

Carotid Body Tumor Mimicking Chronic Paroxysmal Hemicrania: A Case Report

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Abstract

Carotid body tumor (CBT) also known as chemodectoma/glomus tumor is paraganglioma of the head and neck which is uncommonly identified on routine evaluation. They comprise 0.5% of all head and neck tumors; however, most common among head and neck paragangliomas. The entity is a diagnostic dilemma due to its asymptomatic and uncommon presentations. A middle-aged female patient presented with unilateral episodic pain over the head, neck and facial regions for the last 1 year, associated with periodic orbital pain and ocular congestion, which is similar to the presentation in chronic paroxysmal hemicrania. She had a consultation elsewhere for which she was on routine non-steroidal anti-inflammatory drugs i.e., indomethacin 25 mg. The case was sent for neck ultrasound for a palpable right submandibular lump and was also further evaluated with cross-sectional imaging techniques where almost certain diagnosis of CBT was made. Surgical resection was done, and it was histopathologically confirmed.

Keywords: Carotid body, Hemicranias, Paraganglioma

INTRODUCTION

Carotid body tumors (CBTs) are rare neoplasms; however, they represent nearly 65% of all head and neck paragangliomas.¹ They have neural crest origin and are related with body response to fluctuating concentration of oxygen. The most common clinical presentation of CBTs is as an asymptomatic anterior triangle neck mass; however, features of cranial nerve involvement viz. hypoglossal, glossopharyngeal, or spinal accessory nerve, or involvement of sympathetic chain can be seen in nearly 10%.² Rarely these tumors can present as unilateral continuous/discontinuous headache with accessory features supporting the clinical picture of chronic paroxysmal hemicrania (CPH).

CPH is a unique headache syndrome in which patients have multiple short-lived headaches per day.³ The pain is

always one-sided, very severe associated with symptoms of watering of the eye and eye redness; even drooping of the eyelid has been reported.

CASE REPORT

Our patient was a 35-year-old female with a medical history that included right sided hemifacial pain, headache and neck swelling from last 3 months. There was associated ocular congestion and pain associated with watering from eyes. On physical examination, there was a vertically fixed right sided palpable lump in the submandibular region. She reported having approximately 4 or 5 severe unilateral headache episodes before this hospital visit.

Earlier events consisted of similar clinical symptoms, but there was no visible neck swelling and palpable lump. She had previously undergone a computed tomographic (CT) scan of the brain without contrast material enhancement which was unremarkable. The patient reported that the events were not linked to her position and occurred at different times throughout the day. She had no history of seizures or syncopal attack. She was on treatment for headache with non-steroidal anti-inflammatory drugs

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for approximately 2 months but of no relief. However, symptoms were increasing in frequency.

On day one, several diagnostic tests were performed, and she was sent for neck ultrasonography with color Doppler. Results from duplex Doppler ultrasonography revealed normal carotid and vertebral arteries; it also demonstrated a 2.5 cm nodule that was near the submandibular gland but situated between the bifurcation of the right common carotid artery into the right internal and external carotid arteries (Figure 1). Next day the patient was called for contrast-enhanced magnetic resonance imaging of the neck for lesion characterization using Gadolinium based contrast agent i.e., Gadoversetamide (Optimark - 10.0 ml; 500 mM conc.). The mass demonstrated characteristic "salt and pepper" appearance on T1-weighted image (Figure 2) with intense contrast enhancement and splaying of right internal and external carotid arteries "lyre sign"^{4,5} (Figures 3 and 4). Fat planes with carotid vessels were maintained making the lesion amenable to surgical resection and categorization in Group 1 of Shamblin classification, vide infra.

Surgical resection of the tumor was recommended. At resection, it showed a highly vascularized, solid mass resembling a tuft of capillaries. It was sent to a pathologist for histological analysis. Histological findings were typical of a CBT, with cells palisaded by surrounding blood vessels. There were numerous cell clusters in zellballen formation, separated by a prominent vascular stroma that is pathognomonic for CBTs (Figure 5).

DISCUSSION

CBT is a type of paraganglioma that is a rare tumor arising from the neural crest cells in the carotid body. Other types include based on origin or location are jugular paraganglioma (at jugular bulb), tympanic paraganglioma (arising from the tympanic plexus), and vagal paragangliomas. It is highly vascularized tumor commonly diagnosed at fourth-fifth decades with relative female predilection.^{5,6} There are three different types of CBTs been described in the literature viz., familial, sporadic and hyperplastic. Sporadic form is the most common accounting for 85% of all. Familial form is 10-50%, more common in young individuals. Hyperplastic form is associated with chronic hypoxia and seen in people living at high altitude.⁷ About 5% of CBTs are bilateral, and 5-10% is malignant, but these rates are much higher in patients with inherited disease.⁸⁻¹⁰ Chronic hypoxic conditions, such as patients living at high altitudes or those having chronic obstructive pulmonary disease or cyanotic heart disease, can overburden the carotid bodies and subsequently lead to hypertrophy and neoplasia of the chief cells.¹¹ This condition is seen in the hyperplastic type of CBTs. Familial CBTs are usually

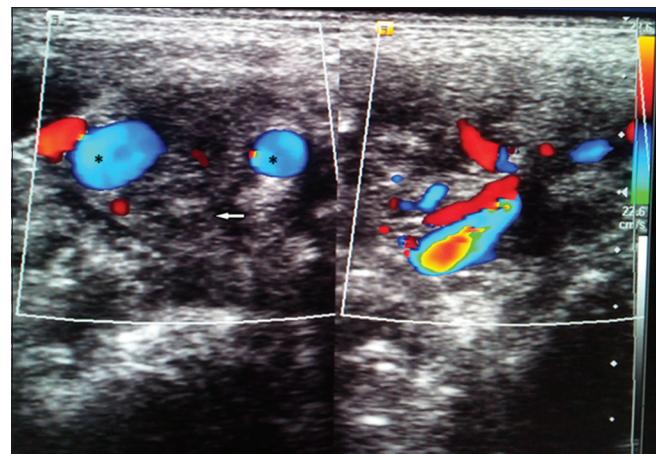


Figure 1: Color Doppler image showing mass (white arrow) at the bifurcation of right common carotid artery causing splaying (*)



Figure 2: Axial T1-weighted image showing the right carotid triangle mass with heterogeneous "salt and pepper" appearance

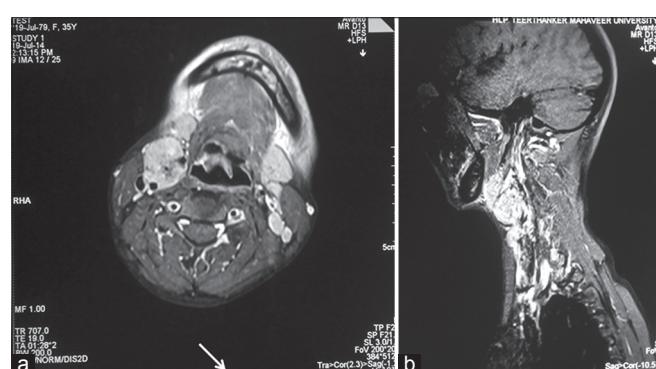


Figure 3: (a) Axial, (b) sagittal post gadolinium magnetic resonance imaging scan showing intense enhancement of the mass

multicentric and are associated with multiple endocrine neoplasia, phakomatoses and Carney's triad.

They are commonly asymptomatic slow growing masses in the anterior triangle of the neck. The doubling time (T_D) of CBTs using sequential imaging, was 7.13 years with a

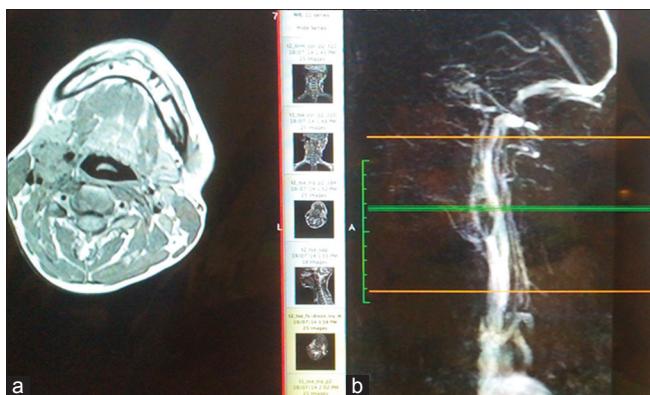


Figure 4: (a) Axial T1-weighted magnetic resonance imaging (MRI), (b) 3D reconstructed MR image showing right-sided mass with splaying of carotids “lyre sign”

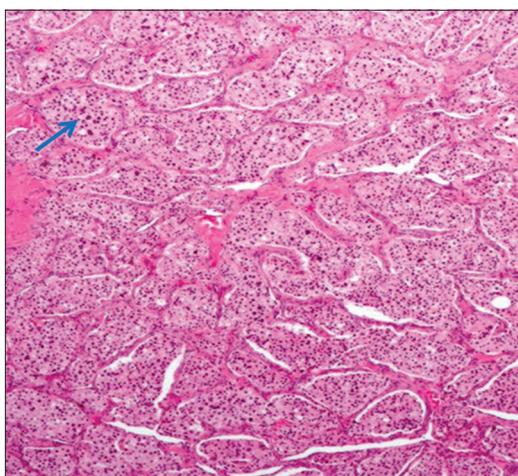


Figure 5: Histological analysis revealed numerous chief cells in the pathognomonic zellballen cluster formation (arrow), separated by a vascular stroma

median growth rate of 0.83 mm/year.¹² On examination, they are vertically fixed because of their attachment to the bifurcation of common carotid artery. Approximately 10% of the cases present with cranial nerve palsy with paralysis of the hypoglossal, glossopharyngeal, recurrent laryngeal, or spinal accessory nerve, or the sympathetic chain involvement,² therefore, they may be associated with pain, hoarseness, dysphagia, and Horner syndrome. As the tumor enlarges and compresses the carotid artery and the surrounding nerves, other symptoms such as pain, tongue paresis, hoarseness and dysphagia may occur. CBTs may rarely present with fever and may be one of the causes of pyrexia of unknown origin.¹³ Functional CBTs may manifest with hypertension and diaphoresis simulating pheochromocytoma.

CBT presenting as CPH is an unusual phenomenon where it presents as short-lived unilateral neuralgic headache with conjunctival injection and tearing. The mechanisms responsible for pain in CPH remain unknown. A history of

head or neck trauma is reported in about 20% of cases, but these findings are similar to those for chronic hemicrania or migraine. There is no familial predisposition.

The relationship between the CBT and its adjacent vessel walls can be predicted by Shamblin¹⁴ classification system developed in 1971 which has three groups of tumors. This helps to predict surgical outcome on the basis of angiographic findings. Higher progression in group correlates with an increased probability of locoregional nerve involvement and operative complications.¹⁵ Group 1 - These tumors shows maintained fat planes with the vessel walls and helps easy resection. Group 2 - These tumors closely abuts vessel walls but do not encase them. Group 3 - These tumors are usually intramural and encase carotid arteries as well as regional nerves.

This classification system is still used in the assessment and management of CBTs. Currently, surgical resection is the mainstay of management if the patient's surgical risk is acceptable, and resection should be performed early so that tumor size does not increase the risk.^{16,17} Despite the documented degree of surgical difficulty, successful resection of Shamblin Group 3 tumors has been reported, although these procedures involved vascular reconstruction with synthetic grafts or autologous vein grafts.¹⁸ Accurate preoperative diagnosis with modern surgical techniques yields excellent results with minimal complications.^{19,20} Without treatment, CBTs may become life threatening because of their increasing size and associated effects.¹⁹ This case has been reported not only because of its rarity but because of its variable presentation mimicking other diseases and misleading treatment options. The case also alerts us to comment upon the relationship of the neck vessels to the lesion that is an important diagnostic marker.

CONCLUSION

CBTs are rare but important masses from diagnostic as well as clinical point of view because they can be picked up on imaging with their typical imaging manifestations and on application of literature described signs. They can present as a diagnostic challenge for physicians because of their unusual presentations like syncope and hemicrania and can mimic other clinical disorders; hence it requires prompt evaluation and management.

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