

## **Carotid Body Tumour (Paraganglioma) Treated Surgically. A Case Report & Literature Review**

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### **Abstract-**

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 are rare neoplasms arising from the chemoreceptor cells of the carotid bulb. We report a case of carotid body tumour in a 46-year-old male, who presented with painless, pulsatile, gradually progressive lateral neck swelling. The diagnosis is suspected on the basis of history, clinical and radiological examination findings and a successful surgical excision of the tumour is performed. Histopathological examination confirms the diagnosis of carotid body tumour. (Paraganglioma) In this article we present brief about carotid body tumours in terms of its clinical and imaging presentation, evaluation, and management (pre & post surgical management).

**Keyword:** - carotid body tumour, carotid bulb, paraganglioma

## 1. Introduction

The carotid body is a structure of elliptical shape, 3 to 4 mm in size, which is located at the bifurcation of the common carotid, at the level of its adventitial layer. Its functions are chemo reception and baro reception and because of this tumours that originate from it are called “chemodectomas”, (that is chemoreceptor tumors<sup>1, 2</sup>.) Carotid body tumours are neoplasms that arise from paraganglion cells and although they are well delimited, they are unencapsulated tumours and are highly vascularised by external carotid artery and their “vasa vasorum”. The CBT is usually benign with the incidence of malignant tumours below 10%. The majority of these tumours are asymptomatic and initially noticed by inspection and palpation of neck swelling during the physical examination, or more commonly as incidental findings on radiological imaging studies. There is no sex prediction for CBT, occurred in usually in 4<sup>th</sup> & 5<sup>th</sup> decayed of life<sup>3</sup>. Symptoms are varied, they can be asymptomatic or manifest as a slow-growing tumour that is painless and pulsatile, in the side of the neck (cervical region), close to the angle of the jaw, and occasionally lead to complaints of hoarseness, deglutition problems and symptoms of carotid sinus syndrome. In this case report our patient c/o pain, dysphasia, and slow growing mass in right side of face since 3 month. In carotid body tumour most commonly involved are hypoglossal nerve, glossopharyngeal nerve, vagus nerve and sympathetic chain / trunk .(injury to sympathetic trunk lead to ptosis<sup>4,5</sup>).

Clinical history, examination, and radiological diagnosis are the keystones to diagnosis and management. Ultrasound, Computed tomography (CT) and Magnetic resonance imaging (MRI) are useful radiological tools in diagnosis<sup>6, 7</sup>. Yet Angiography is essential to study about the vascular anatomy (we didn't perform this in our case) .Thus, in order to prevent local invasion and metastasis, early surgical excision is considered a primer curative treatment option for the treatment of CBTs. In this paper, we report a case of CBT which was successfully treated with complete surgical excision. In this case we did CECT NECK / MRI NECK. A 46-year-old male patient presented with right side neck swelling that persisted for 1 year. On physical examination, a pulsating firm painless mass measuring about 3.1\*\*AP\*\*4.6\*\*TR\*\*5.4CC in size was found on the right side of his neck, near the angle of the mandible in the right jugulo carotid region There was no pressure symptom and it was more mobile transversely than vertically. Pulsations were felt on deep palpation and a faint bruit was heard on auscultation.

- Doppler Ultrasonography s/o: - a well-defined hypo echoic mass with increased vascularity at the bifurcation of the common carotid artery right side.

CECT NECK S/O:-

- Well defined ovoid lobular lesion of muscle density on non –contrast images is seen in right sided carotid space, posterior to the carotid vessels. It is seen to displace ICA and ECA antromedially (non –splaying) with the IJV displaced postero –laterally. The large lesion is seen to indent upon the oropharynx-medially and SCM laterally .The lesion is seen just above the level of hyoid bone at C 5 level and seen to extend rostrally up to C 1 & C 2 level it measure 3.1 \*4.6\*5.4 (cc) cms.

MRI –S/O:-

- Well circumscribed ovoid mass in the right posterior carotid space measuring 5\*3.7\*6.5 cm

Mass is located posterior to the right carotid vessels, displacing the right internal & external carotid vessels anteromedially.

The right IJV is displaced & compressed postero-laterally. Medially mass is indenting the right side of oropharynx and laterally indenting the right SCM. Posteriorly, mass indents the right Para-spinal muscles.

## 2. Procedure

The neck was explored by a vertical incision under general anaesthesia. A homogeneous tumour with a smooth contour and approximately measuring 5\*3.7\*6.5 cm long was observed at the carotid bifurcation. It was classified as a grade-2 tumour according to the Shamblin classification system. The common, internal, and external carotid arteries were isolated. En bloc resection of the tumour and carotid bifurcation was performed with CTVS team. Small arterial branches arising from the bifurcation had to be ligated. Tumour was closely adherent to vagus nerve & hypoglossal nerve. In this case we sacrifice right side vagus nerve and hypoglossal nerve. Dysphasia, hiccups, blur speech and aspiration was occurring in the immediate post operative period. For this we placed Nasogastric tube (NG/OG) in patient for 10 days. We educate the patient for swallowing & speech with the help of trainer / therapist.

- Post op recovery was good after 10 days .patient improve swallowing but some complication's still persist like blur speech and ptosis of right eye.(due to thermal /or/ traction injury of sympathetic trunk of right side )

## 3. Discussion

According to Shamblin<sup>15,16</sup> et al. carotid body tumours can be classified into three groups depending on their circumference and degree of adherence: Group I – small tumour with no adherence to vessels that can be resected without damaging neighbouring structures; Group II – intermediate tumour with minor adherence to vessels, which is more difficult to dissect and may require revascularization; Group III – large tumour with infiltration of vessels, which is almost impractical to dissect, making resection en bloc with the carotid bifurcation and revascularization with saphenous vein or prosthetic graft necessary(Figure-. The carotid body is the largest collection of paraganglia in the head and neck and is found in the carotid space (Table 2). Carotid body was 1st described by von Haller in the year 1743. It is a reddish-brown, well circumscribed, highly specialized round organ, located in the adventitia of the carotid bifurcation, supplied by the feeding vessels run primarily from the ascending pharyngeal branch of the external carotid artery, and innervated through the glossopharyngeal and vagus nerves<sup>17, 18</sup>. (Table 1)

The normal carotid body measures 2-6 mm in diameter but is often larger in people living at higher altitudes. It functions as a chemoreceptor organ which is stimulated by acidosis, hypoxia and hypercapnea, and plays a role in the autonomous control of blood pressure, heart rate, respiration, and blood temperature in response to changes in these parameters by increasing sympathetic flow. CBT can be found at any age and is frequently seen between 50 and 70 years old (range, 18–94 years), with slightly higher prevalence in women than men (male-to-female ratio of 1:1.9). Bilateral disease is significantly more frequent in familial (31.8% of cases) than in non-familial CBT (4.4%).

In this case, the patient is 46 yr male with the tumour on one side (right). The blood supply of CBT is abundant, which is mainly from external carotid artery and branches. Blood supplies from internal carotid artery, vertebral artery, ascending pharyngeal artery and superior thyroid artery have also been reported<sup>19, 20</sup>. CBTs are slow-growing hyper vascular tumours which represent approximately 0.03% of all neoplasms. More tumours (57%) were on the right side, whereas 25% were on the left, 17% were bilateral, and 10% were malignant. Notably, the lesion in the present case is in the right side of neck (most common side) and does not have malignant characteristics. CBTs usually manifests as an asymptomatic anterior neck mass. In larger tumours, they can be associated with the myriad of presenting symptoms of a space-occupying lesion in this location, such as fullness, pain, dysphasia, odynophagia, hoarseness, and strider. The tissue is often rubbery, firm, and noncompressible. CBT may produce neuroendocrine secretions causing catecholamine-related symptoms, such as palpitations, headaches, hypertension, tachycardia, or flushing. The lesion in our case showed an asymptomatic neck mass with rubbery, firm, noncompressible, and more mobile transversely than vertically<sup>21</sup>.

CBTs have diagnostic and management difficulties since there is a lack of guidelines in the literature for their diagnosis and treatment. If a diagnosis of CBT is suspected following a detailed physical examination, the diagnosis is almost always established by radiological imaging methods such as 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 tumour. Angiographic methods allow the evaluation of the vessels supplying the tumour and preoperative embolization. On account of the hypervascularization and proximity to various vascular and nervous structures of these tumours, biopsy as a diagnostic method is contraindicated since it presents a risk of massive haemorrhage and dissemination and can lead to pseudo aneurysm formation and carotid thrombosis as well.

Surgical excision still remains the gold standard therapeutic modality for the treatment of CBTs. Radiotherapy is an alternative treatment modality which may decrease the tumour size or stop its growth. It is recommended for patients who cannot undergo surgery on account of extensive involvement, multiple tumours, and high operative and anaesthetic risk. Additional considerations specific to CBT resection include routine vein mapping, and preparation of one thigh into the operative field for potential saphenous vein harvest should be needed for reconstruction. Additionally, the choice of the incision may vary depending upon the extent of the tumour. Incision choices include a curvilinear incision along the midportion of the tumour, a “hockey stick” incision along the anterior border of the sternocleidomastoid, and in the case of a very high tumour, a modified radical neck T incision<sup>23</sup>. Regardless of patient age and tumour size, early surgical removal of CBTs is advised in order to prevent the development of larger and more advanced tumours, which are related to higher morbidity and mortality.

If the size of CBT is more than 5 cm, mortality is 1% - 3% after surgical intervention. In initial stage surgical excision of CBTs is a very effective and safe procedure

with a low rate of major neurovascular complication and mortality. Early detection and complete surgical removal of CBTs improve the outcome. In our case tumour is classified as a grade-2 tumour according to the Shamblin classification system and it is removed by a vertical incision under general anaesthesia.

### **Differential diagnosis:-**

Differential diagnosis of CBTs should be distinguished from masses in the neck or the lesions originating from the carotid space. The most important differential diagnosis include aneurysm or pseudo aneurysm of the carotid artery, hematoma, glomus vagale tumour, and vagal schwannoma, carotid body hyperplasia<sup>21,22</sup>. In this case Tumour was closely adherent to vagus nerve & hypoglossal nerve. In this case we sacrifice right side vagus nerve and hypoglossal nerve. Dysphasia, hiccups, blur speech and aspiration was immediate post operative complication .For this we placed Nasogastric tube in patient for 10 days. We educate the patient for swallowing & speech with the help of trainer / therapist. Post op recovery was good after 10 days. Patient is improve swallowing but some complication's still persist like blur speech and ptosis of right eye.(due to thermal /or/ traction injury of sympathetic trunk /2<sup>nd</sup> cranial nerve of right side ) for ptosis we suggest the patient to take methylcobalamin/vit.B12 for 90 days . Methylcobalamin is required for maintain integrity of myelin, neural function, proper red blood cells formation, it help's in regeneration of axonal nerves.

### **Conflict of interest-**

The authors declare that they have no conflict of interest."

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