case Report

A 29-year-old woman was referred to our clinic for progressive left-sided hearing loss and tinnitus for 2 years. She also experienced both spontaneous and position-evoked vertigo that lasted for 1 hour. There was no history of otorrhea. The otoscopic examination was normal on both sides. There was no spontaneous, positional, or gaze-evoked nystagmus. Romberg testing was negative. The remaining cranial nerve, neurologic, and head and neck examinations showed normal findings. An audiogram revealed a left-sided sensorineural hearing loss of 33 dB on all frequencies (Figure 4), with a 60% speech discrimination score. The tympanogram of the patient exhibited a bilateral type A pattern and positive acoustic reflexes. An electronystagmogram revealed total canal paralysis on the left side. On the auditory brainstem response (ABR) testing, prolonged I–V waveform responses were observed for the left ear (Figure 5). Gadolinium-enhanced MRI of the internal acoustic canals (IACs) showed an enhancing lesion of 5 × 4 mm in size in the left vestibule. The lesion was slightly hyperintense on T1-weighted MRI. According to MRI findings, the diagnosis of left vestibular schwannoma was made. After five doses of weekly intratympanic...
steroid treatment, the hearing loss improved to 18 dB (Figure 6). Each dose consisted of enough dexamethasone to completely fill the middle ear (about 0.3 cc). During the follow-up of the patient for 15 months, the hearing remained stable.

Discussion

Intralabyrinthine schwannomas can arise from the distal end of any division of the eighth cranial nerve. About 60 patients with intralabyrinthine schwannomas have been reported in the literature. Previous reports suggested that the cochlear division of the nerve may be involved with the tumour more commonly than other branches. It is also stated that the basal turn of the cochlea was the most commonly involved anatomic site. Some cases involved only the vestibular system. Cochlear lesions have been hypothesized to cause a cochlear pattern of progressive sensorineural hearing loss by direct compression of the organ of Corti. Vestibular and semicircular canal lesions have been hypothesized to cause fluctuating hearing loss owing to endolymphatic hydrops through compression of the ductus reuniens and saccule within the vestibule. Progressive or sudden sensorineural hearing loss with poor discrimination can be caused by direct compression of the cochlear nerve or the labyrinthine artery.

Vestibular schwannoma must be differentiated from labyrinthitis, which has the same clinical findings. Labyrinthitis of viral or autoimmune origin causes sensorineural hearing loss, tinnitus, and vertigo, which is clinically indistinguishable from vestibular schwannomas. Both entities are associated with an enhancement in the labyrinth on MRI of the IACs. In labyrinthitis, MRI shows...
Aronzon and colleagues reported 
Nedzelski and colleagues 
and colleagues described a case of 
This improvement is thought to be due to a 
Patients with a 
< 37x4; The Charlesworth Group 
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vestibular schwannoma owing to neurofibromatosis type 2 
Gaffney and McShane reported on a patient with bilateral 
labyrinthine bleeding is hyperintense on T1-weighted MRI. 
Labyrinthine fibrosis owing to chronic labyrinthitis is hypointense on T2-weighted MRI. 
Labyrinthitis and vestibular schwannomas can be differentiated from each other using MRI with contrast. 
Meniere disease should also be included in the differential diagnosis of vestibular schwannomas. In our case, the symptoms and the audiometric pattern of hearing 
loss showed marked resemblance to Meniere disease. The diagnosis of vestibular schwannoma could be made with 
total canal paralysis on caloric tests, prolonged I–V waveform response for the left ear on ABRs, and 
gadolinium-enhanced MRI of the IACs. 
Treatmet of intralabyrinthine tumours is based on the 
age of the patients, the severity of the symptoms, and the 
location of the tumour. Intractable vertigo and severe, 
progressive hearing loss, especially in young patients, are 
the most common indications for surgery. 
Patients with a pure-tone average over 50 dB or a discrimination score of 
less than 50% without severe vertigo can be observed with 
serial audiograms and MRIs. 
Nedzelski and colleagues emphasized the rate of tumour growth as an important 
factor for determining the type of management. 
If the conservative management pathway is preferred, the 
patients must be seen twice yearly, and each visit should consist of a thorough neurotologic examination and high-definition computed tomography or MRI. 
If the clinical course and rate of tumour growth remain unchanged over 
a 3-year follow-up, annual assessments are recommended. 
A rate of growth equal to or exceeding 0.2 cm per year constitutes an indication for tumour removal. 
Surgery may be indicated when intralabyrinthine schwannomas begin 
to show evidence of extension into the IAC. The approach 
used in intralabyrinthine schwannomas depends on the 
location of the tumour. Transmastoid labyrinthectomy is 
the procedure of choice for tumours involving the 
vestibule or semicircular canals. When the IAC is involved, 
opening the dura is necessary. 
Corticosteroids have been used in a variety of inner ear 
disorders to improve hearing; however, only a few reports 
described recovery from sensorineural hearing loss with 
corticosteroid use in the presence of vestibular schwannoma. 
Berenholtz and colleagues described a case of 
sensorineural hearing loss in an only hearing ear with a 1.5 cm vestibular schwannoma, with recovery to normal levels 
after steroid therapy on four parenteral injections. 
Gaffney and McShane reported on a patient with bilateral 
vestibular schwannoma owing to neurofibromatosis type 2 
with sensorineural hearing loss that was responsive to 
corticosteroid therapy. 
Aronzon and colleagues reported 
pure-tone reception threshold improvements ranging from 
10 to 50 dB in at least two frequencies or more in seven 
patients with acute hearing loss secondary to an acoustic 
neuroma with oral prednisone (1 mg/kg for 1 to 2 weeks) therapy. 
This improvement is thought to be due to a reduction in tumour mass by the antiedema effect of corticosteroids. 
Berg and colleagues described only one of 
four patients with vestibular schwannoma who had 
improved hearing with corticosteroid treatment. 
However, these represent somewhat random cases, and 
some studies have demonstrated that the hearing loss in 
vestibular schwannomas tends to be progressive, despite 
steroid treatment. Systemic corticosteroid therapy was 
used in all of these studies. Our literature review did not 
reveal any studies mentioning the use of intratympanic 
steroid therapy for vestibular schwannomas. Patients with 
persistent vertigo despite medical treatment may benefit 
from intratympanic gentamicin application. 

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References 
Authors Queries

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