

Case Report

Otologic Manifestation of Hidden Vestibular Schwannoma

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Citation: Bogosavljević V, et al. Otologic Manifestation of Hidden Vestibular Schwannoma. J Neuro Onco Res. 2025;5(3):1-5.

<https://doi.org/10.46889/JNOR.2025.5307>

Received Date: 06-11-2025

Accepted Date: 26-11-2025

Published Date: 03-12-2025



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Abstract

Vestibular Schwannomas (VS), also known as acoustic neuromas, are benign and usually slow-growing tumors of the eighth cranial nerve. They commonly present with unilateral sensorineural hearing loss, tinnitus, dizziness or may be found incidentally during MRI imaging. Although benign, they may cause compression of intracranial structures and rarely undergo malignant transformation. We present a case of a hidden vestibular schwannoma with special attention to diagnostic accuracy, management options and post-radiosurgical imaging interpretation.

Keywords: Vestibular Schwannoma; Acoustic Neuroma; Hearing Loss; Gamma Knife Radiosurgery; Magnetic Resonance Imaging (MRI)

Introduction

Acoustic Neuromas (AN) or Vestibular Schwannomas (VS), represent benign, often slow-growing tumors of the eighth cranial nerve that may present with hearing loss in one ear, tinnitus, dizziness, or, increasingly, as incidental findings on radiological imaging that include Computed Tomography (CT) and Magnetic Resonance Imaging (MRI). Although benign, these type of tumors can make compressive effect on brainstem and produce intracranial hypertension symptoms due to mass effect and in small amount of cases produces a risk of malignant transformation [1]. VS is one of the most prevalent tumour of Cerebropontine Angle (CPA) with a clinical incidence of about 1 per 100,000 persons [2,3]. Advances in imaging have

made it possible to identify small, asymptomatic neuromas. The early detection of developing neuromas offers new challenges in determining the best method of treatment [4,5]. VS management depends on the age of the patient, tumor size and presenting symptoms. Surgical management of VS should take into account tumor size and morphology and patient's symptoms, comorbidities and preferences. A case of VS with special attention to diagnostic accuracy for detecting this lesion.

Case Report

The patient, a 37-year-old man, presented with hearing loss, tinnitus, episodic vertigo and ear fullness in the left ear. The slow progression of ringing in ear (tinnitus) and hearing loss was present for more than 15 years and worsened during the last year. He was otherwise healthy and active in sports. On admission, clinical examination revealed left-sided deafness, with no facial weakness or other cranial nerve deficits. Otorhinolaryngological findings were normal. Pure-tone audiometry showed profound sensorineural hearing loss, tympanometry was type A bilaterally and stapedial reflexes were absent on the left side (Fig. 1). MRI of the cerebellopontine angle confirmed a vestibular schwannoma approximately 2.5 cm in diameter (Fig. 2,3). The tumor was discussed at a multidisciplinary board and the patient underwent gamma knife radiosurgery. Follow-up MRIs showed early post-radiation changes with intratumoral necrosis and subsequent reduction in tumor volume of more than 50% after one year (Fig. 4,5).

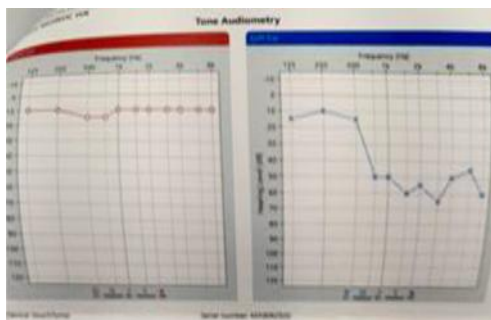


Figure 1: Audiometric findings: Type of diagram that represents profound sensorineural hearing loss on the left side of ear.

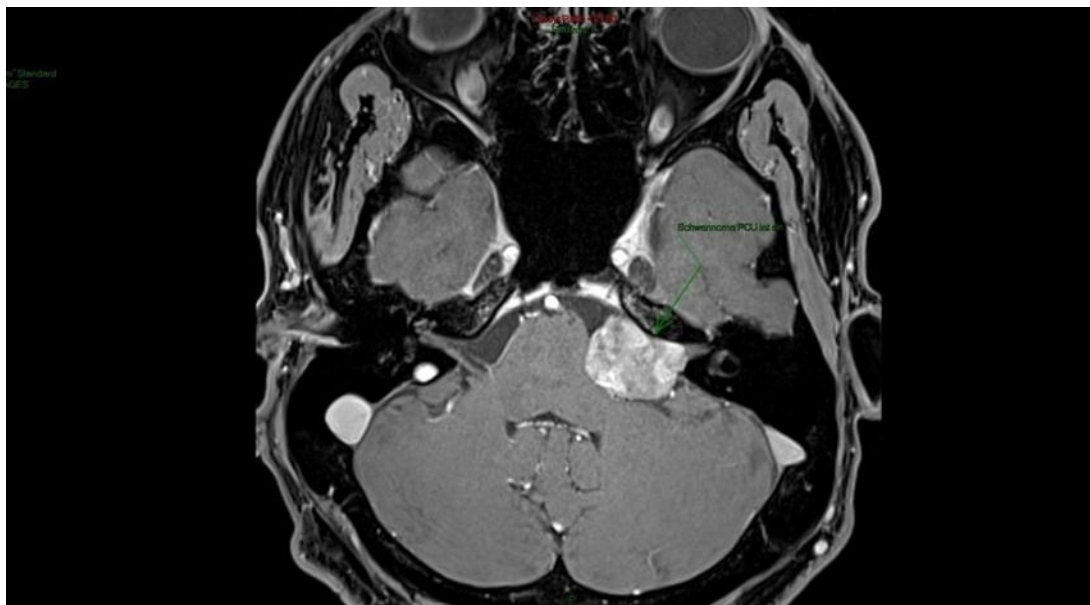


Figure 2: T1W VIBE fs 3D postcontrast (17. Jul. 2023) Left PCA expansive extraxial lesion, which remodels MAI, has intrameatal extension, without intracochlear and intravestibular extension. The longest longitudinal is 17x24x18 mm (APxLLxKK). The greatest compressive effect is pronounced on CN (VII and CN VIII) and the pons. Koss grade scale 4.

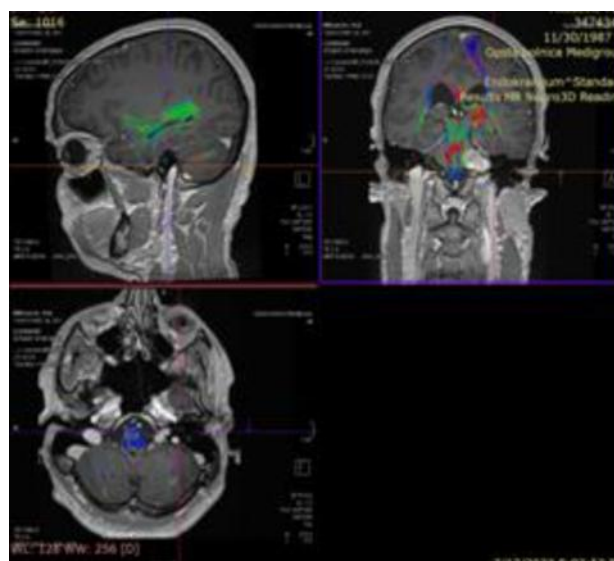


Figure 3: DTI tractography is prepared with DTI 220 DFC tensor, 30 directions. There is compression of the tract upward and mediosagittal, especially pontine craniocaudal and laterolateral (L-R) tracts. The CN VII and CN VIII is not separated. Deep intrameatal in the fundus, MAI noticeable, gracile neural structures in comparison with the contralateral, probably due to chronic compression.

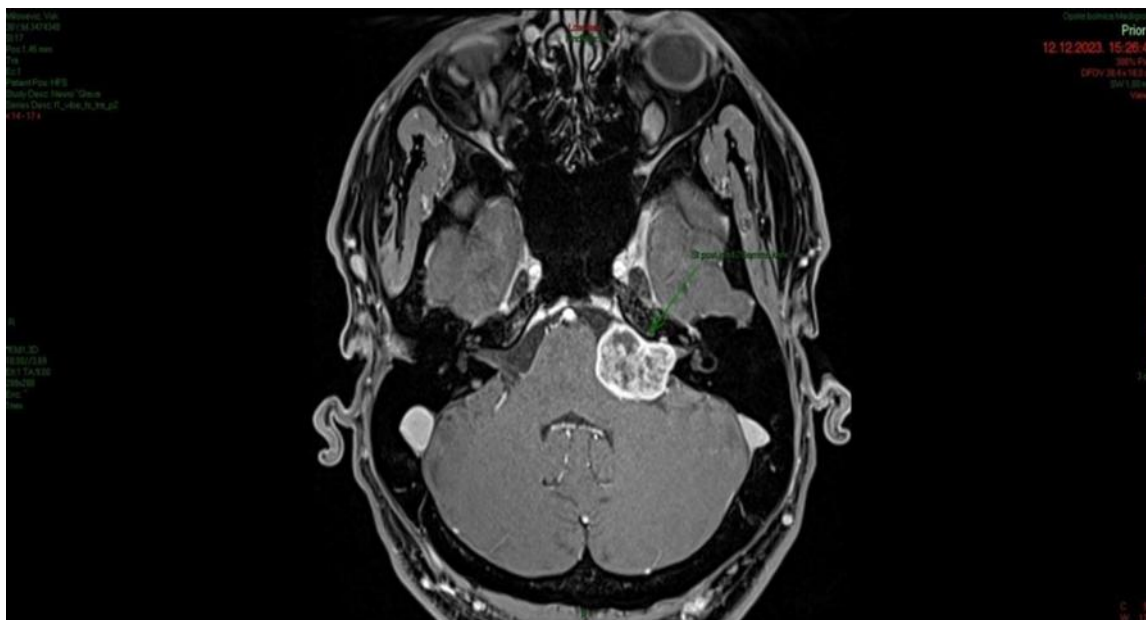


Figure 4: T1W vibe fs 3D postcontrast. Condition after stereotaxic radiosurgery tu left PCU. (TD 12.5 Gy/50% isodose; Sept 2023.) Dec .2023. Relatively early post-radiation changes, which are dominated by changes in IS and the appearance of necrosis zones without a significant reduction in the diameter of these changes compared to MRI before radiation.



Figure 5: Dec. 2024. Postirradiation reduced diameter. extensive zones of cystic necrosis, the longest longitudinal diameters 8.8x17.4x9.2mm (was 17x24x18mm, APxLLxKK). Postirad. Koss gr 3. Reduction in the diameter of the tumor by more than 50% PR.

Discussion

Otologic manifestations of VS include vestibular and cochlear symptoms. Most patients present unilateral sensorineural hearing loss (94%) and tinnitus (83%), while vestibular symptoms vary widely. Larger tumors are associated with neurological dysfunction such as facial pain, headache and ataxia. Studies shows that 20% of patients presenting to Ear, Nose and Throat (ENT) clinics have some of symptoms suggesting a lesion at the cerebellopontine angle [9]. Although benign, VS represent a risk of compression due to mass effect on brainstem, potentially causing hydrocephalus [10]. If the VS are detected bilaterally, it's a commonly associated with Neurofibromatosis type II, but in most of cases these type tumor have sporadic form (nearly 95% of cases). VS grow from the inferior vestibular nerve in most of the cases with rare cases that arise from cochler portion of the nerve or the superior vestibular nerve [11]. Our case emphasizes the importance of early audiometric screening and MRI evaluation. MRI with contrast-enhanced T1-weighted and high-resolution T2-weighted sequences is the gold standard for diagnosis, staging (Koos scale) and follow-up after radiosurgery. In our patient, MRI CPA was a gold standard for identifying VS. With Magnetic

Resonance Imaging (MRI) we can see the site and extension of the lesions as well as the characteristic signal [12]. Several studies have proposed protocols for diagnosing VS based on clinical manifestations. In cases with suspected VS, the first test is tonal audiometry. Imaging methods are the most effective examination for early diagnosis and a better choice of VS treatment. The Koos grading scale is one of the most common used system to classify tumour size with respect to extrameatal propagation and sing of brainstem compression (Fig. 6,7) [13]. It was proposed that MRI is the method of choice for diagnosis due to its high sensitivity and contrast resolution in assessing the structures of the posterior fossa. American Academy of Otolaryngology and Head and Neck Surgery (AAO-HNS) protocol, recommends MRI screening for patients with a mean asymmetry of ≥ 15 dB at frequencies of 0.5 kHz-3 kHz [5]. Also, there are some examples of authors believe that using audiometric and clinical data, deep learning-based analyses failed to produce an adequately model for early detection of patients with VS. It was pointed out that a “golden ticket” for diagnosing VS based on a patient’s clinical and audiometric data [6]. A new study approached the longstanding challenge of identifying which patients require MRI screening for VS using a novel machine learning method to predict the presence of VS based on clinical and audiometrical studies [6]. MRI is the method of choice for the identification of suspected VS, with contrast-enhanced T1-weighted 3D scans for the initial evaluation and postirradiation assessment of recurrence or residual tumors is gold standard. The MRI protocol should include standard T1- and T2 (heavily)-weighted 3D sequences, a fluid-attenuated inversion recovery sequence and Diffusion-Weighted Imaging (DWI).

Koos grade	Description
I	Intracanalicular
II	Extension into cerebellopontine angle, < 2 cm
III	Occupies cerebellopontine angle, no brainstem displacement, < 3 cm
IV	Brainstem displacement, > 3 cm

Figure 6: The Koos grading scale for Vestibular Schwannomas (VS).

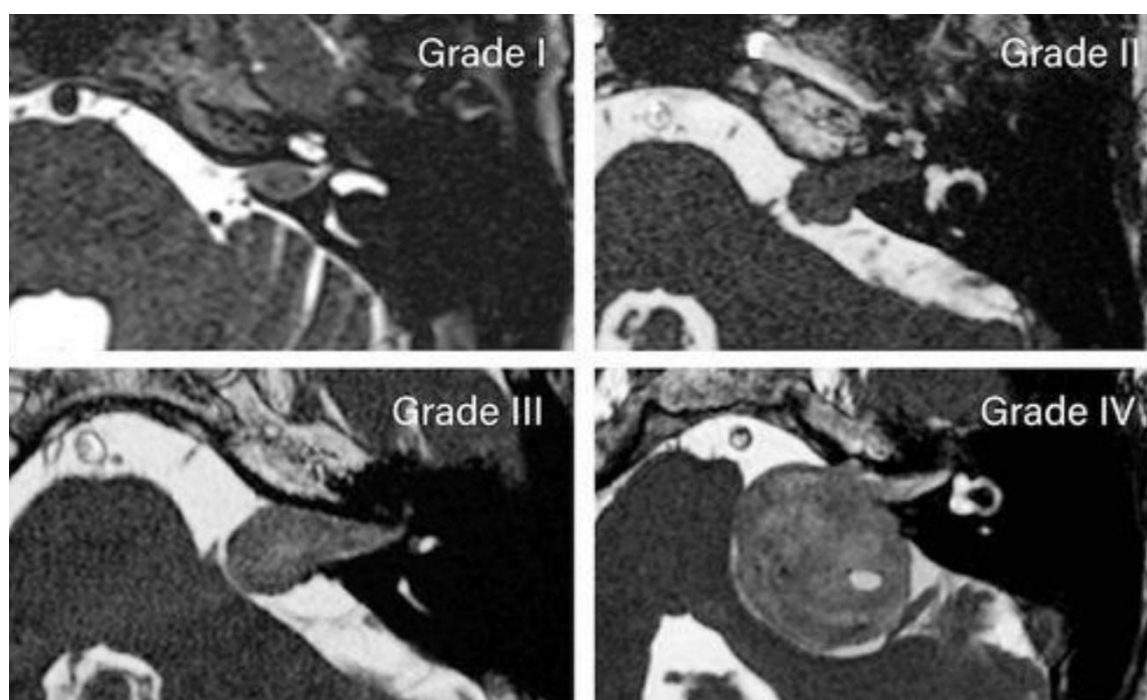


Figure 7: Example of Koos grading scale based on MR of pontocerebellar angle on T2W sequences.

Generally accepted recommendations include four steps

1. Patients with asymmetric SNHL should undergo VS screening with MRI. This is a strong recommendation with a evidence from many studies
2. In asymmetric SNHL, MRI should be performed regardless of ABR results. Strong recommendation
3. Normal results of vestibular tests do not exclude the need for MRI, but vestibular tests should be conducted in patients with suspected VS as first step in diagnosis
4. There is insufficient evidence in patients with asymmetric SNHL that should undergo ABR testing alone, without needing an MRI
5. Recommending an early post-radiation MR examination can anticipate the response to GKSR
6. We agree with the already accepted definition of tumor control as tumor volume at 3 years lower than baseline

Conclusion

Vestibular schwannomas often present with subtle otologic symptoms, which may delay diagnosis. MRI remains the most precise radiological imaging for diagnosis and follow-up. Post-radiation changes must be carefully interpreted to distinguish treatment effects from tumor progression. Early recognition and multidisciplinary management are key to optimizing outcomes.

Conflict of Interest

Authors declare they have no conflict of interest related to this paper.

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