

# A Giant Vagal Schwannoma presenting as Parotid Swelling with Obstructive Hydrocephalus

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## ABSTRACT

Schwannomas are benign tumor arising from Schwann cells. 25- 40 percent of schwannomas are present in head and neck. They generally present as a benign swelling in the neck but may masquerade as other tumour due to presentation at unusual site. We here by presenting a case of 50 year female who presented with parotid swelling which turned out to be Schwannomas arising from vagus nerve with significant intra-cranial extension.

**KEYWORDS:** Vagal Schwannomas, Parotid Tumour, Obstructive Hydrocephalus

## INTRODUCTION

Schwannomas are benign tumours arising from the nerve sheath (Schwann cells). The most common site in head and neck is Para pharyngeal space but may present as at other sites. We here by present a case of giant vagal schwannoma which clinically presented as parotid mass and had a significant intra-cranial extension resulting in compensated obstructive hydrocephalus.

## CASE REPORT

A 50 year lady presented to our outpatient department with the chief complaint of an asymptomatic swelling in the left parotid region for eight years .The swelling started insidiously and gradually progressed in size over the period of three years. There was no history of pain , fever, facial asymmetry or sudden increase or regression in the size of the swelling.

On enquiry the patient also gave a history of noticing a change in her voice in the form of hoarseness since last six months. She also complained of dysphagia mainly to solids without any pain on swallowing for the past 4 months. Interestingly, the patient had hemifacial spasms of the left half of the face which had started only 20 days back.

On external examination, there was a 6x5 cm swelling in the left parotid region extending behind the angle of the mandible and postero-inferior to the pinna. ( Figure 1)The swelling was firm in consistency, non tender and mobile, with well defined margins. There was no local warmth and the skin over the swelling was unremarkable. No other similar swellings were palpable in any region of the neck. On examination of the oral cavity and oropharynx, the patient had a left parapharyngeal bulge and deviation

of the tongue to the left (Figure 2 and Figure 3). The tongue musculature also revealed muscle wasting. The patient had diminished gag reflex. She did not have any facial palsy, however she had left hemifacial spasms which were more conspicuous while speaking. On an indirect laryngoscopy the patient was found to have left vocal cord palsy.



Fig 1: Left Parotid region swelling Fig 2: Left hypoglossal palsy



Fig3: Left parapharyngeal bulge

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An ultrasonography (USG) of the lesion revealed a hypoechoic mass approximately 5.8 x 5.3 x 5.4 cm in dimension pushing the normal parotid gland laterally and the submandibular gland and the vessels medially.

On computed tomography, there was a homogenous mass enhancing on contrast extending from the neck upto the skull base eroding the jugular foramen and hypoglossal canal and abutting the brainstem. The mass extended medial to the mandible along its posterior border pushing the normal parotid gland laterally. There was splaying of the carotid vessels thereby raising a possibility of a vagal schwannoma (Figure 4, Figure 5).

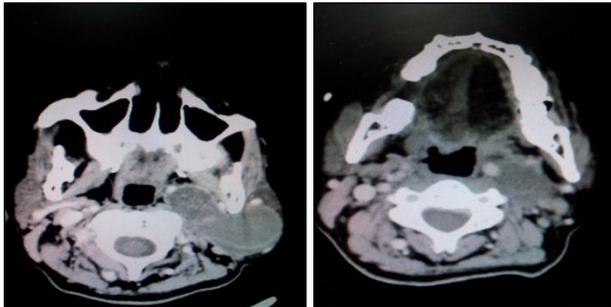


Figure 4. CT (axial cuts) showing tumour splayin the internal jugular vein from internal carotid artery

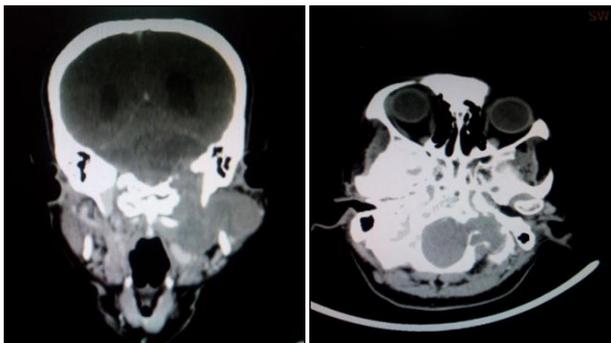


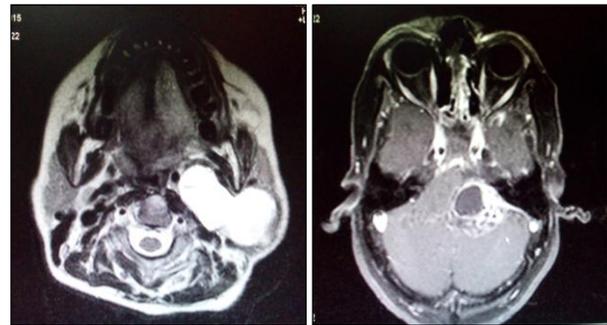
Figure5 .CT (coronal and axial) shoing the tumour eroding the skull base with widening of jugular foramen and obliteration of hypoglossal canal.

On magnetic resonance imaging (MRI), the lesion was found to be T1 isointense, T2 hyperintense and brilliantly enhancing with contrast extending from the neck upto the brainstem, abutting it. The mass was seen to cause a pressure effect over the communication between the third and fourth ventricle, thereby leading to a compensated obstructive hydrocephalus (Figure 6).



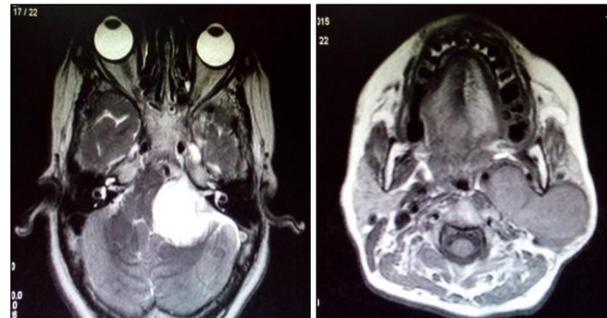
(a)T2 coronal

(b) T1 coronal



(c)T2 axial

(d)T1 axial with contrast



(e) T2 axial

(f) T1 axial

Figure 6(a-e): MRI showing the extent of tumour including the intracranial part

An ultrasonography guided fine needle aspiration cytology was done (FNAC) was performed which showed abundant myxoid material containing spindle cells in a haemorrhagic background suggestive of spindle cell neoplasm.

The patient was referred for radiotherapy for shrinkage of intracranial tumour part.

## DISCUSSION

Schwannomas are benign encapsulated tumors which arise from schwann cells. These tumors were first described by Verocay in 1910. 25-40% of all schwannomas are found in head and neck area. These tumors are slow growing tumors which may manifest as a painless, smooth regular mass. The retrostyloid compartment of the parapharyngeal space is the most common site of origin.<sup>1</sup>

Schwannomas are benign tumours which grow very slowly with speed of about less than a centimeter in an decade. They have tendency to grow along the nerves. With increase in size, the neurological deficit of the nerve of origin may also set on which is not a case with small schwannomas. These schwannoma may cause pressure injury resulting in neuropraxia or neurotmesis of adjacent nerves if they increase in size enormously. In our case, this tumour had an extensive length which reached intra-cranially. The tumour seems to arise from vagus nerve as it splayed the internal jugular vein from internal carotid artery in contrast to schwannomas arising from cervical sympathetic chain which push both the great vessels anteriorly rather than splaying them.<sup>2,3</sup> This tumour has then progressed from the carotid sheath

superiorly towards the skull base, in the process, it pushed the parotid laterally. This displacement of parotid gland resulted pressure on axons of facial nerve giving rise to multiple twitching of facial musculature continuously. The tumour further had grown resulting in erosion of skull base. The jugular foramen was dilated resulting in palsy of glossopharyngeal nerve with paresis of spinal accessory nerve. This tumour had also eroded the hypoglossal canal via eroding through the postero-medial wall of jugular foramen resulting in hypoglossal nerve and deviation of the tongue on the same side due to paralysis of all the external muscle of same side. The tumour further progressed and compressed upon fourth ventricle and brain stem which resulted in gradually developing compensated obstructive hydrocephalus.

The treatment option for schwannomas is surgical excision but radiotherapy is used in cases of inoperable tumours or tumours at the site where surgical access is very difficult and risk of injuring important structures. In this patient, we had to undertake a major surgery including neurosurgical team and head and neck surgery team with chances of risking great vessels, brainstem and spinal nerves in a 50 year old lady, for complete excision which would have been a great morbidity if any of these structures was damaged. Hence we took a decision to give a low dose radiation to tumour to arrest its further growth.

To the best of our knowledge, this the second case in world literature, where vagal schwannoma is

masquerading as parotid swelling. The first case of similar nature but less extensive was reported by Shetty et al in 2006.<sup>4</sup>

## CONCLUSION

Swellings in infra-auricular region are generally arising from parotid but schwannoma should be kept as a differential diagnosis. Radiological investigation like contrast enhanced CT or MRI is must for complete evaluation of the extent of the tumour as well as the site of origin. The risk of surgery should always be weighed in terms of morbidity and post-operative quality of life.

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