



## Huge dumbbell shaped glossopharyngeal schwannoma

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### ABSTRACT

Schwannomas arising from the 9th, 10th and 11th cranial nerves (also called jugular foramen schwannoma) without associated neurofibromatosis are relatively uncommon [1].

Schwannomas arising from the glossopharyngeal nerve are rare and fewer than 50 cases have been reported in the literature [1-4].

Glossopharyngeal schwannoma presenting in childhood is extremely rare and to our knowledge only 4 cases have been reported before [2, 5-7].

### Case Report:

We present a 17 years old boy with a history of slow growing neck mass from eight years ago. He mentioned no change neither in the sense of hearing nor in the force or sense of his extremities and had no complaint of vertigo, dizziness, headache or vomiting. There was also no complaint pertaining to other organ systems and his sole problem was his neck mass.

There was no similar problem in his family.

Examination revealed a 7\*5 cm mass in the right anterolateral part of the neck (beneath the sternocleidomastoid muscle and angle of the mandible) without any sign of inflammation such as redness, warmth or tenderness. He has some limitation of rotating the head and lateral flexion in the same site due to the mass. Function of the cranial nerves was normal and audiologic tests showed normal hearing.

Patient advised brain and cervical magnetic resonance imaging (MRI) and temporal bone computerized tomographic (CT) scan.

Brain MRI revealed a huge and lobulated extra-axial low T1 and T2 signal enhancing in right posterior fossa, causing pressure effect on adjacent pons and medulla oblongata and extending to the right parapharyngeal region. This mass looks to be dumbbell shaped with a large right posterior fossa and large right parapharyngeal components. The mass was enhanced on contrast administered T1-weighted MRI (Fig.1, 2&3). His right internal carotid artery was deviated to anteromedial side by the mass.

The patient was operated for a lower cranial nerve schwannoma in two sessions. In the first session, cervical and foraminal components of ninth cranial nerve schwannoma was resected by infratemporal approach (Modified Fisch A). In the second session which was done one month later intracranial component of the tumor was totally resected by retrosigmoid approach.

Histopathology was consistent with schwannoma.

Postoperatively, the patient had no significant complaint or deficit.

Discussion:

Jugular foramen schwannomas comprise only 2.9% of all intracranial schwannomas [3].

Schwannomas arising from the glossopharyngeal nerve are rare and fewer than 50 cases have been reported in the literature [1-4].

Childhood presentation of this tumor is extremely rare and to our knowledge only 4 cases have been reported before [2, 5-7].

Clinical presentation of intracranial schwannoma is usually characterized by local cranial nerve dysfunction.

However, since the posterior fossa is a small compartment, multiple cranial nerves may be affected simultaneously. Palsies of the ninth cranial nerve are unusual and symptoms of ninth nerve dysfunction may not become apparent until there is bilateral involvement. Furthermore, this neurinoma usually grows toward the cerebellopontine angle and initially affects the facial-acoustic nerve complex. Therefore, hearing loss is the most common symptom in 90–93% of cases [8].

The second most common symptom was related to increased intracranial pressure and cerebellar dysfunction, each occurring in 39% of cases [3].

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 dumbbell shaped tumors with both intra- and extracranial extension. Samii et al. [9] classified tumor extension, in relation to the radiological and surgical features, into Types A, B, C and D. Type A tumor is primarily located at the level of cerebellopontine angle with minimum enlargement of the jugular foramen, type B tumor is primarily in the jugular foramen with intracranial extension, type C is primarily an extracranial tumor with extension into the jugular foramen, and type D is a dumbbell shaped tumor with both intra- and extracranial components.

The radiological findings of this tumor are fairly typical, but not characteristic. Schwannoma of the jugular foramen usually appears as a sharply demarcated, contrast-enhancing tumor, which is typically centered or based in an enlarged jugular foramen with sharply rounded bone borders having a sclerotic rim [10].

Despite its accuracy, neuroimaging is not diagnostic of a ninth nerve schwannoma. The diagnosis is usually made when the tumor arising from the ninth nerve is seen at surgery.

In our case, an accurate pre-operative diagnosis of lower cranial nerve schwannoma was made based on clinical presentation and radiological appearance but the exact nerve of origin was found only in the surgery.

The surgical approach to remove glossopharyngeal schwannomas should be selected according to the location and degree of extension of the individual tumor. The primary goals of surgical and management of these lesions are the preservation and restoration of function of the lower cranial nerves, and of hearing and facial nerve function. Although, some authors may choose a single-stage operation but others may constitute two step operations [4, 7]. Our preferred approach for this case was also a two stage surgery which had an excellent result.

#### Conclusion:

Glossopharyngeal schwannoma may devoid of clinical symptoms and neurological signs. Magnetic resonance imaging may play a key role as an accurate diagnostic tool. A favourable option of approach and appropriate planning of surgical strategy should be the goal of operation for this benign tumor. Glossopharyngeal schwannoma may be radically and safely resected without creating additional neurological deficits and other complications.

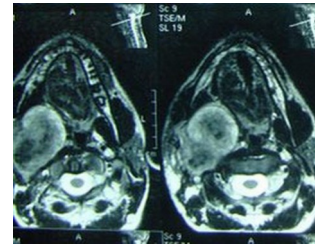


Figure 1: Axial T1-weighted magnetic resonance image shows cervical component of the lesion with a low signal inhomogeneous appearance in the right parapharyngeal space.

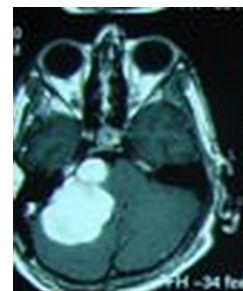


Figure 2: Axial gadolinium-enhanced T1-weighted magnetic resonance image shows enhanced intracranial component of the lesion in right posterior fossa, causing pressure effect on adjacent pons and medulla oblongata.

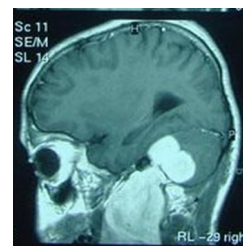


Figure 3: Sagittal gadolinium-enhanced T1-weighted magnetic resonance image shows enhanced foraminal, intracranial and extracranial components of the lesion.

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