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Author: Dr Devender Singh

Company/University: Nizams institute of medical sciences

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Title: Carotid body tumour in a patient with sinus inversus totalis:

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a case report

Dr Devender Singh Dr Pinjala RK, Dr Ramamurthi S, Dr LRC Reddy,

Department of Vascular surgery and Radiology

Nizam's Institute of Medical Sciences, Hyderabad

ABSTRACT:

The carotid body tumour is a rare neoplasm that has generated much literature over the last century, and for which continued controversy exists regarding natural history, biologic behavior, proper technique of excision, and the risk of morbidity & mortality. Resection of the carotid body tumours (neck paragangliomas) carries inherent risk of injury to the cranial nerves and other structures as well excessive blood loss. Pre operative embolisation has been used to lesson the morbidity in tumours that are larger than 2cm in diameter. Our patient of situs inversus totalis, presented with a large asymptomatic tumour of 5°i6cm, over the left neck. She was already operated twice, thinking in terms of an enlarged lymph node. The patient had preoperative angiography that revealed the feeding arterial vessels so that successful embolisation could be accomplished with PVA particles. Success was judged by diminution of the angiographic blush. Patient had an uneventful surgical excision, with the carotid body tumour being able to be resected periadventitally without damage to either the external or internal carotid artery. The cranial nerves were preserved and the blood loss was minimum, not requiring any transfusion. We conclude that pre operative embolisation is an important adjunct in treating patients with large carotid body tumours. The surgical exploration precedes much smoother, the blood loss minimal & the patients have minimal morbidity. Also sinus inversus totalis, seems to be an incidental finding.

Key words: Carotid body tumour, paraganglioma, embolisation, radical excision

INTRODUCTION:

Carotid body tumour is not a common disease but it should be considered in the differential diagnosis of neck masses at the mandibular angle. The Carotid body tumour is a rare neoplasm, which can be caused by a genetic predisposition or by chronic hypoxic stimulation. This usually presents as an asymptomatic mass & many a times being attempted for surgery, considering it to be an enlarged lymph node. Diagnostic modalities include ultrasound, color Doppler, angiogram, CT scan & MRI. Surgical excision is the widely accepted modality of treatment, whereas pre operative embolisation has been used to lessen the morbidity in large tumour. Radiotherapy is useful in patients with unresectable, locally recurrent or partially resected tumours.

We present a patient of sinus inversus totalis who developed carotid body tumour, which progressed to a large size & attempted a couple of time for excision by local surgeons. In view of large size & hypervascularity, pre operative embolisation was done, followed by excision of the tumour in toto.

Case report:

31 years old lady, a school teacher was referred with a large swelling of 5°i6cm in the anterolateral aspect of the neck for the past seven years. She was complaining of giddiness off & on. There was no neurologic deficit & also clinically no symptoms suggestive of endocrinological activity. Physical examination revealed, well circumscribed large mass at the angle of jaw, which was mobile in the transverse plane. There was no palpable bruit over the tumour. There was no family history of such swelling. She was operated a couple of time by the local surgeons, considering it to be

an enlarged lymph node & both the time incision was closed in view of excessive bleeding. At one time the biopsied material was reported as reactive lymphadenopathy. However after second surgery, in view of bleeding a CT angiogram was done, which was suggestive of carotid body tumour at

the bifurcation of the left common carotid artery. At this point of time she was referred to us for further management. During evaluation she was found to be a case of sinus inversus totalis, with good cardiac function. Because of the large size, previous manipulations & hypervascularity, we planned for pre operative embolisation. Her left CCA angiogram reveals hyper vascular tumour in the region of carotid bifurcation. Lesion is deriving arterial feeders from the branches of ECA i.e ascending pharyngeal & occipital arteries. Embolisation of the same done, with PVA particles (255-350microns) & there was an appreciable reduction in the hypervascularity (40%). After 72 hours she was taken up for the surgery, for the radical excision of the tumour. Total tumour of 40 grams was excised successfully via the peri adventitial plane, with minimum blood loss. The tumour was engulfing the bifurcation & left CCA (Shamblin $\beta\pm$). There was no requirement of blood transfusion. There was no peri operative adverse event (stroke etc.). In the immediate post operative period she developed features of transient palsy of the seventh nerve (i.e deviation of the angle of the mouth), which later completely recovered. Her histo pathological examination of the specimen was suggestive of the paraganglioma (benign) of the carotid body.

DISCUSSION:

The carotid bodies are reddish brown, ellipsoid structure, lying embedded in the adventitia of the carotid artery bifurcation^{1,2}. Described first by Haller in 1743, they are derived from epitheloid cells of the neuroectodermal origin³. Tumour of this tissue were originally described as chemodectoma by Mulligan; now they are considered as a part of the widely described group of tumours known as paraganglioma. Carotid body tumours are very rare neoplasm, constituting less than 0.5% of all the body tumours⁴. An incidence of 0.012% of all surgical specimens has been reported by a hospital based study⁵. Sporadic forms are more frequent, while familial forms accounts for about 10% of cases in most series⁷. The tumours are bilateral in 30% of the familial, but only 5% of the sporadic cases⁸. Carotid body tumour are usually benign, however malignant potential with possible metastasis has been estimated to be around 2% to 9%. Carotid body tumours can be a diagnostic

challenge for the clinician and lack of pre operative diagnosis has been reported in upto 30% cases in different series^{6,9}. The usual presentation is a slow growing mass at the angle of the mandible. Sometimes these tumours are confused with enlarged lymph nodes, branchial cyst, salivary gland or neurofibroma. The patients are consequently subjected to unwise attempts at biopsy or explorative surgery. Our patient also had similar history. Additional investigations are mandatory before a surgical exploration is attempted. Exploration & biopsy can be disastrous & should be avoided in the management of carotid body tumour, while FNAC is usually inconclusive^{7,8}. Colour duplex & angiography are the mainstay of diagnosis. The colour duplex, a non invasive procedure has high specificity & sensitivity & can be used for the diagnosis, follow up & also for screening of high risk patients^{6,7}. Angiography is the gold standard & it shows a highly vascularised mass causing typical splaying of the external & internal carotid arteries (Lyre sign). Angiography should be performed on both sides, as in 5-10% cases the tumour is bilateral. CT scan & MRI are usually performed to evaluate the intracranial extent & involvement of the cranial nerves^{6,8}. Catecholamine screening can be done if endocrinological activity in the tumour is suspected on the clinical grounds.

Although resection of carotid body tumour had long been considered a surgical challenge, surgery is the treatment of choice for these tumours. Initial attempts at surgical resection yielded devastating results with very high rates of mortality, cerebrovascular accidents & cranial nerve palsies. First surgical excision was reported by Maydl in 1886, but unfortunately the patient became hemiplegics & aphasic after the operation¹. First successful excision of the carotid body tumour was done by Albert in 1889¹. The persistent high morbidity & mortality associated with the surgery led Martin et al to state in 1957 that these tumours should only be subjected for biopsy. With the advancement in the field of vascular surgery, good peri operative monitoring & preoperative evaluation, the scenario has changed reducing the mortality to zero & stroke to 1%-3%. Cranial nerve palsies still remains one of the major complications, which can occur in 10-40% cases⁶. Tumour size is important because those greater than 5 cm in diameter have a markedly higher incidence of complications ®C

67% vs 15% for less than 5cm (McCaffrey 1994). A classification system based on size & difficulty of resection has been developed (Shamblin 1971), group 1 tumours are small & easily dissected away from the vessels. Group 11 includes paraganglioma of medium size but are separable with careful sub adventitial dissection. Group 111 tumours are large & typically encase the carotids, requiring partial or complete vessel resection & replacement. Our tumour belonged to group 11. Pre operative embolisation of the carotid body tumour has been reported in the literature. The advantage of this therapy is that the devascularised lesion can be removed relatively bloodlessly, which facilitates visualization surrounding structures & lessens the need for transfusion of biological products^{6,10}. This is particularly true for the large tumours. Most authors have concentrated on embolisation with particulate matters (PVA, gelfoam) or glues. Another possible role of angio-embolisation might be palliative use for unresectable tumours¹. Another unresolved therapeutic issue is the benefit of radiotherapy in these tumours. Radiotherapy has been used in patients with metastatic lesions or tumours presumed to be malignant in certain sites⁸. As it is very slow growing tumour with the growth rate of less than 0.5cm per year, small tumours in old patients, with significant risk factors for surgical interventions can be managed by observation only ⁸.

CONCLUSION:

We conclude that pre operative embolisation is an important adjunct in treating patients with large carotid body tumours. The surgical exploration precedes much smoother, the blood loss minimal & the patients have minimal morbidity.

The association of situs inversus totalis & the carotid body tumour seems to be incidental; however with the extensive search in the literature, we have not found such a case.

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