Case Report:

Surgery of the carotid body tumour: a case report and review of literature

C. Jayaprakash,[1] H. Narendra,[1] P. Rajagopal,[2] K. Radhika,[3] B. Vijayalakshmi[4]

Departments of [1]Surgical Oncology, [2]Cardiothoracic Surgery, [3]Pathology, [4]Radiolgy, Sri Venkateswara Institute of Medical Sciences, Tirupati

ABSTRACT

The carotid body tumour is the most common head and neck paraganglioma arising from the carotid bodies located at the bifurcation of the common carotid arteries. These are very rare tumors with an incidence of 0.012% in general population. They present as slow growing, asymptomatic neck masses of varying duration. They occur in sporadic, hereditary and hyperplastic variants. The hereditary forms tend to be bilateral, functional and carry higher risk of malignancy. The angiography is the modality of choice for diagnosis and treatment planning. The surgical resection is the mainstay of treatment which can be performed with acceptable complication rates in experienced centres. We report the case of a 37-year-old lady who presented with a slowly growing mass on the right side of the neck for which she under went surgery and histopathological examination confirmed the diagnosis of carotid body tumour.

Key words: *Carotid body tumour, Paragangliomas*

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INTRODUCTION

The paragangliomas are rare tumours that arise from the neural crest. They may be found in the abdomen, thorax or head and neck regions. Of all the paragangliomas, only 3% occur in the head and neck region and two-third of them are carotid body tumours (CBT).[1,2] Their characteristic clinical and radiological features help in establishing the diagnosis. The primary modality of treatment is surgical excision and can be challenging both for the surgical and anaesthesia team. Here we report the occurrence of carotid body tumour in a 37-year-old lady which we resected successfully.

CASE REPORT

A 37-year-old lady presented to our outpatient clinic with a history of slow growing mass on the right side of the neck of five years duration associated with occasional pain. There were no history of dysphagia, dyspnoea, dysarthria or voice change. There was no history of palpitation or dizziness or syncopal attacks. On examination she had a single non tender, firm spherical pulsatile mass with a smooth surface of about 4 cm diameter in the upper third of neck deep to the muscular plane (Figure 1). The

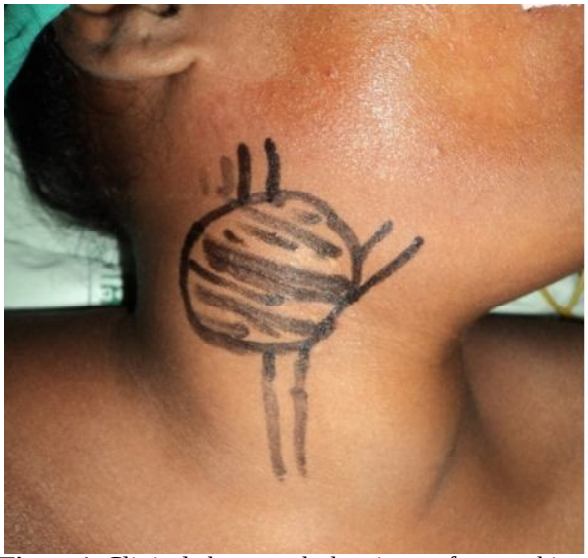

Figure 1: Clinical photograph showing surface marking of the tumour in relation to carotid vessels

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Corresponding author: Dr H. Narendra, Department of Surgical Oncology, Sri Venkateswara Institute of Medical Sciences, Tirupati, India.
e-mail: drnarendrah@yahoo.co.in

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mass could be moved horizontally, while the vertical mobility was restricted. On auscultation a bruit could be heard over the mass. Ultrasonograph revealed a heterogeneous lesion with increased vascularity and splaying of the carotid bifurcation suggestive of a CBT. Contrast enhanced computed tomography (CECT) showed intense enhancement with splaying of the carotid vessels (Figures 2A and 2B) extending from carotid bifurcation upto the angle of mandible. The carotid angiogram confirmed the presence of a CBT and showed encasement of external carotid artery by the tumour. The angiogram also showed patency of the circle of Willis (Figure 2B). At the time of surgery, a 6 × 5 cm sized CBT was noted at the carotid bifurcation encasing whole of the external carotid artery (ECA) and displacing the internal carotid artery (ICA). The carotid sheath was dissected, proximal and distal vascular control obtained with the vascular slings (Figure 3A). The effort to separate the ECA from the tumor in a sub-adventitial plane did not succeed and resulted in a tear at the carotid bifurcation. The ECA was ligated at origin and the tumour was resected (Figure 3B). The ICA was re-anastomosed to common carotid artery and wound was closed. In the postoperative period patient developed a small hematoma on the second day which was drained. There were no neurological deficits. The histopathological examination revealed polygonal cells with vesicular nuclei, prominent nucleoli and plenty of cytoplasm arranged in characteristic *zellballen* pattern with sustentacular cells in fibrous septae (Figure 4) suggestive of an extra adrenal paraganglioma and confirming it as a CBT. The probable intraoperative and postoperative complications have been summarized in the Table 1.

DISCUSSION

The carotid bodies are small, reddish-brown, oval structures, located at the posteromedial aspect of the carotid artery bifurcation in the peri-adventitial tissue. They detect changes in partial pressure of arterial oxygen (PaO_2) and carbon dioxide ($PaCO_2$), pH and temperature in the peripheral blood. They are composed of two types of cells, namely, type I and type II cells. The type I cells (chief cells) are amine precursor uptake decarboxylation type of cells and undergo hyperplasia in response to hypoxia. The type II cells are sustentacular cells and surround the chief cells. The carotid body is

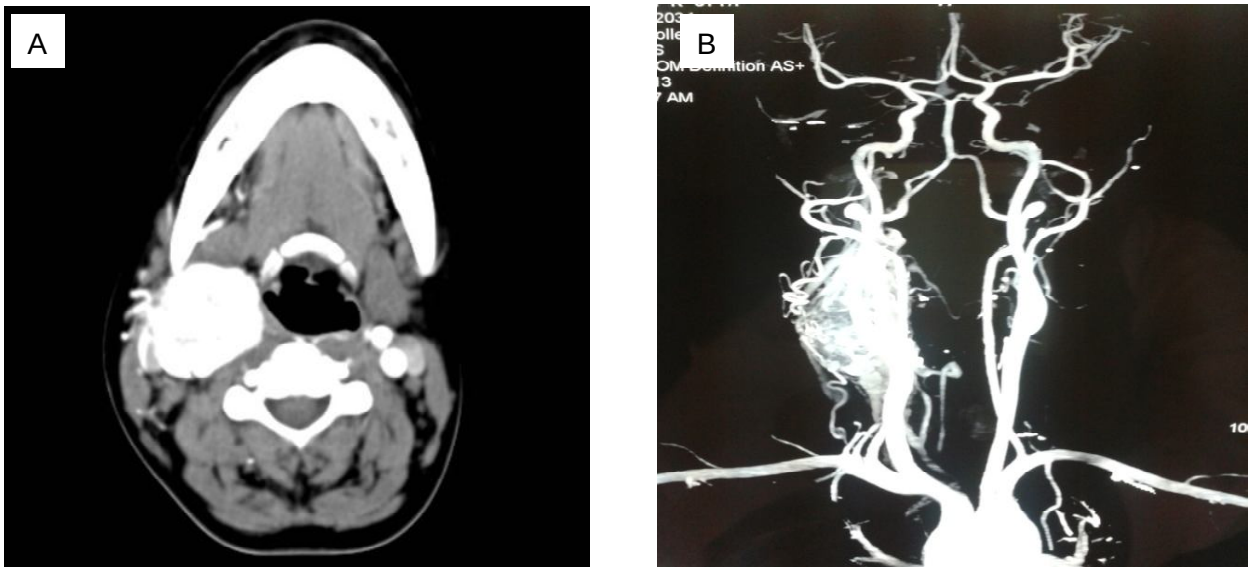
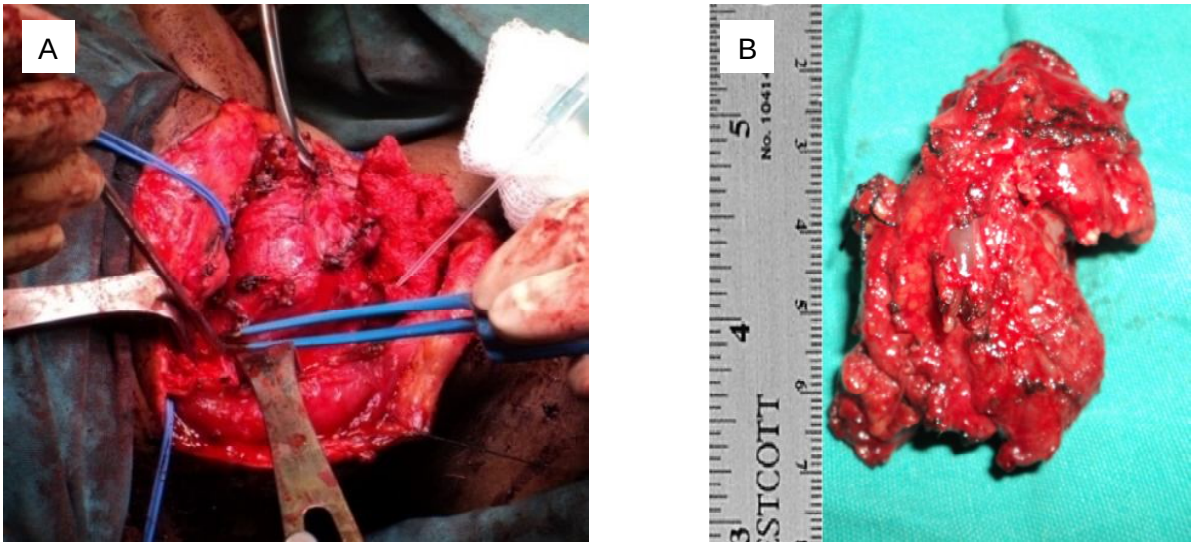


Figure 2: CECT showing intense tumour blush in the region of carotid bifurcation **(A).** Carotid angiogram showing tumour at right carotid bifurcation (Patent circle of Willis can be noted) **(B).**



Figures 3: Intraoperative photograph of obtaining proximal and distal vascular control **(A).** Excised tumour with lobular appearance due to indentation by carotid vessels **(B)**

highly vascular and receives its blood supply from the feeder vessels running through the Mayer ligaments, primarily from the ECA, typically the ascending pharyngeal artery. It is innervated by Hering nerve, a branch of the glossopharyngeal nerve. The chief cells detect changes in arterial oxygen tension and relay it to the respiratory center via IX nerve. The efferents from these centers relay via the X nerve and regulate the respiration and blood pressure.[1,3]

The CBT are seen in middle age with the reported mean age being around 45 years. The hereditary tumours can occur in as early as second decade. These are very rare tumours

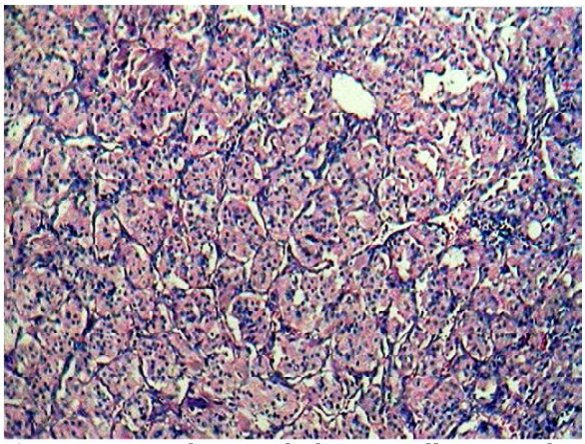


Figure 4: Microphotograph showing cells arranged in characteristic *zellballen* patterns (Haematoxylin and eosin × 100)

with an overall incidence of 0.012% in general population.[4] Overall, 5% of the CBT tend to be bilateral, more commonly in hereditary variants.Though there is no sexual preponderance seen at sea level, an eight-fold increase is incidence in females is noted at altitudes above 2000 meters.[1] The CBT can be sporadic, familial or hyperplastic, the sporadic tumours being the most common type. The familial tumours tend to occur at an earlier age with increased risk of being bilateral and malignant.They can also be associated with multiple endocrine neoplasia II, Von Hippel Lindeau and Neurofibroamtosis I syndromes. The familial forms are associated with mutations in paraganglioma genes that encode for subunits of the enzyme succinate dehydrogenase complex. The defect in this Kreb's cycle enzyme can induce intracellular hypoxia, releasing hypoxia mediators resulting in angiogenesis and hyperplasia.[1] The hyperplastic tumours occur in response to chronic hypoxia like chronic obstructive pulmonary disease, cyanotic heart disease or residing at high altitudes.[1]

Clinically, CBT are asymptomatic, slow growing tumours with estimated growth rate of about 1 mm per year. They can present with

Table 1: Complications of carotid body tumour surgery

Cardiac
Bradycardia
Arrythmia
Hypotension
Neurovascular
Haemorrhage
Vascular injury
Injury to cranial nerves IX-XII
Thromboembolism and stroke
Bilateral resection
Severe postoperative hypertension
Wound
Haematoma
Seroma
Infection

pain, tongue palsy, hoarseness or dysphagia, Horner's syndrome, or shoulder drop, the presence of symptoms indicating either a large tumor or malignancy. These tumours are typically located anterior to the sternocleido-mastoid near the angle of the mandible at the level of the hyoid bone. Characteristically, the tumour can be moved side to side but not upside down, due to location within the carotid sheath (*Fontaine sign*). They are pulsatile and often a bruit can be heard. These tumours can rarely be functional and secrete catecholamines and present with paroxysmal hypertension, palpitations, and diaphoresis.[1-5] The common differential diagnosis would include a metastatic level 2 neck node, salivary gland neoplasms, aneurysm of the carotid vessels, schwannoma of the cranial nerves and branchial cleft cyst.[2] The common temptation of the clinician to put a needle or biopsy the lesion before imaging should be avoided in patients with upper lateral neck swellings situated particularly in the line of carotid vessels with clinical features described above. It is important to keep the diagnosis of CBT in mind in any upper lateral neck swelling as inadvertent incisional biopsy may lead to catastrophic haemorrhage.

The initial evaluation should include an ultrasound, which is a noninvasive and economical test, doppler examination often showing the hypervascularity of these tumors. CECT reveals bright and rapid enhancement of these lesions with splaying of carotid vessels. Magnetic resonance imaging (MRI) shows characteristic salt and pepper appearance of these tumors indicating areas of decreased flow. The carotid angiography or magnetic resonance angiography (MRA) is considered as the gold standard for diagnosis. Angiography shows splaying of the carotid vessels (*lyre sign*) with an intense tumor blush. It also shows the main feeding vessel and facilitates tumour embolization in very large tumours. An additional advantage is that it shows the patency of circle of Willis, which is an important consideration if any intraoperative misadventure occurs as was in our case.[3,7] Whenever resection of carotid vessels is anticipated a carotid balloon occlusion test is done to confirm the patency of the circle of Willis and plan the surgery.

Shamblin classified CBT into three groups based on the relation to adjacent vessels.[2,8] The group I are small tumours that do not involve the vessel wall and donot splay the carotid bifurcation and are easily resectable. The group II tumours are large tumours that splay the carotid vessels and are adherent to the vessel wall but do not encase. The group III tumours encase the carotid vessels and also involve the adjacent nerves. These tumours require vascular resection and reconstruction.[1]

The CBT are treated with surgical resection because these tumours will continue to grow and eventually become symptomatic, some can be malignant, and there is no reliable screening method available.[9] A trans-cervical approach or a cranial basilar approach can be used depending on the proximal extent of the tumor. The obtaining of proximal and distal vascular control is crucial. These tumours are not

encapsulated and cannot be easily shaved off from the vessels. The dissection in a sub-adventitial plane, described as a 'white line' by Gordon is often necessary preferably with bipolar surgical diathermy.[10,11] Care is exercised to prevent injury to X, XI and XII cranial nerves. Intraoperative vigilance on the part of anaesthesia team is needed as bradycardia and hypotension may occur due to stimulation of the baroreceptors at the carotid bifurcation during tumour manipulation. The infiltration of 1% lidocaine in the sub-adventitial plane of the carotid bulb will reverse the bradycardia and hypotension. In majority of the Shamblin type I and II tumours resection can be done safely, whereas type III tumours necessitate vascular resection and reconstruction with either end to end anastomosis as in our case, interposition saphenous vein graft or prosthetic grafts.[2] The most common cause of postoperative morbidity are neurovascular and increase with the larger (> 5 cm) and Shamblin type III tumours. The incidence of permanent neurological damage is 30% to 45% and commonly involves marginal mandibular branch of VII X and XII cranial nerves. The incidence of cerebrovascular events range between 10% to 30%.[10,11] In patients with bilateral tumours, simultaneous resection can lead to severe refractory hypertension due to loss of baroreceptor arc reflex.

The CBT are radiosensitive, but radiation can only prevent further growth of the tumour and is recommended for inoperable disease, high risk patients, and incomplete resections, for metastatic tumours and for multiple paragangliomas. The ionizing radiation can induce arteriosclerosis in the vessel and also second primary tumours. Salvage surgery for any recurrence after radiation is often difficult due to fibrosis. Another alternative strategy in the management includes observation especially for CBT in very elderly and fragile patients with small tumours because of slow growth of these tumours. They are monitored regularly with doppler imaging.[1,9]

About 5%-10% of CBT can be malignant and should be suspected in the presence of local invasion, cranial nerve palsies, regional nodal metastasis and distant metastasis. Like most of the other neuroectodermal tumours, malignancy cannot be histologically diagnosed.[11,12] They are usually treated with surgical resection followed by radiation. There is no proven role of chemotherapy in the treatment of malignant carotid body tumours.

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