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Acoustic Schwannoma Characterized by Asymmetrical Sensorineural Hearing Loss and Ipsilateral Tinnitus: A Case Report

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Abstract

Acoustic neuromas, or vestibular schwannomas, are benign neoplasms originating from the Schwann cell sheath of the vestibular nerve. Approximately 10% of patients complain of atypical symptoms including facial numbness or pain and sudden onset of hearing loss. Moreover, patients with these symptoms are more likely to have large tumors due to delayed assessment.

Herein, we report a 54-year-old female patient with a history of progressive headache, imbalance, tinnitus, and sensorineural hearing loss. There were no abnormalities on tympanometry. Audiogram showed that the left ear had a normal hearing level. However, the right ear presented with minimal to severe high-frequency sloping sensorineural hearing loss. Magnetic resonance imaging was performed, and results revealed right-sided spheric mass forming acute angles with the petrous bone. Hence, the patient was diagnosed with benign neoplasm of the cranial nerves (cerebellopontine angle schwannoma). She underwent craniotomy and tumor excision with external ventricular drain insertion.

Keywords: Acoustic neuromas; Hearing loss; Tinnitus; Reterosigmoid craniotomy; Imbalance; SNHL

Introduction

Vestibular schwannomas (VS), also known as acoustic neuromas, are benign neoplasms originating from the Schwann cells of the eighth cranial nerve and are most commonly observed in individuals aged between 30 and 60 years [1]. The prevalence of VS is high at 1.4-2.1 in 10.000 people [2,3], and VS accounts for 5%-10% of all intracranial tumors and 80%-90% of cerebellopontine angle (CPA) neoplasms [4]. The symptoms of VS include slowly progressive sensorineural hearing loss, feeling of ear fullness, and tinnitus associated with vestibular disorders in 16%-70% of cases. Balance disorders can present with acute rotatory vertigo and/or dizziness [5,6]. Medical history taking, clinical test, and routine audio-vestibular examination alone cannot diagnose VS. The diagnosis must be confirmed via high-resolution intravenous (IV) gadolinium-based contrast-enhanced magnetic resonance imaging (MRI) [7]. Herein, we report a patient with progressive headache, imbalance, tinnitus, and sensorineural hearing loss (SNHL) who was admitted to the hospital and diagnosed with benign neoplasm of the cranial nerves. She then underwent craniotomy and CPA schwannoma excision with external ventricular drain insertion.

Case Description

A 54-year-old female patient presented with hearing loss, which was preceded by tinnitus for a period of time, imbalance, and headache diagnosed as migraine for a long period. She had a history of non-insulin-dependent diabetes mellitus, dyslipidemia, and headache, which was believed to be associated with a visual problem. Thus, she underwent cataract surgery. However, her symptoms did not subside. No other manifestations were observed except for persistent whole-body paresthesia mainly caused by hypercalcemia.

Physical examination

Hearing and balance assessment was performed.

Other evaluations

Tympanometry showed that the volume of the bilateral normal external ear canals, middle ear pressure, and middle ear compliance type A were normal (Figure 1).

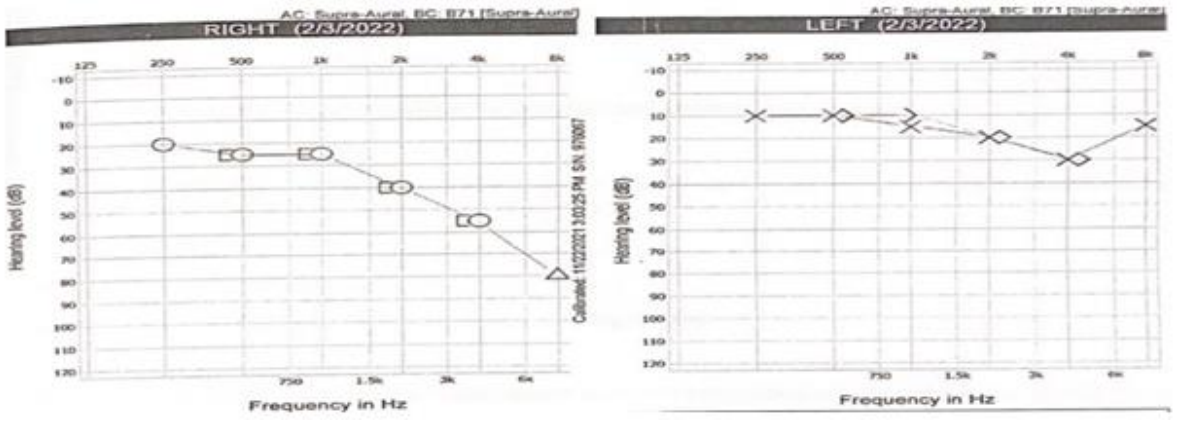

Figure 1: Tympanometry of both ears (type A).

Audiogram revealed that the left ear had a normal hearing level. However, the right ear presented with minimal to severe high-frequency sloping SNHL (Figure 2).

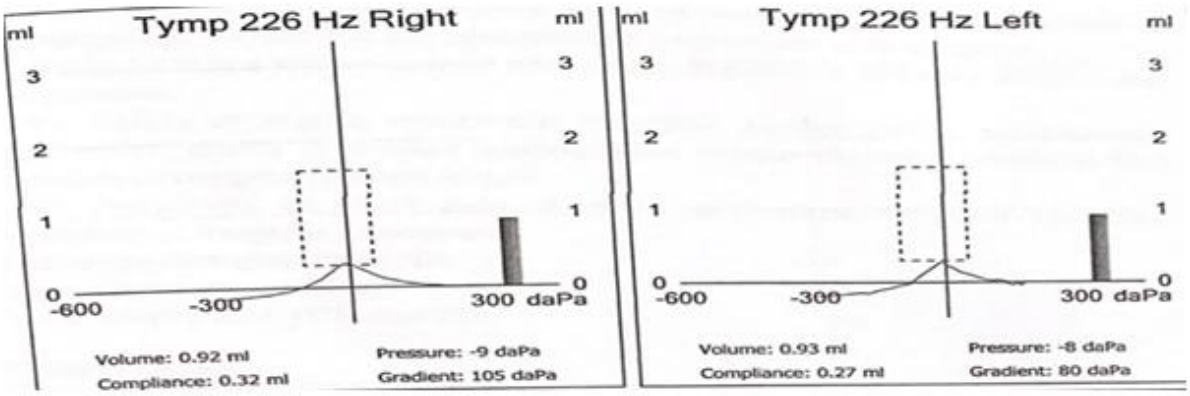

Figure 2: Audiogram showed that the left ear had a normal hearing level. However, the right ear presented with minimal to severe high-frequency sloping sensory neural hearing loss.

MRI was performed, and results showed a right-sided spheric mass forming acute angles with the petrous bone. The mass was located at the center over the internal acoustic canal (IAC), and a funnel-shaped portion extended directly into the widened canal. T1-weighted imaging revealed a slightly heterogeneous mass with an oblong shape. The long axis was located at the center over the IAC, and the tumor extended into the widened canal. Features such as pseudocapsule of the displaced vessels and widening of the ipsilateral CPA and lateral pontine cisterns caused by the displacement of the pons toward the left side indicated an extra-axial location. The mass was bulging and convex toward the brain stem and cerebellum. It was invaginating and deforming the pons and cerebellum, and the fourth ventricle was compressed and displaced causing obstructive hydrocephalus with dilation of the lateral and third ventricle associated with periventricular interstitial edema. The contralateral lateral brain stem cisterns (pontine and medullary) were effaced. On gadolinium-enhanced T1-weighted MRI, the mass had a heterogeneous appearance with multiple cystic structures/degenerations. The MRI features were highly indicative of VS (Figure 3).

The patient had been complaining of symptoms and seeking medical care for 3 years. However, she was not diagnosed during this period. Finally, a diagnosis was obtained now.

Outcome and follow-up

A patient with benign neoplasm of the cranial nerves was admitted to the hospital. She underwent craniotomy and tumor excision with external ventricular drain insertion. In terms of surgical approach, retrosigmoid craniotomy was conducted. The resected tumor, which was in multiple pieces of soft gray, brown tissues measuring 4.5*3*2 cm in four cassettes, was sent to the histopathological department.

Postoperatively, the patient received cefuroxime (Zinacef) 750 mg TDS, dexamethasone 4 mg TDS, Perfalgan 1 g every 6 h, Losec 20 mg IV BD, Zofran 4 mg IV PRN, and morphine 2 mg IV PRN. Facial palsy was observed on the 7th day after the surgery. Balance when standing was fairly static and dynamic postoperatively.

Electromyography was performed 8 days after surgery, and results showed signs of right facial nerve lesion.

The histopathology report was follows. The sections showed lesional fragments of a biphasic tumor with hypo- and hypercellular areas, and the latter areas were predominant. The lesion features intersecting the fascicles of spindle to ovoid cells with mild to moderate regenerative atypia. In addition, hyalinized blood vessels and focal xanthomatous changes/presence of hemosiderin laden macrophages and scattered lymphocytes were noted. Nevertheless, there was no evidence of necrosis or mitosis.

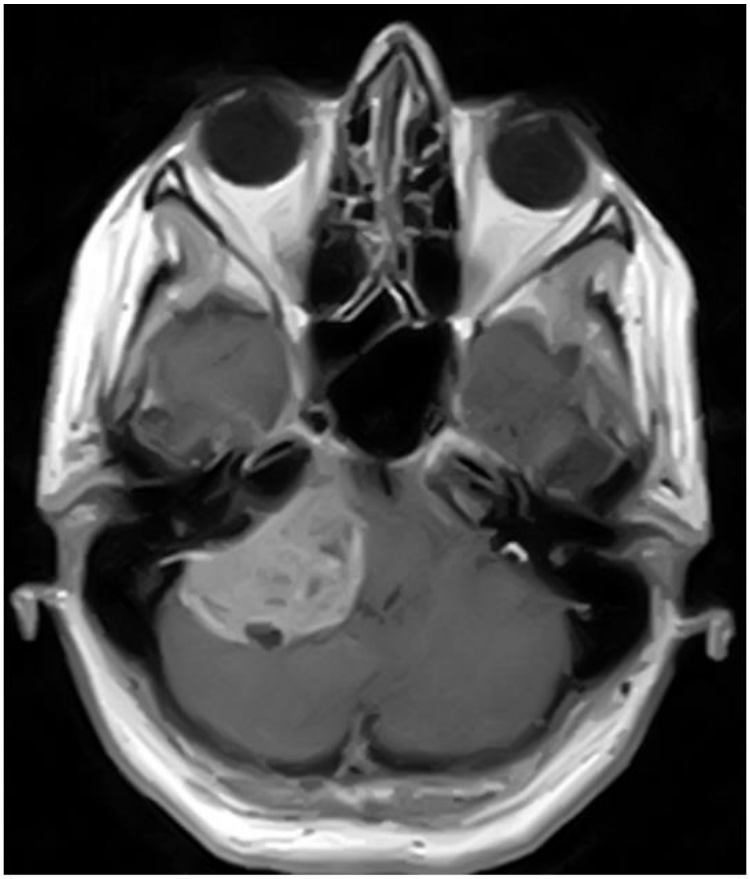

Figure 3: Magnetic resonance imaging revealed a mass located at the center over the internal acoustic canal, and there was a funnel-shaped portion that extended directly into the widened canal.

Discussion

Acoustic neuroma is allied to neurofibromatosis type 2 (defect on chromosome 22) bilateral disease. Previous studies have shown that acoustic neuroma is caused by a specific mutation. Further, radiation exposure may be associated with tumor development [8].

The CPA is a three-sided area in the posterior cranial fossa that is limited superiorly by the tentorium, posteromedially by the brainstem, and posterolaterally by the petrous part of the temporal bone. This area is occupied by the CPA cistern, which houses the cranial nerves V, VI, VII, and VIII and the anterior inferior cerebellar artery [9]. CPA tumors are commonly benign, and they grow gradually [10]. It is challenging to optimally resect tumors in the CPA due to the anatomical complexity of the area. That is, it is deep, narrow, and abundant with vital neurovascular structures [11,12].

VS, meningioma, and epidermoid tumors are the most common tumors found at the CPA, with VS accounts for 75%-85% of CPA tumors, and the incidence is 1 per 100,000 persons in the United States [9,13,14]. Acoustic schwannoma is a tumor that evolves from Schwann cell sheath and can be either intracranial or extra-axial. They originate around the cochlear or vestibular nerve [15]. It involved the facial nerve in approximately 6% of people [16]. Acoustic neuroma is more common among male adults. In addition, it frequently occurs in the right CPA [17]. In another previous study, VS was found to affect the right and left sides with equal frequency in older patients, and that they occur equally in both sexes [18].

The treatment options include surgery, radiation, and observation. The management depends on tumor size, clinical presentation, patient's age, comorbidities, and preference [19].

Stangerup et al. performed a large population-based prospective study with a relatively small fraction of ANs (30% of extracanalicular tumors and 17% of intracanalicular tumors) that grow over several years. Regardless of tumor location or size, the tumor significantly grows within the first 5 years after diagnosis [20]. The growth occurrence or rate was not correlated with sex and age. With a better understanding of the natural an history, the treatment has shifted toward conservative management.

According to morphologies and extensions from the IAC to the CPA, Moffat et al. divided an into, dumbbell, cone, and lollipop-shaped tumors. If the tumor sizes reaches 20 mm, it extends out of the porus acusticus and touches the trigeminal nerve. Therefore, when tumors extend out of the IAC into the CPA, and there will be more involvement of the trigeminal nerve and less involvement of hearing in patients (https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6855845/ - R6) [21]. In particular, involvement of the ophthalmic branch of the trigeminal nerve (VI) is a serious complication due to absence of corneal reflex, which exposes patients to dry eyes and subsequent keratitis particularly if there is grade 4 or higher facial nerve paralysis based on the House–Brackmann classification system.

Moreover, Karkas et al. found a significant association between preoperative trigeminal hypoesthesia and cerebellar peduncle compression on preoperative MRI [22].

Previous studies have found that the median growth rate of an is 1.8-1.9 mm/year, with most tumors only growing within the first 5 years (https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6855845/ - R8) [23].

Therefore, tumors measuring <1 cm are generally managed conservatively with annual MRI for 5 years, followed by twice-yearly MRI for 4 years and once-yearly MRI the last 5 years. Thereafter, no further follow-up imaging is required.

However, Patel et al. found that hearing threshold and speech discrimination could progressively deteriorate even if the tumor size is static (https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6855845/ - R9) [24].

Radiosurgery has favorable outcomes in different studies, with improvement of facial and trigeminal nerve function, preservation of hearing, reduced hospital stay, and better quality of life [25]. However, microsurgery was significantly superior to radiosurgery in treating patients with an who presented with trigeminal symptoms, particularly if the tumor size is >3 cm, because tumor resection and concurrent decompression of the trigeminal nerve can be performed (https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6855845/ - R11) [26].

Conclusion

VS is a benign Schwann cell-derived tumor originating from the vestibulocochlear nerve and is a rare cause of hearing loss. Although benign, it poses a threat to different intracranial structures due to mass effect, and it is associated with a low risk of malignant transformation. Therefore, it represents an important healthcare burden, and it is frequently hypovascular. Even a large VS can be completely resected without major morbidity. However, an unusual hypervascular VS is often complicated by excessive tumor bleeding. Therefore, the preoperative diagnosis of hypervascular VS is important to address surgical risks. This report aimed to review the treatment and outcome of VS in a 54-year-old female patient.

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