Case Report

Multimodality Imaging of Carotid Body Paraganglioma: Radiological Diagnosis and Characterization – A Case Report

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ABSTRACT

Background: Carotid body tumors (CBTs) are rare, highly vascular neuroendocrine tumors originating from paraganglionic cells in the carotid bifurcation.

Case Presentation: We report the case of a 21-years-young female who presented with a painless, gradually enlarging neck mass on the left side of her neck. Imaging studies, including ultrasound and contrast-enhanced computed tomography, confirmed the presence of a well-defined, hypoechoic mass at the left carotid bifurcation, displacing adjacent vessels. A multidisciplinary team planned surgical resection, guided by imaging, resulting in the successful removal of the tumor. Histopathological examination confirmed the diagnosis of a carotid body tumor.

Conclusion: Although rare, carotid body tumors should be considered in the differential diagnosis of neck masses in young patients. Early diagnosis with appropriate imaging and meticulous surgical planning can make promising outcomes in carotid body tumors.

Key words: *Carotid body tumors, Neuroendocrine tumors, Paraganglionic cells*

INTRODUCTION

Carotid body tumors (CBTs), sometimes called carotid body paragangliomas, are uncommon neuroendocrine tumors that originate from the paraganglionic cells of the carotid body, a chemoreceptor situated at the common carotid artery's bifurcation. These tumors, which make up around 0.5% of all head and neck neoplasms, have a mostly benign clinical course and sluggish development are typically benign[1].

Despite their rarity and slow-growing nature, CBTs present significant diagnostic and therapeutic challenges due to their hypervascularity and intimate relationship with critical neurovascular structures, including the internal and external carotid arteries and the lower cranial nerves (IX–XII). The most typical clinical presentation of CBTs is a painless, progressively growing lateral neck lump. While many patients remain asymptomatic, larger tumors may cause symptoms associated with the local mass impact, such as dysphagia, hoarseness, and respiratory compromise[2].

Imaging plays a pivotal role in making the diagnosis, assessing the size of the tumor, and directing surgical planning. Modalities such contrast-enhanced computed tomography (CECT), magnetic resonance imaging, digital subtraction angiography, and color Doppler ultrasonography are especially useful[1,2] . The characteristic splaying of the internal and external carotid arteries, the "lyre sign"—is a pathognomonic finding on cross-sectional imaging.

Here, we report a case of a carotid body tumor in a young female patient, highlighting the key radiological features, diagnostic workup, and successful

surgical management. This case underscores the importance of including CBT in the differential diagnosis of neck masses in young adults to facilitate timely intervention and minimize morbidity.

CASE REPORT

A 21-year-old female presented with a painless neck mass that had gradually increased in size over the past 8 months. She denied associated symptoms, such as difficulty in breathing, swallowing, changes in voice or cranial nerve deficits. There was no history of trauma, weight loss, or systemic illness.
On physical examination, a non-tender, firm, well-circumscribed mass was palpable in the left carotid triangle. The mass was mobile in the horizontal plane but demonstrated limited vertical mobility. No cervical lymphadenopathy was detected. No other significant physical findings were observed, and the patient's vital signs were within normal limits.

Neck Ultrasound revealing a well-defined, hypoechoic mass located at the left carotid bifurcation. Doppler examination demonstrated a solid vascular mass that displaced the internal carotid arteries (ICA) and external carotid arteries (ECA) demonstrating a characteristic splaying sign of ICA & ECA the classic "lyre sign" (Figure 1a, 1b, 1c). No suspicious lymphadenopathy or other abnormal findings were detected.

Subsequent Contrast-enhanced computed tomography (CT) scan of the neck confirmed the presence of a well-defined, avidly enhancing soft tissue mass measuring 4.9 cm (CC) x 3.8 cm (AP) x 2.9 cm (T) in the left carotid bifurcation region with splaying and encasement of left ICA and ECA (Figure 2a, 2b). The lesion demonstrated the characteristic lyre sign, with splaying and partial encasement of the left internal carotid artery (ICA) and external carotid artery (ECA), while maintaining normal distal flow (Figure 2A, 2B). Based on the degree of vessel encasement, the tumor was classified as Shamblin Class II. The mass extended deep to the left sternocleidomastoid muscle but showed no evidence of intracranial extension. The contralateral carotid vessels and intracranial circulation were unremarkable. These imaging findings were highly suggestive of a carotid body paraganglioma.

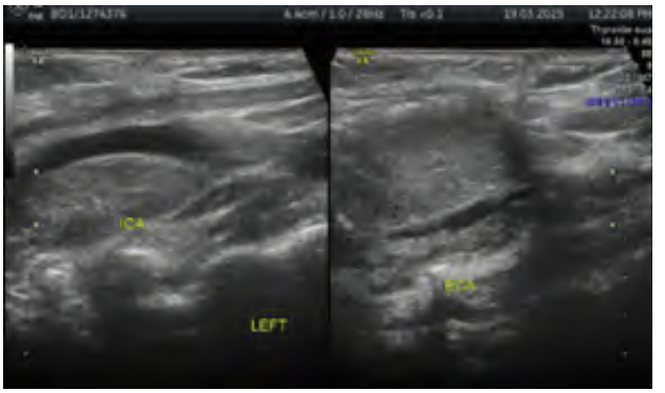

Figure: 1a

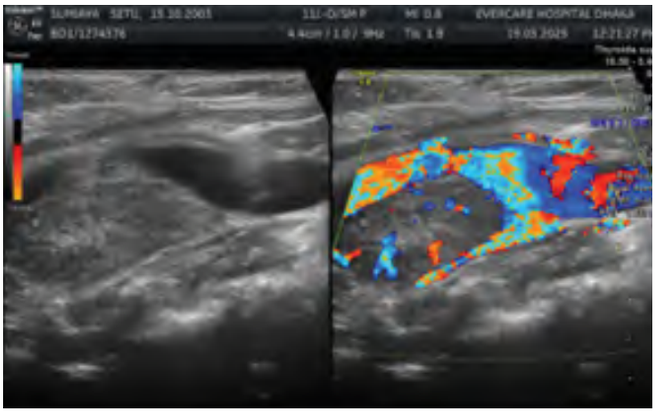

Figure: 1b

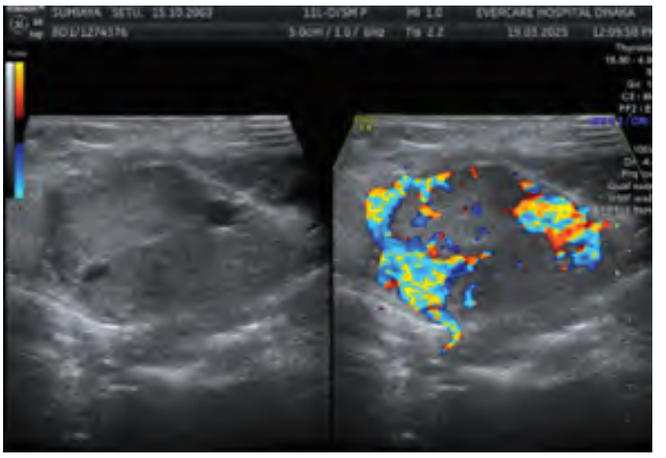

Figure : 1c

Figure: Longitudinal (1a & 1b) and transverse (1c) grayscale and color Doppler views of the neck displaying a solid mass causing displacement of the internal and external carotid arteries demonstrating a characteristic splaying sign of ICA & ECA.

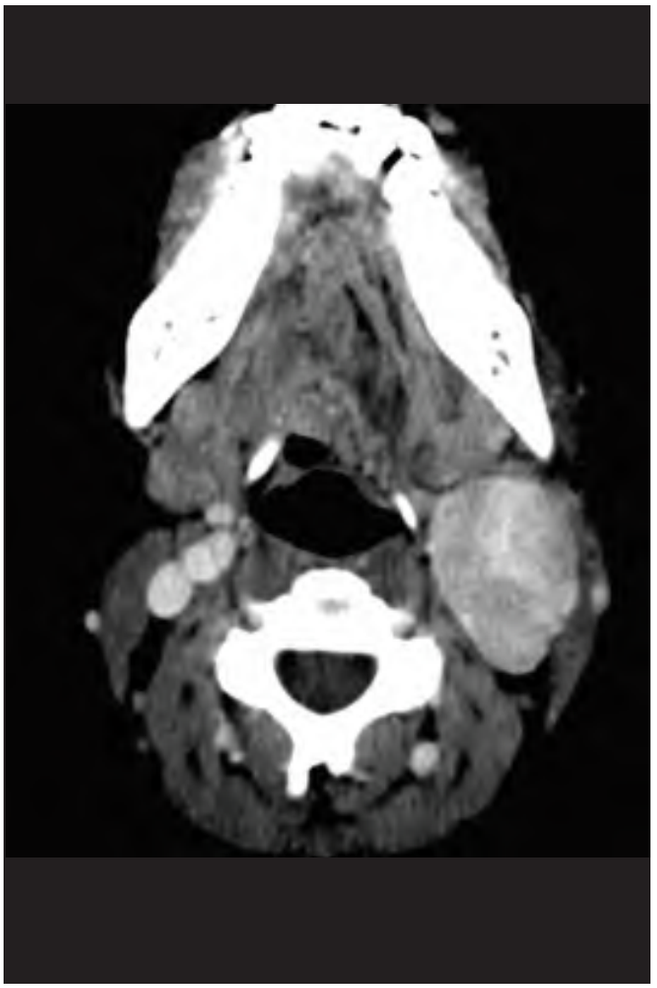

Figure 2a: Axial contrast-enhanced CT image (A) of the neck revealing a prominently enhancing mass located in the notch between the internal and external carotid arteries.

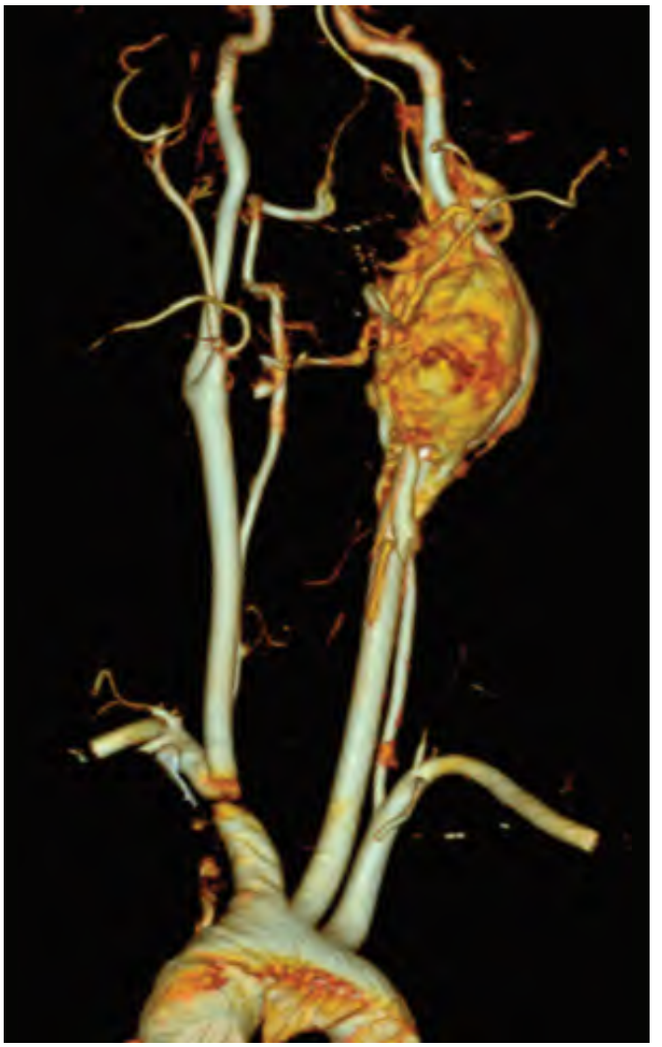

Figure 2b: Three-dimensional volume-rendered CT image (B) illustrating the carotid body tumor.

Following multidisciplinary team (MDT) discussion involving vascular surgeons and head and neck surgeons, surgical resection was planned as the definitive treatment considering the tumor size and vascular nature. Under general anesthesia, excision of the carotid body tumor was performed while preserving the integrity of the carotid artery and other vital structures.

Histopathological examination of the excised tumor shows nests of epithelioid cells arranged in distinctive nests separated by prominent fibrovascular stroma. These findings were consistent with the diagnosis of a carotid body tumor (Figure 3).

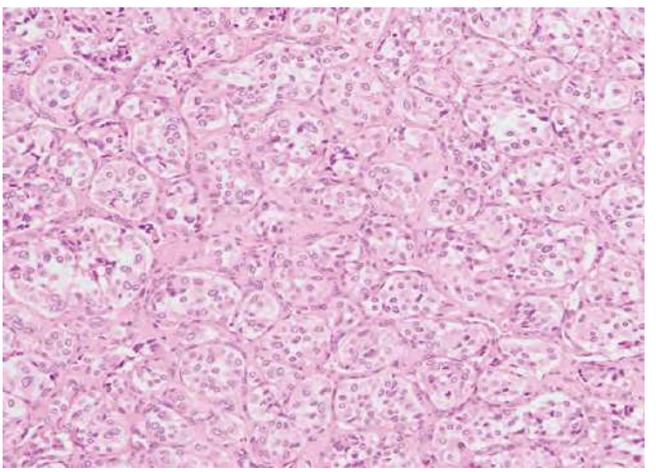

Figure 3: Histopathological picture of tumor composed of cohesive nests of epithelioid cells, embedded within a prominent fibrovascular stroma—findings diagnostic of a carotid body tumor.

DISCUSSION

Carotid body tumors are rare neuroendocrine tumors that originate from the neural crest–derived paraganglionic cells of the carotid body. They represent the most frequent type of head and neck paraganglioma and are typically slow growing but highly vascular lesions. Accurate preoperative diagnosis of carotid body tumors is essential for appropriate management and surgical planning[2]. In our case report, we utilized multiple imaging modalities to confirm the diagnosis and assess tumor characteristics.

Ultrasonography with Doppler is a valuable initial imaging modality for CBTs, offering high specificity and sensitivity. It enables the identification of a well-defined, hypoechoic mass located at the carotid bifurcation, often accompanied by a hyper-vascular appearance and low-resistance arterial flow pattern3. The ability of Doppler imaging to demonstrate hypervascularity and vessel displacement producing pathognomonic "goblet deformity" makes it particularly useful in differentiating CBTs from lymphadenopathy or other solid neck masses. Further evaluation with contrast-enhanced computed tomography (CT) provides detailed anatomical delineation and enables accurate assessment of tumor size, degree of vascular encasement, and its relationship to adjacent neurovascular structures information that is crucial for surgical planning. A characteristic radiologic feature is the splaying of the internal and external carotid arteries, commonly referred to as the "Lyre sign," which strongly supports the diagnosis of a carotid body tumor. In our case, CT imaging demonstrated a well-defined, homogeneously enhancing soft tissue mass at the carotid bifurcation exhibiting this classic vascular splaying pattern[4].

The main radiologic differential diagnoses include vagal paraganglioma, schwannoma, metastatic lymphadenopathy, and carotid artery aneurysm5. Unlike CBTs, vagal paragangliomas typically displace both carotid arteries anteromedially without producing symmetric splaying at the bifurcation.

Although imaging establishes diagnosis, treatment decisions require multidisciplinary input. Surgical resection remains the primary treatment modality for carotid body tumors, aiming for complete excision while preserving the integrity of vital structures, such as the carotid artery and adjacent nerves. However, the use of radiotherapy as a primary treatment approach for carotid body tumors has been debated. Some studies suggest that these tumors are not highly radiosensitive and may exhibit regrowth after suppression. Therefore, surgery is usually preferred for younger, healthier patients, while radiotherapy is considered for elderly patients or individuals who are poor surgical candidates[6,7].

CONCLUSION

This case highlights the pivotal role of multimodality imaging in accurate diagnosis and characterization of carotid body paraganglioma. Doppler ultrasonography and contrast-enhanced CT provide essential information regarding tumor vascularity, extent, and relationship to adjacent neurovascular structures. Early radiological diagnosis, combined with appropriate multidisciplinary management, is crucial for optimizing patient outcomes.

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