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Unwinding the Serpent— Excision of a Unilateral Shamblin 3 Carotid Body Tumour without Vessel Grafting- A Case Report

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Abstract
Carotid body tumour (CBT), is a rare neoplasm, developing within the adventitia of the medial aspect of common carotid artery bifurcation. This case report describes the management of a unilateral carotid body tumour in a 42 year male patient. The relevant literature is also reviewed.
Keywords: *carotid body tumour, Shamblin classification, paraganglioma.*

Introduction

The carotid body, which is a 3 – 5 mm flat brownish nodule in the adventitia of the common carotid artery, is essential for the body to adapt to acute changes in levels of oxygen, carbon dioxide and pH, by releasing various neurotransmitters and hyperventilating when stimulated. This helps prevent hypoxic damage to organs. Blood supply to the tumour is from the external carotid artery, mainly the ascending pharyngeal branch, entering through the *Meyer's* ligament along the postero-medial aspect of the carotid bifurcation.

Carotid body tumour (CBT) constitutes around 65% of the paragangliomas which are of neural crest cell origin in the head and neck region[1] .The most common presentation of CBT is an asymptomatic swelling in the anterolateral neck region. These are slow growing tumours. The doubling time (T_d) of CBT as estimated by Jansen et al using sequential imaging, was 7.13 years with a median growth rate of 0.83mm per year[2].

On clinical examination, a CBT presents as a swelling in the carotid region of the neck, smooth, firm, pulsatile (transmitted pulsations), with horizontal mobility but typically vertically fixed because of it's attachment to the carotid bifurcation (Fontaine sign). A bruit can be heard, though absence of which cannot rule out a CBT.

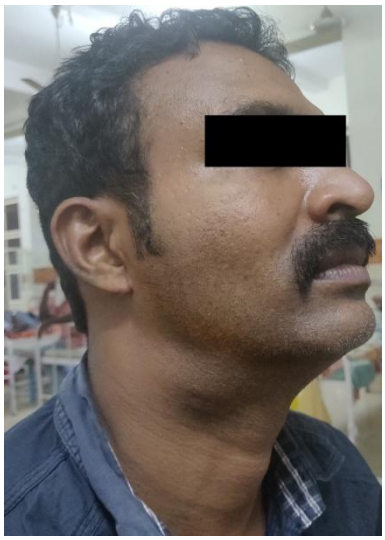
Around 10% of the tumour can present with cranial nerve palsies involving the hypoglossal, glossopharyngeal, recurrent laryngeal, spinal accessory or the sympathetic chain[3].This can cause complaints of pain, hoarseness of voice, tongue paresis, Horner's syndrome and dysphagia. Fever, though uncommon in CBTs, literature has reported it as one of the cause of Pyrexia of Unknown Origin[4]. Symptoms like paroxysmal hypertension, palpitation, diaphoresis similar to those of pheochromocytoma may also be seen.

Case Presentation

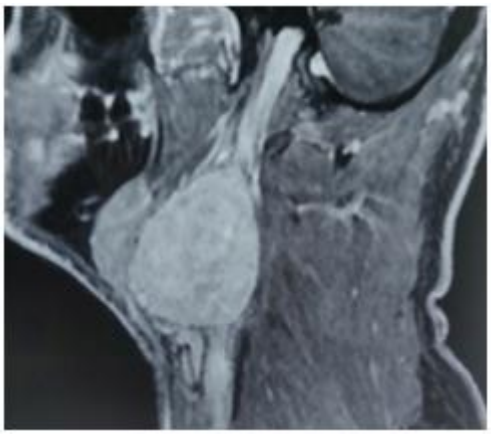
A 42 year old gentleman, who is a motor vehicle mechanic by profession, a resident of an area at sea level, a non smoker, without history of any known co-morbidities nor any significant family history, presented with complaints of a swelling in the right side of neck for over 15 years, with very slow progression in size and without any complaints of dysphagia, dyspnoea, dizziness, syncopal attacks, blurring of vision, hoarseness of voice or loss of weight. On examination, the patient is normotensive, with a normal regular pulse rate. A smooth, firm neck swelling, around 6 x 4 x 1.5 cm is palpable in the antero- lateral aspect of right side of neck with lateral border just anterior to the anterior border of sternocleidomastoid muscle. It is immobile vertically along with transmitted pulsations.

Patient was evaluated initially with an ultrasound neck which showed a solid mass at the region of carotid bifurcation on the right side. Furthermore, on evaluating with a computed tomography angiography and magnetic resonance imaging, a homogenously enhancing mass lesion at the right carotid bifurcation was noted, causing the splaying of internal and external carotid arteries (*Lyre sign)* consistent with the diagnosis of carotid body tumour. On imaging, the tumour was classified to be a Shamblin type 3 tumour.

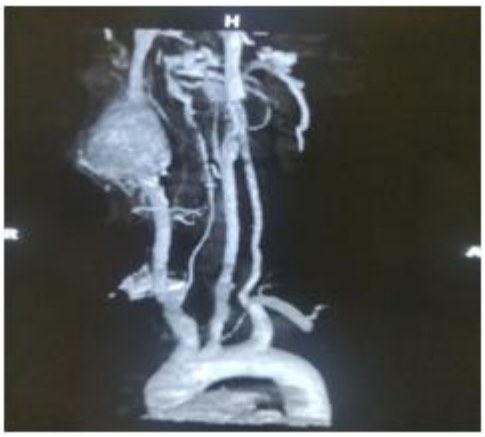
After all pre operative investigations, patient underwent surgical excision of the tumour under general anaesthesia. A 6 cm incision along the anterior border of sternocleidomastoid was put. The dissection continued layer by layer till the tumour was identified. Though, the tumour was highly adherent and vascularised, with gentle blunt dissection and using bipolar cautery, complete excision was achieved without the need of any vascular reconstruction contrary to the general norm of a temporary internal carotid artery bypass channel in a Shamblin type 3 CBT. The specimen was sent for histopathological examination which revealed a paraganglioma with positive staining for synaptophysin, chromogranin, S100 and negative for HMB45. Post-operative period was uneventful, except for a mild transient dysarthria and a minimal right ward deviation of tongue possibly due to neurapraxia of the ipsilateral hypoglossal nerve. Patient was discharged with near complete recovery from all the complaints.



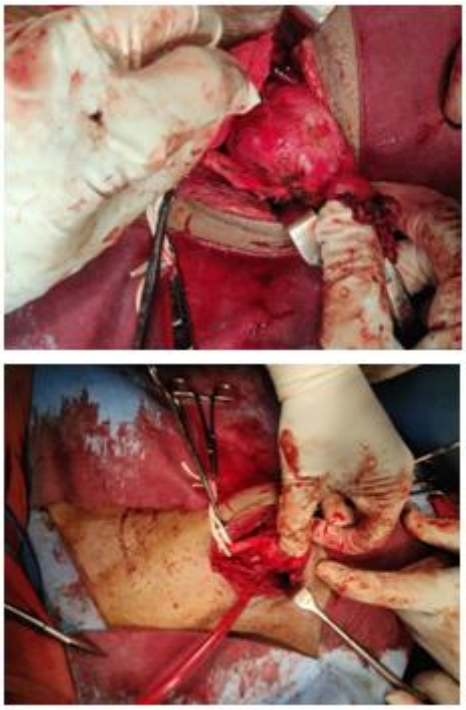
Pre-operative photo of patient showing swelling in right side of neck



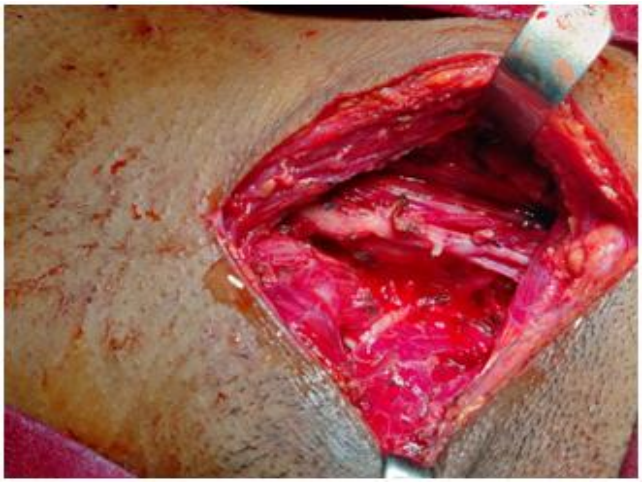
MRI -Sagittal view, T1 -Fat suppression image showing the CBT



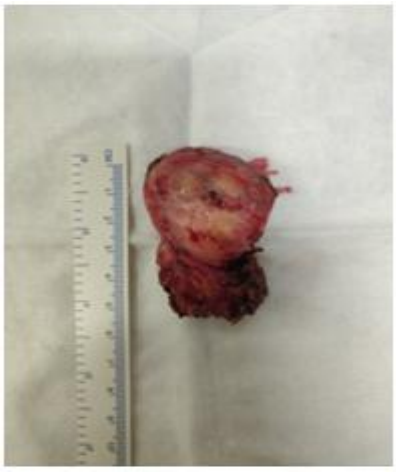
CT-Angiography image showing CBT



Per-operative image showing carotid body tumour at the carotid bifurcation



Per-operative image after the excision of carotid body tumour



Excision specimen in toto

Discussion

Paragangliomas can be divided into sympathetic and parasympathetic with most of the CBTs belonging to parasympathetic and non functional. Only 4-5% of them are sympathetic and have catecholamine production[5].

The 3 different types of paragangliomas that are being described in literature are:

- Sporadic (85%)
- Familial (10-20%)
- Hyperplastic

The sporadic form is the most common type, whereas familial type is more common in young patients. The incidence of hyperplastic type is more in patients with chronic hypoxia like inhabitants of high altitude (5000 feet above sea level), Chronic Obstructive Pulmonary Disease and cyanotic heart disease patients[6].

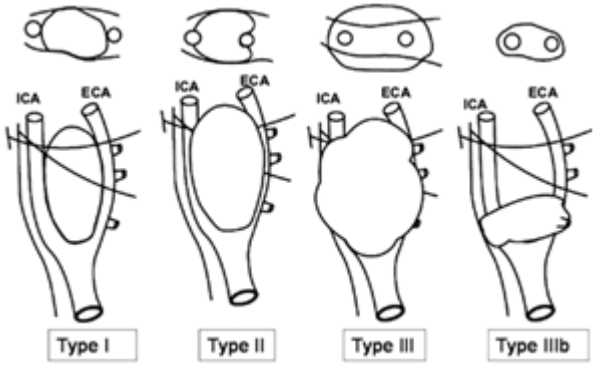
About 30% of paragangliomas have genetic correlation to *Succinyl Dehydrogenase (SDH) gene locus. The SDHA, SDHB, SDHC* genes are particularly associated with head and neck paragangliomas. Head and neck paragangliomas are also associated with Multiple Neuroendocrine (MEN) syndromes MEN 2A and MEN 2B[7].

Histology

Both malignant and benign type of paraganglioma contain the same two types of cells-epithelioid chief cells and supporting sustentacular cells. Together these two cell types form clusters (*Zellbalen)* surrounded by extensive vascular sinusoids. Histological appearances like nuclear atypia, vascular invasion, central necrosis and mitosis are not specifically associated with metastasis or true invasion[7].

Classification

The first classification system was proposed by Shamblin in 1971 based on carotid vessel invasion, modified by Luna -Ortiz to a more detailed system[8].



Shows a schematic diagram of the Shamblin grouping of CBTs into I, II, and III, as well as IIIb (as proposed in the modification to Shamblin's classification by Luna-Ortiz et al[8]). This is based on the relationship of the tumour to the carotid vessels, ICA and ECA. The class IIIb tumours include tumours of any size which are closely adhered to the carotid vessels. The oblique lines shown represent the X and XII nerves, which are intimately related to the tumour

Imaging

It is important to differentiate between secreting and non-secreting paragangliomas preoperatively to prevent an adrenergic crisis intra-operatively. All patients must have negative biochemical results for catecholamine hypersecretion or undergo alpha blockade before older ionic contrast agents are being administered for a CT scan. Current nonionic low- osmolality contrast agents are safe in patients with catecholamine secreting tumours, even without adrenergic blockade[9].

Computed Tomography (CT):

Contrast enhanced CT is excellent in evaluation of CBTs. Typical appearances are:[10]

- Soft tissue density on non-contrast CT (similar to muscle)
- Bright and rapid enhancement – faster than schwannoma.
- splaying of the ICA and ECA (*Lyre sign)*

MRI

- T1
  - Iso to hypointense compared to muscle
  - Salt and pepper appearance in larger lesions because of combination of punctate haemorrhages or slow flow (salt) and flow voids (pepper) [11]
  - Intense enhancement following gadolinium contrast.
- T2
  - Hyper intense when compared to muscle

Management

Symptomatic CBTs need surgical removal, the principles of which include wide surgical exposure, proximal and distal vascular control, identifying and preserving neurovascular structures, and careful dissection from carotid arteries[12]. High rates of post operative cranial nerve dysfunction favour observation of asymptomatic CBTs[13] .

Though, a Shamblin 3 CBT warrants surgical excision.

Conclusion

A carotid body tumour is a rare paraganglioma at the carotid bifurcation which is justifiable to manage conservatively in asymptomatic patients, though around 75% become symptomatic eventually[13]. However, a shamblin 3 CBT in this patient was dealt with surgical management.

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