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Carotid body tumor: Case series and literature review

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Abstract
The carotid body is the largest collection of paraganglion chemoreceptor cells situated at medial aspect of the carotid body bifurcation bilaterally. We report three case of carotid body tumor in a the diagnosis is suspected on the basis of history, clinical and radiological examination findings. Out of three cases one pt agreed for indo vascular embolisation. We also present brief literature about carotid body tumors in terms of its clinical and imaging presentation, evaluation, and management.

Keywords: Carotid body tumor, splaying of common carotid artery

Introduction

Carotid body tumors is also known as paragangliomas or chemodectomas, are rare neuroendocrine neoplasms which arise near the carotid bifurcation. Carotid body is supplied by the feeding vessels primarily from the ascending pharyngeal branch of the external carotid artery, and innervated through the glossopharyngeal and vagus nerves. Incidence of CBTs is 1–2 per 100,000 [1], male-to-female ratio of 1:1.9 accounts for 0.6% of the head and neck tumors in humans & is the commonest location.

Materials and Methods

Patients with palpable neck mass who were referred to Department of Radio-Diagnosis, for further evaluation.

Case Series

Case 1: A male patient aged 45 years presented with complains of palpable mass in the right side of the neck, which was insidious in onset and gradually progressive in size and was associated with headache**.**

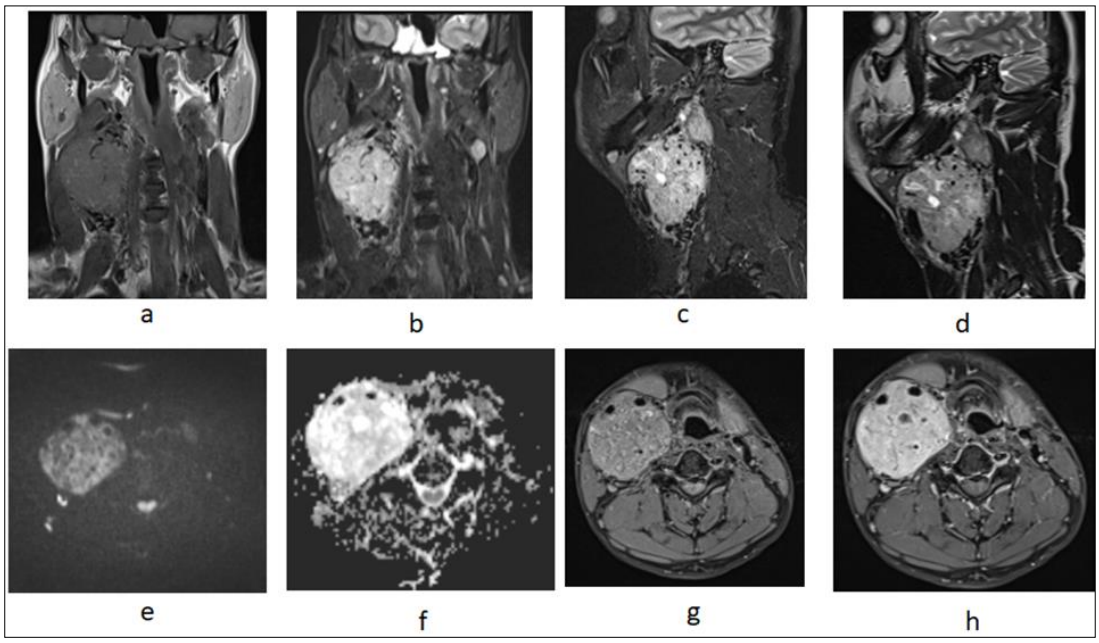

Fig 1

A large bi-lobed well defined altered signal intensity lesion in the supra and infrahyoid regions of the right side of neck appearing heterogeneously hypointense on coronal T1 weighted image (fig- 1a), heterogeneously hyperintense on coronal T2 weighted image (fig – 1b), sagittal T2 weighted image (fig- 1c) and on saggital TIRM sequence (fig- 1d) with few areas of flow voids within. The lesion shows evidence of diffusion restriction on TRACE (fig- 1e) and corresponding ADC maps (fig- 1f). (Fig- 1g) shows pre-contrast T1 fat saturation of the lesion in axial section and

(fig- 1h) shows strong homogeneous enhancement with few non-enhancing areas within which are suggestive of cystic or necrotic areas within.

Case 2: A 52 year old female patient presented with a palpable mass in the right side of the nech which was insidious in onset and progressive in nature.

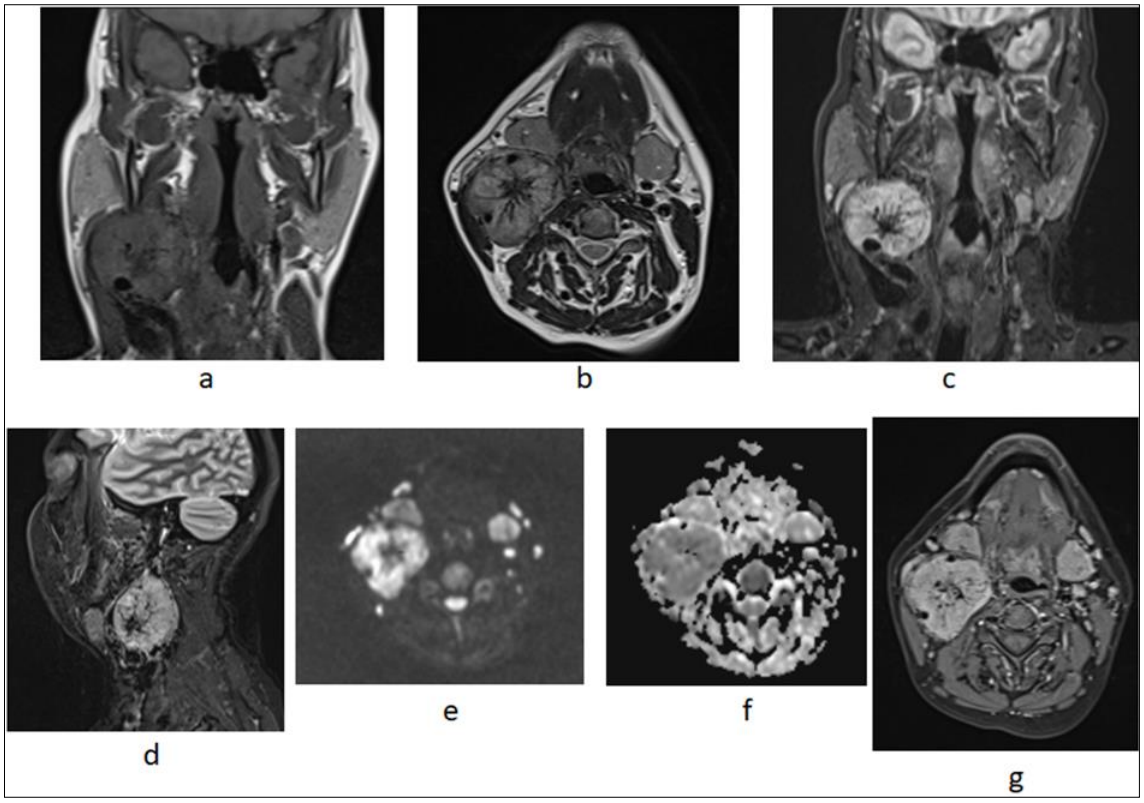

Fig 2

A well defined altered signal intensity lesion in the right carotid space near the bifurcation of common carotid artery which appears isointense on coronal T1 weighted image (fig- 2a) and on axial T1 weighted image (fig- 2b), hyperintense on coronal T2 weeighted image (fig- 2c) and on sagittal STIR image (fig- 2d) with multiple flow voids within. The lesion shows evidence of diffusion restriction on TRACE (fig- 2e) and corresponding ADC maps (fig- 2f). The lesion shows homogeneous post contrast enhancement on axial T1 weighted image (fig- 2g).

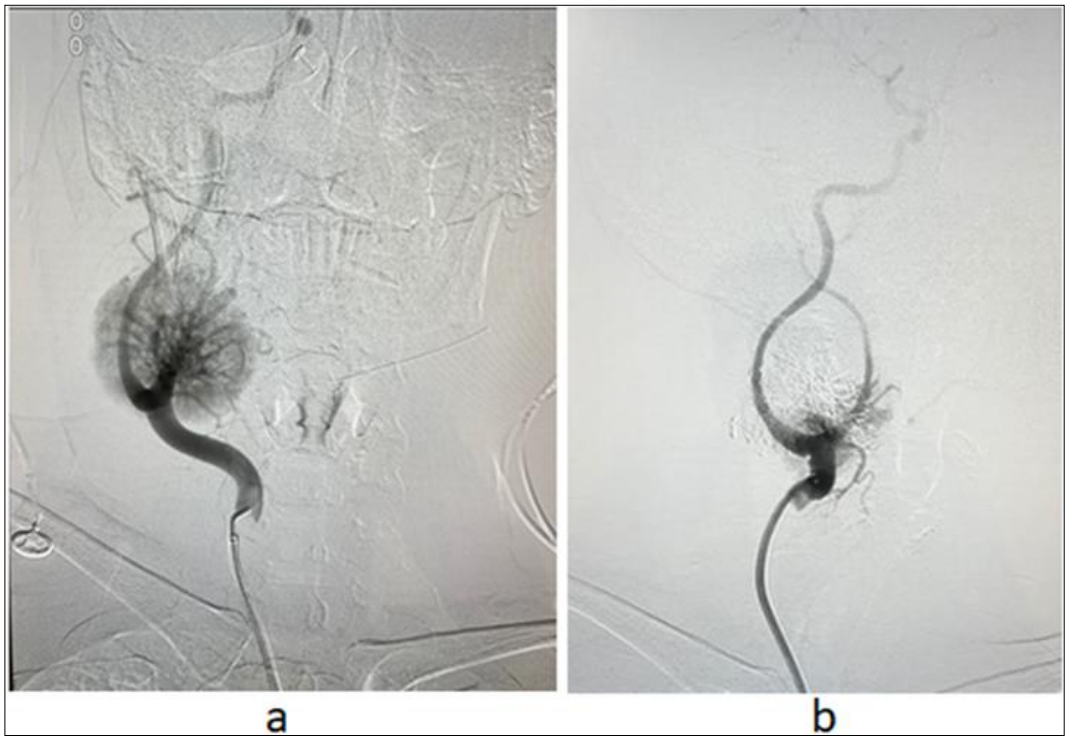

Fig 3

Digital subtraction angiography in the same patient showing intense blush in the tumor (fig- 3a) and splaying of carotid vessels showing Lyre's sign (fig – 3b).

Case 3: A male patient aged 49 years presented with complains of swelling on the left side of neck which was associated with dysphagia.

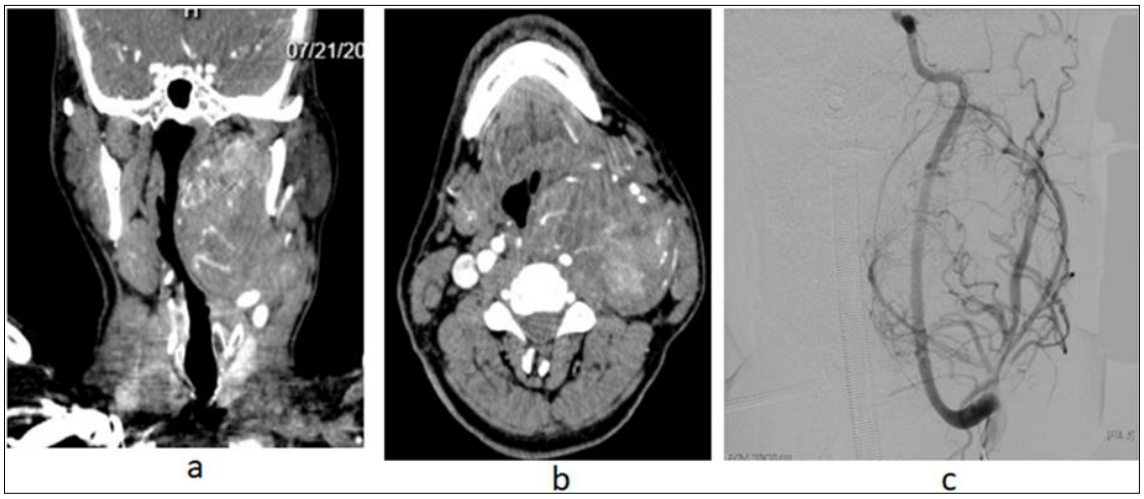

Fig 4

CECT neck coronal section (fig- 4a) and axial section (fig- 4b) showing a well defined soft tissue lesion in the left carotid triangle showing inhomogeneous contrast enhancement, the lesion is aloes seen to push the trachea to the contralateral side. DSA (fig- 4c) shows splaying of the carotid vessels and multiple feeding arteries to the tumor.

Discussion

The carotid body is first described by von Haller [2]. Carotid body 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 [3]. Lesions of the carotid space may arise due to asymmetry of normal vasculature, inflammatory or infectious processes. CBTs usually manifest as asymptomatic anterior neck mass. In large tumors, they can be associated with the myriad of presenting symptoms of a space-occupying lesion in this location, such as fullness, pain, dysphagia, odynophagia, hoarseness of voice, and stridor. Functional CBT may produce neuroendocrine secretions causing catecholamine-related symptoms such as palpitations, headaches, hypertension, tachycardia, or flushing.

Doppler ultrasound, CT, MRI, and angiography play important role in the clinical diagnosis of CBT. Angiography was the "gold standard" diagnostic procedure for CBTs to confirm the diagnosis and provide accurate delineation of the vascular supply. CT angiogram is able to define the size of the tumor and its relation to bony landmarks, which can modify the surgical approach. It can also easily identify contralateral tumors and other head/neck paragangliomas. DWI and dynamic contrast-enhanced MRI, lower ADC value and washout time-intensity curves will support the diagnosis. USG with Doppler shows a soft tissue mass at the bifurcation of common carotid artery causing splaying of internal and external carotid arteries.

Metaiodobenzylguanidine (MIBG) and octreotide scan scintigraphy can be useful for assessing multiple lesions on scintigraphy though non specific [3]. Peptide receptor radionuclide therapy (PRRT) can be a therapeutic option in selected cases of head and neck paragangliomas, including non-resectable tumors, occult, recurrent, malignant, or metastatic lesions.

Hypervascularization and proximity to various vascular and nervous structures of these tumors, biopsy as a diagnostic method is contraindicated since it presents a risk of massive hemorrhage and dissemination and can lead to pseudoaneurysm formation and carotid thrombosis [4].

Embolization of the tumor's main artery usually the pharyngeal ascending artery prior to surgery may help reduce bleeding and other complications associated with the removal of large tumors, facilitating resection.

Differential Diagnosis

Table 1

Diagnosis	Ultrasound findings	CT findings	MRI findings	Angiography/DSA findings
Carotid body tumor	Well-defined hypoechoic mass that splays the carotid bifurcation; hypervascular with low-resistance flow pattern	A mass with soft tissue density; heterogeneously enhancing at the carotid bifurcation in the early arterial phase; strong and rapid enhancement	Nestled between the external and internal carotid arteries; signal intensity is nearly similar to muscle on T1-weighted, and hyperintense signal on T2-weighted; salt and pepper sign appearance	Splaying of the carotid vessels; the ascending pharyngeal artery is the main contributing supply; conventional angiography can demonstrate vascular anatomy
Carotid artery aneurysm	Yin-Yang" sign; pseudoaneurysm or dissection	Change in the diameter of the blood vessel	An oval, irregular, or slit-like cross section of the	Delineate extension, detect stenosis

	can occur		vessel lumen	
Carotid artery pseudoaneurysm	A hypoechoic cystic structure adjacent to the true vessel; color Doppler imaging present “to and fro” flow and bidirectional Doppler waveform in pseudoaneurysm	CT can demonstrate the vessel wall irregularity and irregular outpouching; compatible with rupture and active bleeding	MRI with T1-weighted fat-suppressed sequences allows for evaluation of intraluminal thrombus and pseudoaneurysm sac size	DSA remains the gold standard for evaluation of pseudoaneurysm
Neck hematoma/ thrombus	Dilated and incompressible arterial, intraluminal clot, and no response to arterial maneuver	On non-enhanced CT, hematoma appears as a high-density mass. On enhanced CT, it does not enhance with contrast agent	Hemorrhage on MRI has highly variable imaging characteristics that depend on the age of the blood. On T2-weighted, acute thrombus of the arterial will have a high intensity, whereas subacute arterial thrombus will have a low signal	May locate the active arterial bleed/source of hemorrhage
Glomus tumor	May be seen as a solid heterogeneously hypoechoic lesion comprising of small vascular structures	A mass with soft tissue density; heterogeneously enhancing	Usually low signal on T1-weighted; high signal with multiple flow voids on T2-weighted; intense contrast enhancement on T1-post contrast	Same carotid body tumor but tends to displace the carotid artery anteriorly
Schwannoma	Permitted direct visualization of the vagus nerve, so its position relative to the schwannoma could be examined; can result in an increase of the distance between the artery and vein (separation)	A mass with soft tissue density	Well-encapsulated tumors appear as a round or ovoid mass that is isointense to muscle on T1-weighted, hyperintense on T2-weighted; that may or may not enhance on post contrast	Tends to displace both vessels together rather than splaying them

Conclusion

Color Doppler and MRI imaging modalities have played a crucial role in the accurate diagnosis and preoperative assessment of the tumor. Our case series emphasizes the importance of a multidisciplinary approach, adherence to established guidelines and close collaboration among healthcare professionals to optimize patient outcomes in the management of carotid body tumors.

Conflict of Interest

The authors declare no conflicts.

Patient consent

The authors confirm that patient consent was obtained for the publication of this Case Report.

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