Case Report

Carotid body tumour: A rare case report

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Abstract

Carotid body tumour; also known as chemodectoma or carotid paraganglioma is a rare, highly vascular tumour arising from the paraganglia of carotid body. Head and neck paraganglioma account for 0.5% of all head and neck tumours. We report a case of a 25 year old female patient presenting with a painless, gradually progressive left lateral neck mass, diagnosed as carotid body tumour Shamblin class I by CT angiography and treated with local excision. Patient was followed up for 3 months with no recurrence or neurological complications. We report this case due to its rarity and the high vascularity and proximity to cranial nerves and major vessels make this tumour a surgical challenge.
Keywords: Carotid body tumour, Chemodectoma, Paraganglioma, Shamblin Criteria

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INTRODUCTION

Carotid body tumours also known as chemodectoma or paraganglioma are rare tumours derived from neural crest cells. Head and neck paraganglioma account for 0.5% of all head and neck tumours.[1,2] Carotid body tumours represent more than 50% of head and neck paraganglioma.[3] They are mostly benign and 5-30% of cases are functional or clinically malignant.[4,5,6] The only known risk factors are the presence of chronic hypoxic stimulation and the genetic predisposition.[2,7] About 60 % often remain clinically silent before presenting as a painless, slowly enlarging mass in the lateral neck[8,9,10] They are usually sonographically seen as a solid, well-defined,hypoechoic mass with a splaying of the carotid bifurcation.[8] The external carotid artery (ECA) is usually displaced anteromedially, and the internal carotid artery (ICA) is typically displaced posterolaterally.[11] The confirmation of diagnosis by CT (computed tomography) angiography. The treatment of choice is surgical resection. Good postoperative results can be expected in carotid body tumors of Shamblin Class I and II.[12,13]

CASE PRESENTATION

A 25 years old female presented with history of swelling in left part of the neck for 8 months, which gradually increased in size over the time with no significant past history and family history. On examination a firm swelling of 3x2.5 cm was noted in left side of neck in carotid triangle region which had greater horizontal than vertical mobility as seen in figure 1. The vitals and rest of the clinical examination was normal.

Investigations: CT scan and angiography of head & neck revealed a well circumscribed enhancing mass lesion noted on left side of neck, lateral to scalenus anterior muscle & situated in carotid triangle, deep to the sternocleidomastoid muscle (anterior belly).The mass is forming an angle of 153 degree with internal carotid artery (Figue 2 and 3). There was no evidence of adjacent bony destruction & no similar lesion seen on contralateral side.

Management anf follow up: Patient underwent local excision of tumour with findings as seen in figure 4. Histopathological examination (HPE) was suggestive of paraganglioma with tumour free margins. Patient was followed up for a period of 3 months with no recurrence or sign of neurological deficits.

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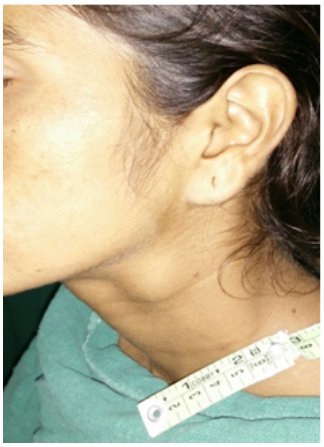
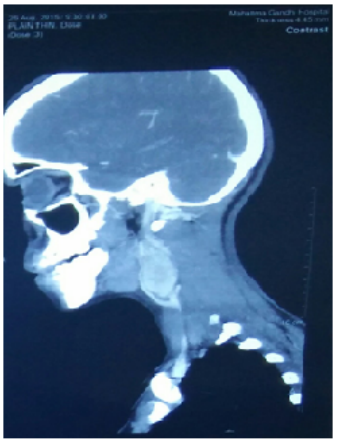


Figure 1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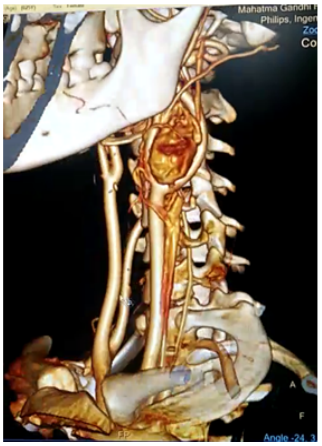
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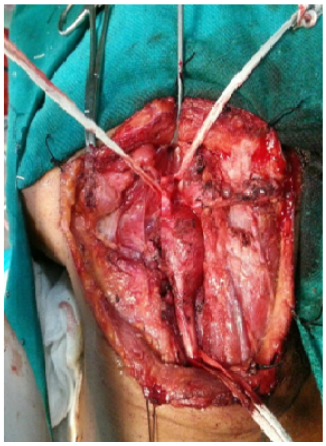
Figure 4

Legend

Figure 1: swelling in the left carotid triangle of the neck

Figure 2: CECT showing a well circumscribed enhancing mass lesion situated in carotid triangle, forming an angle of 153 degree with internal carotid artery.

Figure 3: showing carotid body tumour using 3D CT reconstruction

Figure 4: showing intra-operative findings of carotid body tumour

DISCUSSION

The carotid body, which originates in the neural crest, is important in the body's acute adaptation to fluctuating concentrations of oxygen, carbon dioxide, and pH. The carotid body, first described by von Haller in 1743, is a vascular reddish-brown structure approximately the size of a grain of rice, located within the adventitia posteromedial to the bifurcation of the Common Carotid artery(CCA). Blood reaches the carotid body through a fibrovascular bundle – the ligament of Mayer – that runs from the posterior surface of the CCA to the inferior portion of the carotid body and supplies the normal carotid body with more blood by weight than the brain.

Head and neck PGs are linked to the third branchial arch and the paraganglia of the glossopharyngeal nerve (IX).CNIX gives off a carotid sinus branch, explaining the location of CBTs. The ascending pharyngeal artery plays a large role in the vascular supply to many CBTs.

Carotid Body tumours: Paragangliomas[PG] can occur anywhere neural crest cells migrate from the skull base to pelvis.CBT is the most common type of PG of the head and neck. Other types include (classified by origin or location): Jugular paraganglioma (at jugular bulb), tympanic paraganglioma (arising from the tympanic plexus), and vagal paragangliomas (can arise along the entire course of the vagal nerve)

Etiology: Regarding the molecular genetics, head and neck paragangliomas have been associated with nine susceptibility genes: NF1, RET, VHL, SDHA, SDHB, SDHC, SDHD, SDHAF2 (SDH5), and TMEM127. Hereditary HNPs are mostly caused by mutations of the SDHD gene, but SDHB and SDHC mutations are not uncommon in such patients.Chronic hypoxic stimulation at high altitude is one of the risk factors with a female preponderance.[2]

Clinical presentation: Signs and symptoms are induced by the topography of the anatomical structures from which the tumors originate. 90% of CBTs are spontaneous and usually appears in 3rd and 4th decades.[2] CBTs often remain clinically silent before presenting as a painless, slowly enlarging mass in the lateral neck.[8,9,10] The initial symptoms can also be a pulsating mass in the neck. CBTs can be moved horizontally rather than vertically, a finding known as a positive Fontaine's sign.[8] Larger tumours may induce dysfunction of the vagal nerve and, less frequently, of cranial nerves IX, XI, and XII and occasionally, Horner's syndrome or deficits of the facial nerve.[8,9] Sometimes a carotid bruit or a pulsating mass can be detected.

Differential diagnosis of painless lateral neck mass: Branchial Cleft Cyst, Reactive lymphadenopathy, neoplastic causes such as lymphoma, salivary gland tumors, neurogenic tumors, vascular aneurysms of the carotid artery, hematoma, pseudoaneurysm and paraganglioma.[12]

Imaging modalities: B□mode ultrasonography and color Doppler is the first test for painless lateral neck mass which demonstrates hypoechoic mass that appears hypervascular on color doppler with flow directed upwards.CBTs are often hard to differentiate from other solid masses.

MRI provides very good soft tissue resolution.MRA or MRI with 3D reconstruction imaging demonstrates avid enhancement. On MR or CT angiography, besides the

characteristic tumour blush (lyre sign) in patients with a carotid body tumour, a characteristic distortion of the carotid bifurcation is also visible. The internal carotid artery is generally pushed laterally and posteriorly, while the external carotid artery is displaced anteriorly. Biopsy, including fine needle aspiration, is unnecessary, dangerous and contraindicated in the evaluation of paragangliomas. [15,16]

CT is sensitive for demonstrating bony destruction in other paragangliomas.[17]

Digital subtraction angiography (DSA) remains the gold standard for diagnosis which provides access for preoperative embolization of feeder vessels. Bilateral carotid DSAs should be done to locate other smaller tumors.Preoperative assessment of vasculature allows surgeon to remove arterial supply before venous, diminishing blood loss.[17]

PET18F-DOPA PET is more sensitive than MRI for tumors < 1cm and maybe used to screen patients with hereditary paraganglioma.[17]

Classification of carotid body tumors: Shamblin classification[18]

Class I CBTs - localized tumors with splaying of the carotid bifurcation, but little attachment to the carotid vessels. Complete surgical resection is generally possible with only minimal risk of vascular or cranial nerve complications.

Class II CBTs- partially surround the carotid vessels and complete resection is more challenging.

Class III CBTs intimately surround the carotid.Complete resection is very challenging, and often requires temporary interruption of the cerebral circulation for vascular reconstruction. The risk of permanent vascular and neural defects is significantly higher than for Class I and Class II tumors.[19]

Treatment: Surgery remains the main modality of treatment.Pre-operative embolization is considered by most to be useful in CBTs with majority of blood supply being cut off. The routine use of preoperative embolization is controversial because of the potential neurologic complication associated with the accidental reflux of particulate matter into the ophthalmic or cerebral circulation. Some authors advocate its use before the resection of large tumours because it may decrease the vascularity of the tumour, reducing intra-operative blood loss and transfusion requirements. The apparent benefit of embolization should be weighed against the risk of stroke.[20]

Ferreira *et al.* (2013) from Portugal reported 4 cases of CBTs resected using an ultrasound dissector.They believe that this technique improves the safety of excision, decrease the technical difficulties, lower blood loss and shortens the operation time.[21]

Radiotherapy as primary treatment modality has been advocated only in unresectable tumours or where surgery is contraindicated with complications such as necrosis of the mandible, carotid artery and larynx.[2,20]

Prognosis: For Shamblin type I and type II CBTs, the peri- and postoperative risk of neurovascular complications is low. Complication rate increases strongly for Shamblin type III CBTs.[12,13] The current literature shows serious postoperative complications only in 0–2.3% of patients with Shamblin type I and II tumors, while this figure increases to 7–35.7% for patients with a Shamblin type III tumor.[12,13,22]

Guidelines on Management: There is considerable amount of controversy regarding choice of treatment modality. According to Hussen et al and Suarez et al, surgery is considered by many authors as the standard therapy as it provides an immediate and complete removal of the tumour.[23.24] Some recommend the wait and watch policy since it is mostly benign and has indolent process.[2] But the majority of studies recommend surgery as the treatment of choice.

CONCLUSION

A 25 year old female patient presenting with a painless, gradually progressive lateral neck mass with positive Fontaine sign was diagnosed by CT and CT angiography as carotid body tumour Shamblin class I and treated with local excision with no post operative complications on follow up period of 3 months. Thus to summarise, carotid body tumour is one of the uncommon tumours of head and neck region. This case is being reported for its rarity.

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