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**Received,** 2 August, 2011

**Accepted,** 19 August, 2011

**S**traight forward surgical removal of Carotid body tumor that presents as a clinically benign neck mass, without prior adequate imaging backup can land a novice overzealous surgeon into soup due to unexpected encounter with haemorrhagic neoplasm. Often the surgery may have to be deferred or even abandoned in despair. This case report highlights imaging findings of a unilateral sporadic carotid paraganglioma that presented as a neck mass.

### Case Report

A 44 years otherwise healthy female was referred for ultrasound of a soft tissue mass palpable on the left side of her neck. She told that she could palpate the mass since last four months but had neglected it until now when she could not remove the persistent thought of it being present there from her mind. It was insidious in onset and slowly progressive. There was no history of trauma, fever, weight loss or any other palpable mass in her body. Grey scale ultrasound scan (**Figure 1**) demonstrated a single 2.6 x 3.4 cm sized isoechoic mass just at left carotid bifurcation causing splaying of internal (ICA) and external (ECA) carotid arteries. Color Doppler imaging (**Figure 2**) showed the hyper vascular nature of the mass and peripherally splayed ICA and ECA. Digital Subtraction Angiography (DSA) demonstrated dense blush of the hyper vascular tumor situated at the left carotid arterial bifurcation (**Figure 3**). A provisional imaging based diagnosis of a left carotid body tumor was made and patient was referred for surgery. Under light microscope; the Hematoxylin and Eosin stained section of the excised mass shows numerous polygonal tumor cells arranged to form balls separated by fibrovascular stroma, confirming it to be a carotid paraganglioma (**Figure 4**).

## Unilateral Sporadic Carotid Paraganglioma

Carotid body tumor is one of the last differentials of a unilateral neck mass. But it is the most common type of paraganglioma that manifests as an asymptomatic mass in the antero-lateral aspect of the neck. This case report highlights imaging findings of a unilateral sporadic carotid paraganglioma in a 44 years female who presented with a painless progressive neck mass. It can be clinically confused with cervical lymphadenopathy. Prior imaging is therefore essential before subjecting the patient to surgery and to save the surgeon from the perils of unexpected torrential bleed.

**Key words:** carotid body tumor, paraganglioma.

### Discussion

The carotid body is a round; reddish-brown structure found in the adventitia of the common carotid artery, attached to the posteromedial wall of the vessel at its bifurcation by "Mayer's ligament" through which the feeding vessels run (primarily from the external carotid). Normal carotid body measures 3-5 mm in diameter. It may be enlarged in people living at higher altitudes.<sup>1</sup> Bilateral carotid body tumor resection leads to arterial baroreflex dysfunction, and the normocapnic hypoxic drive is invariably abolished due to peripheral chemo reflex failure.<sup>6</sup>

Although the exact pathogenesis remains unknown, hypoxia and genetic factors are thought to be involved in formation of carotid body tumors.<sup>2</sup> Multiple ganglioneuroma are more common in patients with familial disease.<sup>3</sup> The hereditary form occurs in 7-9% of cases and is seen with equal frequency in both sexes and the inheritance pattern is autosomal dominant. The sporadic form of carotid body paraganglioma is more common than the inherited variety and tends to occur slightly more often in women.<sup>3</sup>

Commonest age group is 50 to 70 years females. It is seen as a non-tender neck mass just anterior to the sternocleidomastoid muscle at the level of the hyoid. It is insidious in onset and slowly progressive. It is mobile medio-laterally but not cranio-caudally. Rarely carotid bruit may be present. Compression on adjacent nerves can cause dysphagia, odynophagia, hoarseness and cranial nerve (IX-XII) deficits. Excess catecholamine production can cause fluctuating hypertension, blushing and palpitations.<sup>5</sup> Due to disturbed function of peripheral chemoreceptors in the presence of carotid body tumors there is increased daytime somnolence.<sup>5</sup>

Ultrasound shows the site and size of mass. Color

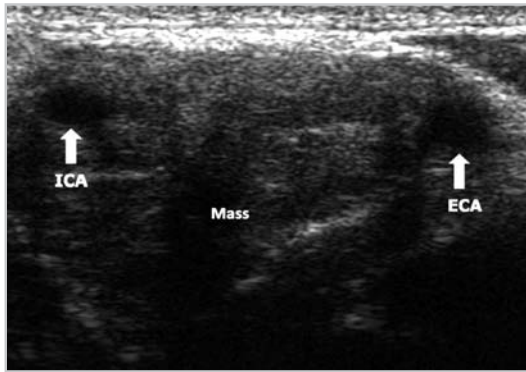


Figure 1: Grey scale ultrasound image demonstrating an isoechoic mass splaying the left internal and external carotid arteries.

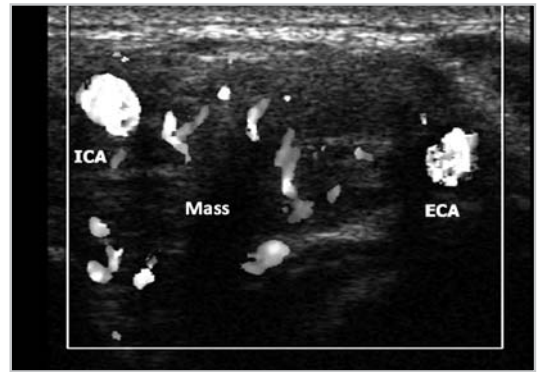


Figure 2: Color Doppler ultrasound image demonstrating vascular mass splaying the left internal and external carotid arteries.

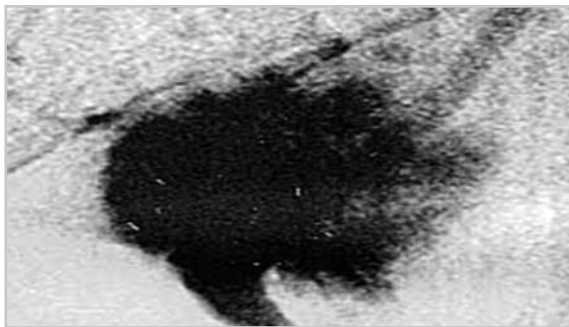


Figure 3: DSA image demonstrating hyper vascular tumor situated at the left carotid arterial bifurcation.

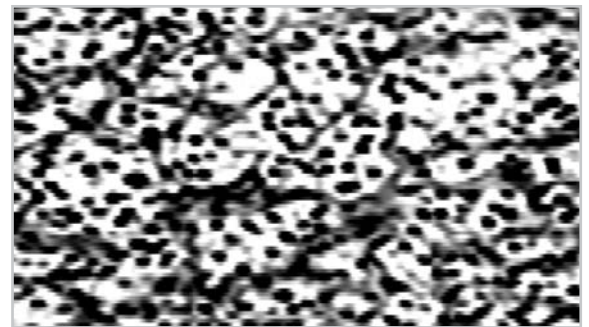


Figure 4: H & E stained section of mass shows fibrovascular stroma separating multiple cell balls (Zellballen) formed as a result of characteristic arrangement of polygonal tumor cells.

Doppler imaging shows the hyper vascular nature of the mass and peripherally splayed ICA and ECA.<sup>7</sup>

Three dimensional imaging modalities like Computerized Tomography as well as Magnetic Resonance Imaging can better delineate the location and number of tumors. DSA is considered the gold standard and it demonstrates dense blush of the hyper vascular tumor.<sup>7</sup>

Cut surface of tumor is dark, purple in color and is well circumscribed, similar to the normal carotid body except that clusters tend to be larger (Zellballen formation). Nuclear pleomorphism and cellular hyperchromatism are common. Malignancy is labeled only when there is local, regional or distant metastasis.<sup>2,3</sup>

Surgery and radiotherapy are the widely used treatment modalities. DSA enables embolization of blood vessels, thereby reducing intraoperative blood loss.<sup>4</sup>

### Conclusions

Carotid paraganglioma is a clinically rare entity in the differential diagnosis of a painless progressive neck mass that can be confused with cervical lymphadenopathy. Prior imaging is therefore essential before subjecting the patient to surgery and save the surgeon from perils of unexpected torrential bleed.

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