Glossopharyngeal Schwannoma: A Case Report and Review of Literature

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Summary
We report a rare case of glossopharyngeal schwannoma whose clinical presentation and the radiological work up suggested an acoustic schwannoma. The diagnosis was made at surgery, once attachment to ninth cranial nerve was seen. The clinical presentation, radiological features and surgical findings of the glossopharyngeal schwannoma are presented along with the review of literature.

Key words: Glossopharyngeal, Schwannoma.

Introduction
Neurinoma or schwannomas represent approximately 7 - 10% of all primary intracranial tumors.1,2 Most frequent site of origin is cerebellopontine angle, more precisely from the 8th nerve. Rarely, neurinoma may arise from other cranial nerves. Other cranial nerves involved, in order of frequency, are 5th nerve, 7th nerve and 12th nerve. Schwannomas arising from the 9th, 10th and 11th cranial nerves (also called jugular foramen schwannoma) without associated neurofibromatosis are relatively uncommon.3,4

Less than 120 cases of jugular foramen schwannomas have been reported in the literature; however, schwannomas arising from the glossopharyngeal nerve are exceedingly rare3 and only 37 cases have been reported.3-15 Since their clinical presentation and radiological findings closely mimic acoustic neurinoma, usually it is very difficult to determine the precise nerve of origin preoperatively. We report a case of glossopharyngeal neurinoma because of its rarity.

Case Report
A 22 year old young man presented in neurosurgery outpatient department with history of progressive hearing loss and tinnitus in the right ear for the last 2 years. He had history of dizziness, which had worsened since last one month. He started having headache, vomiting and double vision, two weeks preceding admission. There was no history of loss of taste, incoordination or difficulty in swallowing. There was no family history suggestive of...
neurofibromatosis-II. On general physical examination, he had no evidence of cafe-au-lait spots or other cutaneous markers. Neurological examination revealed papilledema with bilateral 6th nerve palsy. Hearing was reduced in right ear with bone conduction better than air conduction. Gag reflex was present bilaterally. He had right-sided cerebellar signs. Pure tone audiometry revealed no conduction in right ear. CT scan showed a multiloculated hypo dense mass, which was minimally enhancing on contrast administration, with compression of fourth ventricle, causing hydro-cephalus. No bony erosion or widening of internal auditory canal was noted. MRI revealed a large multiloculated mass in right cerebellopontine angle, which was hypointense on T1WI and hyper-intense on T2WI (Fig. 1 and 2). A provisional diagnosis of cystic acoustic neurinoma was made.

A left ventriculoperitoneal shunt was put to drain the obstructed ventricles. He underwent right retro-mastoid sub-occipital craniectomy, one week later. At surgery, a well-defined cystic mass was found which was soft, suckable and moderately vascular. It was debulked under microscope, 7th and 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. Post operatively, patient did have a transient swallowing difficulty, which resolved within one week. There was no improvement in hearing; audiometry done four weeks later revealed no conduction.

**Discussion**

Glossopharyngeal nerve is closely related to the vagus nerve, sharing common medullary nucleus. Fibers of glossopharyngeal nerve enter and emerge at levels rostral to the rootlets of the vagus nerve, but it is easier to identify, because its fibers form a single compact nerve root whereas fibers of the vagus nerve enter and emerge from brain stem by a number of small rootlets. The glossopharyngeal nerve is a mixed branchiomeric cranial nerve with five functional components (i) general visceral afferent (GVA) fibers, (ii) special visceral afferent (SVA) fibers (taste), (iii) a few general somatic afferent (GSA) fibers, (iv) general visceral efferent (GVE) fibers and (v) a small number of special visceral efferent (SVE) fibers.16,17

Chiro et al18 divided jugular foramen into two compartments: the anterio medial compartment called ‘pars nervosa’ and posterio lateral compartment called ‘pars venosa’ or pars vascularis’. There is controversy regarding the position of cranial nerves in the jugular foramen. Arenberg and Mc Creary8 and Gejrot19 believe that ninth, tenth and eleventh cranial nerves are located in pars nervosa; on the contrary Mountjoy et al9 and Bossy20 argue that only 10th and 11th nerves lie in the pars nervosa.
Jugular foramen schwannoma can arise from proximal or distal part of the ninth, tenth and eleventh cranial nerves, presenting either as intracranial or extra cranial mass. Dumb-bell shaped tumors with both intra and extracranial extension have been known. As with most schwannomas, these tumors 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 predilection. The youngest patient reported in the literature was a four years old boy. Symptoms may not manifest until the tumor attains a fairly large size, hence, they vary from subtle to severe and may extend over years, as was the case in our patient. Intracranial and bone tumors have the longest duration as compared to extracranial tumors.

Sweasey et al analyzed the clinical presentation of the 23 cases in literature and five of their own and found hearing loss to be the most common presenting feature in 25 (93%) of the 27 cases (in one case hearing status was not mentioned), whereas in acoustic schwannomas hearing loss is the initial symptom in about 67% of the patients. Hoarseness and decreased gag reflex were found in only 6 patients (22%). In a series of 14 cases presented by Tan et al, there were 8 patients (57%) with lower cranial nerve palsies at the time of presentation.

Proximal lesions present as cerebellopontine mass and hearing loss is an early symptom. The more distal lesions present as extracranial, cervical or skull base tumors. In the present case, the tumor was entirely confined to the cerebellopontine angle and hearing loss was the presenting symptom and not the lower cranial nerve palsies. The symptoms of glossopharyngeal dysfunction may not become apparent till there is bilateral involvement.

Surprisingly, for such an infrequent tumor, we found three different classifications in the literature. Franklin et al classified these tumors into Class A, B, and D analogously to the categories used for glomus jugular tumors. Samii and Tatagiba classified tumor extension in relation to the radiological and surgical features into Type A, B, C and D. Type A tumor being primarily located at the level of cerebellopontine angle with minimum enlargement of jugular foramen, type B tumor primarily in jugular foramen with intracranial extension, type C primarily being extracranial tumor with extension into jugular foramen and type D, a dumb bell shaped tumor with both intra and extracranial components. The diagnosis of glossopharyngeal schwannoma is usually made once the tumor attachment to the 9th nerve is seen at surgery. Kaye et al classified them into following three types: type A, primary intracranial tumor in cerebellopontine angle with minimum enlargement of jugular foramen and with a small extension into the bone; type B, tumor mainly invading the bone (jugular foramen) with or without an intradural component; and type C, tumors that are primarily extra cranial in location with a minor extension to the bone or into posterior cranial fossa. Pellet et al added Type D, which is a saddle bag shaped tumor with intra cranial and extracranial components. Jugular foramen schwannomas are sometimes difficult to differentiate from acoustic schwannomas as patients frequently present with seventh and eight nerve deficits by virtue of the proximity of jugular foramen to internal auditory canal. This is more common for type A tumors as reported by Kaye et al and also seen in the present case.

Radiological features described in the literature are of a schwannoma in cerebellopontine angle, which is hypodense on plain CT scan with moderate enhancement on contrast administration, with normal internal auditory canal. There may be a sharp edged enlargement of jugular foramen. MRI may show an extension of the mass into jugular foramen and VII VIII nerve complex may be seen anterior to the mass. This is not diagnostic of a ninth nerve schwannoma which may be primarily localized to cerebellopontine angle and may not cause enlargement of jugular foramen. The radiological features in our case were consistent with findings described above, but there was no evidence of the mass going into the internal auditory canal or the jugular foramen.

The excision of the schwannoma was done by retro mastoid suboccipital route, which is the preferred approach for Type A lesions. In Samii and Tatagiba’s series, all type A tumors were operated by a retromastoid approach. Types B, C and D tumors were operated by single stage cervical approach with mastoidectomy. Worsening of cranial nerve deficits following excision have been reported, however, our patient had transient lower cranial nerve palsy which resolved within a week. The hearing loss did not recover as reported in the literature.

**Conclusion**

Glossopharyngeal nerve schwannoma is an infrequent tumor which may not always produce lower cranial nerve deficit and clinical presentation may mimic an acoustic neurinoma. Hence, it is usually not
recognized preoperatively. MRI is useful in diagnosis of this unusual tumor.

References


Accepted for publication : 5th January, 2001.