CASE REPORT

Carotid Body Paraganglioma

Tahir Manzoor, Bashir Ahmed, Atif Najam and Zeeshan Ayub

ABSTRACT

Carotid body tumours are benign neoplasms. This case report describes two patients with this rare tumour with diagnostic workup and treatment options. The first case was a 36-year-old gentleman who presented with 6 months history of painless swelling on the left side of neck. The second patient was 60 years old man who presented with slowly growing swelling on the right side of neck for the last 20 years. Both patients had transmitted pulsations over the swelling. On suspicion of a vascular lesion, a contrast enhanced CT scan and Doppler's ultrasound of neck was advised which suggested the diagnosis of a carotid body tumour. Magnetic Resonance Angiography (MRA) confirmed the diagnosis. Excision in collaboration with vascular surgeon was planned for the first case. The second case was subjected to radiation therapy due to the large size of tumour and the patient's age.

Key words: *Carotid body. Paraganglioma. Cathecolamines.*

INTRODUCTION

Carotid body tumours are rare benign slow growing neoplasms also called paragangliomas. They arise from the carotid bodies located in the adventitia at the posteromedial aspect of carotid bifurcation. These carotid bodies have demonstrated a chemoreceptor role by modulating cardiovascular and respiratory functions in response to fluctuations in arterial pH, carbon dioxide and oxygen tension historically treated by excision.[1] Carotid body tumours are the commonest paragangliomas of the head and neck region. The treatment of choice for these tumours is surgical excision alone or in combination with carotid artery resection.

We share our experience of two cases with different presentations and management protocol.

CASE REPORT

Case 1: A 36-year-old serving soldier presented with 6 months history of painless swelling on the left side of neck. Patient incidentally noticed the swelling that was initially small but gradually increased in size. There was no history of fever, headache, palpitations, dysphagia or hoarseness of voice. Examination of the neck revealed a well-circumscribed swelling measuring 3 x 2 cm, in the upper 1/3rd of neck along the anterior margin of sternocleidomastoid on the left side. The swelling was firm in consistency non-tender, had transmitted pulsations with mobile overlying skin but was adherent to the underlying structures. Keeping in view, the pulsatile nature of the swelling, Doppler ultrasound of neck along with ultrasound guided Fine Needle Aspiration Cytology (FNAC) of the lump was advised. FNAC was inconclusive and revealed a haemorrhagic aspirate. Doppler's ultrasound showed a well-demarcated mass measuring 3 x 3 cm overlying the left carotid bifurcation and splaying the internal and external carotid arteries. Findings were consistent with that of a carotid body tumour. To confirm the diagnosis, a Magnetic Resonance Angiography (MRA) was done, which showed characteristic splaying of the left internal and external carotid arteries. There was no luminal narrowing and blood flow in the distal vessels was normal (Figure 1). Based on strong radiological evidence, final diagnosis of carotid body tumour was made and excision in collaboration with vascular surgeon was planned. Pre-operative work up included 24 hours urinary Vanillylmendallic Acid (VMA), CT scan of chest and ultrasound abdomen was done to exclude a secretory tumour and coexisting multicentric tumour. All investigations were within normal limits.

Peroperatively, the tumor was partially adherent to the internal carotid artery and was completely encasing the external carotid artery. Proximal and distal control of the vasculature was achieved by passing loose tapes around the carotid vessels. Tumor was dissected free from the internal carotid artery. External carotid artery was ligated and resected along with the tumor after identifying and preserving the vagus and hypoglossal nerves. Patient was followed up till 9 months after surgery and there were no neurological complications.

Case 2: The second patient, a 60-year-old male, presented with 21 years history of painless swelling on the left side of neck. There was no history of hoarseness, dysphagia, epistaxis, headache and facial flushing. On examination, a 5 x 5 cm swelling was seen in the upper 1/3rd of the neck located in the submandi-

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bular region and extending upto the posterior triangle of the neck. The swelling was firm, non-tender and had transmitted pulsations over it. The swelling was adherent to the underlying structures with mobile overlying skin. His ENT and systemic examination was unremarkable. Keeping in view the long-standing history, ultrasound guided Fine Needle Aspiration Cytology (FNAC) along with Doppler ultrasound of the neck was advised. FNAC was inconclusive and revealed suspicious spindle shaped cells. Doppler study showed a mass along the common carotid artery pushing the great vessels anteriorly. To exclude a schwanoma or a paraganglioma, an MRI was advised, which revealed a 5 x 5 cm mass at the bifurcation of left common carotid artery causing splaying of the external and internal carotid artery. The mass was adherent to the internal and external carotid vessels on the right side. A smaller 2 x 1 cm mass was also seen at the bifurcation of right common carotid artery, suggesting the presence of a rare bilateral carotid body paraganglioma (Figure 2). In order to confirm the diagnosis and assess the patency of carotid vessels, MRA was advised. MRA revealed bilateral splaying of the carotid vessels more on the left side with normal distal blood flow in both carotid vessels. A search was made for secretory tumour and a synchronous pheochromocytoma but no abnormality was found. Keeping in view the age of the patient and extensive disease on the left side, he was given 45 Grays (Gy) of external beam radiotherapy covering both sides of the neck. Eight months after treatment, the left sided neck swelling did not increase in size. There is still no clinical evidence of swelling on the right side.

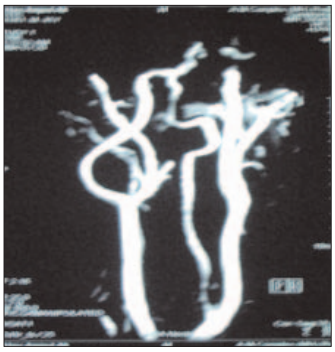

Figure 1: Magnetic resonance angiography showing splaying of left internal and external carotid arteries.

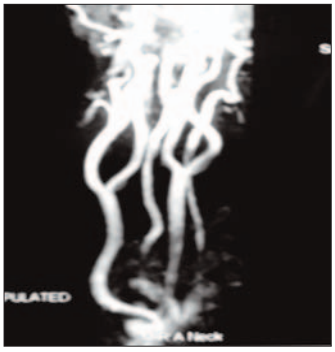

Figure 2: Magnetic resonance angiography showing bilateral carotid body paraganglioma.

DISCUSSION

Carotid body was first described by an anatomist Von Haller in 1743. Von Lushka in 1862 and Marchand in1891 reported carotid body tumours for the first time. Scudder in 1903 successfully removed a carotid body tumour.[1] Majority of these tumors are sporadic but 10% of these tumours are multicentric and 7-9% are familial. These tumours have a reported higher incidence at higher altitudes and a slight female preponderance.[2] One to 2% of these tumours demonstrate secretory activity by releasing catecholamines.

Malignancy has been reported in all sites of paragangliomas determined by local or distant metastasis proven on biopsy. About 6-10% of carotid body paragangliomas are known to be malignant.[3]

Clinically, these tumours present as a lateral cervical mass located in the upper 1/3rd of neck. Mean age of presentation is between 45 to 55 years. The consistency varies from soft and elastic to firm. They are non-tender and pulsatile. A bruit may also be heard by auscultation which might disappear with carotid compression. Swelling is often less mobile in the cranio-caudal direction because of its adherence to the carotid arteries. Tumour enlargement may cause symptoms like dysphagia and hoarseness of voice due to involvement of vagus nerve. These tumours may present as pharyngeal mass pushing the tonsils anteriorly and medially.[4]

Radiological imaging is the mainstay of diagnosis and imaging modalities include Doppler ultrasound, contrast enhanced CT scan, MRI, MRA and carotid angiography. MRA is exceedingly being utilized in place of carotid arteriography. Carotid angiography is the gold standard for diagnosing these tumours and shows splaying of carotid vessels along with tumour blush called the Lyre's sign.[5] Currently, this invasive modality is used for detection of feeding vessel and pre-operative embolization.

Fine needle aspiration cytology of these tumours can help in pre-operative diagnosis.[6] Incisional biopsy is contraindicated in these tumours. Pre-operative work up must include a 24-hour urinary VMA and serum catecholamines levels, which if raised, merits an abdominal CT scan to exclude a coexisting pheochromocytoma.

Surgical excision is the treatment of choice for these tumours. Radiation therapy and simple observation and follow up are other modalities that can be used in elderly patients. Shamblin in 1971 classified these tumours into three types based on the difficulty of resection. Surgical options include resection of the tumour alone or in combination with carotid artery. Davidovic *et al.* successfully operated upon 12 cases over a 21 years time with no significant neurovascular complications.[7] Pre-operative embolization is reserved for tumors more than 5 cm in size. Operative complications include haemorrhage, injury to carotid arteries, vagus and hypoglossal nerves. Das *et al.* operated 10 patients with carotid body tumours and only one patient with Shamblin type III tumor had difficulty swallowing and nasal regurgitation postoperatively that improved with time.[5] Historically, these tumours are known to be radioresistant. Radiotherapy is reserved for large inoperable tumours, metastatic tumours and patients unfit or not

willing for surgery. Stereotactic radiotherapy is an evolving modality that delivers focussed dose to the tumour site. Russell *et al.* treated 121 paragangliomas of the head and neck with radiotherapy and have reported local control rates of 95% with minimal complications.[9] Surgical excision, because of its proven safety and long-term control, still remains the treatment of choice for these tumours.[8] However, with evolutions in stereotactic radiotherapy, further trials are needed before the two forms of treatments can be compared.

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