# An asymptomatic huge intracranial glossopharyngeal/vagal schwannoma in a very young male: A case report



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**Abstract:**
A case of huge intracranial glossopharyngeal/vagal schwannoma presenting incidentally without any symptom and only sign was mild deviation of uvula. The treatment was by surgical excision of the schwannoma with leaving a small portion of the tumour that was inseparable and intermingled with lower cranial nerves, following which the patient made good recovery after an eventful postoperative course. To our knowledge, this is one of the largest reported asymptomatic glossopharyngeal/vagal schwannoma in a very young patient.

***Key words:*** *Asymptomatic, huge intracranial glossopharyngeal/vagal schwannoma*

**Introduction:**
Schwannomas are benign tumours arising from the Schwann cells of the nerve sheath. Glossopharyngeal or vagal schwannomas are rare entities and can arise from either the intracranial, extracranial or jugular foramen portion of the cranial nerves. Intracranial schwannomas usually present as a painless cerebellopontine angle mass with a number of symptoms and signs due to cranial nerve palsy or from pressure over brain stem. We describe a case of rare asymptomatic huge intracranial glossopharyngeal/vagal schwannoma in a young man with only deviated uvula.

**Case report:**
A 25-year-old young male presented to a physician for painful tongue ulcer. During routine examination of the oral cavity, he found the deviated uvula. So he advised for a magnetic resonance imaging (MRI) of brain. Then he referred the patient to us. There was no history that is suggestive of intracranial posterior fossa or cerebellopontine angle or foramen magnum lesion. On examination, everything including the nervous system was absolutely normal except that there was a deviated uvula toward the right. The MRI of brain showed contrast-enhancing huge left cerebellopontine angle mass, distorting and deviating the brainstem to the opposite cerebellopontine angle **(Fig.1)**. The tumour transversely reached almost to the opposite petrous apex. A large portion of the tumour was in the fourth ventricle. Proximally, it extended up through the tentorial hiatus and distally up to the lower

border of the posterior arch of axis. After appropriate counseling, the tumour was operated through the retrosigmoid suboccipital craniectomy with removal of posterior hemi-arch of atlas. During operation, we found that a portion of the tumour was intermingled and firmly attached with glossopharyngeal, vagus and accessory nerve rootlets, but the hypoglossal nerve seemed to be free from tumour. The origin of the tumour could not be ascertained. The tumour was also strongly adhered to the brainstem and separation of the tumour from the brain stem seemed dangerous. So a small portion of the tumour was left to prevent unacceptable damage of the lower cranial nerves and brain stem; rest of the tumour was removed using microsurgical technique, **(Fig. 2)**.

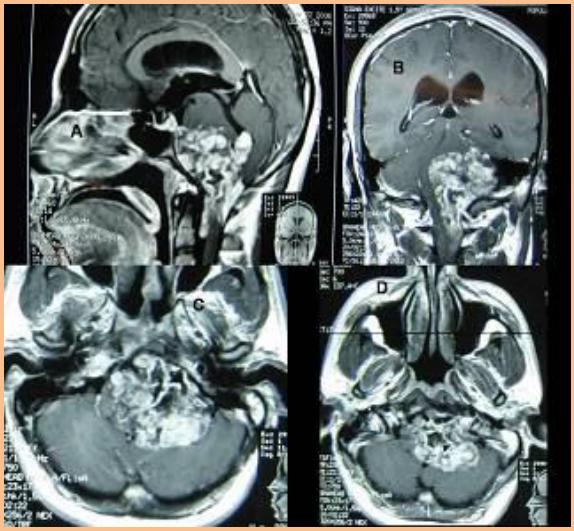


**Fig. 1**

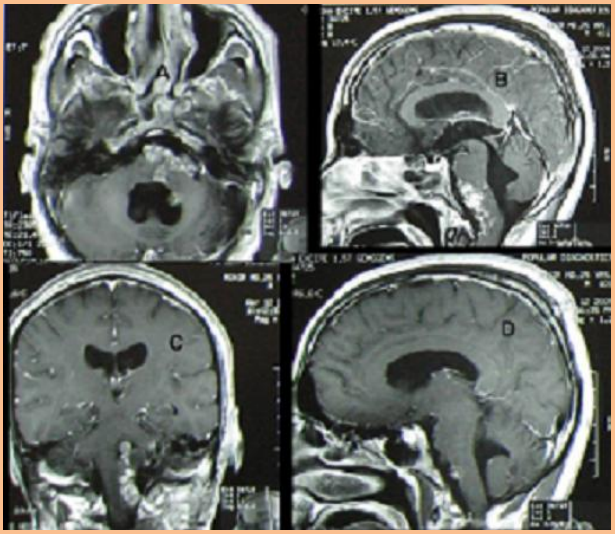


**Fig. 2**

**Figure 1: MRI of brain showed contrast-enhancing huge left cerebellopontine angle mass, distorting and deviating the brainstem to the opposite cerebellopontine angle.**

**Figure 2: Rest of the tumour removed using microsurgical technique**

Immediately after operation, the patient recovered fully from anaesthesia. All limb movements were normal, but there was hoarseness of voice with palatal deviation. From the first postoperative day, the patient began to deteriorate. He became drowsy and unable to maintain oxygen saturation, for which he was intubated and placed on ventilator.

His right-sided limbs became progressively weak to MRC muscle power grade 1, but sensory perception from right-sided limbs seemed to be present. On the 9$^{th}$ postoperative day, tracheostomy was done and the patient was nursed in intensive care unit with respiratory support for another 2 weeks. In this period, he was neurologically static. From the end of fourth week after operation, the patient began to improve and subsequently became fully conscious, but unable to maintain O2 saturation. Right-sided muscle power began to return very slowly. By the end of 2$^{nd}$ month, he was able to maintain respiratory function by himself. By the end of three months, tracheostomy was closed and he was able to stand with support and feeding was through nasogastric feeding tube. By the end of fourth month, he could walk with support and began to eat after removal of nasogastric tube, although, there was difficulty in opening mouth due to temporomandibular joint stiffness. By the end of six months, he was able to walk independently (MRC muscle power on right-sided limbs 4/5), but his voice was husky. Contrast MRI showed small residual tumour in the jugular foramen, foramen magnum area and left side of brain stem.

**Discussion:**

Schwannomas arising from the lower cranial nerves (IX-XI) are rare, constituting only 3% of all intracranial schwannomas unassociated with neurofibromatosis.[8] A great majority of these tumours present as jugular foramen lesions and less commonly they occur along the extracranial course of these nerves.[8] An intracranial hugely extended lesion is extremely rare.

The vagus is affected more commonly than any other nerve. They mainly occur in middle-aged adults and are more common in women than men.[6] Nerve sheath tumours in the head and neck are a common manifestation of neurofibromatosis.[6] Although neurofibromas are often found in association with von Recklinghausen's disease, schwannomas can also occur in these patients. Malignant transformation of these tumours is rare. A schwannoma is distinguished histologically from a neurofibroma by the characteristic Antoni A and Antoni B cellular pattern compared to the loosely arranged stroma of a neurofibroma. Antoni A areas consist of compact tissue with a high cellularity whereas Antoni B areas are composed of less cellular, loose reticular tissue. An ancient schwannoma shows atypical hyperchromatic nuclei.[6] Among the intracranial schwannomas, the acoustic neuroma is by far the most common, although involvement of the V, VII, X, XI and XII cranial nerves have been described, together with the jugular foramen neurinomas[1,5], schwannomas of the glossopharyngeal or vagus nerve, both the intracranial and cervical portions, are rare tumours. The clinical picture of intracranial glossopharyngeal or vagal schwannomas invariably shows a progressive neurological deficit with hearing loss, tinnitus, vertigo, balance disturbances, visual disturbances, pharyngeal and facial hypoaesthesia.[3] In contrast, the cervical tumours tend to present as a persistent, nontender, parapharyngeal mass pushing the tonsil and pharynx medially. They are less likely to cause nerve deficit, although they can cause dysphagia, hoarseness, shoulder drop, tongue atrophy and gagging dysphagia.[6]

Cough reflex on performing fine-needle aspiration in vagal nerve schwannoma[7] has been described and glossopharyngeal neuralgia with syncope, hypotension, bradycardia has been associated with neck masses.[2]

In most cases, a thorough radiological investigation can make a more precise diagnosis. In the case of a schwannoma, the computed tomography (CT) scan will disclose a hypodense tumour mass[4] with little or no enhancement after contrast administration. Bony window can show canal enlargement. Magnetic resonance imaging has been reported to be more specific than CT scan in the diagnosis of intracranial schwannoma.[9] The treatment of choice of nerve sheath tumours is complete microsurgical excision. In some cases, it is impossible to carry out total excision because of the proximity and inseparable adhesion of vital structures to the tumour. Repeated debulking of these cases has been described in neurofibromas without significant risk of malignant transformation.[6] Our case is unique for its huge size with severe distortion and displacement of brain stem without symptom. Operative decision in such a young asymptomatic patient with huge size is not straight forward as it might be associated with significant risk of mortality and morbidity. Here though the patient recovered nicely from anaesthesia but subsequently deteriorated, for which long intensive care unit support was needed. Proper and prolonged postoperative care was associated with good outcome though still there is some morbidity. Postoperative deterioration in such patients is probably due to sudden decompressive brain stem oedema, which should be properly evaluated and managed, otherwise, the outcome is fatal.

**Conclusion:**

In large lower cranial nerve tumour, proper preoperative evaluation, counseling, appropriate microsurgical management, readiness to face complications, and intensive postoperative care is very essential for a favourable outcome.

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