Case report

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The role of ultrasonography in the diagnosis of glomic tumors: a two case report

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Abstract

A carotid body tumor is a rare presentation of an extra-adrenal phaeochromcytoma. Patients commonly present with asymptomatic neck masses and careful preoperative evaluation is required to find out the functional and vascular status of these tumors.

We report two cases of carotid body paraganglioma, their evaluation, ultrasonographic aspect, management and the final outcome.

Keywords: paraganglioma, carotid body tumor, phaeochromocytoma, ultrasonography

Rezumat

Tumorile de glomus carotidian constituie o formă rară de feocromocitom extraadrenal. Pacienţii prezintă mase laterocervicale asimptomatice, iar evaluarea atentă preoperatorie este necesară pentru a preciza starea funcţională şi vasculară a acestor tumori.

În acest articol prezentăm 2 cazuri clinice de tumori glomice carotidiene, evaluarea acestora, aspectul ecografic si atitudinea terapeutică.

Cuvinte cheie: paragangliom, tumora de glomus carotidian, feocromocitom, ecografie

Introduction

Glomic tumours are rare vascular tumours originating in the paraganglionar cells of the external layer of the carotid artery. The tumors develop in the bifurcation point of the common carotid artery in internal and external carotid arteries. These tumours were described for the first time in 1762 by Haller and renamed chemodectomas in 1950 by Mulligan in order to suggest their origin in the chemoceptor cells. In 1974 they were called paragangliomas (PGL) by Glenner and Grimley, because of their histopathological structure. They represent 0.6% of head and neck tumours, respectively 0.03% of total cancers, with maximal incidence between 45 and 50 years; they do not have sex predilection. In most of the situations, they become noticeable by their dimensions, before being symptomatic. They are unilateral in most cases, only 10% being bilateral. Most of them are benign, but 5 to 10% are malignant. Diagnosis is made by ultrasound, CT scan or IRM [1,2].

Carotid body tumors (CBT) are the most common PLG of head and neck [1], but their incidence is very low (0,012%) [2]. Most of these tumors are benign with a small potential of malignancy [3]. PGL is a rare tumor of the head and neck, which arises from the neural crest cells. Histopathologically similar to the adrenal gland neoplasm (phaeochromocytoma), they are usually benign and non-functional, but since the paraganglionic cells contain very small amounts of catecholamine, clinically-significant catecholamine release is possible, though rare [1]. This kind of tumor grows and expands slowly and rarely metastasizes [1].

A carotid body tumour could be also a rare presentation of an extra-adrenal pheochromocytoma. Patients

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commonly present with asymptomatic neck masses and careful preoperative evaluation is required to evaluate the functional and vascular status of these tumours [1].

We report two cases of carotid body paraganglioma, their evaluation, management and the final outcome.

Cases presentation

Case 1

A 22-year-old woman, without any previous diseases, presented with an asymptomatic right laterocervical mass, with a slow growth overthe last year. A family history of PGL was negative. On clinical examination, we found a 3x3 cm palpable, pulsatile mass in the upper third of the right sternocleidomastoid muscle, mobile in the lateral plane and with any audible bruit. She was admitted to the Head, Neck, and -Throat department for preoperative evaluation of the mass. The Doppler colour ultrasound examination confirmed a solid tumoral mass located in the carotid bifurcation, dislocating both carotid branches, hyperechoic, slightly inomogenous, well delimitated, highly vascular, measuring 3x4 cm (fig 1).



Fig 1. Transverse scan above the right common carotid artery bifurcation (arrow – internal carotid artery, open arrow – external carotid artery), power Doppler. The tumour dislocate the carotid branches.

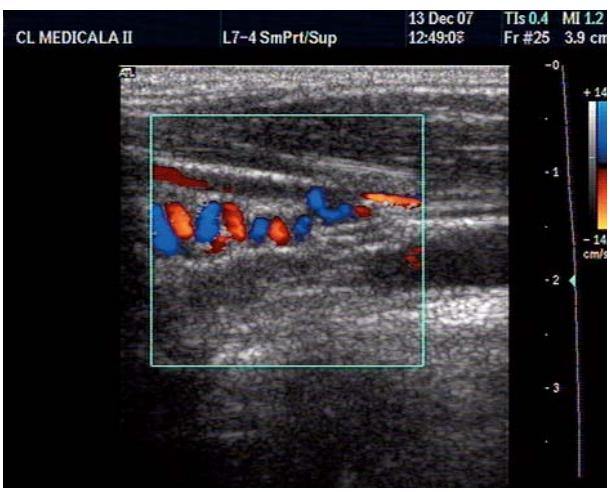


Fig 2. The feeding artery of the tumor – a sinuous artery, emerging from the common carotid artery (color Doppler).



Fig 3. Transverse scan above the left common carotid artery bifurcation (arrow – internal carotid artery). The tumor is highly vascularized (power Doppler).

Blood flow of the tumor was supplied by a sinuos artery, emmerging from the common carotid (fig 2).

At the bifurcation of the left common carotid artery we found another tumor with the same ultrasonographic pattern (fig 3).

These features were mostly consistent with a carotid body tumor. Regarding the fact that the patient was asymptomatic (no pain, no Horner syndrome or laryngeal palsy were present), surgery was postponed and the patient was only monitored. At 6 months, the clinical and ultrasonographic examinations revealed no significant changes in shape and size of the tumor.

Case 2

A 25-year-old woman presented with a gradually painless swelling of the left laterocervical region. As in the first case, no prior disease history, personal or familial, was recorded. The ultrasonographic aspect was very similar to the first case, but the tumor was unilateral, hypoechoic and the vascularization was assured by an artery from the external carotid (fig 4, fig 5).

In view of the asymptomatic tumor and because the patient had no significant complaints, surgical excision of the tumor was postponed. At 8 months, the mass had the same diameter.

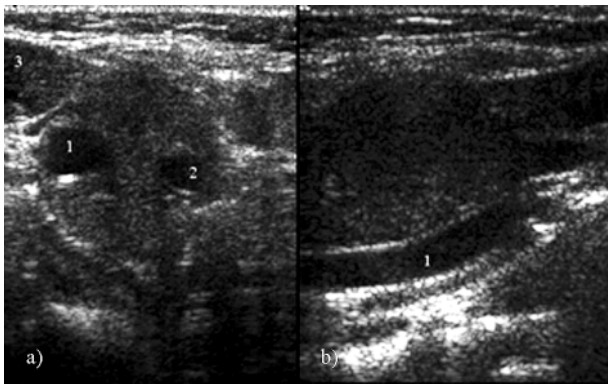


Fig 4. Glomic tumor: *a)* transverse scan – the tumor is growing between the internal carotid artery (1) and external carotid artery (2); *b)* longitudinal scan – the tumor is well define from the artery' wall. 3 – sternocleidomastoid muscle

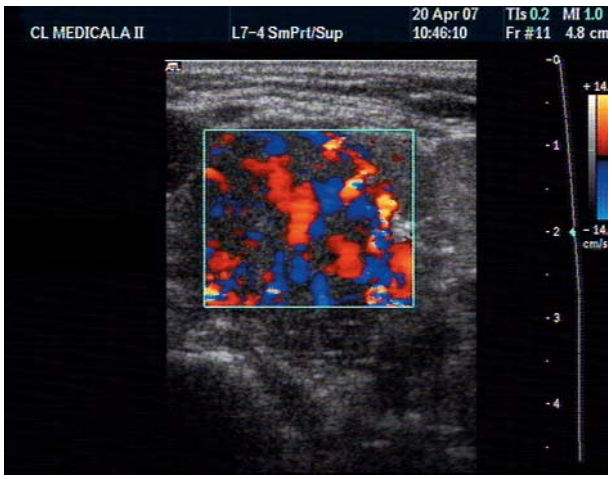


Fig 5. Color Doppler examination – the tumor is hypervascularized

Discussion

The neoplasms originating from the neural crest cells, referred to as PGLs, are associated with parasympathetic nervous system and are found near arteries and cranial nerves of the branchial arches, with a cervicocephalic distribution. PGLs are named after their site of origin and they most commonly occur at the carotid bifurcation where they are known as carotid body tumors. Additional sites of origin include the jugular bulb (jugular paraganglioma), the vagus nerve (vagal paraganglioma) and within the middle ear mucosa (tympanic paraganglioma) [3], but also in the orbita, nasopharynx and the paranasal sinuses [2]. CBT are the most common variety of paraganglioma of the head and neck, and is the only pathology to affect the carotid body. The etiology of CBT is unknown. There have been reports of increased incidence of CBT in patients with chronic obstructive pulmonary disease and in people living at high altitudes because the cells in the carotid body typically detect changes in partial pressure of oxygen, partial pressure of carbon dioxide, and pH levels. CBT may be sporadic or familial [3]. Both of our patients presented the sporadic form, as they did not have familial history for this pathology.

Phaeochromocytomas are located extra-adrenally in 10% of cases. 90% of these extra-adrenal PGLs are located in the abdomen, mainly in chromaffin cells of the organ of Zuckerkandl. The remaining 10% of the extra-abdominal PGLs including the head and neck [1, 4]. The carotid body is a vascular reddish-brown structure about the size of a grain of rice located within adventitia posteromedial to bifurcation of the common carotid artery. It acts as a chemoreceptor responding to variations in oxygen tension and carbon dioxide concentrations, henceforth alternatively named chemodectomas [1, 5].

CBT usually present in the fourth to sixth decades of life (age range: 3 months–89 years). Our patients were both younger persons.

The average tumour size is 4.5×3.5×3 cm. The largest tumor described in literature exceeded 15 cm in diameter and weighed 200 g [6]. The incidence of bilateral CBT vary from 10% to 25% in different series [1].

Most of these tumours are benign, a fact suggested in our patients by the clinical evolution, but malignant lesions can be found in 6–12% of cases. The diagnosis of malignancy is reserved for tumours with local, regional and distant metastasis, because there are no histological features for the distinguishing of the benign from the malignant lesions [2,7]. In a series of 24 cases published by Gaylis and Mieny, the differentiation between benign and malign tissue was not possible, so the diagnosis of malignancy was made only by microscopic proof of the presence tumoral cells in lymph nodes or other organs in 7 patients (~30%) [8]. Because of the favorable evolution of our patients, no surgery and no histological examination were performed.

Between 10% and 50% of PGLs are hereditary (autosomal dominant) [9].

Patients with CBT typically present with a painless mass in the angle of the jaw that may be partially covered by the sternocleidomastoid muscle. These vascular tumours may transmit pulsations from nearby carotid arteries or may be pulsatile inherently [1]. This was in fact the clinical presentation of our cases: a painless but pulsatile tumor. Clinical examination has reduced reliability for pulsatility assessment, being difficult to establish if this phenomenon is due to pulsations from a highly vascular tumor or to pulsations from nearby arteries. The tumor is typically mobile in the lateral plane but its mobility is restricted in the cephalocaudal direction (Fontaine sign).

At the time of diagnosis, cranial nerve palsies are present in 10% of cases, with the X, XII, V and VII cranial nerves commonly involved, while the involvement of cervical sympathetic produces ipsilateral Horner's syndrome. Carotid sinus syndrome with bradycardia, hypotension and unconsciousness can occur spontaneously or secondary to head movement or pressure on the tumour. Episodic symptoms of phaeochromocytoma are rare and are seen in 1–3% of cases, with noradrenaline being the usual product and hypertension the most frequent finding. Lack of awareness of these functional tumours can cause disastrous consequences during embolisation or surgery [6].

Diagnosis is made by means of ultrasonography (gray-scale, Doppler), CT scan, IRM and angiography [2].

The differential diagnosis includes cervical lymphadenopathy, carotid artery aneurysm, brachial cleft cyst, laryngeal carcinoma and metastatic tumor [3].

The size of the tumour is important not only for the clinical manifestations, but also for a decision on treatment strategy. Generally, tumors larger than 4–5 cm tend to have partial or complete encirclement of the carotid arteries. Larger tumor sizes have been associated with higher risk of bleeding and cranial nerve injury complications with surgery. Pre-operative tumor embolization has been employed to shrink the tumor size and thereby decreasing the complications. Shamblin classification system based on tumor size was proposed in 1971. According to this classification:

- group I tumors are relatively small tumors minimally attached to carotid vessels; the surgical excision is not difficult;
- group II tumors are large tumors with intimate, but moderate attachment to the carotid vessels, sometimes exercing compressive effect; these tumors are amenable to careful surgical subadventitial excision;
- group III tumors are very large tumors, encasing the carotid vessels and often require arterial resection and grafting [3, 10].

Our cases were included in group I, being of relatively small size, not having an attachment to the carotid arteries or any compressive effect.

Biochemical assays are routinely employed for the diagnosis of a functioning tumour, irrespective of symptoms. The usual methods are measurement of urinary free catecholamines and its metabolites VMA and MNs in a 24-hour urine sample.

Our patients had no clinical and biochemical elements suggestive for a functioning tumor and this was the reason why we did not perform measurements for plasmatic or urinary free catecholamines or vanil-mandelic acid.

The ultrasonographic pattern found in these patients was typical, therefore we considered that CT scan and IRM examinations were not necessary for diagnostic in this stage of evolution. Ultrasonography is useful in monitoring and surveillance of these tumors, as it is an accessible method, with a good cost-efficiency.

In **conclusion,** we have reported two cases of CBT with similar clinical and ultrasonographic presentations, their evaluation and management. Differences occurred regarding the location of these tumors, bilateral for the first patient and unilateral for the second.

The age of these patients was inferior to the mean age described in larger series.

Long-term follow-up is mandatory as the time interval for local recurrences and distant metastasis can vary from months to years after the initial diagnosis.

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