

Carotid Body Tumor: A Radiological Case Study and Literature Analysis

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Abstract: Carotid bodies are collections of Paraganglia noted at the bifurcation of the common carotid artery on either side of the neck. Neoplasia involving the carotid body even though a rare occurrence is of significant importance due to its high risk of neurovascular complications, and difficulty in surgical resection due to its location. Early tumor detection, meticulous evaluation, and multidisciplinary approach are vital for the successful management of these tumors. Here we report a case of a 32 - year - old male who presented with complaints of painless, pulsatile lateral neck swelling at our institute along with a brief literature review.

Keywords: Carotid body tumor, Paraganglioma, lyre sign, Shamblin classification

1. Introduction

The carotid body is a large collection of Paraganglia noted in the neck, which are derived from neural crest cells [1]. It is usually found posterior to the Common carotid artery in its adventitia at the bifurcation of external and internal carotid arteries on either side of the neck. The Carotid body is mainly an arterial chemoreceptor that is sensitive to hypoxia [3] and, to a lesser extent to hypercapnia and acidosis and brings about changes in heart rate, blood pressure, and respiration by increasing sympathetic outflow.

Carotid body tumors (CBTs) account for less than 0.5% of head and neck tumors which are usually non - familial. They are slow - growing, hypervascular tumors that typically present unilaterally in a majority of patients. CBTs are usually benign, with less than 10% showing malignant transformation [5].

The etiology of carotid body tumors is not well understood, but chronic hypoxia [3] is said to play a key role, this explains the higher incidence of CBTs in people living at higher altitudes [4] and in patients suffering from COPD. Bilateral presentation is seen more frequently in familial than in non - familial CBTs [6]. Familial or inherited CBTs usually show a genetic mutation in SDH genes, mitochondria complex genes, etc. [8].

Due to the location of carotid body tumors, any attempt at biopsy or exploratory surgery usually leads to disastrous outcomes, and fine needle aspiration cytology often leads to inconclusive findings due to difficulty in obtaining an adequate sample. Therefore, imaging studies such as contrast - enhanced CT, MRI, and digital subtraction angiography (DSA) play a vital role in not only diagnosing the lesion but also in delineating anatomical relations of the lesion, which is invaluable for better surgical outcomes.

2. Case Report

We present a case of a 32 - year - old male referred to our department with complaints of painless neck swelling for 1 year. No other neurological complaints were noted, and there was no significant family history or personal history.

On examination, a firm, non - tender mass was observed at the angle of mandible, anterior to the sternocleidomastoid muscle on right side of the neck. The lesion exhibited horizontal mobility but could not be moved up and down (i. e., Fontaine's sign positive). No signs of inflammation, such as pain, redness, or tenderness, were seen at or near the lesion. Auscultation revealed a faint bruit, suggesting vascular origin.

The patient underwent diagnostic imaging tests, such as color Doppler Ultrasonography, contrast - enhanced CT, and MRI to further confirm and validate the diagnosis.

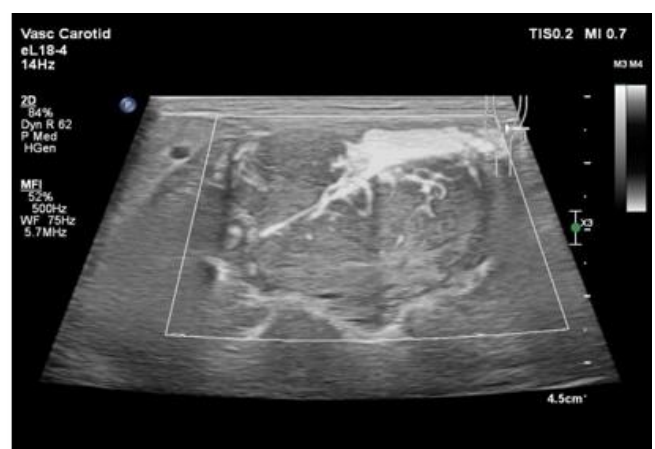


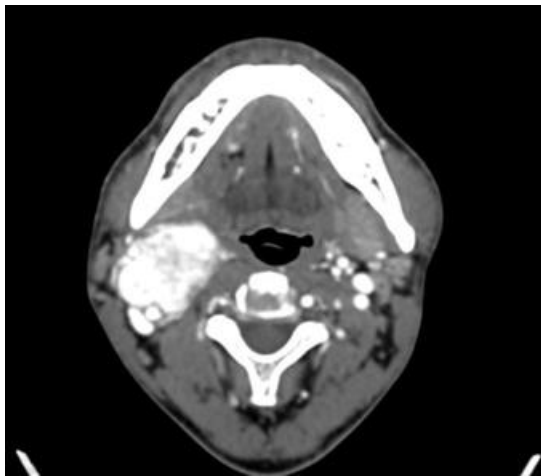
Figure A: USG showing well - defined hypoechoic lesion



Figure B: Doppler Usg showing splaying of internal and external carotid artery

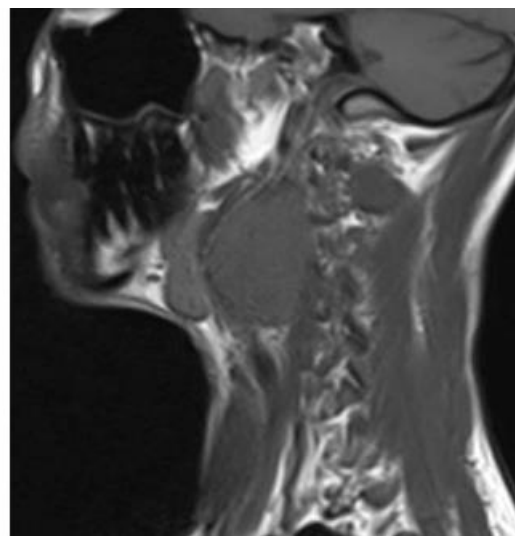
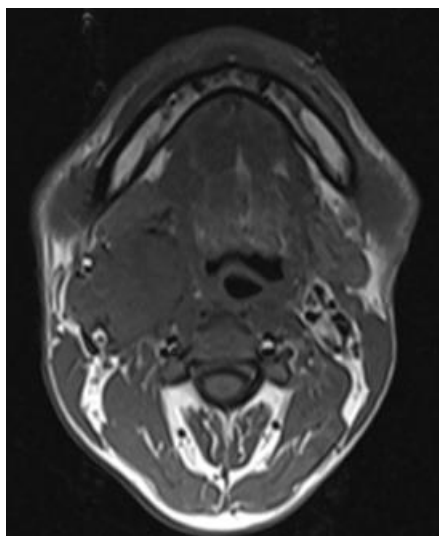
On Doppler ultrasonography, well defined predominately hypo - echoic (fig A) lesion of size measuring approx 37mm in the long axis and 28mm in the short axis, with internal vascularity, was seen at the bifurcation of common carotid artery on the right side causing significant splaying of the internal and external carotid artery (fig B) (suggestive of Lyre sign) in close relation to right submandibular gland. Surrounding arteries showed normal caliber and color flow on CFM.

On CT scan, A well-defined predominantly iso - dense lesion was observed which exhibited bright and rapid enhancement in arterial phase (fig C - D).



Figures C and D: Axial and sagittal section of Arterial phase showing rapid contrast uptake and splaying of internal and external carotid arteries.

The lesion was seen at the bifurcation of the common carotid artery causing splaying (D) of internal and external artery. Both ICA and ECA were normal in caliber and contrast opacification with no intraluminal filling defect.



Figures E and F: Axial and Sagittal T - 1 weighted images showing hypointense lesion at carotid bifurcation.

MRI studies showed a distinct lesion measuring approx (3.6 x 2.9 x 4.1) cm in right carotid space at the carotid bifurcation. The lesion appears to be hypo - intense (fig E, F) in

comparison to muscle on T1 but shows a heterogeneously hyper - intense signal on T2 - weighted (fig G, H).

DW images show evidence of restricted diffusion. The angle between the lesion and the internal carotid artery was found to be between 180 and 270 degrees, with mild encasement.

Thus, categorizing it as group II in the Shamblin group system.

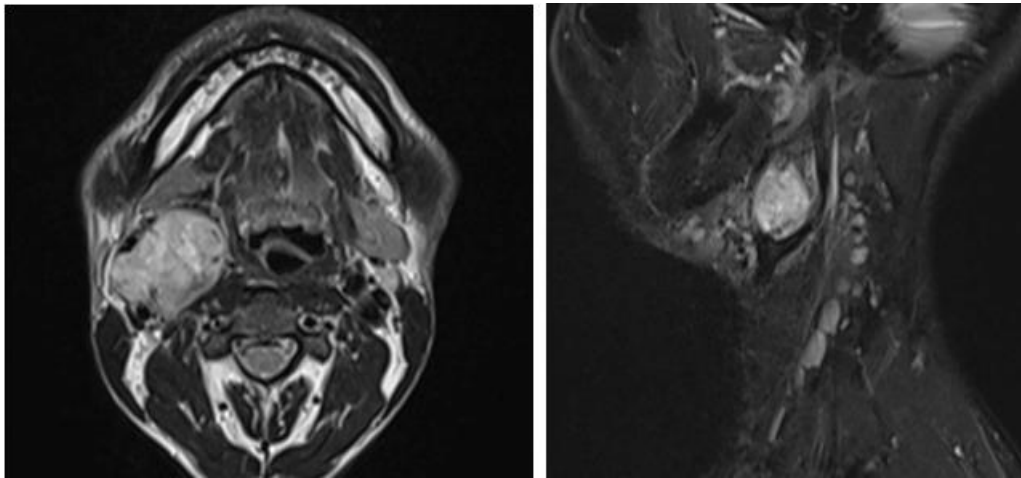


Figure G and H: Axial and Sagittal T - 2 weighted images showing heterogeneously hyper - intense signal intensity lesion.

Contrast study with gadolinium shows intense heterogeneous enhancement immediately post - contrast with evidence of multiple flow voids within giving a distinct salt and pepper appearance.

Splaying of the external and internal carotid artery was seen on enhanced MR angiography maximum - intensity projection image (MIP).

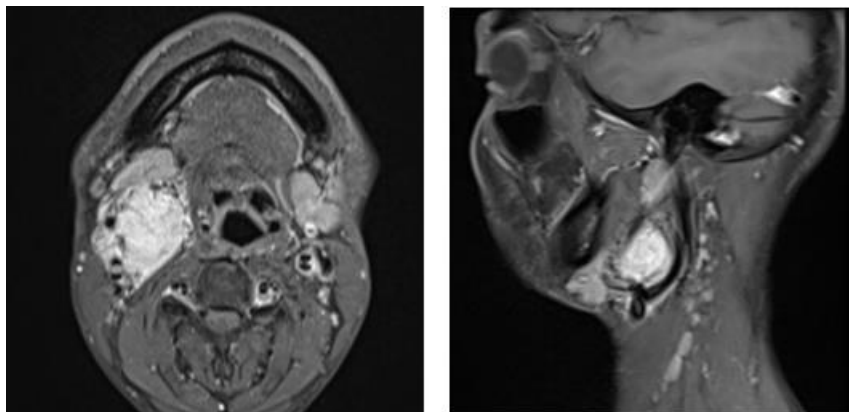


Figure I and J: Post - contrast T - 1 Weighted axial and Sagittal sections showing salt and pepper appearance.

3. Discussion

Carotid body tumors are slow - growing, hypervascular, usually solid benign lesions noted in relation to the angle of mandible, typically diagnosed between the third to sixth decades of life. They are usually asymptomatic for years before medical attention is sought.

Despite a majority of CBTs being benign in nature, they sometimes exhibit aggressive local spread, causing pressure symptoms and local infiltration into surrounding nerves, leading to neurological symptoms in later stages.

CBTs initially present with painless neck swelling, later on, symptoms such as dysphagia, hoarseness of voice, and rarely repeated episodes of syncope is seen due to compression of the internal carotid artery.

Local spread into surrounding nerves causes neurological

symptoms such as hoarseness of voice, dysphagia, Horner's syndrome, etc due to the involvement of the vagus nerve, glossopharyngeal nerve, and cervical sympathetic chain. Usually, 15 to 20% of CBTs show cranial nerve involvement with the vagus nerve being most commonly affected [7].

CBTs are mostly non - functional in nature, but sometimes catecholamine - secreting tumors are occasionally encountered manifesting as excessive sweating, headache, tachycardia, or episodes of uncontrolled hypertension. The Manifestation of these symptoms warrants suspicion of a functional tumor, requiring extensive preoperative evaluation and management before any intervention. This includes urinary or plasma fractionated metanephrine assay, which are metabolites of catecholamines.

B mode imaging with color Doppler remains the first - line investigation for screening and early detection of CBTs. Contrast - enhanced CT and MRI helps to determine the

exact location of the tumor, its extent and its relationship with neighboring structures.

Digital subtraction angiography is considered to be gold standard [10] [11] in the diagnosis of CBTs and plan treatment. In addition to providing details such as size, location, tumor extension, and its relations with neighboring structures, it also helps to locate the dominant vessel thereby allowing for preoperative embolization.

FNAC is rarely done due to dreadful complications such as hemorrhage, and injury to surrounding vessels and nerves, and 18F - DOPA positron emission tomography (PET) is indicated if multiple Paragangliomas are suspected in genetically predisposed individuals [9] [13].

4. Management

Treatment modalities for CBTs are controversial due to the slow - growing nature of the tumor. Surgical excision forms the primary treatment of choice which mainly depends on the size of the tumor while Shamblin's classification is used to assess difficulty in resection.

Shamblin class I lesions consist of tumors easily isolated and dissected from the carotid vessels while Class II lesions are more adherent to the adventitial layer and partially encircle the vessels at the bifurcation; and class III lesions are more densely adherent to the carotid vessels and almost completely encircle the vessels where surgical risks outweigh the benefits [14].

Postoperative complications include nerve injuries, cerebrovascular accidents, and wound complications. Pre - operative embolization may be considered to simplify operation and reduce blood loss but is controversial due to the risk of stroke and neurological complications. Recent studies show that large CBTs can be resected safely with or without preoperative embolization [12].

Principal indications for the use of radiotherapy as primary treatment for carotid Paragangliomas include extensive tumor spread where resection would result in significant morbidity as well as patient - related factors such as age and medical condition.

Surgery is considered by many as the standard therapy as it provides an immediate and complete removal of the tumor. However, the risk of cranial nerve injuries and stroke associated with surgery is significant. At the same time slow growing nature and the fact that most of them are benign and rarely cause death by themselves, encourages some to adopt to a more conservative approach.

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