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Successful Treatment of a Rare Glossopharyngeal Schwannoma: A Case Report

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Abstract

Background: Glossopharyngeal schwannomas are rare tumors, often presenting with symptoms that mimic more common vestibular schwannomas. This case report describes a 64-year-old male with a history of hypertension, who presented with progressive right-sided hypoacusis, signs of increased intracranial pressure, and dizziness over a four-month period. Neurological examination revealed a static cerebellar syndrome and vestibular dysfunction. Imaging, including CT and MRI, identified an extra-axial mass in the right posterior cranial fossa, consistent with a glossopharyngeal schwannoma. Surgical resection was performed using a right-lateralized midline suboccipital approach, leading to complete excision of the tumor. Histopathological examination confirmed the diagnosis. Postoperative recovery was uneventful, with no complications such as facial paralysis or swallowing difficulties. This case highlights the importance of recognizing rare glossopharyngeal schwannomas in differential diagnoses and demonstrates successful management through surgical intervention, with favorable outcomes for the patient.

Subject Areas

Surgery & Surgical Specialties

Keywords

Glossopharyngeal Schwannomas, Cranial Nerve Tumors, Jugular Foramen Tumors, Cerebellopontine Angle

1. Introduction

Glossopharyngeal (that is, cranial nerve IX) schwannomas are extremely rare

nerve sheath tumors that frequently mimic the more common vestibular schwannoma in their clinical as well as radiographic presentation [1].

The clinical presentation of glossopharyngeal schwannomas often overlaps with other more prevalent cranial nerve pathologies, especially vestibular schwannomas, which affect the vestibulocochlear nerve (cranial nerve VIII).

In this article, we report the case of a 64-year-old male patient diagnosed with a glossopharyngeal schwannoma following a thorough clinical evaluation and detailed imaging studies. This case highlights not only the rarity of these tumors but also the diagnostic and therapeutic challenges they present.

2. Patient and Observation

We report a case of 64-year-old male with hypertension, treated with amlodipine, who has been complaining of progressive right-sided hypoacusis for the past 4 months, associated with signs of increased intracranial pressure and dizziness. His symptoms had gradually worsened, prompting further investigation.

Upon admission, the patient is conscious, with a supple neck. Neurological examination reveals a static cerebellar syndrome associated with a vestibular syndrome. Ophthalmological examination shows a visual acuity of 8/10 in the right eye (OD) with bilateral stage 2 papilledema. Audiometry shows right-sided conductive hearing loss.

Initially, the patient underwent a cerebral CT scan (**Figure 1(a)** & **Figure 1(b)**), which revealed an extra-axial tissue mass in the right posterior cranial fossa (PCA), which enhances heterogeneously after contrast injection, with hypodense areas, without extension into the internal auditory canal (IAC) or widening of the IAC.

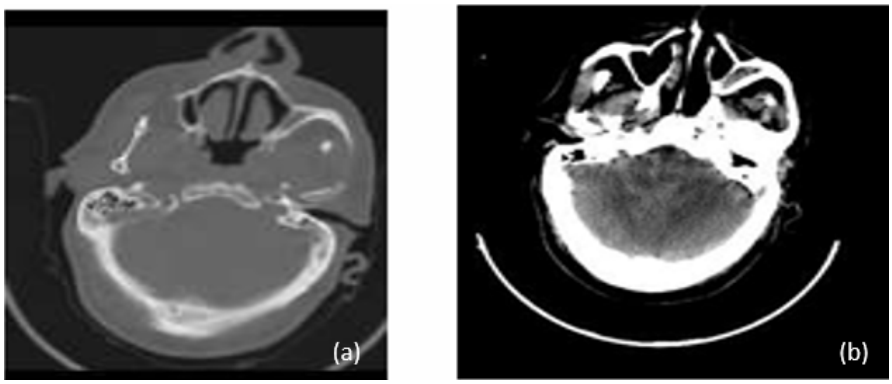

Figure 1. CT scan of the brain with a bone window shows no widening of the right internal auditory canal.

Brain MRI (**Figure 2**) shows an extra-axial process in the right ponto-cerebellar angle, appearing hypointense on T1, heterogeneously hyperintense on T2 and FLAIR, and enhancing moderately and heterogeneously, with cystic areas. The mass infiltrates the fourth ventricle and exerts a mass effect, resulting in triventricular hydrocephalus.

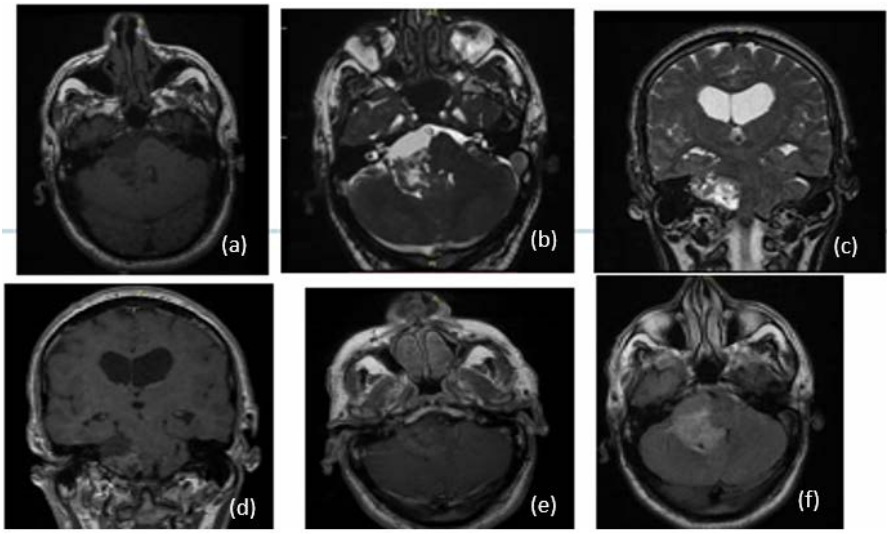

Figure 2. (a) Brain MRI shows an extra-axial process in the right ponto-cerebellar angle, hypointense on T1 (a) (d) (e), heterogeneously hyperintense on T2 (b) (c), enhancing moderately and heterogeneously, with cystic areas (f).

Upon the initial presentation of the patient, the clinical symptoms—including progressive right-sided hypoacusis, signs of increased intracranial pressure, dizziness, and the results from the imaging studies prompted a thorough differential diagnosis for cerebellopontine angle (CPA) tumors. Several potential diagnoses were considered, given the overlapping clinical features of these tumors. These included: Vestibular Schwannoma, Meningiomas, Epidermoid Cysts.

The patient was operated on via a right-lateralized midline suboccipital approach.at surgery, a well-defined cystic mass was found which was soft, suckable and moderately vascular (**Figure 3**). It was debulked under microscope, 7th and

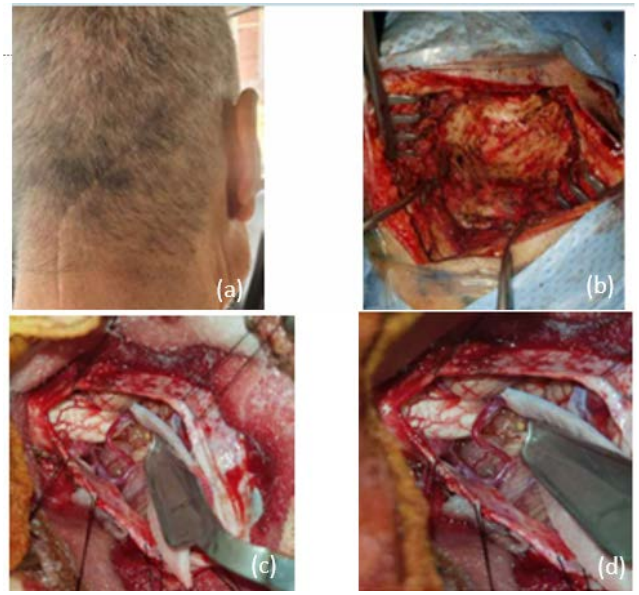

Figure 3. (a) right-lateralized midline suboccipital approach, (b)-(d) intraoperative image showing a cystic mass arising from 9th cranial nerve.

8th nerve complex was seen pushed up and anteriorly and tumor was seen to be arising from 9th cranial nerve (glossopharyngeal nerve). Total excision of the mass was done. Histopathology was consistent with schwannoma.

Postoperatively, the patient did not present a facial paralysis, swallowing difficulties, or any other signs of mixed nerve damage.

The patient's postoperative course was uneventful; the patient's neurological status remained good. Postoperative MRI (**Figure 4**) confirmed total removal of the lesion he stayed in the intensive care unit for two days after the operation and was then transferred to the Neurosurgery Department without post-operative complication.

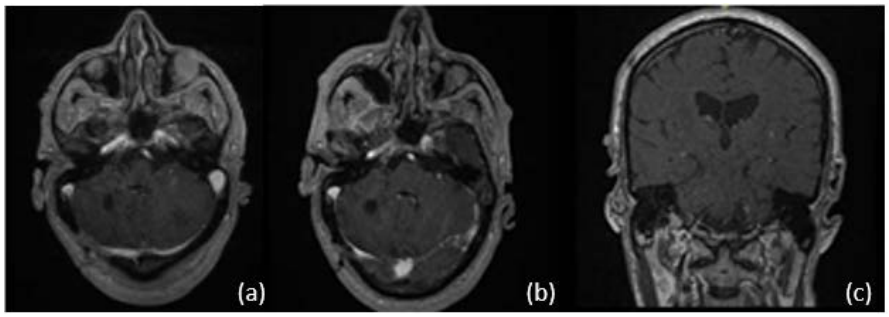

Figure 4. Postperative axial T1-weighted postcontrast (a) (b), postcontrast sagittal T1-weighted (b) postcontrast T1-weighted coronal (c) confirmed complete excision of the tumor.

3. Discussion

Intracranial schwannomas represent approximately 7% - 10% of all primary intracranial tumors, with vestibular schwannomas making up the majority, and non-vestibular schwannomas accounting for around 0.8% - 8% of these tumors. Among non-vestibular schwannomas, the most commonly affected cranial nerves are the 5th, 7th, and 12th, in decreasing frequency. Glossopharyngeal schwannomas are relatively rare and are often grouped with jugular foramen schwannomas. These tumors most commonly affect men, and their age of presentation typically ranges from the third to the fifth decade, with the youngest reported case being a four-year-old male [2].

Clinically, intracranial schwannomas are usually characterized by local cranial nerve dysfunction. Due to the confined space of the posterior fossa, multiple cranial nerves may be involved simultaneously. Palsies of the ninth cranial nerve are unusual, and symptoms of ninth nerve dysfunction may only appear with bilateral involvement. These schwannomas often grow toward the cerebellopontine angle, initially affecting the facial-acoustic nerve complex, which causes hearing loss in 90% - 93% of cases. Hoarseness and a decreased gag reflex are also common symptoms [3] [4].

Radiologically, schwannomas in the cerebellopontine angle are typically hypodense on plain CT scans with moderate enhancement following contrast administration, and the internal auditory canal remains normal. There may be sharp-edged enlargement of the jugular foramen, and MRI scans can show the mass

extending into the jugular foramen, with the VII-VIII nerve complex visible anterior to the mass. However, this is not conclusive for a ninth nerve schwannoma, as such tumors can remain confined to the cerebellopontine angle without causing enlargement of the jugular foramen. In the case presented, the radiological findings were consistent with these descriptions, though there was no evidence of the mass extending into the internal auditory canal or the jugular foramen [5].

Total surgical removal is the standard treatment for these tumors. However, the management of these lesions is complicated due to their complex anatomical location and potential postoperative complications. Samii and Tatagiba classified tumor extension into four types based on radiological and surgical features: Type A (located at the cerebellopontine angle with minimal enlargement of the jugular foramen), Type B (primarily in the jugular foramen with intracranial extension), Type C (extracranial tumor extending into the jugular foramen), and Type D (dumbbell-shaped tumor with both intra- and extracranial components) [5]. Kaye *et al.* further classified them into three types, while Pellet *et al.* added a Type D, which is described as a saddlebag-shaped tumor with both intracranial and extracranial components. Type A tumors are the most common, primarily located at the cerebellopontine angle with minimal enlargement of the jugular foramen. In such cases, the diagnosis of glossopharyngeal schwannoma is usually confirmed during surgery when the tumor's attachment to the ninth nerve is observed [5]-[7].

Surgical excision remains the preferred treatment for glossopharyngeal schwannoma, with the goal of preserving the ninth nerve. If resection is incomplete, clinical and radiological follow-up is essential, as these tumors may continue to grow. Gamma knife radiosurgery is a viable alternative for patients with small tumors and preserved lower cranial nerve function, as well as for those with residual or recurrent tumors following microsurgery [8] [9]. The prognosis for glossopharyngeal schwannomas is generally favorable, especially when the underlying nerve is preserved [10].

4. Conclusion

Glossopharyngeal nerve schwannomas are exceedingly rare, and their clinical and radiological features often resemble those of acoustic schwannomas.

Conflicts of Interest

The authors declare no conflicts of interest.

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