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Glossopharyngeal schwannoma: diagnostic and therapeutic aspects

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ABSTRACT

Among posterior fossa tumours, schwannomas arising from glossopharyngeal nerves are extremely rare, and only 39 cases of glossopharyngeal schwannomas have been described. The clinical and imaging features of glossopharyngeal schwannomas closely resemble that of acoustic schwannomas. Despite its accuracy, magnetic resonance imaging is not diagnostic of a ninth nerve schwannoma. This is because the schwannoma may be primarily localised to the cerebellopontine angle and may not cause enlargement of the jugular foramen. The diagnosis is possible only at surgery, once attachment to the ninth cranial nerve is seen. Because of the different surgical implications and management, preoperative recognition will help in determining the operative approach. We review the relevant literature and discuss the clinical presentation, radiological features and surgical findings in a 52-year-old woman with glossopharyngeal schwannoma.

Keywords: cerebellopontine angle tumour, cranial nerve tumour, glossopharyngeal schwannoma, jugular foramen tumour, nerve sheaths tumour

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INTRODUCTION

Neurinomas or schwannomas represent approximately 7%–10% of all primary intracranial tumours.[1,2] The cerebellopontine angle is the most frequent site of origin and the eighth nerve is the most commonly involved. Rarely are other cranial nerves involved, and descending order of frequency is the fifth nerve, seventh nerve and twelfth nerve. Schwannomas arising from the ninth, tenth and eleventh cranial nerves are relatively uncommon.[3,4] Less than 120 cases of jugular foramen schwannomas have been reported in the literature.[4,5] Schwannomas arising from the glossopharyngeal nerve are exceedingly rare and only 39 cases have been reported.[3-16] The symptoms of glossopharyngeal schwannomas are non-specific, and include hearing loss, tinnitus, vertigo, gait ataxia, headache, nystagmus, hoarseness of voice, palate-pharyngeal and facial hypoaesthesia, and rarely, papilloedema and dysphagia, depending on the tumoral growth patterns. Their clinical presentation and radiological findings closely mimic acoustic schwannomas, and the identification of the nerve of origin of the tumours often cannot be established preoperatively. The diagnosis is usually accomplished only at surgery. We review the relevant literature and discuss the clinical presentation, radiological features and surgical findings in a case of glossopharyngeal schwannoma.

CASE REPORT

A 52-year-old woman presented with progressive difficulty in walking, with noticeable swaying to the left side, and also experienced decreased hearing in the left ear, of three months' duration. She was also complaining of mild headaches not associated with nausea, vomiting or blurring of vision. There was no history of focal weakness, dysphagia or nasal

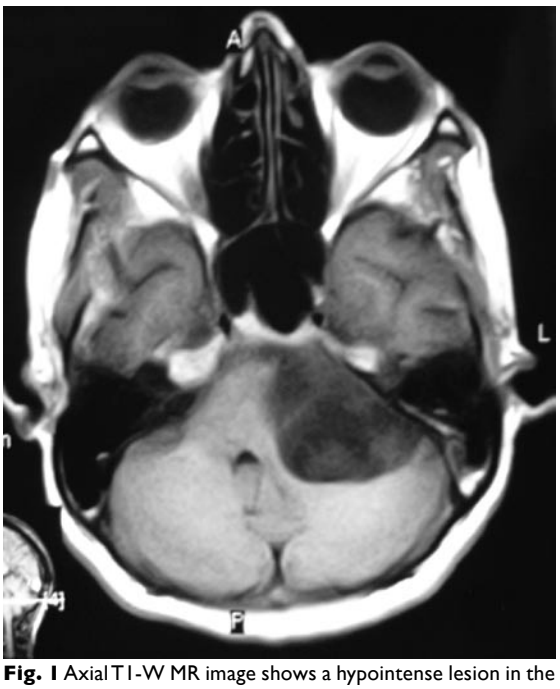

Fig. 1 Axial T1-W MR image shows a hypointense lesion in the left cerebellopontine angle, with compression and distortion of the brain stem and cerebellum.

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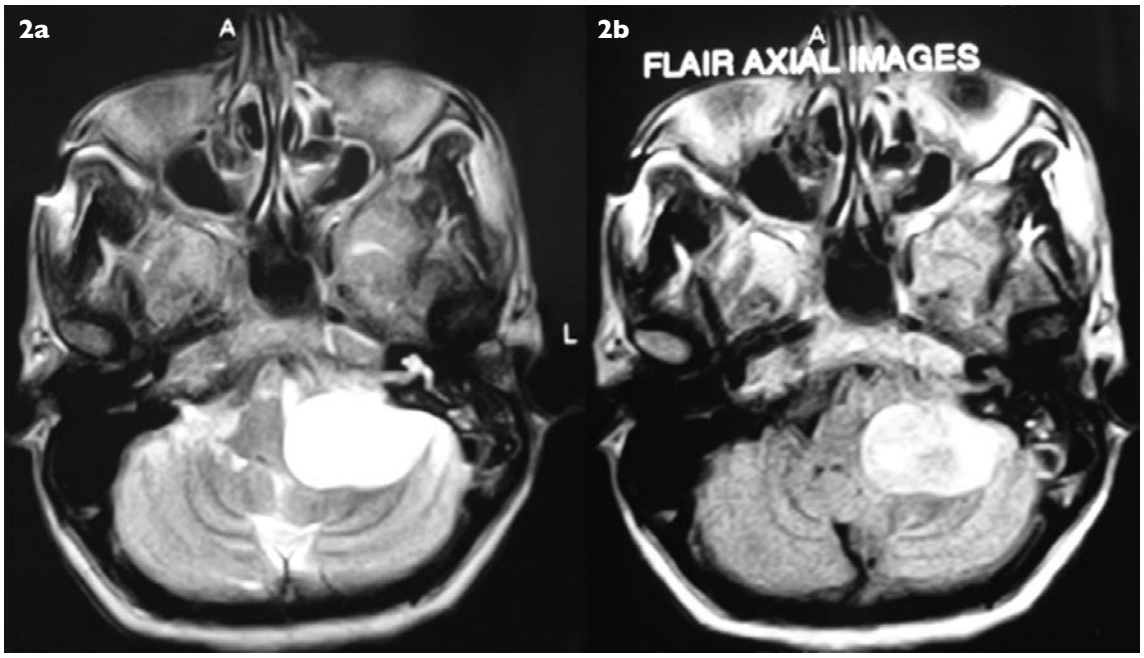

Fig. 2 Axial (a) T2-W and (b) FLAIR MR images show that the tumour is hyperintense and the eighth nerve complex can be seen anterior to the lesion.

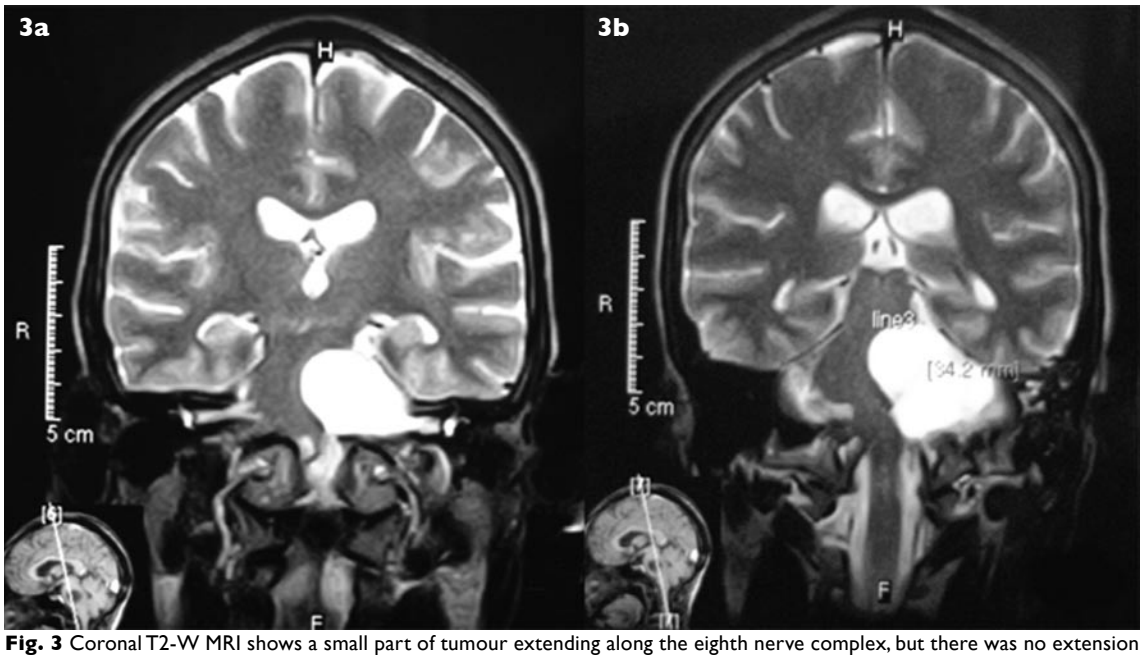

Fig. 3 Coronal T2-W MRI shows a small part of tumour extending along the eighth nerve complex, but there was no extension into the jugular foramen. However, at surgery, the tumour was well separated from the nerves by the arachnoid plane.

regurgitation. She was a known asthmatic and hypertensive for the last eight years, and was on regular treatment. She had no evidence of cafe-au-lait spots or other neurocutaneous markers. She had bilateral papilloedema with normal visual acuity. Neurological examination revealed slurring of speech. Corneal reflex was absent on the left side. She had left side grade I facial weakness but no loss of taste. Hearing was reduced in the left ear with bone conduction better than air conduction. Gag reflex was present bilaterally. All deep tendon reflexes were exaggerated. There were no motor or sensory deficits. She had left cerebellar signs and gait ataxia with swaying to the left side. Pure tone audiometry revealed

mild sensorineural deafness on the left side with normal hearing on the right side.

She was investigated with magnetic resonance (MR) imaging which revealed a large mass (Figs. 1–3) in the left cerebellopontine angle, which was hypointense on T1-WI and hyperintense on T2-WI and fluid attenuated inversion recovery (FLAIR) images (Figs. 2 & 3). Based on clinical and imaging features, a provisional diagnosis of cystic acoustic schwannoma was made. She underwent left retromastoid suboccipital craniectomy and near total excision of the tumour. At surgery, a large, soft, suckable, moderately vascular, cystic mass with a very fragile capsule was found occupying the left cerebellopontine angle. It was debulked under microscope, and the eighth nerve complex was seen pushed anteriorly. The tumour was attached to the ninth cranial nerve (glossopharyngeal nerve), and it was densely adherent to the brainstem. Histopathology showed Antoni A and Antoni B areas typical of schwannoma. Postoperatively, the patient had a transient increase in facial weakness, which resolved over the next two days, and there were no other deficits. She also experienced improvement in hearing. Cerebellar signs and speech quality also improved with time.

DISCUSSION

Jugular foramen schwannoma can arise from proximal or distal parts of the ninth, tenth and eleventh cranial nerves, presenting either as an intracranial or extracranial mass, or as dumb-bell shaped tumours with both intra- and extracranial extension. These tumours are classified according to the radiological and surgical features. Franklin et al classified these tumours into Classes A, B, and D, which were analogous to the categories used for glomus jugular tumours.[14] Samii and Tatagiba classified tumour extension, in relation to the radiological and surgical features, into Types A, B, C and D. Type A tumour is primarily located at the level of cerebellopontine angle with minimum enlargement of the jugular foramen, type B tumour is primarily in the jugular foramen with intracranial extension, type C is primarily an extracranial tumour with extension into the jugular foramen, and type D is a dumbbell-shaped tumour with both intra- and extracranial components.[3] Kaye et al classified them into the following three types: type A: a primary intracranial tumour in the cerebellopontine angle with minimum enlargement of jugular foramen and with a small extension into the bone; type B: a tumour mainly invading the bone (jugular foramen) with or without an intradural component; and type C: a tumour that is primarily extracranial in location with a minor extension to the bone or into the posterior cranial fossa.[17] Pellet et al added type D, which is a saddlebag-shaped tumour with intracranial and extracranial components.[18] Type A tumour is the most common, and is primarily located at the level of the cerebellopontine angle with minimum enlargement of the jugular foramen. However, the diagnosis of glossopharyngeal schwannoma in these cases is usually possible once the tumour attachment to the ninth nerve is seen at surgery, as in the present case.[3,14,17]

Glossopharyngeal schwannomas usually present in the third to fifth decades of life, the age at diagnosis ranging from 14 to 63 years (average 37 years), with a slight female predisposition.[4,6] Symptoms may not manifest, until the tumour attains a fairly large size, and they vary from subtle to severe, and may extend over several years.[19,20] A 'jugular foramen syndrome' has been described with tumours arising from the region of the jugular foramen, but this entity seems to be present only in a few cases.[17,19,20] Lesions growing into the cerebellopontine angle are difficult to differentiate clinically from acoustic schwannomas, as patients frequently present with seventh and eighth nerve deficits by virtue of the proximity of the jugular foramen to the internal auditory canal and their growth pattern.[5,16,17,20-22] There may be impairment of other cranial nerve functions in the cerebellopontine angle region, depending on the extent of the tumour, sometimes in the absence of signs of the lower cranial nerve involvement.[5,17,23] In the present case, the bulk of the tumour was in the cerebellopontine angle and it was extending superiorly to the tentorium and medially distorting the brainstem, pushing the seventh and eighth cranial nerves anteriorly, and responsible for the absent corneal reflex, mild facial weakness, mild sensorineural deafness, cerebellar signs and pyramidal tract involvement. The symptoms of glossopharyngeal dysfunction may not become apparent until there is bilateral involvement.[19] Glossopharyngeal schwannomas may be distinguished by an observable history of hoarseness, demonstration of abnormal palatal function and absence of expected findings, upon standard radiographical examination of the petrous bones and internal auditory canal.[22]

While it is not possible to distinguish between glossopharyngeal schwannoma and schwannomas arising from the tenth or eleventh cranial nerves until surgery, it is often possible to separate schwannomas arising from the ninth, tenth or eleventh nerves from those arising from the eighth nerve on imaging (particularly MR imaging). Arteriography was the diagnostic tool of choice before the advent of CT

and MR imaging, but nowadays, it is only indicated to exclude a glomus tumour. On CT (including bone window), these tumours are hypodense on a plain scan with moderate enhancement on contrast administration, with a normal internal auditory canal. There may be a sharp-edged enlargement of the jugular foramen. MR imaging will better delineate the extension of the mass, its relation to posterior fossa structures, any extension into jugular foramen and relation to the seventh and eighth nerve complexes, which can be seen anteriorly to the tumour. In our case, the more posterior location of the tumour on the axial image with well-visualised seventh and eighth nerve complexes, would have given a clue that this lesion could be arising from the lower cranial nerves. Post-gadolinium T1-weighted images will further delineate that there is no significant extension into the internal auditory canal (which could not be performed in our case due to financial constraints). Also, if the mass has an extension into the jugular foramen even without enlarging it, this by itself is a clue that the schwannoma is probably arising from the ninth, tenth or eleventh nerve. Acoustic schwannomas do, on rare occasions, extend into the jugular formen, but they generally do not do so. Despite its accuracy, MR imaging is not diagnostic of a ninth nerve schwannoma, which may be primarily localised to the cerebellopontine angle and may not cause enlargement of the jugular foramen.[1,3-9,11-16,24] Because of its different surgical implications, such a rare condition must be clearly recognised preoperatively.[16]

In Samii and Tatagiba's series, all type A tumours were operated via a retromastoid approach, which was also used with this case. Types B, C and D tumours were operated by a single-stage cervical approach with mastoidectomy.[3,5] During surgery after an initial intracapsular debulking, the dissection of the tumoral capsule from the lower cranial nerves and brainstem, is the most difficult and challenging step of the entire procedure. Total removal of the tumour requires resection of some nerve rootlets without damaging the main glossopharyngeal trunk, the other cranial nerves and brainstem.[3,5,17,19,24] Worsening of cranial nerve deficits following excision have been reported in literature. Due to this concern, and the presence of good preoperative function of the lower cranial nerves, it is justifiable to leave behind any tumoral capsule that is tightly adherent to the nervous trunks and brainstem, and accept a subtotal removal without worsening the brainstem and cranial nerve dysfunction.[3,5,17,19,24] In our patient, part of the tumour which was adherent to the brainstem, was left behind, and she did not have lower cranial nerve palsy or brainstem dysfunction. The hearing also improved as was reported in the literature.[5] However, in cases of incomplete removal, clinical and radiological follow-up are necessary as these tumours can grow larger.[5] Gamma knife radiosurgery is an effective alternative to microsurgical resection for patients who have small tumours and intact lower cranial nerve function. It is also effective for patients who have residual or recurrent tumours after microsurgical resection.[25-27] Excellent preliminary tumour control rates and a favourable toxicity profile support the effectiveness of linear accelerator stereotactical radiosurgery for patients with nonacoustic schwannomas.[26,27] Compared to historical controls treated with surgical resection, radiosurgery appears to have less treatment-associated morbidity for nonvestibular schwannomas, especially for schwannomas involving the lower cranial nerves.[26,27]

In summary, glossopharyngeal nerve schwannomas are infrequent tumours and preoperative diagnosis can be difficult as specific signs of glossopharyngeal nerve involvement can be absent in most of the cases. Though MR imaging is a more accurate diagnostic tool and will give some clues towards the nerve of origin, the exact identification of the nerve of origin of the tumour can be accomplished only at surgery. Total removal should be the goal of the operation for this benign tumour; however, a partial resection could be an option in the presence of large tumours with dense adhesions to the other cranial nerves and brainstem. This option is to prevent postoperative morbidity, and an option of radiosurgery can be considered subsequently.

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