International Surgery Journal
Shankar M et al. Int Surg J. 2017 Jun;4(6):2067-2069
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: http://dx.doi.org/10.18203/2349-2902.isj20171992

Case Report

A large carotid body tumour: a rare case report

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Received:19 April 2017
Accepted: 24 April 2017

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ABSTRACT

Carotid body tumour (CBT) is one of the most common jugular paraganglioma involving the carotid body chemoreceptors. We report the case of a 22 year young man who presented with complain of large painless progressive swelling in right upper neck for 3 years duration. On examination, swelling appeared pulsatile and arising from carotid vessel. MRI Neck and MRA performed revealed large well defined hypervascular mass lesion encasing right common carotid artery, its bifurcation and ECA, with feeders from ECA. Patient was undertaken for surgical excision (Shamblin Grade-III) through exposure in neck, extended by midline sternotomy and mandibulotomy approach, tumour was completely dissected. Patient recovered well from surgery without any sequale. HPE revealed CB paraganglioma. At two years follow up, he is recurrence free. The surgical removal of CBT has a good result.

Keywords: Carotid body tumour, ECA, Paraganglioma, Shamblin grade

INTRODUCTION

The carotid body (CB), described for the first time by Albrecht Von Haller in 1743 is a small structure 12 mg in weight, located in the adventitia of carotid bifurcation serving as a chemoreceptor. It has both ectoderm and mesoderm tissue elements and consist of 2 Types of Glomus cells: Type I (Chief or Paraganglion cells) and Type II (Sustantecular cells).

The carotid body receives its blood supply mainly from the external carotid artery (ECA) and has afferent innervations via the glossopharyngeal nerve. A rare highly vascular mostly benign tumour may arise from the CB paraganglia previously known as Chemodectoma or CB Paraganglioma. As the paraganglia are widely distributed in the body (in association with the sympathetic and parasympathetic chains from the skull base to pelvic floor); paragangliomas may arise in different locations such as the abdomen, retroperitoneum, mediastinum, head and neck and best named by their location.[1] Carotid Body Tumour (CBT) is the commonest head and neck paraganglioma. The exact etiology of CBT is unknown, although its higher incidence among people living at high altitudes or those with chronic obstructive pulmonary disease suggests a role of chronic hypoxia. The tumour mostly occurs in sporadic form while minority of patients had a familial type which is thought to be related to genetic factors.

The tumour is usually non-functional but occasionally tumours capable of catecholamine secretion are diagnosed with symptoms similar to pheochromocytoma such as hypertension and tachycardia. Carotid arteriography used to be the gold standard for diagnosis, although Duplex US scanning and magnetic resonance arteriography (MRA) are favored by many authors nowadays. CBT has a characteristic histopathological appearance.[2]

It represents a surgical challenge due to its location, high vascularity and potential morbidity and mortality.

Surgery is the standard therapy as it provides an immediate and complete removal of the tumour. However, the morbidity of cranial nerve injuries and stroke associated with surgery is significant. On the other hand, the indolent and very slow growth rate of CBT and the fact that most of them are benign and rarely cause death by themselves encourage some authors to adapt the conservative or the so-called (wait and scan) policy. A third group of workers recommend radiotherapy as a primary mode of management of CBT to achieve a tumour growth control while avoiding the potential morbidity of surgical intervention.

CASE REPORT

In the present study a case of 22-year young male, who presented with painless progressive swelling in right upper neck of 3 years duration was reported. On examination swelling appeared pulsatile and arising from carortid vessel. MRI Neck with MRA performed revealed well defined hypervascular mass lesion approx. 7.8 cm x 7.4 cm x 7 cm encasing right common carotid artery, its bifurcation and external carotid artery, with feeders from external carotid artery. Patient was undertaken for surgical excision (Shamblin Grade-III) through exposure in neck, which was subsequently extended by midline sternotomy and mandibulotomy approach to expose all length of carotid vessel from arch of aorta to the base of skull in view of extent of tumour involving carotid from its origin at arch of aorta to base of skull.

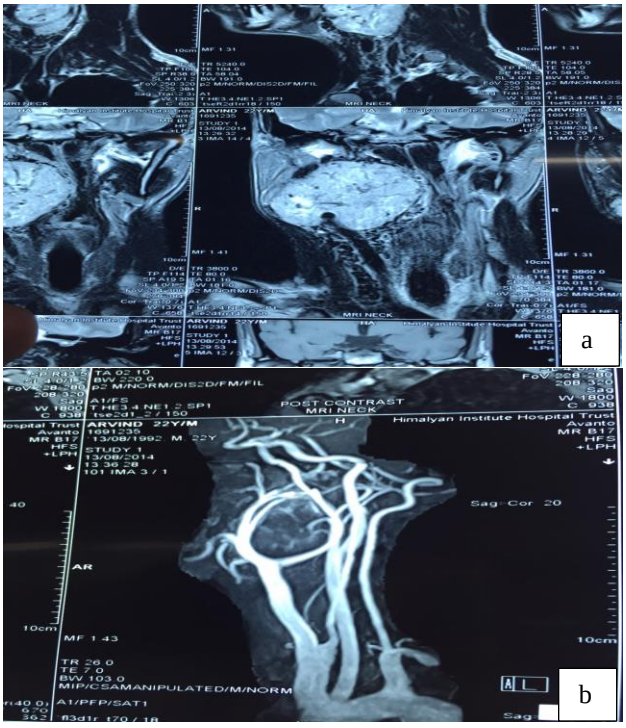

Figure 1(a and b): MRI neck with MRA revealed well defined hypervascular mass lesion approx. 7.8 cm x 7.4 cm x 7 cm encasing right common carotid artery, its bifurcation and external carotid artery, with feeders from external carotid artery.

Tumour was dissected in subadventitial plane, without any arterial bypass, minor perforations were repaired. External carotid artery was sacrificed. Patient recovered well from surgery without any sequale. Histopathology of tumour revealed carotid body paraganglioma. At two years follow up, on clinical evaluation and colour Doppler study he is recurrence free with normal blood flow through the carotids. Patient developed hypertrophic scar at neck incision sites.

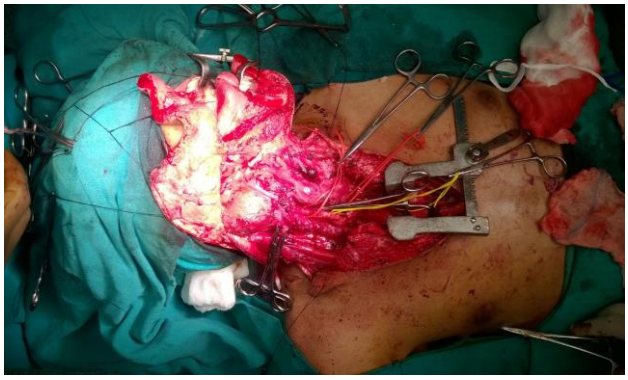

**Figure 2: Surgical excision through exposure in neck, extended by midline sternotomy and mandibulotomy approach to expose all length of carotid vessel from arch of aorta to the base of skull in view of extent of tumour involving carotid from its origin at arch of aorta to base of skull. tumour was dissected in subadventitial plane, without any arterial bypass, minor perforations were repaired.
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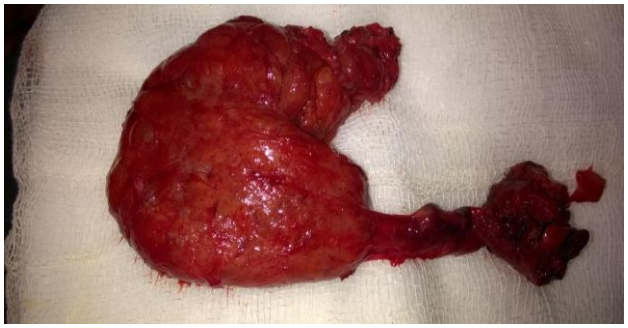

Figure 3: Completely excised carotid body tumour.

DISCUSSION

CB paragangliomas are slow growing painless masses localized in the neck, anterior to the sternocleidomastoid muscle at the level of the hyoid bone. As the tumour grows, dysphagia, odynophagia, dysphonia, and symptoms due to compression of cranial nerves 9 to 12 may be seen. The most commonly involved cranial nerve is the vagus, up to one third of all cases will show cranial nerve palsies. Physical examination of patients with CBT characteristically reveals a rubbery, painless mass in the upper neck along the anterior border of the sternomastoid muscle and the lesion is more freely movable laterally

than vertically (fontaine`s sign). Occasionally the tumour mass may transmit the carotid pulse or demonstrate a bruit or thrill.

Carotid arterial angiography is the most valuable diagnostic technique. It can detect multiple lesions, tumour size and vascularity, and the major vascular tributaries perusing the tumour. Pathognomically, a specific perfusion increase (Lir sign) at the carotid bifurcation can be seen due to the tumor. Arteriographic imaging should be carried out bilaterally because of the possibility of bilateral tumors. In cases of bilateral tumours, serum catecholamine levels as well as urinary vanilmandelic acid and metanephrine levels should be assessed. MRI and CECT scans are additional noninvasive diagnostic tools. With MRI, paragangliomas smaller than 0.8 cm in diameter can be detected. Biopsy is contraindicated in cases of paraganglioma. Paragangliomas are generally sporadic in character. Patients with a family history and hypertensive palpitation attacks with flushing should be assessed.[3]

Surgery is the first choice of treatment. Radiotherapy has been used as the sole treatment in a few cases, and the short-term results are disputable. Radiotherapy may be considered as an alternative modality for an elderly patient in poor general status, whereas surgical treatment is preferred in young patients.[4]

Tumour classification (Shamblin) is based on the size and the difficulty of surgical resection-

Grade-I: small and easily resected from vascular elements.
Grade-II: medium-sized tumour closely associated with vascular structures, which can be resected by careful subadventitial dissection.
Grade-III: large and enfolded by the carotid arteries; it can be resected only by partial or total vascular resection may also require vascular replacement.[5]

A transcervical approach is usually preferred, but in superiorly localized carotid paragangliomas, a cranial basilar approach can be used. The most important principle is to preserve the integrity of the internal carotid artery. If necessary, the external carotid artery can be sacrificed. Defects in the common or internal carotid arteries must be repaired immediately and graft replacement may be necessary.

CONCLUSION

Surgical excision is the first choice of treatment in younger patients. The diagnosis of CBT at an early stage minimizes the risk and complications associated with surgical removal and lead to good post-operative results.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Shankar M, Saini S, Pattanayak M. A large carotid body tumour: a rare case report. Int Surg J 2017;4:2067-9.