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Case Report

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Atypical presentation of a large cervical schwannoma

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ABSTRACT

Hypoglossal nerve schwannomas are rare benign tumors arising from the schwann cells covering the hypoglossal nerve. These tumors are slow growing and their symptoms are usually due to pressure effects over the adjacent structures. We present a rare case of a large intracranial and cervical schwannoma arising from the hypoglossal nerve. We discuss the atypical presentation and vague symptoms with which a patient of hypoglossal schwannoma may present. Surgery in such cases requires the joint efforts of the team of Neurosurgeons and Otorhinolaryngology and Head and Neck surgeons.

Keywords: Hypoglossal nerve schwannoma, Cervical schwannoma, Atypical presentation

INTRODUCTION

Tumors of the parapharyngeal space (PPS) are rare tumors of the head and neck, comprising less than 0.5% of all head and neck tumors. They mainly affect the salivary glands and the lower cranial nerves. Schwannomas are the commonest neurogenic tumor of the schwann cells in the PPS. They are benign, slow growing neoplasms originating from the myelin producing schwann cells in the peripheral nervous system. They arise commonly from vagus followed by cervical sympathetic chain.[1] Schwannomas arising from the hypoglossal nerve are rare and account for 5% of all nonacoustic intracranial schwannomas.[2] They usually originate intracranially but can also extend extracranially through the hypoglossal canal in a dumb bell fashion. They are slow growing tumors and their symptoms are usually due to the pressure effects over the adjacent structures.[3] We present a rare case of large intracranial and cervical schwannoma arising from the hypoglossal nerve with an atypical presentation.

CASE REPORT

A 37 year old lady presented to the ENT department with complaints of unsteadiness of gait and giddiness for past 01 month. She also had a history of headache for past 03 months and a slowly progressive swelling on right side of upper neck for past 3 years. On examination, a firm swelling was palpable on right side of the neck, measuring 7 cm×4 cm, extending from the angle of mandible to the thyroid notch. The swelling could be moved horizontally but not vertically. Paresis of right hypoglossal nerve was also found.

A CT and MRI scan was done which showed the presence of a mass lesion in the neck, involving the right parapharyngeal and carotid spaces and the right jugular foramen. The lesion was found to be extending into the posterior cranial fossa, communicating with it through the hypoglossal canal which was widened and tumor was extending intracranially to posterior fossa upto cerebello pontine angle in a dumb bell fashion (Figure 1-3). The patient was planned to be operated in two stage

procedure. In the first phase, the intracranial part of the tumor was tackled by the neurosurgeons by posterior cranial fossa approach. Post-operative, the patient had palsy of 7th, 8th, 9th, 10th, 11th and 12th cranial nerves. In the second stage, the extracranial part of the tumor was addressed by ENT surgeons. A trans-cervical approach was used to remove the cervical portion of the schwannoma which was engulfing vagal and hypoglossal nerves (Figure 4). The tumor was found to displace the carotids anteriorly and the internal jugular vein was found coursing laterally by the tumor. The submandibular gland was removed and the digastric tendon was cut to approach the part of tumor which was inferior to mandible and extending to skull base upto jugular foramen. The tumor was closely involving the vagus nerve which was non-functional and was therefore removed along with the tumor. The tumor was dissected in plane with bipolar cautery after retracting ICA superomedially (Figure 5). The styloid process was palpated and tumor was dissected bluntly above it from the skull base, a vascular clip was placed at its base and tumor was removed in toto (Figure 6). The patient recovered uneventfully and has no signs of recurrence after 6 months.

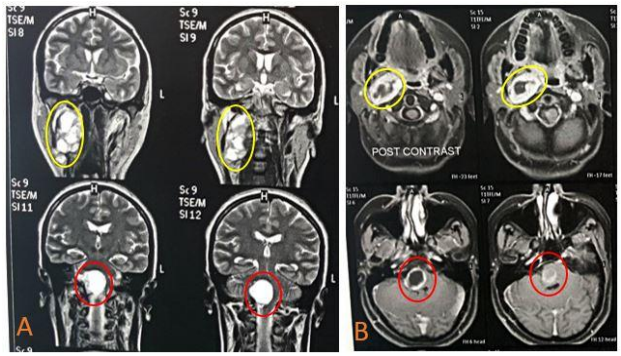

Figure 1: (A) T2W images (coronal sections) showing large hypoglossal schwannoma with solid and cystic component displacing carotids anteriorly and IJV posterolaterally. Intracranial portion marked with red circle can be seen compressing the brainstem. Cervical component is marked with a yellow circle. (B) T1W post contrast images (axial sections) of the same patient.

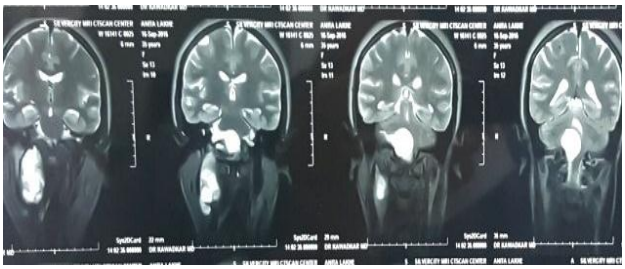

Figure 2: T2W MRI showing the intracranial and extracranial portion of hypoglossal schwannoma appearing as a dumb bell shape.

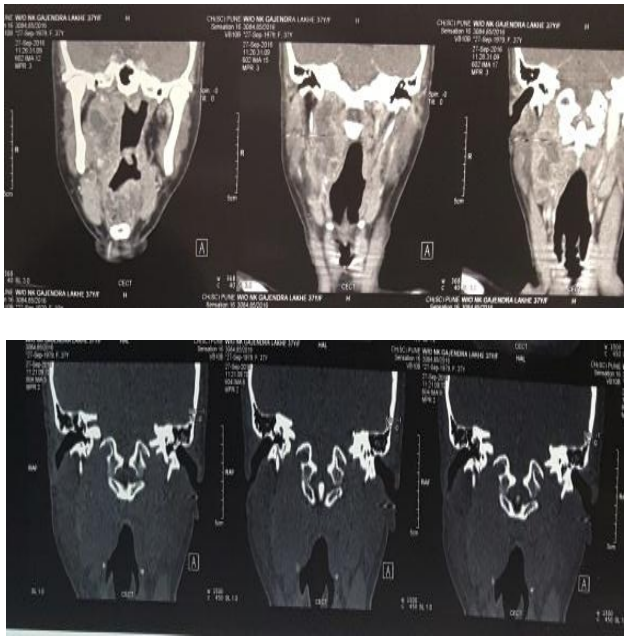

Figure 3: Widening of the hypoglossal canal.

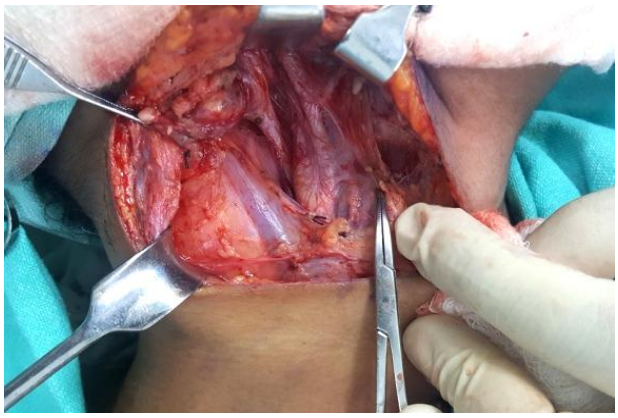

Figure 4: Tumor engulfing vagal and hypoglossal cranial nerves and displacing carotids anteriorly and IJV laterally.

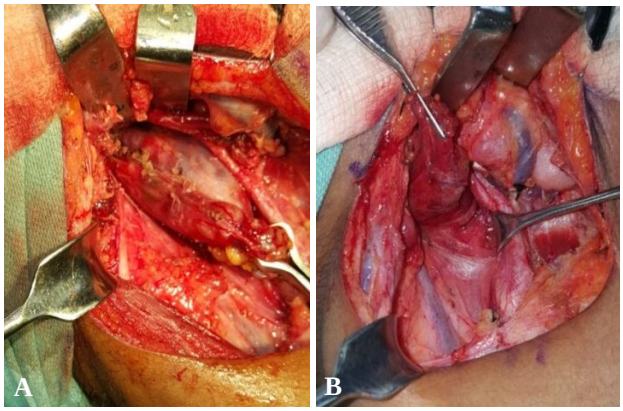

Figure 5: (A) Intraop appearance of the hypoglossal schwannoma in the neck. (B) ICA retracted anteromedially.

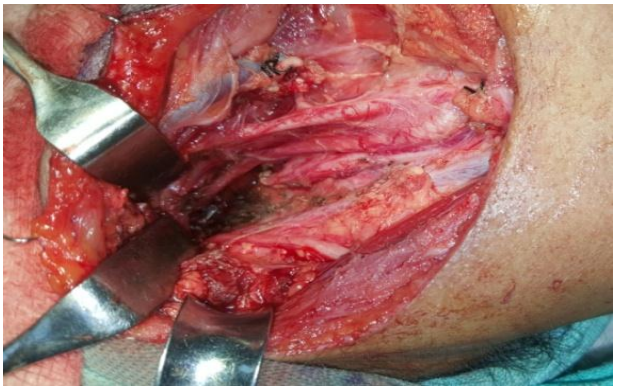

Figure 6: Parapharyngeal space exposed and clip at jugular fossa area at skull base. Vagal nerve resected.

DISCUSSION

Schwannomas of the hypoglossal nerve are rare and slow growing benign tumors. Their symptoms are caused mainly due to the pressure effect over the adjacent tissues. Patients usually present with symptoms like hoarseness, dysphagia, neck swelling, swelling over the posterior pharyngeal wall and hypoglossal nerve palsy.[4] MRI and CT scan are done preoperatively to determine the extent and origin of the tumor and to help in surgical decision. MRI provides better preoperative information and relationship with surrounding structures. CT gives a better idea about bone involvement.[5] Surgical excision is the treatment of choice. The chances of recurrence are rare. The survival for benign parapharyngeal tumors has been found to be 100% at end of 5 and 10 years and for malignant tumors, the survival rate is 93% at end of 5 years and 57% at end of 10 years.[6]

Hypoglossal nerve schwannomas are rare tumors of the head and neck. The disease presents with symptoms which are often misleading and result in a long delay between the initial symptoms and definitive diagnosis. It often results in lower cranial nerve palsy and long tract signs due to compression at cervico-meduallary junction. The presence of ipsilateral hypoglossal nerve palsy is seen in about 85% of patients with hypoglossal schwannoma and has a diagnostic value.[7] Hypoglossal nerve palsy resulting in ipsilateral hemiatrophy and weakness of the tongue is the commonest sign seen in hypoglossal nerve schwannomas.[8] It usually precedes the other symptoms caused by the lesion.

Our patient presented with unsteadiness of gait for past 1 month along with headache for past 3 months and a neck swelling of 3 years duration. On presentation, she was found to have paresis of the hypoglossal nerve. The tumor was found to originate from the trunk of the hypoglossal nerve, extending extracranially and presenting as a slow growing mass in the upper neck. Hypoglossal nerve schwannomas have a slow rate of growth and this can account for the late presentation and slow progression of the symptoms in our case. MRI showed the intracranial part of the tumor to be compressing upon the brainstem. This can account for the recent development of headache and unsteadiness in the patient. In view of the intracranial involvement, the tumor was first operated by the neurosurgeons, followed by a second stage surgery of the extracranial component of the tumor by the team of Otolaryngology - Head & Neck surgeons.

Our case presented with symptoms due to pressure from the intracranial portion of the tumor over the brainstem. It illustrates the vague symptoms with which a patient of hypoglossal nerve schwannoma may present. Surgery for schwannomas from the cranial nerves carries the risk of post-operative cranial nerve palsy and the patient should be counselled about the same pre-operatively.[3]

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