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# Unilateral Carotid Body Tumor: Case Report and Implications Report and Pre-operative Management

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## Abstract

Carotid body tumors are rare and slow-growing neurogenic tumors for which surgery is currently the standard of care. However, because of the rare presentation, little is known about the pre-operative management. This case report describes a 72-year-old patient presenting with a swelling in the neck. The relevant literature is also reviewed.

## Introduction

Carotid Body Tumors (CBT) are rare neoplasms originating from neural crest cells, referred to as paragangliomas (extra-adrenal). They are not to be confused with pheochromocytomas, which are intra-adrenal neoplasms, due to the different implications for associated neoplasms, malignancy risk and genetic testing. They can be divided in sympathetic and parasympathetic paragangliomas [1]. Most CBTs are parasympathetic and non-functional. Only four to five percent of them are sympathetic and have catecholamine production [2]. These extra-adrenal parasympathetic paragangliomas are found near arteries and cranial nerves of the branchial arches, with a cervicocephalic distribution. Paragangliomas are named after their site of origin.

They most commonly occur at the carotid bifurcation where they are known as carotid body tumors. The tumor is typically mobile in the lateral plane with restricted mobility in the cephalocaudal direction (Fontaine sign) [3]. Additional sites of origin include the jugular bulb (jugular paraganglioma), around the vagus nerve (vagal paraganglioma) and within the middle ear mucosa (tympanic paraganglioma).

If the tumors produce catecholamines, patients may complain of symptoms such as episodic headache, fluctuating hypertension and palpitations [4].

Because of its close proximity to the carotid vessels and cranial nerves (X-XII), enlargement of the tumor may cause progressive neurologic symptoms such as odynophagia, dysphagia or hoarseness of voice. Compression or erosion of surrounding tissue alone is not an acceptable proof of malignancy. Evidence of malignancy is only accepted when there is metastasis to non-neuroendocrine tissue, such as cervical lymph nodes, lung, liver and skin [5]. Local recurrence or lymph node metastasis after total resection of the primary mass, or by the detection of distant metastasis is also seen in malignant CBTs [6].

For long time whether malignancy in CBTs can be seen on histologic characteristics. One study stated that the following histologic characteristics are suspicious for malignancy: central necrosis of the clusters, invasion of the vascular spaces, and mitoses [7].

However, others have considered that histology alone is unreliable to differentiate between benign and malignant CBTs. Only the presence of metastasis in lymph nodes or distant organs is acceptable proof of malignancy [8,9]. If CBT metastasize, there is a very high rate of regional confinement (94%) [10]. Bone and lung metastasis are the most frequently reported distant metastases.

## Case Presentation

A 72-year-old female presented to the otolaryngology clinic with disturbing pain in the right

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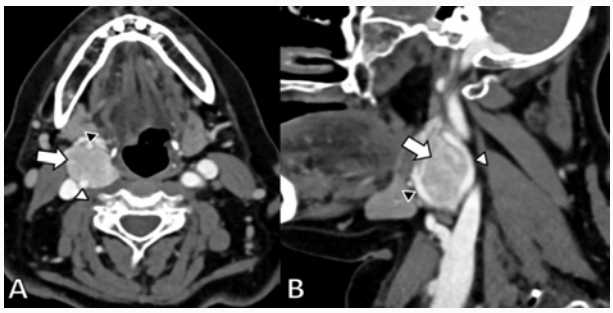


**Figure 1:** a) Axial reformatted CT angiography images showing a well-delineated intensely homogenously enhancing mass (arrow) at the bifurcation of the common carotid artery. The mass is closely related to the internal (white arrowhead) and external (black arrowhead) carotid arteries. b) parasagittal reformatted image demonstrates splaying of the ICA and ECA, the so-called lyre-sign.

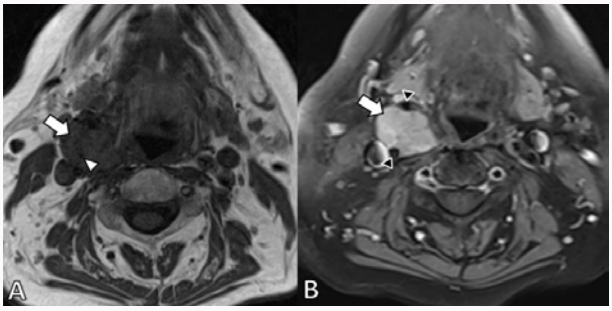


**Figure 2:** a) On T1-weighted image the lesion (arrow) is isointense compared to muscle. Note the small flow void (white arrowhead) within the mass. b) T1-weighted image with fat suppression after intravenous administration of gadolinium contrast. There is homogenous enhancement of the lesion.

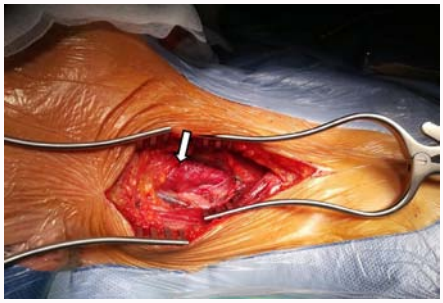

**Figure 3:** Per-operatively: Carotid body tumor (arrow) surrounding the bifurcation.

side of the neck for about six months. She also complained of a sore throat and general fatigue. Physical examination showed a 2 cm to cm swelling situated just below the mandibular angle, lateral to anterior border of sternocleidomastoid muscle. Fontaine sign, in which the swelling is more mobile in the horizontal plan compared to the vertical plane, was positive. The mass was non-tender, non-pulsatile with no increase in size with act of coughing or straining. No murmur was noticed on auscultation. Examination of all cranial nerves was normal.

Due to the swelling and her general malaise an ultrasound was preformed demonstrating a solid mass at the region of carotid bifurcation. Further imaging evaluation with computed tomography angiography (Figure 1) and magnetic resonance imaging angiography (Figure 2) showed a homogenously enhancing mass in the crotch of the left carotid artery bifurcation. The mass causes splaying of the internal carotid and external carotid arteries, in keeping with the diagnosis of a carotid body tumor. On imaging the tumor was classified as Shamblin type II. Referral to the vascular surgeon for removal of the carotid body tumor was made.

Hormonal check-up showed no abnormalities in catecholamines.

Computed Tomography Angiography (Figure 1) and Magnetic Resonance Imaging angiography showed a homogenously enhancing mass in the crotch of the left carotid artery bifurcation. There is splaying of internal carotid and external carotid arteries, in keeping with the diagnosis of a carotid body tumor. On imaging the tumor was classified as a Shamblin type II (Figure 2).

After all this pre-operative investigations, the patient underwent surgery under general anesthesia. An incision was made along the anterior border of Sternocleidomastoid Muscle (SCM) and after careful incision of the platysma and the carotid sheath; the tumor was found (Figure 3).

The tumor was highly vascularized and therefore removed with the use of bipolar coagulation. Repeated nerve stimulator tests were essential and performed to ensure nerve preservation. Complete removal was obtained and no vascular reconstruction was needed (Figure 4). 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 with no neurological deficits except some mild complaints of hypoesthesia on the right side of her neck.

## Discussion

### Therapy strategies

It is important to distinguish between secreting and non-

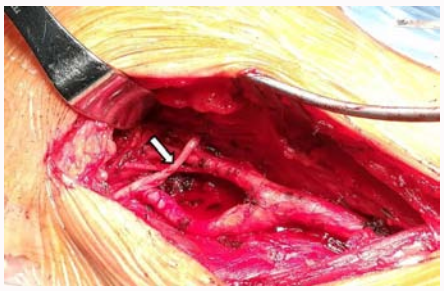

**Figure 4:** Complete removal of the tumor with preservation of the hypoglossal nerve (arrow) and carotid vessels.

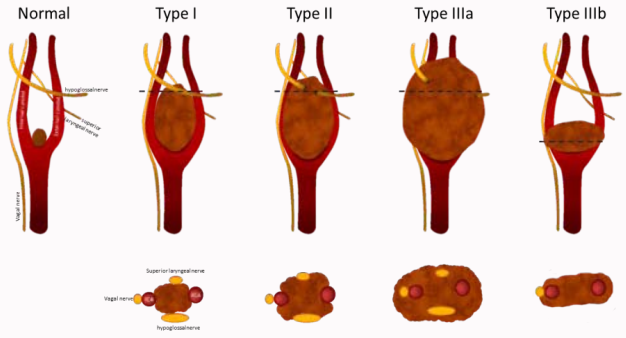


**Figure 5:** Shamblin classification as proposed by Luna Ortiz et al. [25] in which type IIIa represents the original type by Shamblin and type IIIb including I, II, III, but differently in partial or total infiltration.

secreting tumors. A hormonal check-up for catecholamine should be under taken to avoid precipitating a catecholamine crisis. All patients should have negative biochemical results for catecholamine hypersecretion or undergo alpha blockade before older ionic contrast agents are administered for a CT scan. Current nonionic low-osmolality contrast agents are safe in patients with catecholamine secreting tumors, even without adrenergic blockade.

The majorities of CBT appear to be sporadic or can be hereditary (one-third to one-half of the cases) [11]. 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 [12].

Genetics will not be further discussed in this article, but germline mutation testing is advised for all patients with paragangliomas in the current literature [13].

### Surgical removal

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 [14].

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 favors observation rather than resection for asymptomatic tumors [15].

Therefore, a conservative approach in an asymptomatic CBT is justifiable, although they eventually cause symptoms in nearly 75% of patients due to local growth, which makes removal more challenging.

### Radiation therapy

Another option is radiation therapy. It is commonly performed in patients who are poor candidates for surgical excision or embolization, secondary to their age or co-morbid conditions. Complications of radiotherapy include inflammation of the external auditory canal and middle ear, osteoradionecrosis, cranial nerve neuropathies, carotid stenosis and direct injury to the brain tissue. Radiotherapy also makes subsequent head and neck surgeries highly challenging.

Despite limited experience, radiotherapy for paragangliomas appears to be helpful in cases with unresectable lesions, in high-risk patients, and as an adjunct to surgery for incompletely excised tumors or metastases. Earlier reports even have opted for the use of radiation therapy as the first choice of treatment [16,17].

### Pre-operative techniques

Fine needle aspiration or an incisional biopsy is of little value for paragangliomas. Aspirates can easily be mistaken for many other neoplasms, and the procedure itself brings a high risk for hemorrhage at the operative site [18].

### Imaging

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 [19].

Furthermore pre-operative tumor embolization has been used to shrink the tumor size and disconnect the tumor from its feeding vessels, decreasing complications [18].

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 color Doppler. Displacement of the Internal (ICA) and External Carotid Arteries (ECA) is an important clue and should prompt further image evaluation [20].

Both CTA and MRA appearances are diagnostic. On CT, after intravenous contrast administration, carotid body tumors present as a solid homogenous intensely enhancing mass, located within the carotid bifurcation (Figure 1a) [21]. Seldomly heterogeneous pattern of enhancement can be seen due thrombi or hemorrhage in larger tumors. As a result of their hyper-vascularity, contrast enhancement is rapid and contrary to the gradual enhancement of nerve sheath tumors [21]. Larger CBT will displace the internal and external carotid arteries with splaying of the vessels, the so-called lyre sign (Figure 1b) [22]. While CT has a better special resolution, MRI better depicts the soft tissue components.

On T1-weighted images (T1WI) CBT are hypo- to isointense compared to muscle (Figure 2a) and hyperintense on T2-weighted sequences [22]. A characteristic finding is the so called "salt and pepper" appearance in larger lesions (>2 cm) on T1-weighted images. The "salt" phenomenon, although uncommon, is due to multiple punctuating hemorrhages inside the lesion. These subacute hemorrhages present as high signal intensity spots. The low signal intensity "pepper" component (Figure 2a) is a result of multiple flow voids from intratumoral blood vessels. Similar to CT, there is vivid enhancement after contrast administration (Figure 2b) [22].

### Classification systems

The first classification system based on invasion of the carotid vessels was proposed in 1971 by Shamblin [23].

Because this classification does not predict neurological damage but only prognosticate surgical time and perioperative bleeding, the modified Shamlin's classification proposed by Luna-Ortiz is a more detailed classification system (Figure 5). According this classification, group I tumors are relatively small tumors minimally attached to carotid vessels. The group II tumors are more adherent to the adventitia but still removable without reconstruction of the vessels. The group III tumors encase the carotid vessels and often require arterial resection and grafting. Frequently type III tumors are larger, but this does not mean there is a direct relation between these two specifications. A smaller tumor can be classified as type II or III according to vessel involvement [15,24]. Luna et al. [24] suggest that aCBT of any size, if intimately adherent to the vessels, should be classified as modified Shamblin class IIIb, whereas IIIa represents the original III as described by Shamblin. In many studies a cut-off size of four centimeters to classify between type I and type II or III tumors, because a correlation is known to exist between larger tumors, and neurologic damage [25].

Arya et al. [25] proposed pre-operative criteria to predict the Shamblin classification of CBTs on imaging by measuring the angle of contact from the center of the ICA to tumoral-edge as an assessment of vascular encasement. They defined type I: less than or equal to 180°; type II: greater than 180° and less than 270°; and type III: greater than or equal to 270°. The degree of circumference of contact of the tumor with the external carotid artery or common carotid artery was not incorporated in this classification [25].

Recently in 2017, a new article stated that distance to the base of the skull is more predictive for cranial nerve lesion and the amount of perioperative blood loss than tumor volume, whereas tumor volume is significant correlated with blood loss alone [24,26].

## Conclusion

A carotid body tumor is a rare neuroendocrine tumor located at the carotid bifurcation. They are mostly benign but are often linked with a genetic syndrome. Therefore, genetic testing is advised in every patient presenting with a CBT. Malignancy cannot be confirmed by histopathological investigation, but can only be proven by metastasis into other tissue organs.

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