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***Case Report***

# AN INTERESTING CASE OF VAGAL PARAGANGLIOMA

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## ABSTRACT
Paragangliomas is not a rare entity but those commonly encountered are carotid body tumours or glomus tumours. A paraganglioma arising from the course of vagus nerve is extremely rare with very few cases reported in literatures till date. Standard approach to dealing with such tumours is complete surgical excision with a careful attempt to preserve the lower cranial nerves and great vessels of the neck. We report a case of a 19 year old female patient who presented to our outpatient department with complaint of right sided neck swelling below the mandible angle which was progressively increasing in size since last 7 months. Based on clinical examination and radiographic investigations a provisional diagnosis of benign neurogenic tumour was made. Gross total excision of the tumour was done by transcervical approach and histopathological reports confirmed the above diagnosis. The rare occurrence of vagal paraganglioma and threats encountered during surgical excision due to proximity to vital structures have prompted us to report the case.

***Key Words:*** *Vagal Paraganglioma, Parapharyngeal Space, Transcervical, Carotid Artery*

## INTRODUCTION
Vagal paragangliomas are benign neurogenic neck tumours with just 200 cases reported in the world literatures (Netterville, 1998). They arise from the neural crest cells located on the vagus nerve in the retrostyloid compartment of parapharyngeal space. Growth of these tumours is very variable, some grow upwards towards the skull base, and others extend down into the neck. They are seen with a slight more frequency in 4$^{th}$ decade females and can be functional (catecholamine hypersecretion) or non functional (Groblewski, 2004). Malignancy reported in 10 -19 % cases. They are ranked as least frequently occurring head and neck paraganglioma, highest being carotid body tumours followed by glomus jugulare (Urquhart, 1994).

## CASES
A 19 year female patient presented with painless, asymptomatic right sided neck swelling below the mandible angle which first appeared 7 months ago. It progressively increased in size of a peanut to present size of a large lemon. There were no associated complaints of dysphagia, dysnoea, hoarseness of voice, palpitations, excessive sweating or syncopal attacks. Patient's medical history was unremarkable. On examination a firm, discrete, oval swelling of size 4x3.5x5 cms was found (Figure 1) with associated transmitted pulsations. Fluctuation, reducibility, compressibility, movement with deglutition or protrusion of tongue was absent. No cervical lymphadenopathy noted. USG Doppler revealed a vascular mass lesion on right lateral neck and Computed tomography (CT) showed a well defined mass lesion extending superiorly upto styloid process and inferiorly upto lower margin of right submandibular gland at level of carotid vessel bifurcation (Figure 2).

### *Management*
Tumour was excised by trans-cervical approach, with platysma cut and SCM retracted for adequate exposure. It was seen arising from right vagus at the level of carotid fork. However difficult identification of great vessels was due to distortion of regional anatomy, internal carotid artery and jugular vein displaced forwards and backwards by the tumour respectively was preserved. Excision was done by cutting the nerve first at the level of carotid fork, then at the level of skull base. Adequate haemostasis was achieved. Post operative right vagal nerve palsy was inevitable. In addition patient suffered from

hypoglossal nerve paresis with deviation of tongue to right. Rest of the post operative period was uneventful.

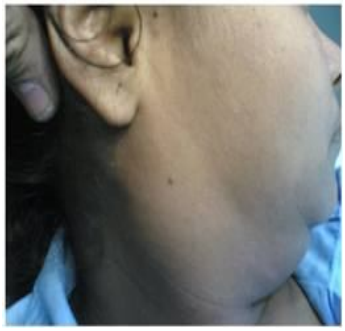

**Figure 1: Image of patient showing right sided neck swelling below the mandible angle**

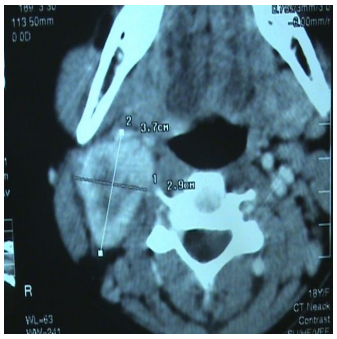

**Figure 2: CT scan showing well defined mass lesion at the level of carotid artery bifurcation**

***Surgical***

Control CT taken at early postoperative period confirmed the removal of the tumour. (Fig3 and 4)The patient was discharged on the 7th day with hypoglossal nerve showing slight recovery. Follow up after a period of a month showed complete recovery in tongue deviation. To relieve discomfort due to vagal nerve paralysis, patient is to be posted for type 1 thyroplasty after 6 months from the operative date.

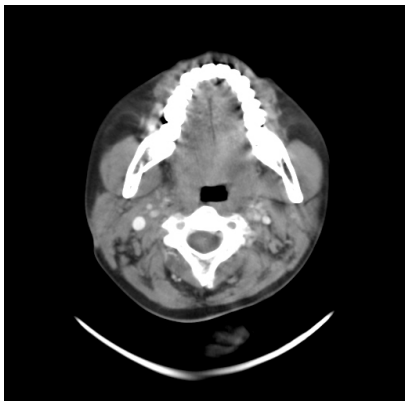

**Figure 3: Post operative CT scan showing no evidence of tumour**

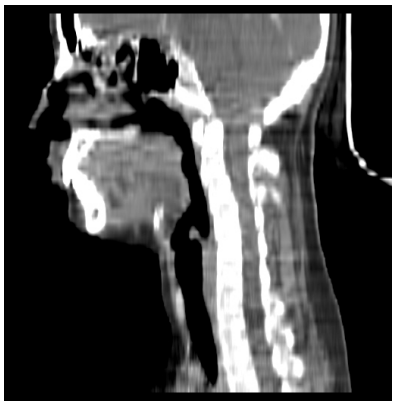

**Figure 4 : Post operative CT scan showing no evidence of tumour**

***Histopathological Diagnosis***

Gross – Weight of the tumour was 38 grams and size 4*3.5*5cms. It was tan pink coloured, partly encapsulated and well circumscribed.

Microscopically – Slides showed predominant monomorphic cell population comprising of neuroectodermal cell clusters were seen. Also areas of well defined nests and lobules separated by highly vascular septae (Zellballen pattern) were found randomly in the stroma. Features were suggestive of paraganglioma.

## DISCUSSION

Vagal paragangliomas are rare benign neck neoplasm accounting for only 2% of all head and neck paragangliomas. It can be solitary, bilateral, and either unilateral or bilateral associated with multiple paragangliomas (Myers, 1966). The incidence of multifocal tumours is more in those with a positive family history. Familial cases account for 40-50 % and diagnosis of these tumours is made at a younger age (Endicott, 1980). Recent studies show development of the disease has a genetic basis with an autosomal dominant inheritance.

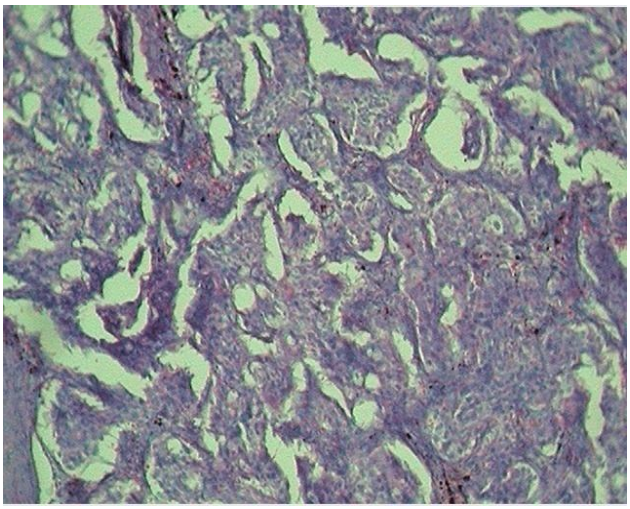

**Figure 5: Histopathology (10x) showing a characteristic Zellballen pattern**

These tumours usually present as an asymptomatic painless mass in the lateral aspect of neck. Large tumours may present with pressure symptoms like dyspnoea,dysphagia or may even be associated with cranial nerve palsies (9,10,11,12th), as seen in 38 % cases. Functional tumours which secrete catecholamines present with symptoms of palpitations, sweating, headache, hypertension and raised serum catecholamines and urinary vanilyl mandelic acid levels (Moore, 1986). Advances in imaging like CT scan, USG colour Doppler help in basic diagnosis of the tumours. MRI is a gold standard tool for skull base lesions and PET scan for evaluation of metastasis (Chen, 1985). FNAC and open biopsy are contraindicated. Benign and malignant tumours can be distinguished only on basis of local spread and distant metastasis, and not on HSP (Moore, 1986). Management option is local excision of tumour. Radiation therapy appears to be of minimal help and increases the risk of neurological deficits.

Morbidity associated with these tumours is unpredictable and is associated with loss of vagal function (Heinrich, 1995). Both our experience and that reported in literature show that vagal function cannot be preserved even when the nerve is anatomically intact but there is necessity to prevent additional cranial nerve deficits. The age of the patient has to be weighted against the size of the tumour, predicted growth and cranial nerve function. It may be reasonable to adopt a watchful waiting policy in elderly patients who might not adapt well to acute neural deficits (Sykes, 1996).

Nevertheless, even with high risk of injury to cranial nerves and great vessels, surgery still remains the preferred method of treatment for these tumours (Wetmore, 1981). Our case shows that such masses can be removed successfully and that with meticulous dissection and care, the cranial nerves and carotid arteries can be preserved.

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