

## Case Report

# A case of successfully resected Shamblin type III carotid body tumour without vascular reconstruction

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## ABSTRACT

Carotid body tumours (CBT) are rare vascular neoplasms arising from the bifurcation of the common carotid artery, and are benign tumors with low incidence, slow growth, and low rate for malignant transformation. A rare functional CBT may produce neuroendocrine secretions causing catecholamine-related symptoms, such as palpitations, headaches, hypertension, tachycardia, or flushing. These tumours have classic radiographic features. These lesions splay apart the internal (ICA) and external carotid arteries (ECA), and as it enlarges, it will encase, but not narrow the ICA and ECA-Lyre's sign. If a diagnosis of CBT is suspected following a detailed physical examination, the diagnosis is almost always established by radiological imaging methods such as duplex ultrasonography, computed tomography (CT) angiography, magnetic resonance (MR) angiography, and digital subtraction angiography. These tumours were classified by Shamblin, according to the gross tumor vessel relationship and was based on the radiology, intraoperative findings and postoperative specimen examination. Group 1 tumors were minimally attached to the vessels and easily resectable. Group 2 tumors seemed to partially surround the vessel and were more adherent to vessel adventitia. These tumors were difficult to dissect but amenable to careful resection. Shamblin group 3 tumors had an intimate adherent relationship to the entire circumference of the carotid bifurcation and are usually inoperable. Surgical excision still remains the gold standard therapeutic modality for the treatment of CBTs. Radiotherapy is an alternative treatment modality which may decrease the tumor size or stop its growth. It is recommended for patients who cannot undergo surgery on account of extensive involvement, multiple tumors, and high operative and anesthetic risk.

**Keywords:** Carotid body tumour, Paraganglioma, Shamblin, Lyre's sign

## INTRODUCTION

A carotid body tumor (CBT), or a paraganglioma, is a chemodectoma. It is a rare neck tumor located at the adventitia of the common carotid artery bifurcation and are benign tumors with low incidence, slow growth, and low rate for malignant transformation.<sup>1</sup> A rare functional CBT may produce neuroendocrine secretions causing catecholamine-related symptoms, such as palpitations, headaches, hypertension, tachycardia, or flushing.<sup>4</sup> These lesions splay apart the internal (ICA) and external carotid arteries (ECA), and as it enlarges, it will encase, but not narrow the ICA and ECA.<sup>2</sup>

If a diagnosis of CBT is suspected following a detailed physical examination, the diagnosis is almost always established by radiological imaging methods such as duplex ultrasonography, CT angiography, MR angiography, and digital subtraction angiography.<sup>4</sup> Surgical excisions still remains the gold standard therapeutic modality for the treatment of CBTs.<sup>5</sup>

More widespread and frequent use of sensitive imaging modalities enhances early tumor detection. This may explain the decreasing rates of radical carotid artery resection and the declining rate of nerve injury.

Higher Shamblin class tumors are associated with greater blood loss, longer operative times, higher incidence of nerve injury, and the need for vascular sacrifice and reconstruction.<sup>5</sup>

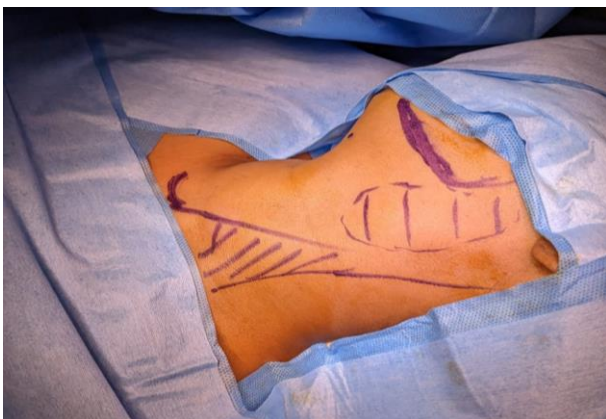
Here, we present a case of a Shamblin type III CBT which was successfully resected surgically without need of vascular reconstruction and the patient had an uneventful recovery following the surgery.

## CASE REPORT

36-year-old lady came to the outpatient department with complaints of recent increase in size of a swelling on left side of the neck below the jaw which she was having since past few years. She never had any complaints of pain over the swelling. There was no history of headache, dizziness, excessive sweating, palpitations, syncope, restlessness, visual disturbances, voice change, dysphagia, weakness of limbs. She was a known hypertensive for which she is on regular medications and is under control.

On examination, she was conscious and alert, moderately built and nourished, her vitals were stable and systems were found to be within normal limits.

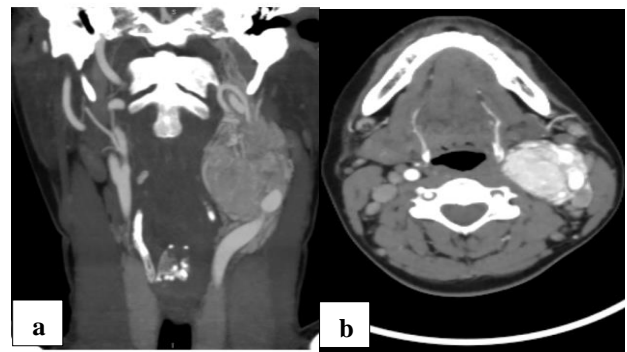
There was a palpable mass on the left side of neck in the jugulo carotid region, of approximately 6x4 cm, anterior to medial border of sternocleidomastoid, posterior to ramus of mandible and extending below the angle of mandible. The swelling was non tender, with well-defined margins and rubbery consistency, and showing transmitted pulsations. There was no sinus, ulceration or any other skin changes over the swelling. Fontaine's sign was positive, i.e., swelling could be displaced laterally but not vertically and with lateral displacement of the swelling, there was displacement of carotid pulse in the same direction. There was no bruit over the swelling.



**Figure 1: Tumour location and surface markings.**

She was provisionally diagnosed as having a non-functional left carotid body tumour, based on the clinical findings and her normal catecholamine levels. She was then referred to the radiologist for CT angiography.

CT angiogram of neck showed a well-defined ovoid soft tissue density lesion with avid heterogeneous enhancement measuring 3.0x4.0x4.7 cm, involving the left carotid space at the left common carotid bifurcation and splaying the internal and external carotid arteries. Superiorly the lesion was extending to C2 vertebra and inferiorly limited by carotid bifurcation. The distal left common carotid artery and the left carotid bulb were encased by the lesion. There was near complete encasement of external carotid artery in its proximal portion and complete encasement with mild narrowing of cervical internal carotid artery - which were features suggestive of carotid body paraganglioma (Shamblin type III).



**Figure 2: (a) and (b) CT angiogram of neck showing carotid body tumour of left side encasing the distal common carotid, proximal external and internal carotid arteries.**

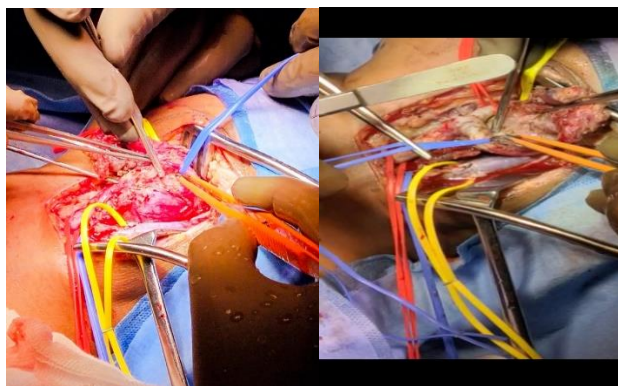
Patient and her attenders were counseled about the nature and complexity of the disease, possible risks of a surgical resection, need for vascular re-construction procedures and the potential complications that may follow the surgical procedure. After anesthetic evaluation and clearance and with informed written consent, she was planned for surgical resection of the tumour under general anesthesia, with carotid shunt, ePTFE grafts and patch as standby in case if arterial wall reconstruction was required due to tumour invasion.

An oblique incision was made anterior to medial border of sternocleidomastoid muscle and the platysma was divided. Sternocleidomastoid was retracted laterally. Spinal accessory nerve was identified and safe guarded.

Intraoperatively the findings were, 5x4 cm tumour over left jugulocarotid region, arising from the bifurcation of common carotid with multiple feeding vessels. Tumour was found to be encasing left common carotid, internal and external carotid artery and indenting left internal jugular vein. Tumour was densely adherent to left external carotid artery, internal carotid and common carotid, left glossopharyngeal and left hypoglossal nerve.

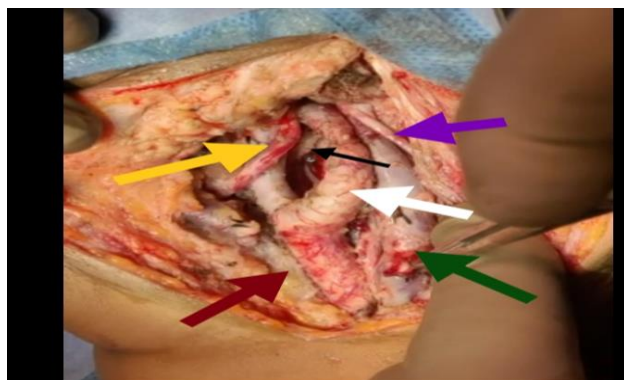
Left internal jugular vein isolated and left common facial venous trunk ligated and divided. Dissection started proximally where the tumour was adherent and to and

encasing the common carotid. Dissection started at sub-adventitious level of common carotid artery. Circumferential control of proximal common carotid artery was achieved. Maintaining the plane, dissection progressed superiorly. Superior thyroid artery was visualized, ligated and divided.



**Figure 3: Intra operative images showing early and late phases of dissection.**

Hypoglossal and glossopharyngeal nerves were gently dissected off the tumour. All nerves and other neck vessels were identified and isolated. The feeding vessels were ligated and divided. Tumour carefully dissected out completely from the internal and external carotid arteries, leaving the great vessels intact.



**Figure 4: Post-operative image showing the intact cranial nerves and great vessels (red arrow- CCA, white- ICA, green- IJV, black- IXth CN, yellow- XIIth CN, purple- XIth CN).**

She tolerated the procedure well. There were no intraoperative fluctuations of vitals and was extubated uneventfully after the surgery. She was then shifted to surgical high dependency unit for further monitoring in view of possible cardiovascular or neurological complications due to handling of great vessels. She was not allowed oral diet as there was a concern regarding neuropraxia and aspiration. On post-operative day second, she was shifted to general ward as she was found to be clinically stable and was allowed oral clear liquids maintaining propped up position. She developed

hoarseness of voice post operatively - vagal nerve stretch- for which ENT opinion was sought. No vocal cord palsy was demonstrated and she was managed conservatively. She was discharged on post-operative day four as she was clinically and hemodynamically stable, symptomatically better and was tolerating orally. On post-operative day five and day seven she was called back for review, there was no evidence of cardiovascular or cerebrovascular compromise and no signs of cranial nerve injuries.

Her histopathology report showed a circumscribed neoplasm composed of cells arranged in Zellballen pattern and organoid pattern with the neoplastic cells having eosinophilic granular cytoplasm with round to oval nuclei granular chromatin. There was no evidence of vascular invasion, increased mitotic activity or necrosis.

## DISCUSSION

The carotid body is the largest collection of paraganglia in the head and neck and is found on the medial aspect of the carotid bifurcation bilaterally. A CBT, or a paraganglioma, is a chemodectoma and the most common type of paraganglioma derived from the neural crest located at the adventitia of the common carotid artery bifurcation.

CBTs are benign tumors with low incidence, slow growth, and low rate for malignant transformation, which are not significantly associated with age.<sup>1</sup> A rare functional CBT may produce neuroendocrine secretions causing catecholamine-related symptoms, such as palpitations, headaches, hypertension, tachycardia, or flushing.<sup>4</sup>

In the head and neck region the normal paraganglia are associated with the parasympathetic nervous system and paragangliomas arising from these parasympathetic sites account for up to 70% of extra-adrenal paragangliomas. The most common site is the carotid body and have classic radiographic features. These lesions splay apart the internal (ICA) and external carotid arteries (ECA), and as it enlarges, it will encase, but not narrow the ICA and ECA- Lyre's sign.<sup>2</sup>

Recent literature suggests a molecular basis for the development of some paragangliomas, i.e. germline mutations. Six genes have been identified and are thought to contribute to the development of pheochromocytoma/ paraganglioma. These include RET, VHL, NF1 and SDH subunits SDHB, SDHC, and SDHD. SDHD and SDHB mutations account for a significant percentage of head and neck paragangliomas.<sup>2</sup>

CT angiography reveal a hypervascular mass with enlarged feeding arteries (typically the ascending pharyngeal or ascending cervical artery), intense tumor blush and early draining veins. Indium-111 octreotide, which is a somatostatin analog, is a nuclear medicine imaging study that is useful in the evaluation of paragangliomas, since these are neuroendocrine neoplasms that have surface receptors for somatostatin.



A focal area of early intense radiotracer uptake will be seen in the region of the paraganglioma and is sensitive for detecting tumors greater than 1.5 cm.<sup>2</sup>

Shamblin et al in the 1970s suggested a surgical classification of CBTs into 3 groups.<sup>2</sup> This classification was according to the gross tumor vessel relationship and was based on the radiology, intraoperative findings and postoperative specimen examination.

Group 1 tumors were minimally attached to the vessels and easily resectable. Group 2 tumors seemed to partially surround the vessel and were more adherent to vessel adventitia. These tumors were difficult to dissect but amenable to careful resection. Shamblin group 3 tumors had an intimate adherent relationship to the entire circumference of the carotid bifurcation and are usually inoperable.<sup>3</sup>

If a diagnosis of CBT is suspected following a detailed physical examination, the diagnosis is almost always established by radiological imaging methods such as duplex ultrasonography, CT angiography, MR angiography, and digital subtraction angiography. Nowadays, ultrasonographic examination is widely used for screening because it is an easily available and non-invasive imaging modality. CT and MRI help to assess the size, degree, and invasiveness of the tumor. Angiographic methods allow the evaluation of the vessels supplying the tumor and preoperative embolization.<sup>4</sup>

Surgical excision still remains the gold standard therapeutic modality for the treatment of CBTs. Radiotherapy is an alternative treatment modality which may decrease the tumor size or stop its growth. It is recommended for patients who cannot undergo surgery on account of extensive involvement, multiple tumors, and high operative and anesthetic risk.

More widespread and frequent use of sensitive imaging modalities enhances early tumor detection. This may explain the decreasing rates of radical carotid artery resection and the declining rate of nerve injury.

Higher Shamblin class tumors are associated with greater blood loss, longer operative times, higher incidence of nerve injury, and the need for vascular sacrifice and reconstruction.<sup>5</sup>

## CONCLUSION

CBTs, also known as paraganglioma or carotid body chemodectoma, are rare neck tumours that arise from the adventitia of common carotid artery at the bifurcation. These tumours are typically painless and slow growing. A thorough clinical examination followed by radiological imaging methods such as duplex ultrasonography, CT angiography, MR angiography, and digital subtraction angiography helps in diagnosis of the tumour. Surgical resection is the treatment of choice, but these neoplasms are very vascular making resection challenging. In carotid body tumours, the histological features are more readily identified and the differential diagnosis includes other neuroendocrine tumours such as medullary thyroid carcinoma and neuroendocrine carcinoma.

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