Carotid Body Tumors: A Case Series and Review of the Literature

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Background: Paragangliomas of the head and neck are rare vascular tumors derived from the paraganglia tissues originating from the neural crest. They are usually benign and hypervascularized. Diagnosis is relatively easy in condition to consider it in evaluating every lateral neck mass.
Methods: We made a retrospective study of the records of 10 patients who presented with carotid body tumors at the Department of Vascular surgery of the Military Hospital Avicenne in Marrakech during the period between 2008 and 2013. Epidemiologic, etiologic, diagnostic, and therapeutic features were analyzed.
Results: The average age of our patients was 35.4 years (26–55 years), with a male predominance (sex ratio = 2.33). We noted 7 cases of isolated carotid locations and 3 cases of multiple locations. A slow-growing neck mass was the main clinical presentation. Other signs were pain, dysphonia, dizziness, headache, and tinnitus. Physical examination showed, in most cases, a neck nontender mass with side to side mobility. Imaging techniques included Doppler ultrasound, computed tomography (CT) scan, magnetic resonance imaging, and catheter arteriography. Urinary analysis for metanephrine was carried out in 1 case. The clinical presentation and imaging results strongly suggested the diagnosis of carotid paraganglioma in all cases. Treatment was surgical excision in all cases associated with a preoperative embolization in 1 case and a postoperative radiotherapy in 2 cases. Pathology confirmed the diagnosis, and a lymph node metastasis was suspected of malignity in 1 case. The evolution was favorable in all our patients.
Conclusions: Carotid body tumor requires early diagnosis and an adequate multidisciplinary team. The diagnosis must be considered in the case of any pulsatile cervical mass. Surgery is the treatment of choice despite its risks especially in large tumors. The therapeutic indication should, ideally, be set in a multidisciplinary consultation.

INTRODUCTION

Paragangliomas of the head and neck are rare vascular tumors derived from the paraganglia tissues originating from the neural crest and associated with autonomic ganglia along the sympathetic trunk.[1–3] Carotid paraganglioma or carotid body tumors (CBTs) represent 60–70% of paraganglioma of the head and neck.[4–6] Diagnosis is relatively easy; however, it should be considered in evaluating every lateral neck mass. Monro said in 1950: ''The first step in diagnosis of carotid body tumors is to think of it''.[7] Many imaging techniques are available for assessing paraganglioma. Treatment is based primarily on surgical excision difficulty of which is variable but always higher than for other neck tumors.

In light of a series of 10 cases and a review of the literature, we will discuss the epidemiological, clinical, imaging, and therapeutic features of this rare disease.

MATERIALS AND METHODS

We made a retrospective review of the medical records of 10 patients diagnosed with CBT at the

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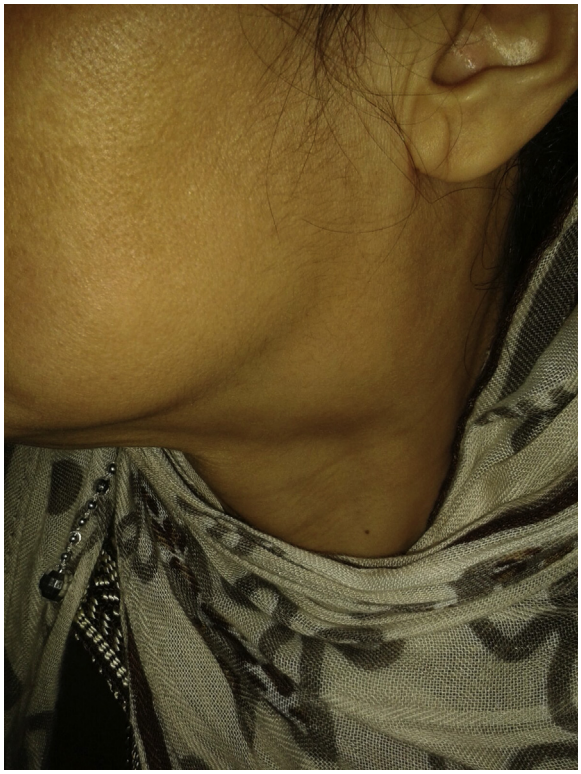

Fig. 1. Female patient with a large left carotid body tumor.

Department of Vascular Surgery of the Military Hospital Avicenne in Marrakech during the period between 2008 and 2013.

RESULTS

The average age of our patients was 35.4 years, ranging from 26 to 55 years. Seven patients were male (70%) and three were female (30%), the sex ratio M/F was 2.33. Two patients were from the same family (father and son). The location was on the right side in 5 patients (50%), the left side in 4 patients (40%), and bilateral in 1 case (10%). We noted 7 cases of isolated CBTs (70%) and 3 cases of multiple locations (30%; 1 case of bilateral CBT associated with a tympanic location, 1 case associated with a thoracic location, and 1 case with history of a tympanic paraganglioma treated previously).

Slowly growing mass of the neck was the main reason for seeking medical attention. However, in the case of 1 patient, the evolution was fast during 6 months prior to admission. We also noted 2 cases each of vertigo, headache, dysphonia, and painful mass and 1 case of hypoacusia due to a tympanic location.

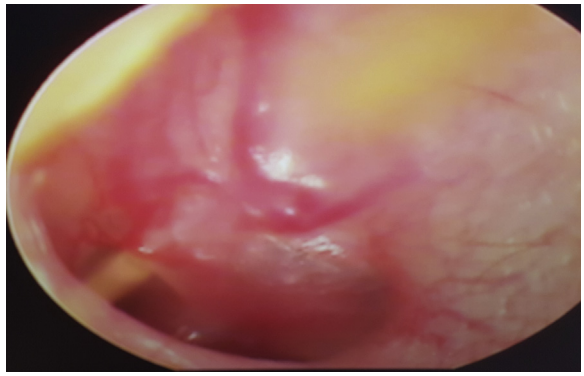

Fig. 2. Otoscopy revealing a reddish tumor behind eardrum characteristic of tympanic location.

Physical examination revealed, in most cases, a nontender mass along the anterior border of the sternocleidomastoid muscle (Fig. 1). Although, in 1 case of a patient who underwent a biopsy by mistake in another hospital, a 2 cm scar with inflammation was observed. Ear, nose and throat (ENT) examination was normal except in a case with an associated tympanic location (Fig. 2).

Doppler ultrasonography was performed in 8 patients (80%) which revealed a vascularized solid tumor: homogeneous in 5 cases, heterogeneous in 3 cases, and surrounding the carotid vessels in 3 cases (Fig. 3). CT scan was performed in 7 patients (70%), showing a solid tumor, that was highly enhanced after the administration of contrast (Fig. 4). Five magnetic resonance imaging (MRIs) (50%) were performed allowing a better analysis of the relation with environmental structures (Fig. 5). Catheter angiography was performed in 4 patients (40%), highlighting their vascular nature, with a tumor blush and a splaying of the carotid bifurcation.

Urine analysis was made in 1 case, demonstrating no excess of metanephrines.

Tumors were classified according to Shamblin criteria: group 1 in 2 cases (20%), group 2 in 6 cases (60%), and group 3 in 2 cases (20%). All patients underwent surgical excision. Embolization was performed 48 hours before surgery in 1 case only. The tumor was dissected in the periadventitial plane and resected completely without vascular or nerve sacrifice in 8 cases (80%; Figs. 6–8). In 2 cases (20%), the resection was partial due to extension to the skull base, we proceeded to the marking of residual tumors by clips in both cases. Ligation of the thyroid artery was necessary in a case with resection and anastomosis of the external carotid artery (ECA) and transposition of the internal carotid artery (ICA). A ligature of the ECA, without sacrifice of the ICA, was necessary in 1 case. We performed a neck dissection in 1 case due to the presence of