CAROTID BODY CHEMODECTOMA AND CAROTID ANEURYSM – A CASE ILLUSTRATION OF DIAGNOSTIC PROBLEMS.

By

[1]DR OLUWOLE A. OGUNLEYE, [2]PROF. TAIWO A. IJADUOLA, [3]DR ADEREMI A. ADEOSUN, [4]DR ABIODUN O. ADEYINKA, [5]DR MOJISOLA ATALABI,

[1,2,3]Department of Otorhinolaryngology, [4,5]Department of Radiology, College of Medicine, University of Ibadan, and University College Hospital, Ibadan, Nigeria.

Correspondence:

Dr A.O A. Ogunleye
Department Of Otorhinolaryngology,
University College Hospital,
P.M.B. 5116, Ibadan, Nigeria.
234-02-2410088; 2411099 Ext 2620; 2773
Fax: 234-02-2411768 E-MAIL: aawole@yahoo.co.uk

ABSTRACT

Carotid body tumours are very rare and occur at the carotid artery bifurcation between the internal and external carotid arteries. The carotid bodies are reddish brown ellipsoid structures, lying embedded in the adventitial of the carotid artery bifurcation.

The usual presentation is a slow growing mass at the angle of mandible. Sometimes these tumours are confused with enlarged lymph nodes, brachial cysts, salivary glands, neurofibromas and carotid aneurysms especially. Thus carotid body tumours can be a diagnostic challenge for a clinician resulting in patients being consequently subjected to unwise attempts at biopsy or explorative surgery.

We present the diagnostic problems of carotid body tumour and carotid aneurysm as seen in a Nigerian African.

KEY WORDS: *carotid body, chemodectoma, carotid aneurysm*.

INTRODUCTION

Carotid body tumours are very rare and occur at the carotid artery bifurcation between the internal and external carotid arteries. The carotid bodies are reddish brown ellipsoid structures, lying embedded in the adventitial of the carotid artery bifurcation. [1, 2,

Specifically they detect changes in arterial partial pressure of oxygen and carbon dioxide and changes in P^H and other blood-borne factors. Accordingly it can increase or decrease stimulation to the respiratory centres of the brainstem, which affects various cardiopulmonary functions including blood pressure, heart rate and respiration.

Haller introduced glomus tumours of the head and neck into the medical literature in 1762 when he described a mass at the carotid bifurcation that had a glomus body – like structure and these carotid bodies are derived from the epitheliod cells of neuro-ectodermal origin.[3]

Tumours of this tissue were originally described as chemodectomas by Mulligan in 1950; now they are considered as a part of the widely described group of tumours known as Paragangliomas on the basis of its anatomic and physiologic characteristics.[1]

The actual aetiology of these tumours is not known but they are noted to be more common in people living at high altitudes and some studies have shown that long exposure to high altitudes appear to be correlated with a 10-fold higher incidence of carotid body tumours but no increase in the incidence of paragangliomas at other sites.[4] An insidious link appears to exist between oxygen deprivation and glomus tumour incidence. Other studies are underway to explain the effects of smoking and other sources of long-term anoxia.

Carotid body tumours can be a diagnostic challenge for the clinician and lack of pre-operative diagnosis has been reported in up to 30% of the cases in different series.[4,5]

The usual presentation is a slow growing mass at the angle of mandible. Sometimes these tumours are confused with enlarged lymph nodes, brachial cysts, salivary glands, neurofibromas and carotid aneurysms especially.

The patients are consequently subjected to unwise attempts at biopsy or explorative surgery [6].

The aim of this article is to present the diagnostic problems of carotid body tumour and carotid aneurysm as seen in a Nigerian African.

CASE ILLUSTRATION

A 53-year-old male taxi driver, presented to our E.N.T. Department with a 4-year history of progressive, painless, pulsatile left lateral cervical swelling. There were no associated traumas, fever, heat/cold intolerance or tremors.

No associated ear, nasal nor throat symptoms. Patient is not a known hypertensive and has no significant history of alcohol consumption or smoking. A month prior to presentation, he developed progressive blurring of vision of the left eye (ipsilateral to the cervical mass).

Attempt at biopsy in a private hospital prior to presentation was haemorrhagic and non-representative. General examinations were essentially normal including the vital signs.

Ear, Nose and Throat examinations appeared normal.

The neck examination revealed a left lateral cervical oblong – shaped 14cm x 12cm mass occupying the posterior triangle of the neck and extending to the left anterior triangle in its horizontal dimension.

It spans vertically from the supraclavicular fossa to the angle of the mandible. The surface is smooth with scar at the mid point (following biopsy), pulsatile with scattered bruit, no differential warmth and neither moves with swallowing nor tongue protrusion. It has free palpable margins above and below.

The cervical mass move freely in the horizontal plane but not in the vertical plane. All the cranial nerves appeared normal except the left optic nerve (there was blurring of vision), which was diagnosed by the Ophthalmologist as secondary optic atrophy.

The general investigations that included full blood count, electrolyte/urea and chest radiograph were within normal limit.

Doppler ultrasonography revealed normal artery on the right but with an irregular saccular dilation at the terminal portion of left common carotid artery with evidence of

aliasing flow pattern and a well circumscribed homogenous solid hyper echoic mass encompassing the carotid at region of dilation with a conclusion of aneurismal dilation of left common carotid artery (Dissecting aneurysm).

Enhanced C-T scan of the neck revealed left carotid aneurysm with intra and extra mural massive haematoma.

However, spiral computed tomographic scan with angiography of the neck revealed a large left soft tissue mass (7.9cm x 6.6cm) of the neck extending from the neck root to the mandibular angle (figure 1). The mass is seen to have splayed the left internal and external carotid arteries at the point of bifurcation, thus distorting both vessels with numerous collateral formations around it (figure 2). There appears to be pooling of contrast within the mass. The right carotid vessels showed normal caliber, course and branches. The circles of Willis series for both cerebral and cerebellar hemispheres are within normal limit and thus concluding that all these findings were highly suggestive of carotid body tumour.

Following the above diagnosis and considering the size of the tumour, and the likely risk of vascular damaged to the speech centre following surgery, the risk patient declined, patient was offered radiotherapy of 45 Gy in 20 fractions over 4 weeks. Post radiotherapy period had been satisfactory with regression of the tumour size considerably.

DISCUSSION

Carotid body tumours are very rare neoplasms constituting less than 0.5% of all body tumours.[7] Sporadic forms of carotid body tumours are more frequent while familial forms account for about 10% of the cases in most series and the tumours are bilateral in 30% of the familial, but only 5% of the sporadic cases. [8,9]

Carotid body tumours are usually benign but the malignant potential with possible metastasis has been estimated to be around 2% to 9% and the usual histologic criteria for malignancy (i.e. nuclear atypia and nuclear to cytoplasmic ratio) do not apply. [10,11]

The true proof of malignancy is the presence of lymph node or distant metastases, which may not become evident even years after the original resection. Familial, bilateral or multiple paragangliomas in young patients are more prone to aggressive behaviour and these tumours can metastasize to the lungs.[7]

Most of the carotid body chemodectomas are asymptomatic in the early clinical phase. Eventually at least 75% of the patients develop symptoms such as enlarging neck mass, neck pain, hoarseness or syncope and the physical examination usually reveals a pulsatile mass below the angle of mandible which can be moved laterally but not vertically (Fontaine's sign)[7]

In this case illustration, the condition presents as a painless, slow growing, enlarging mass, pulsatile and with bruits and these features are pointing towards carotid aneurysm. The only feature that was against the diagnosis of carotid aneurysm was the presence of Fontaine's sign, which is a characteristic of carotid body chemodectoma.

Furthermore all the specific investigations done (i.e. Doppler ultrasonography, enhanced CT scan) were pointing strongly to carotid aneurysm except the spiral CT angiography findings that actually revealed features that were highly suggestive of carotid body tumour in this case, thus creating diagnostic problems between carotid aneurysm and carotid body chemodectoma. The evidence weighed more toward carotid body chemodectoma with all the features seen on the spiral CT angiography of the neck of this patient (Figure 1, 2)

Imaging is the primary investigative modality for glomus tumours of the head and neck. A combination of contrast-enhanced CT, MRI and angiography is ideal for proper diagnosis and localization of the tumours.

Lesions show a characteristic signature on images, which is based on it's location as seen in this case with splaying of both internal and external carotid arteries at its bifurcation (common carotid artery) and intense vascular blushing.

Currently, MRI is frequently the imaging study of choice for primary diagnosis, followed by contrast – enhanced CT imaging. Angiography remains of paramount importance if the diagnosis is obscure or if embolization is contemplated.

The treatment options include surgery, radiotherapy and angio-embolization and the decision to operate depends upon the age of the patient, symptoms, co-morbid conditions, rate of growth and the size of the tumour [12].

Chemodectomas of carotid artery bifurcation are generally managed with surgery, irradiation being reserved for an in-operable, bulky and recurrent tumours, probably due to this pre-treatment selection of patients chemodectomas are anecdotally considered radio resistant tumours, this concept is no longer supported by the recent literature as it has been shown that radiation appears to be effective in achieving long term clinical control of chemodectomas. [13 – 17]

In this case radiotherapy was offered to this patient due to high tumour size with its extension into the internal carotid artery unto the skull base on the left side and the likely risk of vascular damaged to the speech centre following surgery on the tumours, the risk the patient declined. The patient received Forty-five (45) Gray in 20 fractions over 4 weeks; post radiotherapy period had been satisfactory with minimal morbidity and with tumour regression considerably.

It is concluded that a number of diagnostic approaches have been suggested for the management of carotid body chemodectomas, but they continue to pose diagnostic challenges to clinician.

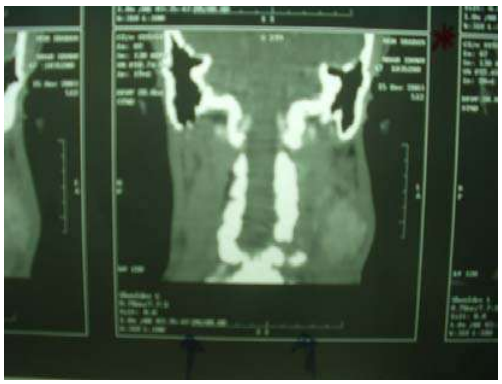

FIG. 1: Spiral computed tomographic scan with angiography of the neck showing a large left soft tissue mass.

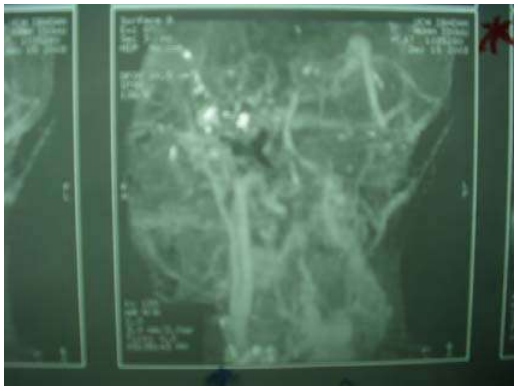

FIG. 2: The mass is seen to have splayed the left internal and external carotid arteries at the point of bifurcation, thus distorting both vessels with numerous collateral formations around it.

REFERENCES

1. McPherson G.A.D., Hallidary A.D. MansField A.O. Carotid body tumours and other cervical paragangliomas: diagnosis and management in 25 patients. Br. J. Surg. 1989; 76:33-6

2. Khan Q., Heath D., Smith P. Anatomical variations in human's carotid bodies. J. Clin. Pathol 1988; 41:1196-9

3. Heath D. The human carotid body in health and disease. J. Pathol 1991; 164:1-8

4. Westerbad A; Amuter G.C., Cintara I.; et al. Current trends in the detection and management of carotid body tumours. J. Vascul Surg. 1998; 28:84-93

5. Saldana M.J., Salem L.E., Travezan R. High altitude hypoxia and chemo dectomas. Hum Pathol. 1973; 4:251-63

6. Joshi R.M.; Tapia A.A.; Wagle P.K., Mavadgi S.M., Bern Z.T. Carotid body tumour. Bombay Hospital Journal. 2000;42: 419-20

7. Coffrell E.D., Smith L.L. Management of uncommon lesions affecting the extra cranial vessels. In: Rutherford R.B., (ed). Vascular Surgery Philadelphia: W.B. Saunders, 1995:1623-27

8. Muhm M., Poltaraner P., Castottner W. et al. Diagnostic and Therapeutic approaches to carotid body tumours. Arch. Surgery 1997; 132: 279 -84

9. Bernard R.P., Carotid body tumours. Am J Surg. 1992; 163:494-96

10. Khiewer K.E., Wen D., Cancilla P.A. et al Paragangliomas: Assessment of prognosis by histologic immuno histochemical and ultra structural techniques.
Hum Pathol 1989; 20: 29 -39

11. Barnes L., Taylor S.R., Carotid body Paragangliomas: a Clincopathologic and DNA analysis of 13 tumours. Arch Otolaryngol Head and Neck Surg. 1990; 116:447-53

12. Dickson P.H., Griffin S.M., Guy A.J. Carotid body tumour: 30 years experience. Br. J Surg. 1986; 73: 14-16

13. Valdagni R., Amichetti M. Radiation therapy of carotid body tumours.
Am J Clin, Oncol. 1990; 13:45-8

14. Schild S.E., Frose R.L., Buskirk S.J. et al. Results of radiotherapy for chemodectoma. Mayo Clin Proc. 1992; 67:537-40
Ogunleye Carotid body chemodectoma

15. NMumber M.P., Grevan K.M. Control of advanced chemodectomas of the head and neck with irradiation. Am J Clin Oncol. 1995; 18:389-91

16. Evension L.J., Mendenhall W.M., Parsons J.T., Cassisi M.J.Radiotherapy in the management of chemodectomas of the carotid body and glomus vagale.
Head Neck. 1998; 20:609-13

17. Hinerman R.W., Mandanhall W.M., Amdur R.J. et al Definitive radiotherapy in the management of chemodectomas arising in the temporal bone, carotid body and glomus vagale. Head Neck. 2001; 23:363-71