

## Case Report

# Chemodectoma: three case-series with review of literature

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Accepted 21 May, 2011

**Chemodectomas (Paragangliomas, Glomus tumours, Carotid body tumours) are extra-adrenal chromaffin tumours. Historically, in the year 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 as a chemodectoma to reflect its origins from chemoreceptor cells. In 1974, Glenner and Grimley renamed the tumour paraganglioma on the basis of its anatomic and physiologic characteristics. Presentation is normally history of a slow growing mass. Most cases are benign with scant record of malignant variants. There is no report of chemodectoma from this centre in literature. We present three cases of chemodectoma seen in this centre within one year.**

**Keywords:** Chemodectoma, paraganglioma, glomus tumour, neck mass, highland, plateau

## INTRODUCTION

Chemodectomas (Paragangliomas, Glomus tumours, Carotid body tumours) are extra-adrenal chromaffin tumours. Historically, in the year 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 as a chemodectoma to reflect its origins from chemoreceptor cells.

## CASES PRESENTATION

### Case 1

HT is a 34- year-old female who came to the surgical outpatient clinic on 10<sup>th</sup> January, 2009 with a 9-year history of a slow 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

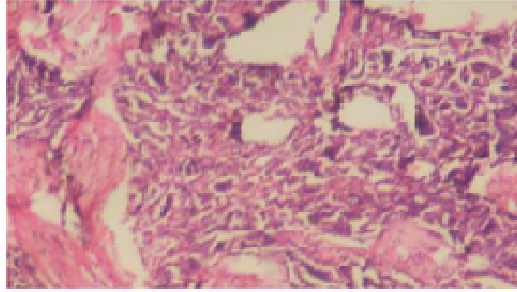
signs were essentially normal. On the left side of the neck was a firm mass of 10 x 6cm dimension with mild tenderness. The mass was not freely mobile but was not attached to the overlying skin. An impression of lymphadenopathy (? Lymphoma) was made. After preliminary laboratory work up, a wedge incision biopsy was done and sent to the histopathology laboratory in formal saline. Histology showed nests of ovoid cells having ovoid to round basophilic nucleus and moderate eosinophilic cytoplasm. There was network of thin walled capillaries within the surrounding fibrous stroma. There were no mitoses. Based on these features, a histological diagnosis of chemodectoma was made. The patient was booked for definitive surgical excision but declined on financial ground and was lost to follow up.

### Case 2

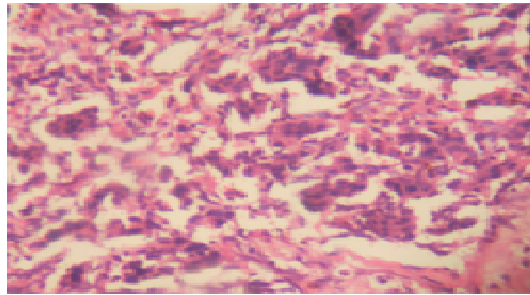
MA is a 29-year-old male with right sided cervical swelling of 2 years duration. The mass was painless and had been increasing in size gradually over the period. No associated fever, no weight loss nor night sweats. He coughed occasionally which was not productive of sputum. Physical examination showed a healthy looking young man with a firm, non tender mass of 5x2cm on the right side of the neck in the anterior triangle. The patient was therefore prepared for excision biopsy which turned out to be a firm encapsulated mass at the carotid artery

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**Figure 1 a:** Nests of ovoid cells delineated by delicate fibrous stroma and vascular network. 20X objective



**Figure 1 b.** Nests of ovoid cells delineated by delicate fibrous stroma and vascular network. Higher magnification. (40X objective)



**Figure 2a.** A greyish-brown ovoid mass (chemodectoma) from the carotid bifurcation in a 29-year-old man



**Figure 2b.** Cut section of the mass in 2a.

bifurcation. The tissue was excised and preserved in 10% formal saline after which it was sent for histopathological review. The histological feature was as in case 1 above (Figures 1a and b). Figures 2a and b show the gross feature of the mass.

### Case 3

She is a 56-year-old woman who was referred to the surgical team with a long (10 years) history of polypoid growth in the left ear that was being managed locally with herbs/medicaments. There was associated tinnitus, vertigo and limited hearing ability from that ear. She decided to come to the hospital (August, 2009) when she noticed ulceration around the ear. Examination revealed an ulcerated mass with destruction of the base of the left ear and erosion of the mastoid bone and overlying skin. She was pale, ill looking and hypertensive (Blood pressure 180/100mmHg). She was therefore, admitted. Incision biopsy showed nests of ovoid cells delineated by delicate fibrous stroma and vascular network. Histological diagnosis of paraganglioma was made and was billed for surgical excision and plastic surgery. However, her children took her home against medical advice on financial ground.

## DISCUSSION AND REVIEW OF LITERATURE

This report shows the presence of chemodectoma in our region, and relatively prevalent with the diagnosis of three cases in one year. Chemodectomas are believed to result from an over response to a change in body homeostasis. There is a relationship between oxygen deprivation and chemodectoma incidence. 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 appears to be correlated with a 10-fold higher incidence of carotid body tumors (Koenigsberg et al., 2008). Our region is a high altitude plateau; however, we have not been seeing cases in our centre before now. Could it be misdiagnosis or are the patients who are mostly from the rural poor setting living with the mostly benign tumours? Two of the three cases actually left without surgical treatment. The first case with the huge neck mass left against medical advice due to financial constraint and was lost to follow up; the elderly lady with poor condition was taken home by the children also on financial ground. Marlowe et al (1993) reported familial cases accounting for 18% of the tumours in their study. 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 (Victor et al., 1975). However, studies have shown that chemodectomas of the head and neck are associated

with 4 primary locations, viz:

**Jugular bulb:** Tumours here are commonly called glomus jugulare tumours (Patel et al., 1994; Pluta et al., 1994; Pemberton et al., 2005; Varma et al., 2006; Li et al., 2007; Henzel et al., 2007). These arise in the adventitia of the dome of the jugular bulb. This is the most common type of chemodectomas of the head and neck.

**Middle ear cavity:** Tumours here are commonly called glomus tympanicum tumours (O'Leary et al 1991; Cheng et al 1997; Coulier et al., 2003; Pemberton et al., 2005; Salami et al., 2008). 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 similar lesion that 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 usual close association with the vagus nerve (Biller et al., 1989; Cheng et al., 2006; Karatas et al., 2008). 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 (Victor et al., 1975; Smith et al., 2006; Piazza et al., 2007; Karatas et al., 2008).

Although glomus tumours usually appear as solitary lesions at one site, multiple lesions at multiple sites are not uncommon (Som et al., 1996). Because they are parts of the neuroendocrine system, these tumours are highly vascularised. Clusters of tumour cells (type I cells interspersed with type II cells), called zellballen, are surrounded by a dense network of capillary caliber 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 (Vogl et al., 1994).

Sajid et al. (2007) reported 4.2% malignant tendency of these tumours. They also along with other researchers reported female preponderance (Gaffey et al., 1988; Sajid et al., 2007). The tumour is rare in children, and has peak age of incidence between 45-60years (Koenigsberg et al., 2008).

Mortality rates in these tumours are said to be in the range of 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 localization of the tumours (Gaffey et al., 1988).

Complete resection is the standard of care of paraganglioma, affording the patient with the best chance of cure since these tumours are relatively resistant to

chemotherapy and irradiation (Lamy et al., 1994). The highly vascular nature of these tumours and strategic anatomical locations make complete resection demanding (Andrade et al., 2003). In most cases surgical approach is tumour site dependent. In some cases these tumours can be removed in a single-stage operation. However, a recent report by (Qedra et al., 2009) described a two-stage approach for resection of a paraganglioma of the mediastinum invading the pulmonary artery and ascending aorta.

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, the proximity and invasion to the great arteries contribute to a high risk of bleeding (Otake et al., 2006). Hormonal-related crises are uncommon but are associated with significant morbidity and mortality (Ericson et al., 2001). Meticulous surgical technique and tight preoperative blood pressure control are the key steps in prevention and management of these complications (Paul et al., 2007).

Prognosis after complete resection is favourable. (Lamy et al., 1994) 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%, mean survival time was  $98.2 \pm 11.7$  months. For patient undergoing complete resection survival was 84.6%, mean survival time was  $125.7 \pm 18.7$  months. For patient undergoing incomplete resection survival was 50.0% with mean survival time of  $71.5 \pm 13.8$  months (Lamey et al., 1994). Complete surgical resection remains the standard of care and is associated with excellent survival. According to Wald and co-workers (2010), Life-long surveillance for local recurrence and metastatic spread is mandatory.

## CONCLUSION

This study shows that chemodectoma is not rare on the highland of Jos, Plateau state. And the literature has shown that complete tumour resection remains the standard of care.

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