A case report of bilateral carotid body tumor.

Dr. Zalak Panchal[1*], Dr. Vipula Goswami[2]

[1] Resident Doctor, [2] Associate Professor , Radiology Department, B.J. Medical college, Civil Hospital, Ahmedabad, Gujarat, India.

Abstract:

A carotid body tumor is a rare presentation of an extra-adrenal paraganglioma which typically present as a slow growing, painless neck mass found along the anterior border of the sternocleidomastoid muscle. These tumors are generally benign but possess aggressive local growth potential. Therefore, definitive treatment requires surgical resection. Carotid body paragangliomas are diagnosed by Doppler ultrasound, carotid artery angiography, cranial computed tomography and magnetic resonance imaging. Here we describe a case of bilateral carotid body tumors in a 46-year-old female presented with a bilateral neck swelling.

Keywords: Carotid body tumor, Neoplasm metastasis, Paragangliomas, Radiology.

Introduction:

Carotid body tumor is an extra-adrenal paraganglioma that may also be termed chemodectoma. It originates from the neural crest tissue in the carotid bifurcation. Usually, it is a solitary occurrence. The majority of cases are considered to be benign, with only 10–20% demonstrating malignant transformation.[1] They are generally presented as unilateral neoplasms that are located at the carotid bifurcation, without distant metastasis.

The present study reports the case of a patient with bilateral carotid body tumours in 46 year old female.

Case Report:

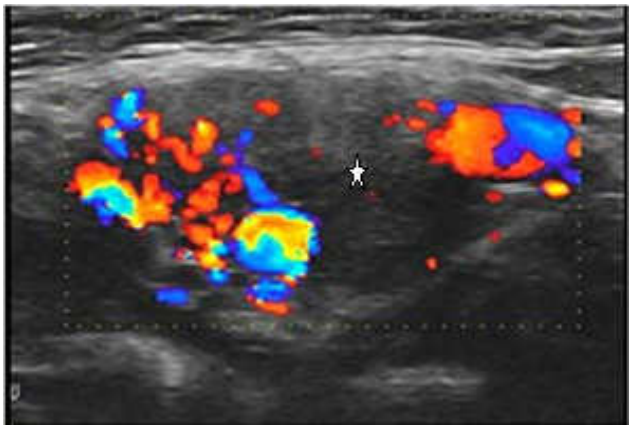
A 46-year-old female patient presented with a bilateral neck swelling which had progressively worsened over few months. There was no history of dysphagia, hoarseness, headache, hypertension crisis, tachycardia or similar history among relatives. On physical examination, pulsating firm painless masses were found on the both sides of her neck, near the angle of the mandible in bilateral juglocarotid region. Pulsations were felt on deep palpation and a faint bruit was heard on auscultation.

Imaging Findings:

An ultrasound and Doppler examination revealed an ill-defined heterogenous echotextured mass lesions with increase vascularity at the bifurcation of the bilateral common carotid arteries.[Image-1] The masses were approximately ~2.3×2.1 cm and ~4.0×2.1 cm in size (right and left side, respectively). No enlarged lymph nodes were identified in the cervical region.

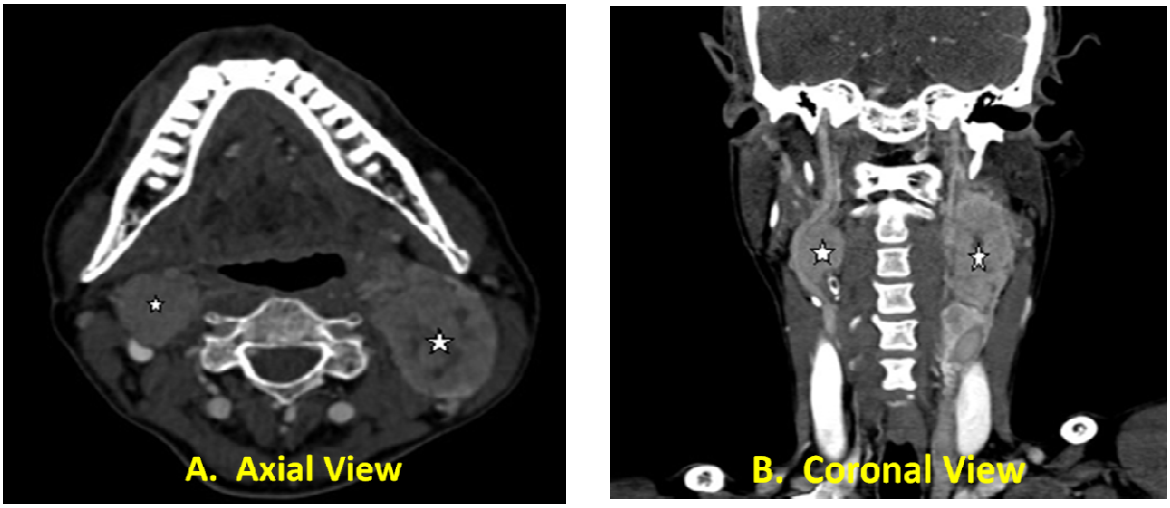
[*] **Corresponding Author:**
Dr. Zalak Panchal
E-mail: zalakpanchal.92@gmail.com

Image 1: Doppler ultrasonography shows an ill-defined hypoechoic mass at the bifurcation of the common carotid artery causing splaying of External & Internal carotid artery.



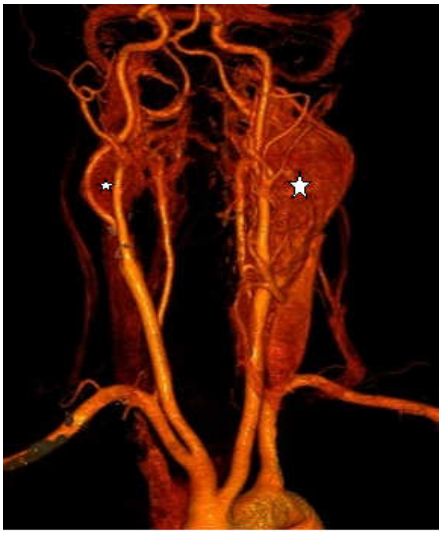
A computed tomography (CT) scan of the neck with contrast enhancement and CT angiography was performed. Contrast-enhanced Computed tomography demonstrated a well-circumscribed lobulated heterogeneously intensely enhancing soft tissue density mass of size 2.5 cm × 2.2 cm × 3.4 cm located at the bifurcation of the right carotid artery causing splaying of external & internal carotid artery, with circumferential encasement of the right proximal ICA (270 degree) and circumferential encasement of right proximal ECA (180 degree) without luminal invasion.[Image-2] No infiltration into adjacent structures is seen.

Image 2 Contrast enhanced CT scan images show intensely enhancing well defined soft tissue density lesions in bilateral carotid spaces. (A. Axial view, B. Coronal view)



Carotid angiography showed a vascular blush at the widened carotid bifurcation. The lesion shows intense heterogenous enhancement on arterial phase and rapid washout on delayed phase. There is also evidence of approximately 4.1 cm × 2.2 cm × 7.7 cm sized similar characteristic mass lesion at the bifurcation of left common carotid artery. [Image-3] On left side, the lesion causes splaying of external & internal carotid artery with circumferential encasement of left carotid bulb, bifurcation, proximal ECA (270 degree) and ICA (360 degree) without luminal invasion. The lesion causes displacement of left internal jugular vein posterio-laterally with severe luminal compression and non-visualisation of approximately 3.5 cm long segment. This finding was considered to be consistent with a diagnosis of glomus caroticum (carotid body tumor).

Image 3: 3D Angiography image of Head and Neck vessels shows masses at the bifurcation of bilateral common carotid arteries causing splaying of external and internal carotid arteries.



Surgical treatment was preferred. On the left side, an oblique incision was made, following the anterior border of the sternocleidomastoid muscle, and extended proximally toward the mastoid process. Surrounding neurovascular structures separated and tumor removed. There was no evidence of neurological deficit after the surgery. Few months later the mass on the right side was resected. Diagnosis was confirmed by the histological analysis of fragments of both masses.

Discussion:

Carotid body tumor is a type of extra-adrenal paraganglioma. It is highly vascular tumor that arises from the paraganglion cells of the carotid body. Usual location of carotid body tumor is at the carotid bifurcation with resultant splaying of the ICA and ECA. Age for presentation of carotid body tumor is 4th and 5th decades of life with notable female predilection.[1,3] They are the most common type of paraganglioma of the head and neck (accounting for approximately 60-70%). They tends to be bilateral in approximately 10% of cases.[1,3] Malignant transformation is encountered in 2-36% of cases with metastases most commonly to bone, lung and liver and regional lymph nodes.[3]

A small number of cases are familial (7-10%), in which they are frequently multicentric (35-50%) .[1,3] When familial, they are usually autosomal dominant in inheritance, and associated with

- Multiple endocrine neoplasia: MEN IIa and MEN IIb
- Phakomatoses : tuberous sclerosis complex (TS), neurofibromatosis type 1 (NF1), Von Hippel-Lindau disease (VHL)
- Carney triad

It usually presents as a slow growing rounded neck mass located anterior to the sternocleidomastoid muscle near the angle of the mandible. The tumor can be moved side to side but not up or down, due to its location within the carotid sheath.[1] Cranial nerves travelling in the carotid sheath (glossopharyngeal, vagus, accessory and hypoglossal nerves) may be involved with resultant symptoms due to their dysfunction.[2] Differential diagnosis

includes branchial cysts, carotid aneurysms, metastatic carcinomas, intravagal tumors, lymphomas and ectopic thyroid.

Carotid arterial angiography is the most valuable diagnostic technique.[5,6] It can detect multiple lesions, tumor size and the major vascular tributaries of the tumor. Arteriographic imaging should be carried out bilaterally because of the possibility of bilateral tumors.[5,7] On CT (Computed tomography) images, tumor is seen as a well-defined soft tissue mass with a homogeneous enhancement that is located within the carotid sheath. Larger tumors are frequently inhomogeneous due to necrotic and hemorrhagic regions.[3] The ECA is usually displaced anteromedially and the ICA is typically displaced posterolaterally, which strongly indicates a diagnosis of CBT.[3]

On MRI (Magnetic Resonance Imaging), the tumor appear iso to hypointense on T1 weighted images, hyperintense on T2 weighted images and show intense enhancement following gadolinium contrast injection. When larger the lesion shows salt and pepper appearance, representing a combination of punctate regions of hemorrhage or slow flow (salt) and flow voids (pepper).[3]

On Angiography, splaying of the carotid vessels (lyre sign) is seen with an intense blush in tumor and early venous drainage due to arteriovenous shunting.[3] The ascending pharyngeal artery is usually the main contributing supply.

Surgical excision is the treatment of choice. The larger the tumor the higher the risk of operative complications.[2] In patients for whom the risk of complications precludes surgery, radiotherapy may be considered.[1,2]

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