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Vestibular schwannoma presenting as unilateral vocal cord palsy - an unusual and atypical presentation

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Abstract

Vestibular Schwannoma (VS) is a slow-growing, benign tumor of the vestibulocochlear nerve. It commonly presents with otologic symptoms, mainly unilateral hearing loss, tinnitus, and vestibular disturbance. Other rare presentations have been reported when the tumor is large and causes compression of structures within the Cerebellopontine Angle (CPA). Advancement in the diagnosis of VS using Magnetic Resonance Imaging (MRI) has led to early diagnosis and incidental identification of the tumor when imaging is done for other non-neurotologic indications. We hereby report the case of unilateral vocal cord palsy in a patient with VS diagnosed on MRI scanning of the brain.

Key words: vestibular schwannoma, acoustic neuroma, rare presentations, vocal cord palsy, radiosurgery.

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Introduction

Vestibular Schwannomas (VS), also called acoustic neuromas, are benign tumors that arise from Schwann cells of the vestibulocochlear nerve.[1-3] They arise from the vestibular nerve typically found within the internal acoustic meatus (intracanalicular). Still, they can also project beyond the petrous portion of the temporal bone (extracanalicular) extending into the Cerebellopontine Angle (CPA).[1] VS are the most common tumors seen in the CPA (80-90%),[2,4-7] and account for 6-8% of all intracranial tumors.[3-6] The incidence of VS has increased over the last few years and this could be due to increased accessibility and widespread use of Magnetic Resonance Imaging (MRI).[1,5,6,8] The annual incidence is reported to be in the range between 0.5 and 1.9 per 100,000 persons.[1,6] The exact etiology of VS is unknown. However, some risk factors such as noise exposure and radiation have been shown to increase the risk of VS.[1] Although the use of cell phones has already been associated with the onset of VS, no clinical association has been demonstrated between cell and cordless telephone use and VS.[9]

The clinical presentation of VS may vary broadly depending on tumor extension and location. While some tumors may progress to cause life-threatening neurologic symptoms, others remain stable.[5] Typically, patients most commonly present with unilateral hearing loss, tinnitus, and vestibular symptoms of variable degree.[1-3,6,7] With further tumor growth, other rare symptoms such as headache,[3,6,7] facial hypoesthesia,[1-3,6,7] facial weakness,[1-3,6,7], ataxia,[6] lower cranial nerves damage,[6] or hydrocephalus[6,7] can manifest. Other atypical presentations of VS have also been reported in the literature.[10-13] Similarly, unilateral vocal cord palsy was reported in a case of jugular foramen schwannoma arising from cranial nerves IX and X[14] as well as in a meningioma of the CPA.[15] However, this was not reported in the case of VS to the best of our knowledge. Due to the rarity of the condition and the atypical nature of the presentation, we hereby report the case of unilateral vocal cord palsy in a patient with an incidental diagnosis of VS in our hospital.

Case Report

A 70-year-old woman was referred to our clinic with a 2-month history of progressive unremitting hoarseness, choking spells, dysphagia, and a finding of unilateral right vocal cord palsy on flexible nasoendoscopy. She had an associated cough but no difficulty with breathing. Further history revealed right intermittent tinnitus and hearing impairment that was not worrisome, for about 10 years. She has no associated vertigo or imbalance. Other Ear, Nose, and Throat (ENT) symptoms were not significant. She is a known hypertensive patient which has been under control with antihypertensive medications. She has no other comorbidities of note and no history of exposure to loud noise or exposure to radiation. The ear examination was normal, but Pure Tone Audiometry (PTA) showed moderate sensorineural hearing loss. Other ENT and systemic examinations were grossly normal.

A Computed Tomography (CT) scan of the head (Figure 1) showed an ovoid pedunculated intensely enhancing hyperdense lesion within the right posterior fossa, measuring about 22.5x22x18 mm. No evidence of mass in the lungs or the neck on

CT evaluation of the neck and chest.

MRI (Figure 2) confirmed the presence of a fairly ovoid-shaped mass in the right CPA extending into the right internal auditory canal, in keeping with VS. It measures about 20.93x18.83x20.38 mm. It appears hypo-intense on T1, hyper-intense on T2, and shows avid enhancement on post-IV Gadolinium contrast administration. The left CPAis clear.

Following a neurosurgical review and detailed discussions about appropriate treatment options based on the tumor size and the aidable hearing loss in the ipsilateral ear, she was referred for stereotactic radiosurgery.

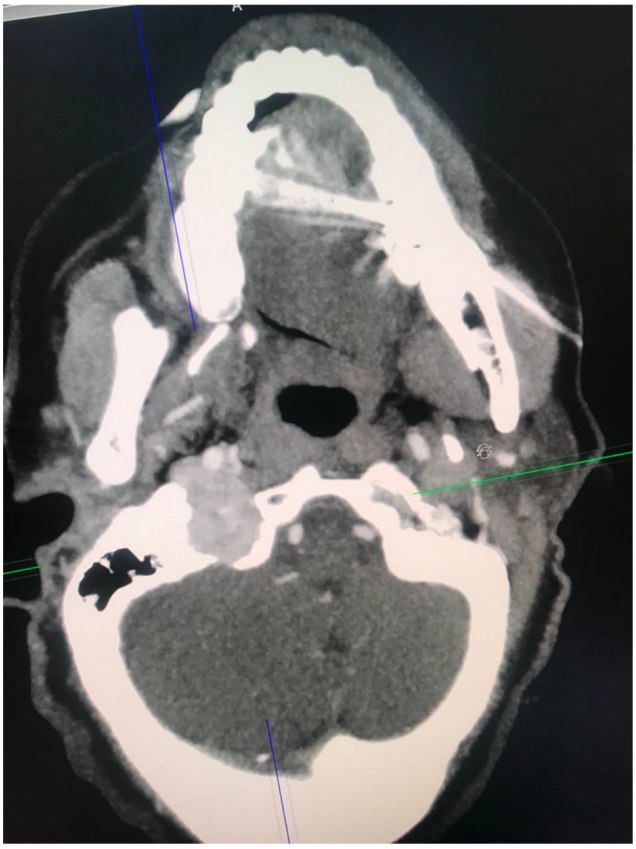

Figure 1. A Computed Tomography (CT) scan of the head.

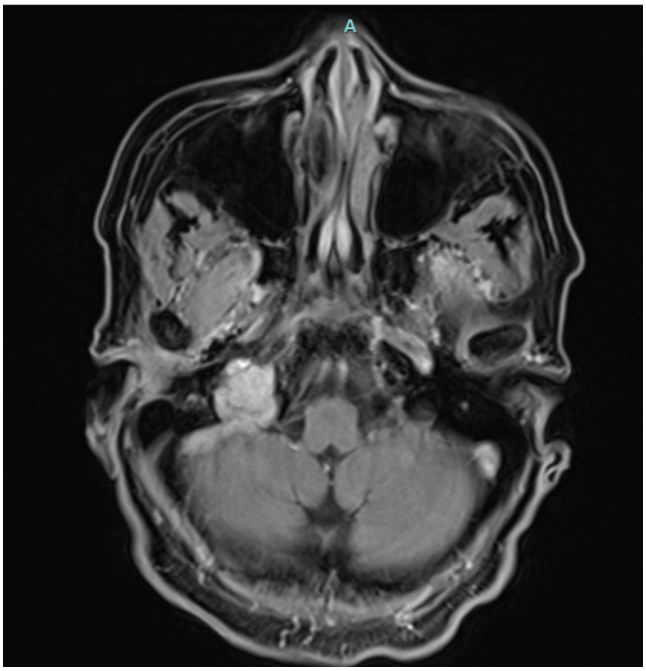

Figure 2. Magnetic Resonance Imaging (MRI) of the head.

Discussion

VSs are the third most common nonmalignant intracranial tumors after meningiomas and pituitary adenomas.[7] They arises from either the inferior or superior division of the vestibular nerve.[6] They remain within the Internal Auditory Canal (IAC) or extend into the CPA, and present with symptoms typically related to compression of adjacent Cranial Nerves (CNs), brain stem, or Posterior Fossa (PF) structures.[4] VS presents at a median age of 50 years and is unilateral in >90% of patients, with an equal incidence on the left and right.[4] The peak incidence is in the age group of 65-74 years, with no difference related to gender.[7] The clinical presentation may vary broadly depending upon tumor extension and site of origin of the tumor.[6] Most commonly, patients present with classic otologic symptoms consisting mainly of unilateral progressive sensorineural hearing loss, imbalance and unilateral tinnitus.[16] About 10-10.7% of patients presents with atypical symptoms[11,16] requiring a high index of suspicion. Once suspected, a patient should undergo Pure Tone Audiometry (PTA) and vestibular testing. Those with abnormalities on either, especially unilateral sensorineural hearing loss, should have MRI with and without contrast.[6]

MRI is currently the gold standard and the preferred technique use in the diagnosis of VS.[4,6] It has been shown to provide exquisite tumor characterization, has a role in surgical planning and in post therapeutic evaluation.[4] Contrast-enhanced CT of the temporal bones can serve as an alternative if the patient cannot undergo MRI. Furthermore, imaging plays significant role in the diagnosis of incidental VS especially in those group of patients whose presentations are atypical and underwent imaging for a non-neurotologic indication or clinical suspicions, accounting to about 12.3% of cases in some study population.[3]

In our index case, the patient presented with atypical symptoms of unilateral vocal cord palsy. History of hearing loss and tinnitus were not significant, except for PTA which showed an ipsilateral moderate SNHL. The diagnosis of VS was based on suspicion from CT scan which was confirmed by MRI. The absence of hearing loss as an early presentation in the patient could be explained by location of the tumor. It has been shown that patients with lateral neuromas generally have small tumors, sometimes only located in the internal auditory canal, and present early subjective hearing loss, while patients with medial neuromas have large tumors which grow without causing significant audiological symptoms.[17] The treatment options of VS include observation with the use of serial MRIs, microsurgical resection, stereotactic radiosurgery, and fractionated radiotherapy.[1] The choice of treatment is dependent on patient age, health status, clinical symptoms, tumor size, and patient preference.[1] Tumor characteristics such as initial size and growth on serial imaging (often >2 mm between scans) are commonly associated with the

decision to start treatment. Surgery is the mainstay of treatment because it removes the tumor, allowing histologic evaluation.[7] Nowadays, preservation of function is considered a primary aim in VS treatment. There is therefore widespread use of radiosurgery in the treatment of VS, since it has the potential to stop the growth of the tumor with minimal side effects and with a very high level of preservation of nerve functions.[6] Following neurosurgical review and discussion of treatment options, stereotactic radiosurgery was recommended to the index patient based on her age, the size of the tumor (<2.5cm) as well as the presence of aidable hearing loss in the ipsilateral ear, as it was associated with minimal morbidity and has better function preservation when compared with open surgical approaches. Due to the lack of this treatment option in our facility and nearby hospitals, she was referred elsewhere and lost to follow up. This is a limitation of this case report as patient's outcome as well as her treatment response could not be ascertained. Since the presentation and natural history of VS varies between patients, the evaluation process, treatment options as well as well follow up should be individualized.

Conclusions

VS has a wide spectrum of manifestations and can present with rare, and sometimes atypical features such as vocal cord palsy. High index of suspicion and thorough clinical evaluation is needed to unravel the diagnosis especially in the absence of the classic otologic symptoms.

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