

Sudden Deafness as a Presenting Symptom of Acoustic Neuroma: Case Report

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DOI: <https://doi.org/10.36347/sasjm.2026.v12i10.018>

| Received: 17.08.2026 | Accepted: 26.10.2026 | Published: 29.10.2026

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Abstract

Case Report

Vestibular schwannoma, also known as acoustic neuroma, is a benign tumor arising from the vestibular nerve. Sudden hearing loss revealing this condition is rare, as the disease often develops insidiously, it is usually above 30 dB, involving three or more contiguous frequencies, and sets in in less than three days. We report the case of sudden right-sided hearing loss in a 31-year-old patient; the finding of ipsilateral vestibular areflexia without true vertigo provided further grounds for performing magnetic resonance imaging, which revealed a Koos stage 3 vestibular schwannoma. This case illustrates the major importance of imaging in early diagnosis and appropriate management of vestibular schwannoma.

Keywords: Sudden Deafness, acoustic neurinoma, imaging, vestibular areflexia.

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INTRODUCTION

Sudden sensorineural hearing loss (SSNHL) is defined as a rapid-onset sensorineural hearing loss of ≥ 30 decibels (dB), affecting at least 3 consecutive frequencies that occurred within 72 h with no identifiable cause [1]. Most pathogenic hypotheses stipulate a viral or vascular origin, but the possibility of acoustic neuroma (AN) should always be kept in mind, especially if it is accompanied by certain arguments such as an extension of latencies on auditory brainstem response (ABR) or a low-grade, ipsilateral vestibular areflexia developed without true vertigo [2]. magnetic resonance imaging (MRI) or ABR are the gold standard for the retrocochlear pathology evaluation of patients with SSNHL according to the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) guidelines and most of authors [3].

CASE REPORT

A 31-year-old man with a history of malaria treated in 2023 following a trip to Central Africa presented to the ENT emergency department with acute-onset hearing loss in the right ear (developing within less than 24 hours), unaccompanied by vertigo or associated

neurological signs. ENT examination was unremarkable, notably showing normal-appearing tympanic membranes and a normal vestibular assessment. Pure-tone audiometry revealed profound right-sided hearing loss with a mean threshold of 100 dB across all frequencies; speech audiometry was consistent with these findings, showing a maximum speech intelligibility score of 30% (**Figure 1**). A diagnosis of sudden sensorineural hearing loss was made, and the patient was hospitalized for further evaluation and treatment.

High-dose corticosteroid therapy (2 mg/kg/day) was initiated, combined with antiviral and vasodilator treatments and hyperbaric oxygen therapy. Further investigations were ordered to complete the clinical assessment; notably, auditory brainstem response (ABR) testing revealed a hearing threshold of 90 dB on the right side with normal hearing on the left side with no latency prolongation (**Figure 2**).

Vestibular testing (video nystagmography and video head impulse test) showed 100% right-sided vestibular areflexia and severely reduced gains, despite the patient reporting no vertigo or balance disorders (**Figure 3**).

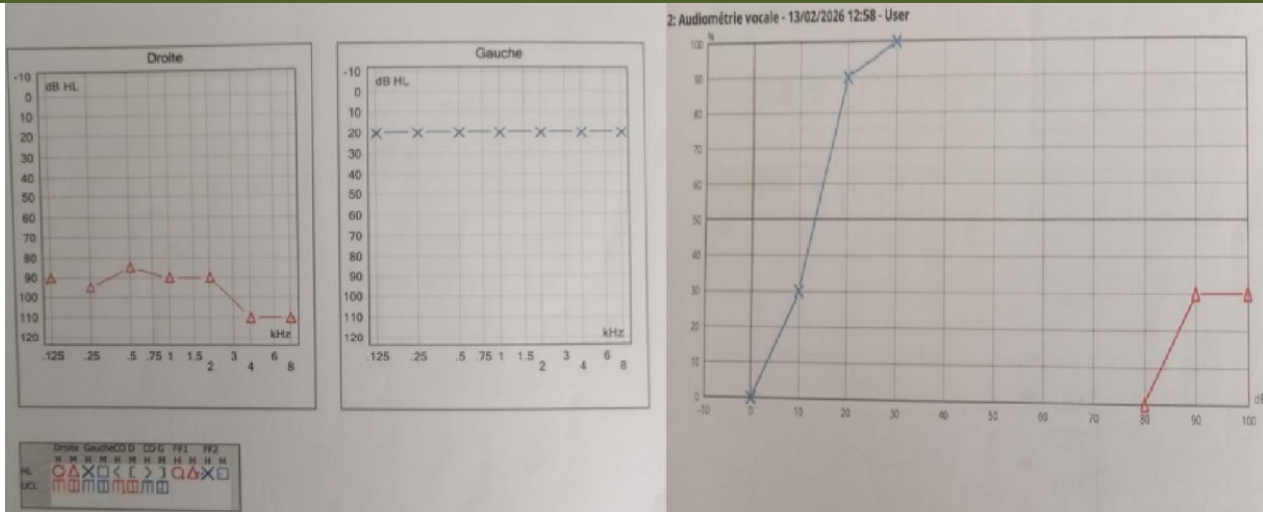


Figure 1: Pure-tone audiometry and speech audiometry showing profound right-sided hearing loss with a mean threshold of 100 dB across all frequencies and a maximum speech intelligibility score of 30%

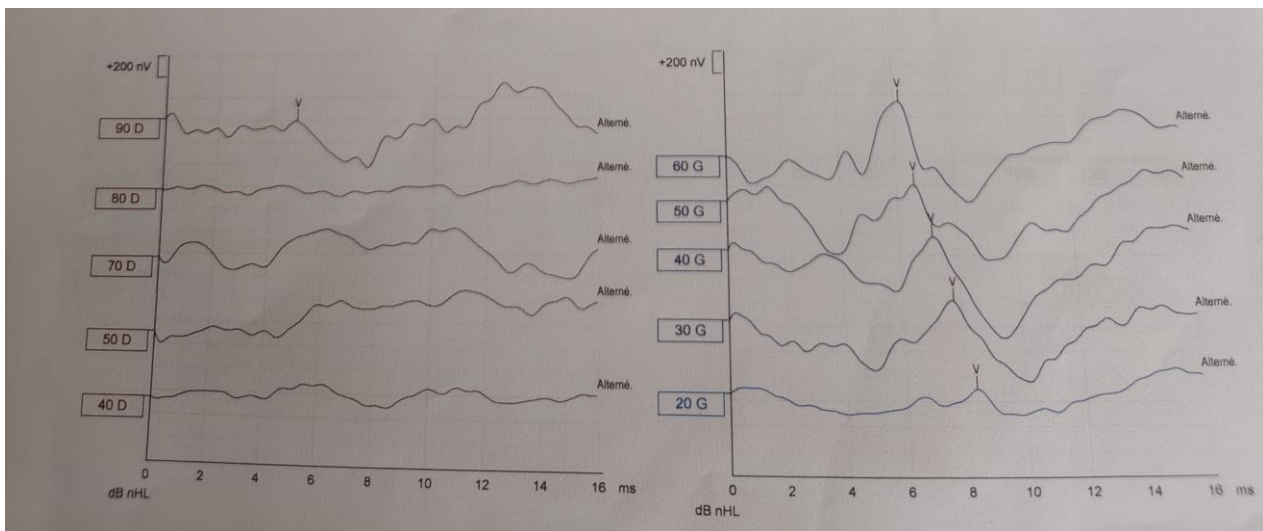


Figure 2: Auditory brainstem response (ABR) testing revealed a hearing threshold of 90 dB on the right side with normal hearing on the left side, latency obtained was normal

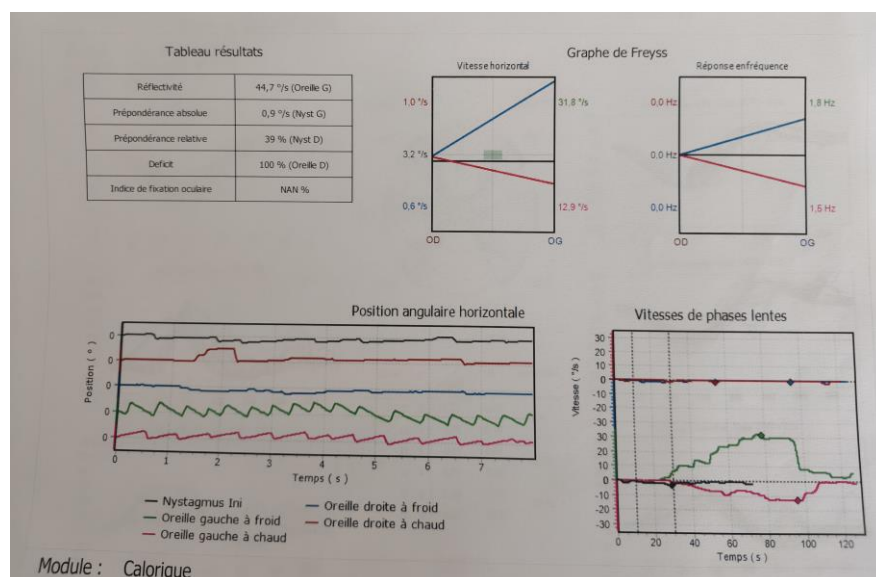


Figure 3: caloric test in video nystagmography showed 100% right-sided vestibular areflexia

The clinical course was stable; magnetic resonance imaging (MRI) was requested and revealed an expansile process in the right cerebellopontine angle that enhanced following gadolinium injection. The lesion showed no diffusion restriction, extended into the internal auditory canal, and exerted mass effect on the pons and the middle cerebellar peduncle. A Koos grade

3 classification was proposed, given the absence of neurological impact (**Figure 4**). All therapies were consequently interrupted, and the patient was scheduled for surgical excision via the translabyrinthine approach, given the size of the tumor and the lack of functional hearing.

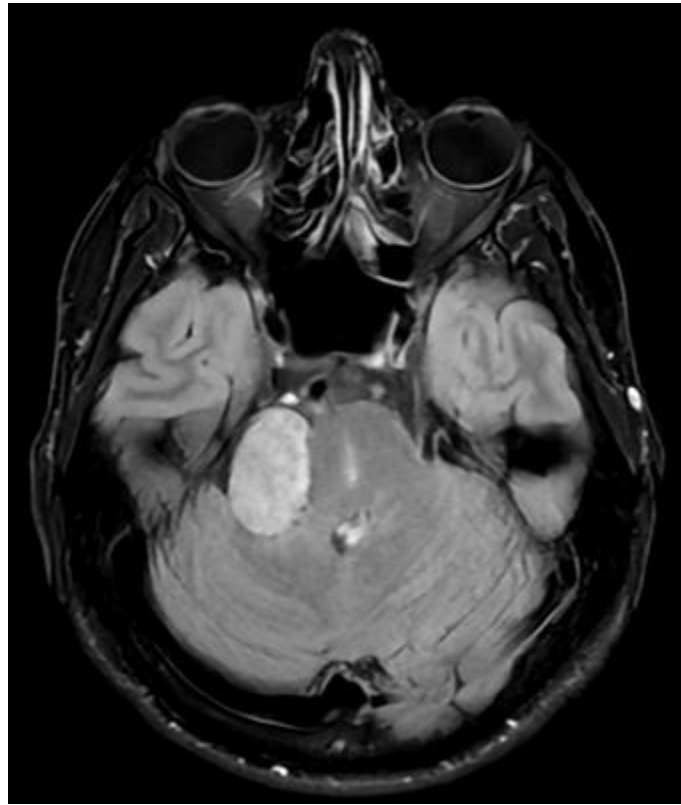


Figure 4: Axial MRI in T1 sequence showing an acoustic neuroma Kroos 3

DISCUSSION

The prevalence of AN in patients presenting with SSNHL has increased from 1.8 to 5.2% given the widespread use of MRI [4]. Diagnosing SSNHL is simple, relying on well-defined criteria (a loss of at least 30 dB across three consecutive frequencies within 24 hours); however, its pathophysiological mechanism remains unknown with viral or vascular theories predominating largely due to the lack of direct examination methods for the inner ear. The main differential diagnoses are acoustic neuroma and vertebrobasilar artery dissections, which can present with sudden hearing loss.

Clinically, unilateral sensorineural hearing loss especially associated with unilateral tinnitus should always raise the suspicion of AN. The presence of associated vertigo is also typical though it might also suggest a diagnosis of Ménière's disease, but the hearing loss predominantly affects high frequencies in SSNHL rather than low frequencies. Audiograms are generally classified into low-frequency ascending, high-frequency descending, flat-type, and profound hearing loss and

some studies found the incidence of a trough-shaped audiogram was significantly higher in patients with acoustic neuroma than in those without and thus suggesting that a trough audiogram is a significant finding in patients with SSNHL and indicates the presence of AN [5]. Song and al found that there was no obvious relation between tumor size and hearing loss [6], a Koos stage 3 or 4 tumor can thus have a minimal impact on hearing despite its size, and vice versa.

The ABR finding remains valid, even though a few cases of neuromas with normal ABR have been described especially those of small size [7]. The sensitivity of this test reported in the literature varies widely and is strongly correlated with tumor size. The key element confirming the diagnosis is MRI with the combination of the two sequences T1- and T2-weighted, this imaging is recommended by most practitioners, and its sensitivity is very higher and varies between 96–100% [8]. Despite its cost, the increasing widespread application of MRI, a higher rate than expected of acoustic neuromas has been detected in patients with sudden hearing loss.

Corticosteroid treatment can, in some cases, lead to hearing improvement, which can sometimes be a source of confusion by not requiring an MRI. Corticosteroids can indeed reduce edema and perilesional inflammatory phenomena, thus improving symptoms [6]. The rate of hearing recovery reported in the literature varies considerably. According to a study by Nageris *et al* [9], hearing recovery in the presence of an acoustic neuroma was observed in 16.7% of cases; all of these tumors were intracanalicular.

CONCLUSION

for patients with SSNHL, ABR and MRI are recommended and should be combined to improve the sensibility of detection of AN and prevent misdiagnosis and missed diagnoses, especially for the smaller one. Therapeutic response to corticosteroid treatment for SSNHL does not exclude the presence of AN and all patients with SSNHL should undergo MRI to prevent misdiagnosis and delays in potential treatment.

DECLARATIONS

Consent for publication: Was provided and signed by the patient.

Funding: no external funding was received.

Conflicts of interest: the authors declare no conflicts of interest.

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