



CAROTID BODY TUMOR - A RARE CASE REPORT

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**ABSTRACT**

Carotid body tumors are rare, slow-growing, hyper vascular neuroendocrine tumors, accounting for 0.5% of all head and neck tumors. They typically present as a slow growing, painless neck mass occurring in the middle-aged people with higher predisposition in females along the anterior border of the sternocleidomastoid muscle. Although these tumors are benign they have tendency for malignant transformation. Complete surgical excision is the gold standard therapeutic modality for the treatment of carotid body tumors. Early surgical removal is recommended to prevent the development of larger and more advanced tumors, which are associated with higher morbidity and mortality. We report a case of a 48-year-old female patient presenting with a painless, gradually progressive left lateral neck mass, diagnosed as carotid body tumor by CECT and treated with local excision. Patient was discharged without neurological complications. We report this case due to its rarity and the high vascularity and proximity to cranial nerves and major vessels make this tumor a surgical challenge.

**KEYWORDS :** Carotid body tumor, surgical excision

**INTRODUCTION**

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. Carotid Body Tumours (CBT) are rare neoplasms arising from the chemoreceptor cells of the carotid bulb. They are also known as, intercarotid Paragangliomas. The majority of these tumours are benign and slow-growing, with an overall incidence of 0.01% of the population<sup>1</sup>. They have a very small chance of becoming malignant. Of these 10% are familial with bilaterality seen in 30% Of the familial form, while only 5% of the sporadic forms display multicentricity.

Once a paraganglioma is diagnosed, complete surgical excision is the treatment of choice, before they become painful and involve cranial nerves. Although surgical resection is the recommended treatment modality, it is associated with a risk of up to 30-40% morbidity and mortality<sup>2</sup>. Currently, the Shamblin Classification system, which is based on degree of encasement of adjacent XCarotid arteries, is used to assess the patient's risk of intraoperative bleeding, Cranial Nerve injury, and potential need for carotid artery resection and revascularisation<sup>3</sup>.

**CASE REPORT**

A 48 Yrs. old female patient presented with complaints of swelling over left side of neck since 1 yr. Initially the swelling was 1 x 1 cm<sup>2</sup>, later gradually progressed to the present size of 4x4 cm<sup>2</sup>, associated with difficulty in swallowing. No h/o trauma, episodes of Transient ischemic attacks, headache, flushing or chest pain. It was not associated with pain.

**Later on, On Examination:**

A spherical pulsatile swelling noted on the left side of the neck.Fig 1.



Fig 1 Neck mass

Later on, Ultrasound of neck shows A well defined heterogenous mass lesion with internal anechoic tubular structures is noted measuring 4.2x2.9x3.2 cm and the lesion is located at the left carotid bifurcation and is seen to splay the ICA and ECA. On applying colour doppler shows intense vascularity within mass lesion. Fig 2.

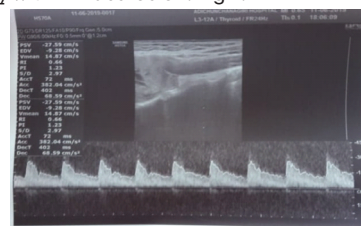


Fig 2 colour doppler

On CECT Neck study reveals: A well defined soft tissue density lesion measuring 3.6x4.3x5.0 cm is seen at the bifurcation of left CCA, splaying ECA and ICA showing homogenous intense post contrast enhancement. The mass lesion is extending from C2-C5 vertebra. The lesion is abutting submandibular gland anteriorly and SCM posteriorly, Medially hyoid bone and parasternal muscles posteromedially with maintained fat planes. Few sub centimetric lymph nodes noted bilaterally, largest measuring 5.5 mm seen in level I and Level II on right side. Features suggestive of left sided carotid body tumour. Fig 3.

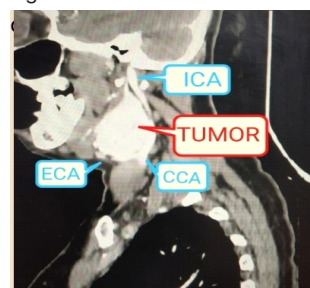
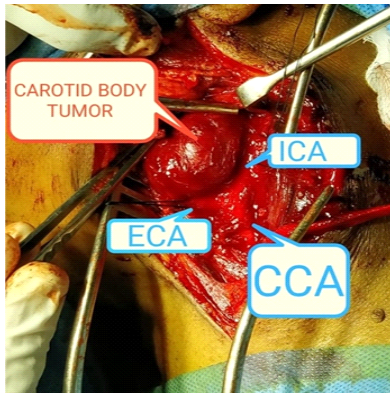


Fig 3 CECT Neck

After all these pre-operative investigations, the patient underwent surgery under general anaesthesia. After ensuring

temporary carotid control with vascular loops, sub-adventitial dissection of the lesion was carried out starting from the common carotid caudally and then progressing cranially towards the bifurcation and its branches. The major vascular supplies were sequentially isolated, ligated and divided. Fig 4.



**Fig 4** intra operative findings

Histopathological examination confirmed the diagnosis of a glomangioma, the histological equivalent of a Carotid Body Tumor (CBT). The postoperative period was uneventful, and the patient was discharged on 7th post-operative day with no neurological deficits. Fig 5.



**Fig 5** Post-operative 7th day

## DISCUSSION

It is important to distinguish between secreting and non-secreting tumors. A hormonal check-up for catecholamine should be under taken to avoid precipitating a catecholamine crisis.

In this case, the tumor was non-functioning/non secreting.

The majorities of CBT appear to be sporadic or can be hereditary (one-third to one-half of the cases) 6. They have been linked to mutations in the genes encoding subunits of the Succinate Dehydrogenase (SDH) enzyme and are also seen in syndromes such as MEN2, Von Hippel Lindau (VHL) and neurofibromatosis type I 7.

When patients are referred for atypical neck swelling, ultrasound is often the first acquired imaging modality. Carotid body tumors appear as a hypoechoic mass at carotid artery bifurcation and display prominent vascularization when examined with colour Doppler. Displacement of the Internal (ICA) and External Carotid Arteries (ECA) is an important clue and should prompt further image evaluation 8. Historically Digital Subtraction Angiography (DSA) has been the golden diagnostic standard. With the advent of high resolution angiographic Computed Tomography (CTA) and Magnetic Resonance Imaging (MRI) DSA has been replaced as the imaging modalities of choice for detection of the carotid body tumors. However, preoperative angiography provides useful information about the vascular anatomy and its collateral circulation. This allows for careful planning if sacrifice of a major blood vessel is deemed necessary during surgery 9. Due to unavailability of CTA and MRA in our institution we couldn't get it done which is the limitation of this article.

Surgical excision has been the standard approach for removal of CBTs, especially when the CBTs are symptomatic. The standard surgical principles for carotid body tumor excision include wide surgical exposure, proximal and distal vascular control, identification and preservation of the neurovascular structures, careful tumor dissection from the external and internal carotid arteries, ligation of the external carotid arteries when necessary, and vascular shunting and grafting wherever necessary<sup>7</sup>.

In this case, the patient had pressure symptoms like difficulty in swallowing, so surgical excision is the best option and also the treatment of choice. Intraoperatively, after ensuring temporary carotid control with vascular loops, sub-adventitial dissection of the lesion was carried out starting from the common carotid caudally and then progressing cranially towards the bifurcation and its branches.

However, a high incidence of postoperative cranial nerve deficits has remained over the years. This raises the question whether the high rate of postoperative cranial nerve dysfunction favours observation rather than resection for asymptomatic tumors<sup>5</sup>.

In this case, patient was discharged on post-operative day 7th without any neurological deficits.

Hence, a conservative management in an asymptomatic CBT must be done, even though CBT eventually cause symptoms in nearly 75% of patients due to local growth, which makes removal more challenging.

## CONCLUSION

Carotid body tumors (CBT) are rare neuroendocrine tumours, located at the carotid bifurcation. They have a very low potential of becoming malignant. Malignancy cannot be confirmed by histopathological investigation, but can only be proven by metastasis into other tissues organs. Improved diagnostic methods are useful for the treatment of CBT, nowadays. The CTA and MRA may identify the border and supply artery of the tumor, leading to markedly decreased bleeding episodes during the surgery. However, in the absence of these investigatory modalities also refined surgical techniques or skills can enable safe resection of the tumor with minimal post-operative complications.

It is evidently seen in this case, that the patient had no neurological deficits after surgery whatsoever and could be discharged safely within 7 days of post-operative period.

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