

CASE REPORT

Chemodectoma on the Highland of Jos, Nigeria: Three Cases with Review of the Literature

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ABSTRACT

Chemodectoma (paraganglioma, glomus tumour or carotid body tumour) is an extra-adrenal chromaffin tumour, which originates from neuroectodermal cells of the autonomic nervous system. To date, this tumour has been a rare diagnosis. Herein, three cases of chemodectoma encountered during a one-year period are presented. A brief review of literature on this topic is also provided. *Biomed. Int.* 2011; 2: 32-35 ©2011 Biomedicine International, Inc.

Key Words: Carotid body, Chemodectoma, Glomus tumour, Nigeria, Paraganglioma

INTRODUCTION

Chemodectomas (Paragangliomas, Glomus tumours, Carotid body tumours) are extra-adrenal chromaffin tumours that develop at the expense of neuroectodermal cells of the autonomic nervous system. In 1743, von Haller first reported a gross description of a structure lying at the bifurcation of the carotid artery, which he termed the "ganglion minutum". The microscopic appearance of this organ was described by von Luschka in 1862, and the first tumour of this body was surgically removed by Riegner in 1880 and described by Marchand in 1891. In 1950, Mulligan renamed this type of neoplasm a *chemodectoma* to reflect its origin from chemoreceptor cells. In 1974, Glenner and Grimley renamed the tumour *paraganglioma* on the basis of its anatomical and physiological characteristics. Presentation is normally history of a slowly-growing mass. Most cases are benign with scant record of malignant variants. There is no report of chemodectoma from this Centre in the literature.

We present three cases of chemodectoma seen in this Centre within one year.

CASE REPORTS

Case 1

HT is a 34-year-old female who came to the surgical outpatient clinic on 10th January, 2009 with a 9-year history of a slowly-growing painless mass on the left side of the neck. The patient came to the hospital because the mass had grown large and had become painful in the last 6 weeks before presentation. On examination, she was not critically ill-looking, not pale, and afebrile. Her vital signs were essentially normal. On the left side of the neck was a firm mass of 10 x 6 cm dimension with mild tenderness. The mass was not freely mobile but was not attached to the overlying skin. The immediate impression was of lymphadenopathy (? lymphoma). After preliminary laboratory work, a wedge incision biopsy was sent to the histopathology laboratory in formalin-saline. Histology showed nests of ovoid cells having ovoid to round basophilic nuclei and moderately eosinophilic cytoplasm. There was a network of thin walled capillaries within the surrounding fibrous stroma. There were no mitoses. On the basis of these features, a histological diagnosis of chemodectoma was made. The patient was booked for definitive surgical excision but declined on financial grounds and was lost to follow up.

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Submitted May 6, 2010; accepted in revised form June 2, 2010.

Advance Access Publication 15 May, 2011 (see www.bmijournal.org)

Case 2

MA is a 29-year-old male with a right-sided cervical swelling of two years' duration. The mass was painless and had been increasing in size gradually over the period. No associated fever, no weight loss and no night sweats were reported. He coughed occasionally but this was not productive of sputum. Physical examination showed a healthy-looking young man with a firm, non-tender mass of 5x2 cm on the right side of the neck in the anterior triangle. The patient was therefore prepared for excision biopsy, which revealed a firm encapsulated mass at the carotid artery bifurcation. The tissue was excised and preserved in 10% formalin-saline, after which it was sent for histopathological review. The histological features were as in case 1 above (Fig. 1 and Fig. 2).

Case 3

She is a 56-year-old woman who was referred to the surgical team with a long (10 year) history of polypoid growth in the left ear, which was being managed locally with herbs/medicaments. There was associated tinnitus, vertigo and limited hearing ability in that ear. She decided to come to the hospital (August, 2009) when she noticed ulceration around the ear. Examination revealed an ulcerated mass with de-

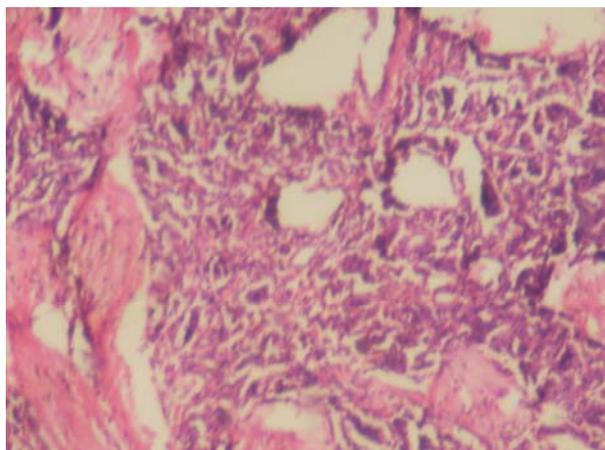


Fig. 1. Nests of ovoid cells delineated by delicate fibrous stroma and vascular network. 20X objective

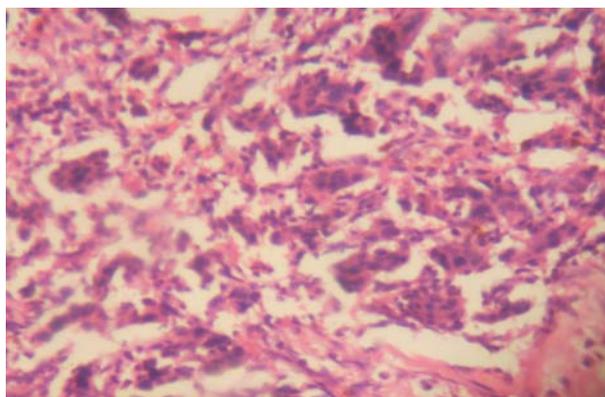


Fig. 2. Nests of ovoid cells delineated by delicate fibrous stroma and vascular network. Higher magnification. (40X objective)

struction of the base of the left ear pinna and erosion of the mastoid bone and overlying skin. She was pale and ill-looking. She was therefore admitted. Incision biopsy showed nests of ovoid cells delineated by delicate fibrous stroma and a vascular network. Histological diagnosis of paraganglioma was made and she was billed for surgical excision and plastic surgery. However, her children took her home against medical advice on financial grounds.

DISCUSSION

Chemodectomas are believed to result from an over-response to a change in homeostasis. There is a relationship between oxygen deprivation and the incidence of chemodectomas. Compensatory hypertrophy of the carotid body is known to occur only in patients with prolonged hypoxia and hypercapnia. Some studies have shown that long exposure to high altitudes is correlated with a 10-fold higher incidence of carotid body tumors.¹ Our region is a high altitude plateau; however, we have not seen cases in our Centre previously. Could it be misdiagnosis, or are the patients - who are mostly from the rural poor setting - living with these mostly benign tumours? Two of the three cases left without surgical treatment. The first case, with the huge neck mass, left against medical advice owing to financial constraints and was lost to follow up; the elderly lady in poor condition was taken home by the children, also on financial grounds. Some authors have reported familial cases, accounting for 18% of the tumours in their study.² Chemodectomas usually arise from the carotid bodies but have also been reported in the glomus jugulare, vagus nerve, ganglion nodosum, mediastinum, lungs, abdomen, ciliary body, femoral canal, mandible, retroperitoneal region, and extremities.³ However, studies have shown that chemodectomas of the head and neck are associated with four primary locations, viz:

Jugular bulb: Tumours here are commonly called glomus jugulare tumors.^{4,5,6,7,8,9} These arise in the adventitia of the dome of the jugular bulb. This is the most common type of chemodectoma of the head and neck.

Middle ear cavity: Tumours here are commonly called glomus tympanicum tumors.^{9,10,11,12,13} They arise from the glomus bodies that run with the tympanic branch of the glossopharyngeal nerve. Glomus tympanicum tumours are the most common primary neoplasms of the middle ear. One of our cases had a similar lesion that had eroded and ulcerated the mastoid, ear and skin due to late presentation.

Vagus nerve: Tumours in this area are commonly called glomus vagale tumours because of their usually close association with the vagus nerve.^{14,15,16} Specifically, they arise infratemporally along the course of the cervical vagus nerve.

Carotid body: Carotid body glomus tumours, also called carotid body tumours, occur at the bifurcation

of the common carotid artery and arise from the tissue of the normal carotid body.^{3,16,17,18}

Although glomus tumours usually appear as solitary lesions at one site, multiple lesions at multiple sites are not uncommon.¹⁹ Because they are parts of the neuroendocrine system, these tumours are highly vascularized. Clusters of tumour cells (type I cells interspersed with type II cells), called *zellballen*, are surrounded by a dense network of capillary-calibre blood vessels. Functioning tumours, which are rare, can increase the risk of mortality. These active tumours secrete catecholamines, which can lead to clinical manifestations of hypertension, headaches, palpitations, and tachycardia.²⁰

Sajid et al. reported 4.2% malignant tendency of these tumours. Along with other researchers, they also reported female preponderance.^{21,22} The tumour is rare in children, and has a peak age of incidence between 45 and 60 years.¹ Mortality rates from these tumours are said to be in the range 9-15%, depending on the location of the tumour and the study. 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 location of the tumours.²²

Complete resection is the standard of care for paraganglioma cases, affording the patient with the best chance of cure since these tumours are relatively resistant to chemotherapy and irradiation.²³ Their highly vascular nature and strategic anatomical locations make complete resection demanding.²⁴ In most cases, the surgical approach is tumour-site-dependent. In some cases these tumours can be removed in a single-stage operation. However, a recent report described a two-stage approach for resection of a paraganglioma of the mediastinum invading the pulmonary artery and ascending aorta.²⁵

The major concerns involving resection of paragangliomas include intraoperative bleeding and catecholamine crises in patients with metabolically active tumours. The highly vascular nature of the tumour, and the proximity to and invasion of the great arteries, contribute to a high risk of bleeding.²⁶ Hormonal-related crises are uncommon but are associated with significant morbidity and mortality.²⁷ Meticulous surgical technique and tight preoperative blood pressure control are the key steps in prevention and management of these complications.²⁸

Prognosis after complete resection is favourable. Lamy et al reported a follow up of 79 patients with middle mediastinal paragangliomas over a period of 180 months. Among these patients overall survival was 62.0%, and mean survival time was 98.2 ± 11.7 months. For patients undergoing complete resection survival was 84.6%, and mean survival time was 125.7 ± 18.7 months. For patients undergoing incomplete resection survival was 50.0% and mean survival time was 71.5 ± 13.8 months.²³ Complete surgical resection remains the standard of care and is associated with excellent survival. Life-long surveillance

for local recurrence and metastatic spread is mandatory.²⁹

CONCLUSION

This study shows that chemodectoma is not rare on the highland of Jos, Plateau State. The literature shows that complete tumour resection remains the standard of care.

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