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Carotid Paragangliomas: Report of 3 Cases and Review of the Literature

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Abstract
Introduction: Carotid Paragangliomas (CP) are relatively rare tumors and usually benign. They appear as a cervical mass of slow and painless evolution, which is often responsible for diagnostic and management delays. Surgery remains the recommended treatment, although there is a risk of neurological and vascular sacrifice for large tumors. The objective of this study is to report our experience and to review the literature on the clinical features, positive diagnosis and management of CP. Methods: It is about a retrospective study of 3 patients with CP who had undergone a surgical resection as a treatment of CP over a period of 5 years from 2013 to 2017. Results: The study concerns 3 female patients without any significant family history. The mean age was 63 years. The tumor has evolved on average 13 years before its diagnosis. The patients were admitted for a firm and incompressible lateral cervical mass on the right side in 2 cases and on the left side in one case. The surgical treatment consisted on a total resection of the tumor in all cases with a vascular graft interposition in one case. A sacrificed of hypoglossal nerve was required in another case. Pathological examinations of the 3 tumors revealed a benign histology of paragangliomas. The follow-up was simple in 2 cases while one patient presented a transient peripheral facial paralysis. Conclusion: The diagnosis of CP should be evoked towards any firm and painless lateral cervical mass having evolved for several years. Surgical treatment is curative for benign lesions, despite its risks especially in advanced tumors.

Keywords: Carotid Paraganglioma, Surgical Treatment, Preoperative Embolization, Rare Case Report

INTRODUCTION

Paragangliomas are extra-adrenal tumors derived from the chain of neuroectodermal tissue, occurring from the skull base to the pelvic floor. In the head and neck region, they are found at the jugular bulb, the vagal and tympanic nerves and the aortic glomus. Carotid paraganglioma (CP), commonly known as carotid body tumor is the most frequent parasympathetic form. The majority of these tumors are benign but 2% to 13% of patients may have malignant lesions with metastases that could interest regional lymph nodes, lungs and even bones (1). The history of the disease and the clinical examination are essential to be determined for the diagnosis of these tumors. Because of the topography and the hypervascular character of these tumors, a radiological exploration remains a necessary. Indeed, ultrasound, CT-scan and MRI are crucial imaging tools as they can confirm the diagnosis of CP and eliminate other etiologies of cervical mass (2-4). Different therapeutic approaches are described in the literature, surgery consists the main curative approach. Through three cases report of CP and a
recent extensive review of literature, clinical presentations, imaging tools and therapeutic modalities being used in this pathology are presented and discussed.

PATIENTS AND METHOD

It is a retrospective study conducted within the department of vascular surgery of Mohammed VI University Hospital Center. It concerns three cases of CP that had been hospitalized over a period of 5 years from 2013 to 2017.

1st Case

A 73-year-old woman presented to our department with a palpable right lateral cervical mass. She had a cholecystectomy as personal antecedent without family history of tumors. The onset of the symptomatology dates back to 11 years before her admission, with the occurrence of a right lateral cervical mass gradually increasing in volume, accompanied by intermittent pains without other associated signs. Physical examination revealed a mass that located below the right mandibular angle measuring 5 cm in its great axis. The mass was mobile laterally, firm, incompressible and pulsatile without any inflammatory signs. The rest of the somatic examination was normal.

The ultrasonography demonstrated a solid mass in contact with the right parotid gland. The tumefaction was taking the aspect of a homogeneous hypoechoic tissue, well limited and hypervascularized, measuring 6 x 4,9 x 3 cm. The mass was located within the carotid bifurcation causing a splaying of the carotid bifurcation. The angio-CT scan showed a right lateral cervical mass, centered at the level of the carotid bifurcation, with a tissue density. The mass was intensely and heterogeneously enhanced after injection of the contrast product with a repressing of the right jugular vein without any sign of invasion. Its measurements were 3,8 cm (anteroposterior diameter), 3,7cm (transverse diameter) and 5,6 cm (height). The therapeutic management consisted on surgical treatment of the carotid paraganglioma. A total resection of the tumor, without sacrificing the carotid axes has been practiced. The post-operative follow-ups were simple. The histological result of the resection piece reveals a benign tumor.

2nd Case

A 58-year-old woman was admitted in our department for the management of a painless, palpable mass in the right side of the neck. No family history was reported in the anamnesis. The beginning of the symptomatology goes back to 19 years by the apparition of a lateral cervical mass, gradually increasing in volume, without associated signs. Physical examination revealed a mass located below the mandible angle. The mass was mobile, firm, incompressible and without any inflammatory signs. Ultrasoundography showed a homogeneous tissue lesion. The tumor was centered on the right carotid bifurcation, hypervascularized and fed by arterial branches of the external carotid artery. The CT-scan showed a homogeneous mass that measures 5,6 x 4,3 x 3,7 cm. The mass appears well delimited respecting the adjacent structures and without cervical lymphadenopathy.

A preoperative embolization was performed 24 hours before the surgery. Surgical exploration found a mass invading both internal and external carotid arteries. After releasing the hypoglossal nerve, a total resection of the tumor was performed and vascular graft interposition was performed between the common carotid artery and the distal internal carotid artery with inverted saphenous vein. The external carotid has been ligated and resected with the tumor. The postoperative follow-ups were simples, without any need for transfusion. The histological result of the resected mass was found benign.

3rd Case

A 71-year-old woman with a coronary artery disease, under oral anticoagulation, was admitted for the management of a left neck swelling. The tumefaction appeared 10 years before her admission in our department. The Physical examination had found a firm, incompressible and fixed mass. It extends upwards to the mastoid
bone. The Cervical CT-scan showed a hypervascular tumor being developed on the carotid body in favor of a carotid paraganglioma. Measuring 45 mm in its major axis.

As a received treatment, a surgical procedure revealed a huge tumor towards the carotid bifurcation extending superiorly into the mastoid bone. The mass was adherent to the hypoglossal nerve, and, hence, it had to be sacrificed. The facial nerve was dissected and spread by a soft instrument. A complete resection of the tumor was performed leading to a wound of the internal carotid artery, that was repaired by 7/0 Prolene thread. We noted, as an immediate follow-up, a peripheral facial paralysis with a positive Charles Bell’s sign. This complication was successfully treated by oral corticotherapy (5 days). The Pathological examination of the tumor did not show any signs of malignancy. The evolution was favorable after 3 months of follow-up.

The radiological aspect of CP at the CT-scan:

Figure 1: CT-scan with frontal and 3D view: Hypervascularized mass sitting at the left carotid bifurcation and extended upwards

Figure 2: axial CT-scan: A cervical mass separating the internal and external carotid arteries at the level of the bifurcation. The mass presents an intense homogeneous enhancement after intravenous administration of contrast.
The aspect of CP at the arteriography:

Figure 3: Arteriography that shows the characteristic appearance of carotid bifurcation due to CP (Classic image in lyre)

Figure 4: Arteriography revealing the hypervascular character of CP
Surgical management of CP:

Figure 5: Surgical resection of CP using the pre-sterno-cleido-mastoid incision

Figure 6: Per-operative picture that shows the CP extension

Figure 7: CP after being completely resected
DISCUSSION

Paraganglioma is rare and usually benign. It develops from diffuse neuroendocrine system cells. Cervical paraganglioma accounts for 0.6% of head and neck tumors (1) with incidence of 1/30,000 to 1/100,000 in the general population. Carotid paraganglioma (CP), named also carotid body tumor (CBT), is a hypertrophy of the carotid body tissue. CP remains the most common type of paragangliomas of the head and neck and its incidence is estimated at less than 0.03% (2).

The first description of CP was made by Von Haller in 1743, who described it as a chemoreceptor located in the adventitia of the carotid bifurcation (3). CPs very rare and most vascular surgeons will encounter only few cases during their career. This tumor is rarely seen in children. A recent review of the literature by Georgiadis et al. identified fewer than 20 cases of carotid tumors in children under 14 years old (4). Their etiology is still unknown (5). A high incidence of carotid paraganglioma was noted in patients suffering from chronic pulmonary disease and those living at high altitude (6). The resulting lesions are referred to as non-heritable or
sporadic tumors. These tumors can be bilateral in 32% in cases of familial forms and in 5% in non-familial forms. CP can occur at any age but are particularly common between 30 and 60 years of age. Unfortunately, nuclear pleomorphism, mitotic activity, necrosis, or vascular or perineural invasion cannot predict the biological behavior because all these features may also be found in benign forms of CBTs. Therefore, CBTs are mostly considered benign lesions; however, malignant behavior is often encountered. This diagnosis is reserved for the tumors with local, regional, and distant metastasis. The rate of malignancy is reported to be 6% to 12.5% of all cases (7-9), and 7% to 9% of hereditary cases (10,11).

Clinically, CBT presents as a painless, palpable, slow-growing swelling located in front of the sternocleidomastoid muscle at the level of the hyoid bone. Sometimes, it can cause hoarseness, dysphagia and dyspnea secondary to compression (12). Sometimes, clinical signs of hypersecretion of catecholamines such as hypertension, palpitations and diarrhea can be seen (13). Physical examination reveals a mass which may be pulsatile, located below the mandibular angle, typically laterally mobile but vertically fixed. The mass is usually non tender, rather rubbery, firm, and non-compressible. A sound may be audible (14). Neurologic abnormalities caused by vagal or hypoglossal nerve involvement and Horner’s syndrome are unusual but may be present in some patients (15).

Since these tumors are highly vascularized, a direct biopsy is not recommended and diagnostic imaging modalities are very important for positive and differential diagnosis of this condition. Ultrasonography with B-mode ultrasound and duplex color ultrasound represent the first imaging step for cervical CBT. They are inexpensive and noninvasive diagnostic tools (16). It is a valuable diagnostic modality because it shows a highly vascularized hypoechoic mass located at the carotid bifurcation. CP are easily detected using computed tomographic (CT) imaging and magnetic resonance angiography (MRA). CT angiography depicts the anatomy more accurately than MRA because of its better spatial resolution. However, contrast-enhanced MRA is better suited for screening and detecting multiple lesions (17). On contrast-enhanced CT, the carotid body tumor appears as a hypervascular mass that tends to spread at the bifurcation between the internal carotid artery (ICA) which is posteriorly displaced and the external carotid. On magnetic resonance imaging (MRI), paragangliomas are observed in T1-weighted sequences with low intensity signal and in T2 with a hyperintense signal. After the intravenous administration of contrast material, these tumors exhibit a pattern of intense and homogenous enhancement similar to CT imaging. The angiographic examinations don’t provide additional diagnostic information compared with CT imaging and MRI but may allow a possible preoperative embolization or ICA stenting of Shamblin class III CBT (18,19). In addition, angiography with or without embolization is mostly used to confirm the diagnosis and to identify the supplying artery before surgical resection.

The size of tumor is important to be determined not only for its clinical manifestations but also for treatment. In 1971, Shamblin introduced a classification system based on tumor size (13). He classified small tumors that could be easily dissected away from the vessels as group I. In this case, complete surgical resection is generally possible with minimal risk of vascular or cranial nerve injuries (20). Group II included paragangliomas of medium size that were intimately associated and compressed carotid vessels but could be separated with careful subadventitial dissection. Group III intimately surround the carotid artery, thus a complete resection is very difficult and often requires a temporary interruption of cerebral circulation for vascular reconstruction. The risk of permanent vascular and neural injuries is significantly higher than for Class I and II (21).

The treatment strategy includes conservative management, radiotherapy and surgical resection (22). Surgical resection is the only curative treatment for resectable CBTs. However, the rich blood supply sometimes makes CBT resection difficult. The intraoperative hemorrhage and cranial nerve injury incidence remains considerable (23). The incision is usually done along the anterior border of the sternocleidomastoid muscle. Avoiding the injuries of the vagus, hypoglossal or superior laryngeal nerve during the resection should be a must for every surgeon. External carotid artery could be sacrificed if it was invaded or taken by the tumor. When the internal carotid artery has to be resected, a reconstruction with autogenous great saphenous vein should be performed (24).
Radiotherapy is suggested in cases of extensive involvement, making the tumor not resectable or when there is a high surgical risk (25). It is also recommended for giant and recurrent carotid body paragangliomas and metastatic carotid body paragangliomas of the regional lymph nodes (26).

The role of preoperative embolization is controversial, even though intraoperative hemorrhage may decrease, transfusion requirements are unaffected and the procedure may increase the risk of ectopic embolism. In 1980, Schick et al. (27) first reported preoperative transarterial embolization, which is proved to be effective in decreasing blood loss and operative time by some investigators (28). The aim of the embolization is to occlude the tumor’s feeding vessels avoiding decrease of blood flow to the normal tissues. Vogel et al. (29) demonstrated that embolization reduces morbidity and mortality and shortens the time of surgery by reducing intraoperative bleeding. While Litle et al. (30) compared 11 embolized tumors and 11 non-embolized tumors and showed no differences in blood loss, operative time, transfusion requirements, or perioperative morbidity. In an opposite manner, Liapis et al. (31) concluded that preoperative embolization is helpful by decreasing intraoperative bleeding in patients who underwent embolization before surgical resection of CBTs in comparison to patients who had surgery alone. In patients with vascular tortuous, spasm or small feeding artery, percutaneous approach was preferred since it might have the advantage of obliterating the tumor capillary bed (32). When embolization is carried out, aseptic inflammation might exist and collateral circulation will form gradually. In this case, it is recommended to carry out CBT resection immediately after the embolization to avoid the consequences. Usually the time gap between the procedure of embolization and CBT resection varies from immediately to 3 days.

CONCLUSION

Carotid paragangliomas remain rare, poorly known and often benign. Its diagnosis should be evoked towards any pulsatile latero-cervical mass. The optimal therapeutic approach of the CP requires a meeting of multidisciplinary consultation and depends on several parameters (patient, tumor and expertise of the surgeon). A Complete surgical resection is the only curative treatment option. It is considered the treatment of choice by most experts.

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