

Neck trauma: a red-herring to diagnosis of chemodectoma

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Abstract

Chemodectoma, a neuroendocrine tumour of the paraganglionic cells in the carotid body remains an uncommon tumour. We report the first case from University of Ilorin Teaching Hospital, Ilorin, Nigeria. Though with a red herring history of trauma induced swelling, clinical and radiologic features were characteristic of chemodectoma. Histologic features of the excised lesion are presented.

Keywords: *Chemodactoma; paraganglioma; carotid body tumour; neck swelling; neck trauma*

Résumé

La chémodectome, une tumeur neuroendocrine des cellules para-ganglionnaires du corps carotidien demeure une tumeur rare. Nous rapportons le premier cas provenant de l'Université d'Enseignement Hospitalier d'Ilorin, Ilorin, Nigeria. Bien qu'avec une histoire d'harengs saurs de dilatation induite par le trauma, les caractéristiques cliniques et radiologiques étaient caractéristiques de la chémodectome. Les caractéristiques histologiques de la lésion excisée sont présentées.

Mots-clés: *chémodectome; para-gangliome; Corps de tumeur carotide; dilatation du cou; traumatisme du cou*

Introduction

Von Haller (1743) was credited with the first gross description of the carotid body which he termed ganglion minutum [1,2]. Neoplasm of the carotid body was named chemodectoma by Mulligan in 1950 and paraganglioma by Glinner and Grimley in 1974 [1,3]. In 1862, von Luschkar provided its microscopic description [1].

Rarity of this tumour, as reported in most literature [4-6] seems to spur confusion with other more common causes of neck swelling. However, with the development of advanced non-invasive radiological diagnostic techniques, more precise preoperative diagnosis can be made.

Surgical excision, which remains the treatment of choice, was first performed in 1880 by Riegner though the first successful resection was

done six years later by Madyl but the patient had postoperative aphasia and hemiplegia [1,2]. The first uncomplicated excision was performed by Scudder in 1903 [2].

Case Report

A 28 year old lady who presented with left sided anterior neck swelling of two months duration which was noticed following being hit on that part of the neck during a fight. The swelling was not painful. She had two episodes of exercise-induced syncopal attacks, and a persistent tinnitus. However, there was no impairment of hearing, vertigo, hoarseness of voice, visual impairment, or abnormality of the muscle or sensory performance of the surrounding muscle and skin. There was no history of weight loss and no history suggestive of tuberculosis. She had no similar swelling in any other part of her body.

On examination, a left-sided neck swelling was seen in the anterior triangle; oblong shaped, smooth, firm, non tender, pulsatile, and un-attached to adjacent muscles or skin. It measured 6×8cm in its longest dimensions, and had well defined edges. Fontaine's sign was positive and bruit was heard over it.

Plain radiograph of the neck demonstrated soft tissue swelling over the left lateral aspect. The ultrasound showed a hyperechoic oval mass at the bifurcation of the left common carotid artery which was highly vascularized on Doppler evaluation. The ipsilateral internal jugular vein was displaced laterally. A diagnosis of highly vascularised left carotid body tumour was made. This was confirmed with computerized tomographic angiography [Fig 1].



Fig.1: Post contrast CT angiography showing a well vascularised mass in the left side of the neck (red arrow)

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She had surgical excision via a longitudinal incision along the anterior border of the sternocleidomastoid muscle. Tumour was firmly adherent to the carotid artery particularly at its bifurcation (Shamblin grade III). The external carotid and distal internal jugular vein were completely embedded in the tumor hence were sacrificed. The internal carotid artery was anastomosed to the common carotid artery end-to-end using 4/0 prolene suture in order to maintain cerebral supply. The distal segment of the tumour which extends into the base of the skull and encasing the distal internal carotid artery was left in-situ. A corrugated drain was inserted and was removed on the 10th day postoperative when effluent reduced satisfactorily.

She had a smooth recovery following surgery and was discharged on the 15th postoperative day. She was referred to another teaching hospital for radiotherapy where she completed the third session eight months following surgery. She has been followed up for two years with no evidence of recurrence.

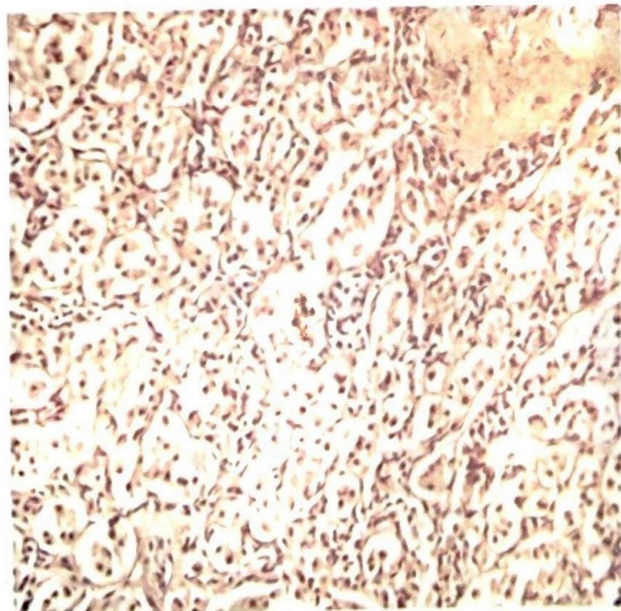


Fig 2a: Chemodectoma showing the alveolar (Zellballen) arrangement of the chief cells and thick fibrous band (H&E x 400)

Histology revealed highly vascularised tumor, essentially composed of the two cell types, chief and sustentacular (supporting) cells arranged in organoid (Zellballen) and peritheliomatous patterns [Fig 2a]. The capsule showed varying degree of thickness and in areas contained nerve fibers. In the organoid pattern the tumour cells were polyhedral, epitheloid cells with anisonucleosis and pale to fairly granular eosinophilic cytoplasm having capillary networks around each group of cells, whereas the

peritheliomatous pattern was composed of smaller cells with darkly stained nuclei and indistinct cytoplasm [Fig 2a]. Immunohistochemically, the sustentacular cells stained positively for S-100 protein while the chief cells component of the tumour was unstained [Fig 2b].

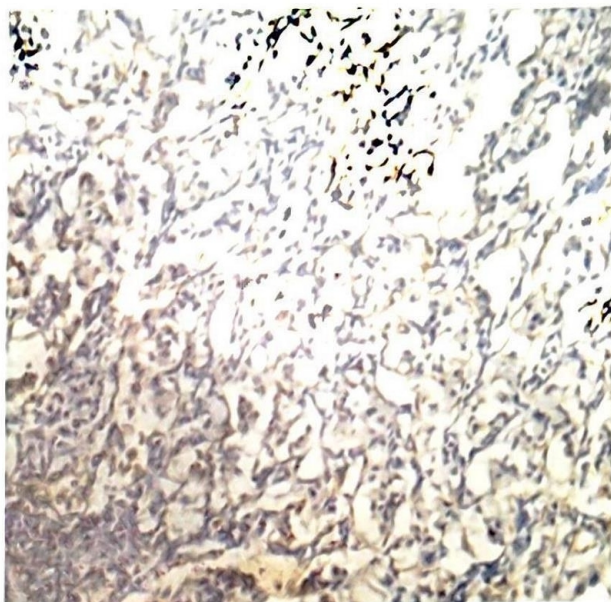


Fig 2b: Chemodectoma showing Sustentacular cells immunostained positively for S-100 protein (Immunohistochemistry x400)

Discussion

Chemodectoma also known as carotid body paraganglioma though a rare tumour, accounting for 0.5% of all tumours; is the most common type of paraganglioma of the head and neck accounting for 60-70% [2,7]. Chronic hypoxia has been identified as major etiologic factor hence sleep apnoea and high altitude abode may predispose to chemodectomas [2,4]. Echejor *et al* reported 3 cases in one year from Jos, which is located in a high altitude region in Nigeria [1]. It is commoner in females with a male to female ratio of 1:2 and present in a wide age range of 10-75 years [7].

Clinically, the presentation is that of a slow growing rounded neck mass located anterior to the sternocleidomastoid near the angle of the mandible at the level of the hyoid bone and usually asymptomatic [8]. Characteristically, the tumour is more freely moveable horizontally than vertically owing to attachment to carotids (Fontaine's sign) [4]. It is usually pulsatile with a bruit as seen in the index patient [7]. Imaging is important in its diagnosis to demonstrate its size, extent and its dense vascularity

On sonography, Chemodectomas present as a well-defined solid hypo echoic mass located at the carotid bifurcation with characteristic splaying of the internal carotid artery and external carotid artery. This

is described as the 'Lyre sign' [9,10] On colour Doppler imaging, the tumour's hypervascularity with a low resistance flow pattern is seen [9].

Contrast enhanced CT is excellent at depicting these lesions with typical appearances being that of a soft tissue dense mass on non-Contrast CT which shows as bright and rapid enhancement on contrast injection [10].

On MRI, the T₁ weighted sequence shows as hypointense to isointense mass when compared to the surrounding muscle. A salt and pepper appearance is seen when the mass is large. This represents a combination of punctate regions of haemorrhage or slow flow (salt) and flow voids (pepper). It also shows intense enhancement on post gadolinium series [10]. T₂ MRI sequence however, shows hyper intense mass when compared to the surrounding muscle and also with salt and pepper appearance. Both CT and MRI provide good information about the extension of the tumour allowing for good surgical planning. Digital subtraction angiography is crucial in showing vascularisation details of the tumour. The splaying of the carotid vessels (Lyre sign) is again identified with an intense blush in the tumour with an early venous phase seen due to arteriovenous shunting [2,10].

Scintigraphy, although may not be specific shows uptake with metabenziodobenzylguanidine (MIBG) can be useful for assessing multiple lesions [10]. Despite presence of characteristic features on routine haematoxylin and eosin staining on histologic examination, the presence of S-100 in sustentacular cells on immunohistochemistry provides a significant feature which differentiates paragangliomas from other forms of small cell carcinoma and neuroendocrine neoplasm such as carcinoid tumour [11].

Surgical excision is the treatment of choice. Although the highly vascular nature and strategic location make complete resection difficult [12]. Shamblin classification is used to predict difficulty in resection [4,6]. Small, minimally attached lesions are in class I and pose minimal challenge to resection. Class II lesions are larger, moderately adherent and particularly encircle the carotid bifurcation. Class III lesions, as seen in this case are larger, densely adherent and encase the carotid vessels. In this later group, resection is difficult and often complicated. Ligation and resection of the external carotid artery with the tumour is recommended once there is transmural invasion of the vessel as we opted to do [6]. Re-establishing continuity of the internal carotid artery using saphenous vein graft could not be performed in this case due to presence of tumour in its distal segment [6] hence referral for radiotherapy

which is recommended in cases of incomplete resection or in patients in whom the risk of complication preclude surgery [13].

Though earlier report suggest that chemodectomas are non-metastasizing, [3] recent documentation indicate malignant transformation rates of up to 50% with possible metastasis to the lungs, lymph nodes, liver and bones [2]. Life long surveillance for fear of recurrence and metastatic spread is therefore mandatory.

We therefore highlight the uncommonness of chemodectoma and vigilance with thorough examination is necessary particularly with a history swaying from the diagnosis.

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