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CASE REPORT

Intracochlear Schwannoma Presenting with Vertigo

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Objective : A schwannoma , arising de novo from the cochlea, is considered rare tumor. Only few prior cases were described in literature. We describe a 53 years old female who presented to our hospital with sudden evoked positional vertigo.

Materials & Methods: The audiogram demonstrated a unilateral, left sided –down-sloping sensorineural hearing loss (SNHL) with PTA of 35 dB and discrimination score of 64%. Magnetic resonance imaging (MRI) revealed a 3mm lesion in the left cochlea, in keeping with the diagnosis of cochlear schwannoma.

Results: The diagnosis of cochlear schwannoma in this patient was mostly an "incidenteloma". Management should consider the fact that hearing cannot be preserved for patients with serviceable hearing and growing tumors.

Conclusion: Completing an audiologic workup for a dizzy patient, as well as head MRI for unilateral hearing loss is essential.

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Introduction

Intralabyrinthine schwannomas are neoplasms that arise from the distal branches of the cochlear, superior vestibular or inferior vestibular nerves [1]. The term was first described by Mayer [2] in 1917. While most of the intralabyrinthine schwannomas arise from the vestibular nerve, there have been prior cases which reported shwannomas confined to the cochlear nerve, named cochlear shwannomas, however, they are rare tumors [3-5] .

Progressive or sudden sensorineural hearing loss (SNHL) is the most common symptom of cochlear schwannoma, with or without accompanying subjective tinnitus. The hearing loss is thought to be caused by direct compression of the organ of Corti or its vascular supply [6-8].Vertigo, in contrast, is rarely the presenting symptom of isolated cochlear schwannoma [6].

Like the much more common vestibular shwannomas, the diagnosis of cochlear schwannoma is based on clinical suspicion, neurootologic exam, audiometric findings, and proven on gadolinium-enhanced magnetic resonance imaging (MRI).

We present a case of cochlear schwannoma with the unusual presentation of vertigo.

Case report

A 53 year old woman presented to the emergency ward with sudden, positional-evoked vertigo, lasting for several hours. There was no history of recent head trauma, upper respiratory tract infection, nor did she report any hearing impairment or tinnitus. She also did not report prior otologic or vestibular disease. Physical examination revealed normal tympanic membranes and a complete neurotologic exam, including the positioning testing showed normal findings. The patient was admitted to the Otolaryngology Department and was discharged a day later, as symptoms subsided.

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The patient was seen again one week later and she was free of symptoms. An audiogram (figure 1) demonstrated a unilateral, left sided, down-sloping SNHL with a PTA of 35 dB and discrimination score of 64%.

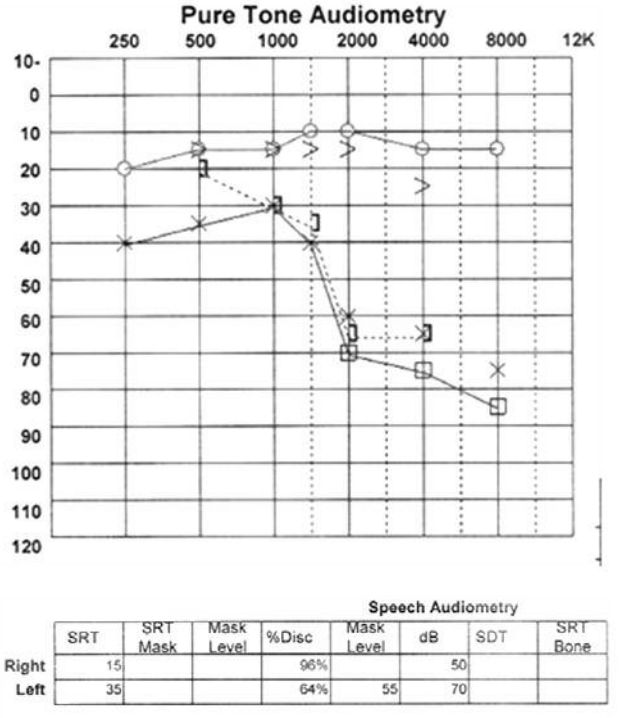

Figure 1. Pure tone audiometry showing a unilateral, left sided-down-sloping SNHL with a PTA of 35 dB and discrimination score of 64%.

MRI (figures 2, 3) of the head demonstrated symmetric and normal cerebellopontine angles and internal acoustic canals, as well as normal appearing facial, cochlear and vestibular nerves. On post- gadolinium T1 images a enhancement was demonstrated in the left cochlea, in keeping with the diagnosis of a cochlear schwannoma.

The patient was advised follow- up with an annual audiogram and MRI. On three years follow up there was no change on imaging and there was a slight decrease in pure tone hearing.

Discussion

A schwannoma, arising de novo from the cochlea, is considered rare [3-5]. Progressive or sudden SNHL is known to be the most common presenting symptom of cochlear schwannoma [6]. Vertigo, on the other hand, is an uncommon presenting symptom compared to hearing loss. Neff et al [6] thoroughly reviewed all published cases and reported that 7 out of the 27 patients with cochlear schwannoma presented with vertigo. The authors offer no explanation to the mechanism of vertigo.

Our patient presented with vertigo and a normal neurotological exam. The cause of her vertigo was unknown, and one can only speculate that the patient may have experienced benign paroxysmal positional vertigo (BPPV), or less likely, vestibular neuritis that resolved rapidly. Alternatively, the incident could have been the first attack of recurrent vestibulopathy or Meniere's disease, but this seems unlikely, as there were no recurrent attacks during the three years follow up. Other causes of a transient-non vestibular dizziness are also possible.

The hearing test, in our patient, was carried out as a part of the standard investigation for a dizzy patient. The patient was unaware of her left sided hearing loss, probably due to the fact that she was right handed and therefore she used the phone with her right ear. Following the diagnosis of unilateral sensorineural hearing loss, the intent of the head MRI was to rule out, the much more common diagnosis of a cerebello-pontine angle tumor.

Before the use of MRI, 14 cases of intralabyrinthine schwannomas were diagnosed at autopsy [2, 4, 9-12]. Moreover, schwannomas were revealed incidentally during labyrinthectomy for Meniere's disease or other intractable cases of vertigo in 11 cases [7, 10-14]. It is possible that the vertigo in our patient was a symptom of the cochlear schwannoma in an unknown mechanism, similar to the previous 7 cases mentioned, however since it was transient, this seems unlikely. We hypothesize, that the vertigo was not directly related to the cochlear schwannoma, but was rather an incidental complaint that warranted an audiogram, as a part of the usual workup. The audiometric finding of unilateral hearing loss, especially with decrease word discrimination led us to perform a head MRI, revealing a cochlear schwannoma in her left ear.

We adopted a *"wait and scan with serial audiograms" approach* since: 1. Any surgical procedure to remove this lesion would sacrifice her remaining serviceable hearing, 2. Growths of intracranial and intratemporal of schwannomas are slow in older patients and are easily followed up by MRI.

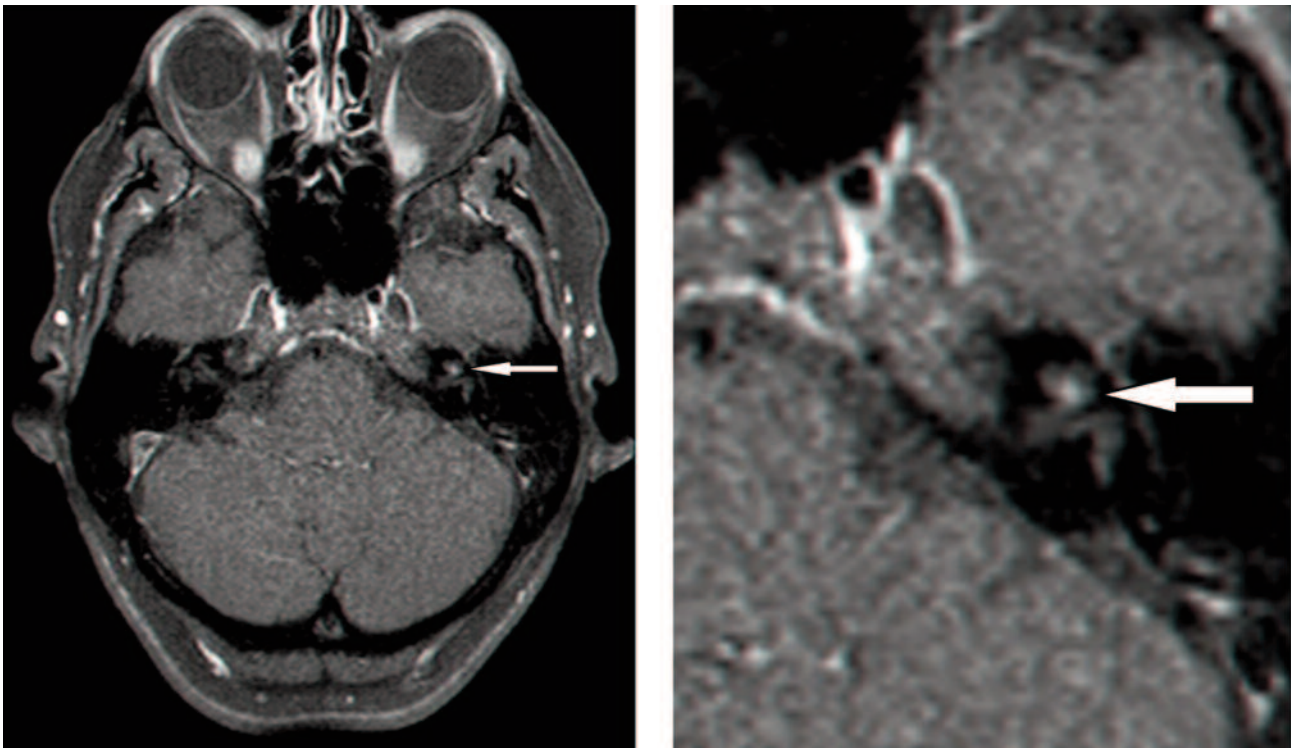

Figure 2a, 2b. T1 spir sequence after gadolinium injection demonstrates an enhanced lesion in the left cochlea (arrow).

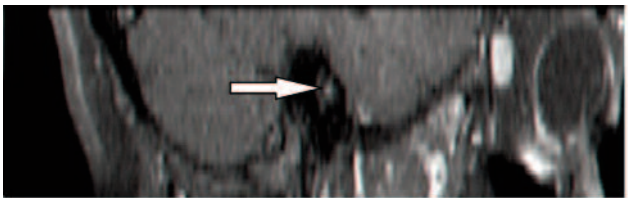

Figure 3. T1 spir saggital reconstruction showing the lesion in the left cochlea (arrow).

In younger patients without serviceable hearing and with evidence of active growth, the tumor would best be removed with a transcochlear approach, sacrificing any residual hearing.

Conclusions

Cochlear schwannomas are rare tumors which may present with hearing loss, tinnitus and rarely, with vertigo. Diagnosis is made on gadolinium-enhanced head MRI. Therefore, completing an audiologic workup for a dizzy patient, as well as head MRI for unilateral hearing loss is essential. Management should consider the fact that surgery cannot preserve hearing and should be reserved for younger patients with no serviceable hearing and actively growing tumors.

Summary

Cochlear schwannomas are tumors, which can present with hearing loss, tinnitus and even vertigo. Diagnosis is made by MRI and treatment depends on tumor progression on the one hand, and clinical symptoms of the patient on the other hand.

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