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An evaluation on management of carotid body tumour (CBT). A twelve years experience*

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SUMMARY: **An evaluation on management of carotid body tumour (CBT). A twelve years experience.**

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Objectives. *Carotid Body Tumor (CBT) is a rare lesion of the neuroendocrine system but it is the most common form of head and neck paraganglioma (PGL). Our objective is to discuss the optimal management of these lesions to provide the best outcome of patients treated by surgical resection.*

Patients and Methods. *A retrospective evaluation was obtained by review of the records of 20 patients with 26 CBT treated at our institution between 2000 and 2012. Primary tumor characteristics, diagnostic protocols, surgical treatment, short and long-term outcomes were collected and analyzed.*

Results. *A total of 26 CBTs resections were performed on 20 patients; the age range was 21-89 years. There was a female prevalence (14 women-80% and 6 men-20%). Familial cases occurred in 6 patients (30%); of these, 3 patients had bilateral lesions and 1 patient multiple paragangliomas. In all cases no lymph node metastasis was found. All lesions were grouped into three groups according to the latero-lateral diameter: Group I < 3 cm; Group II 3<>5cm; Group III >5cm. All patients were managed by surgical resection of the CBT. There were no operative deaths. Overall we found transitory neurological impairment in 15,3% and permanent neurological deficit in 7,6% of cases. No complications occurred in all resections of Group I tumors. In Group II only 1 resection was followed by dysphonia by recurrent nerve palsy (after vagal nerve en-bloc resection). In Group III only 1 resection was followed by permanent vagus nerve palsy.*

Conclusions. *Surgical removal of the tumor is the only treatment that can ensure a complete eradication of the disease. Family screening is of great importance in patients with hereditary forms. Careful preoperative planning of surgical procedure by integrated diagnostic imaging and a full mastery of the surgical technique can minimize the risk of the most common postoperative complications. Lifelong follow-up is mandatory to make early diagnosis of recurrent disease.*

KEY WORDS: CBT - Carotid Body Tumors - Paraganglioma - Embolization - Resection.

Introduction

Although carotid body tumors (CBTs) are rare, with an incidence of 1:30000, they are the most common tumors of extra-adrenal chromaffin tissue and represent more than 50% of head and neck paragangliomas (1). The age of onset is generally between 30 and 60 years (2), although in the literature cases have been described with earlier onset, due to family genetic predisposition (3); no sex prevalence has been reported. Surgical excision is the only radical treatment of the disease because histopathology cannot give a definitive diagnosis of malignancy (4, 5); clinical malignant behaviour is described in 5-30% of cases.

Patients and Methods

Between 2000 and 2012, twenty patients with 26 CBTs were treated by surgical excision at our institution: 14 women (80%) and 6 men (20%), with overall age ranging from 21 to 89 years. The most common clinical presentation was a painless mass of the latero-cervical region (50%); only 8 patients (40%) had incidental diagnosis by ultrasound scan requested for another reason. Nerve palsy as first symptom occurred in 1 patient (5%). All patients have been evaluated by duplex ultrasound as first step. All patients underwent at least a second step examinations such as CT angiography, magnetic resonance angiography, arteriography, PET-CT, I-MIBG. Detailed

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list of imaging performed in all patients is given in Table 1. Familial cases occurred in 6 patients (30%); of these, 3 patients had bilateral lesions and 1 patient had multiple paragangliomas. In all cases no lymph node involvement was found at preoperative evaluation. The lesions were grouped into three groups according to the latero-lateral diameter : 9 tumors (34.6%) in Group I (< 3 cm), 12 (465.1%) in Group II (3-5 cm), and 5 (19.2%) in Group III (>5 cm). Two patients underwent preoperative embolization of the feeding vessels by superselective carotid artery angiography. Preoperative and postoperative laryngoscopy plus phoniatrics evaluations were obtained in all cases. All patients were treated by the same surgeon by complete surgical resection of the CBT through conventional approach to the carotid artery and its bifurcation; all interventions were carried out under general anesthesia

Results

Operative mortality was null and no strokes were observed prior to surgery or in the postoperative days. Only 1 case required early surgical revision for bleeding. The overall incidence of transient peripheral neurologic complications was 15.3%; permanent lesions occurred in 7.6% of cases and only in lesions classified as group 2 and 3. In group 2, in one case out of 12 (8.3%) dysphonia was observed by recurrent laryngeal nerve palsy, this complication arose after the removal of the vagus nerve by vagal paraganglioma of origin and in the same patient (already having undergone bilateral removal of carotid paraganglioma) occurred postoperative hypertension treated with clonidine and β-blockers. Another patient in group 2 presented a mild Horner's syndrome resolved spontaneously on the third postoperative day. In group 3, in only one case out of 5 did postoperative permanent dysgeusia occurred and in another patient dysphagia for solid foods that was resolved spontaneously. (Postoperative complications are listed in Table 2). In 1 of 26 (3,8%) interventions a strong adhesion of the tumor to the posterior wall of the carotid bulb required a resection-anastomosis of the first portion of the internal carotid. In 6 cases a partial or complete encasement of the vagus nerve was found intraoperatively; although this condition increased the complexity of the surgical procedure, the nerve was preserved by a cautious dissection from the tumor. One of the two cases of suspected recurrent tumor was found intraoperatively to be a metachronous primary paraganglioma of the vagus nerve, treated by *en bloc* resection of the nerve with the tumor. The other case was a recurrence found during follow-up. All resected lymphnodes resulted negative for metastasis at histologic examination. The follow-up consisted of yearly clinical evaluation and duplex ultrasound of the neck. No tumor recurrence was seen during follow-up. Only one relapsed tumor was diagnosed during follow-up, in a case with familial disease, 2 months after resection of the primary tumor.

TABLE 1 - DIAGNOSTIC EXAMS.

Duplex Ultrasound	20 (100%)
Angio-CT scan	15 (75%)
Angio-MRI scan	12 (60%)
Angiography	3 (15%)
PET-CT scan	1 (5%)
MIBG scan	1 (5%)
Genetic Analysis	1 (5%)

TABLE 2 - POSTOPERATIVE COMPLICATIONS.

Postoperative complications	*Group 1*	*Group 2*	*Group 3*
Permanent neurological damage	0	0	1
Horner's syndrome	0	1	0
Hypertension	1	1	0
Stroke	0	0	0
Dysphagia/sore throat transient	2	0	1
Dysphonia	0	1	0
Postoperative bleeding	0	1	0

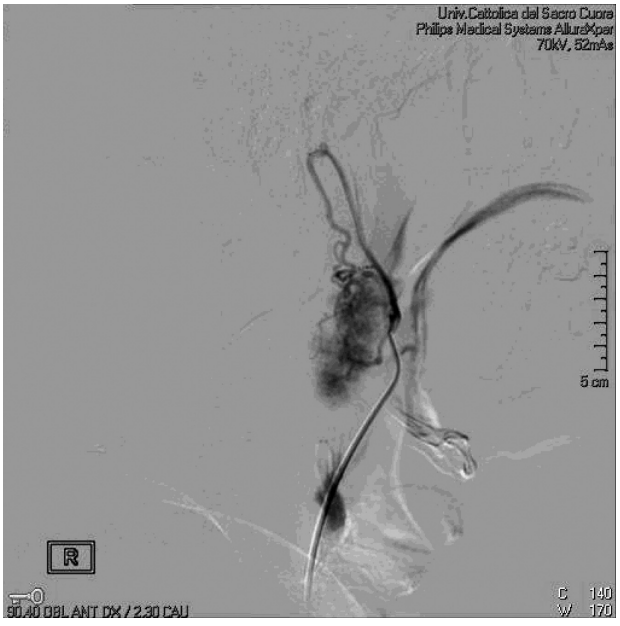

Fig. 1 - Preoperative angiography.

Discussion

CBT are uncommon tumors and some considerations regarding their optimal treatment have emerged from our study of this 12 years period. The incidence of familial CBT in our series (30%) is similar to other series (8, 12, 20); also the finding of painless mass as the most common clinical presentation is in agreement with others

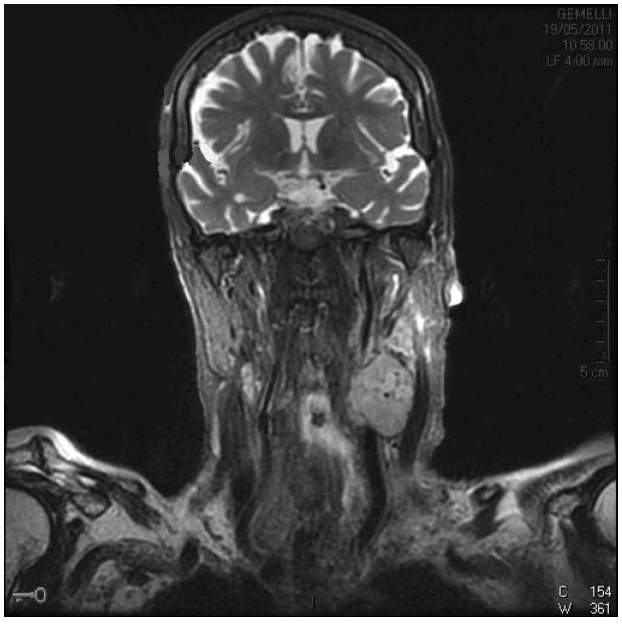

Fig. 2 - MRI. Salt and pepper lesion.

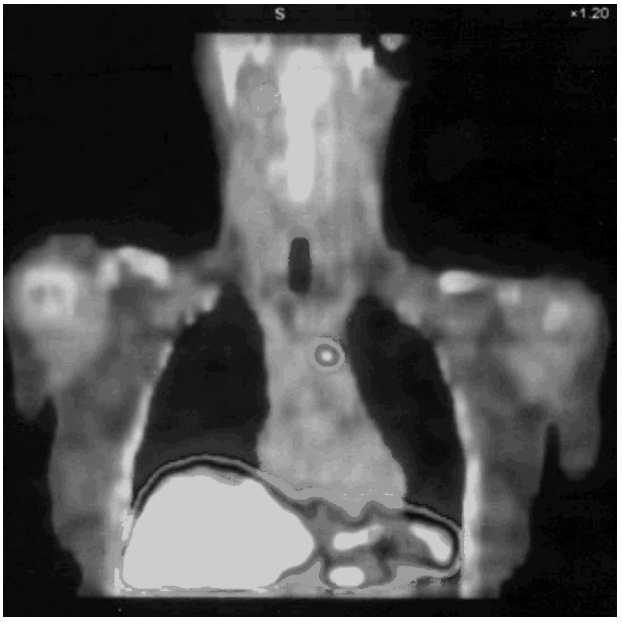

Fig. 3 - MIBG-Scintigraphy.

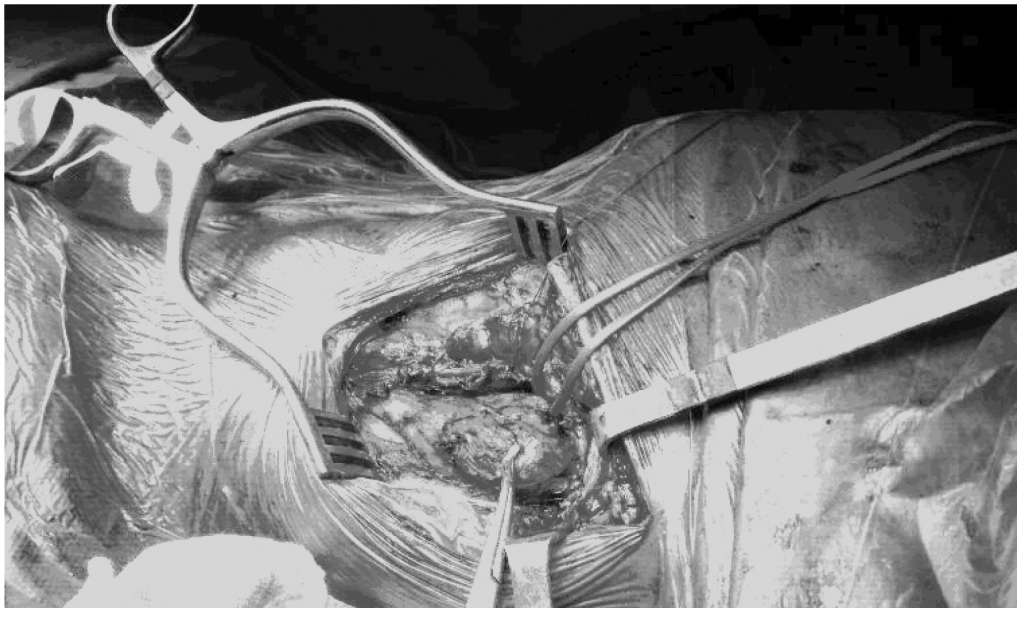

Fig. 4 - Intaoperative image.

authors (6, 7). Preoperative diagnosis is crucial; differential diagnosis is with thyroid nodule, lymphadenopathy, and brachial cysts. Fine-needle biopsy (FNB) has been suggested but most authors do not recommend such procedure because of the hypervascularization of the tumor and its proximity to nervous and vascular structures Moreover, cytologic evaluation cannot differentiate benign from malignant lesions. In our series 3 patients (15%) had FNAB in another hospital. Integrated use of non-invasive imaging techniques, can give correct diagnosis in most cases with adequate sensitivity and specificity. Duplex-scan is usually the first diagnostic step; it can usually provide information such as tumor size, its position and relationships with the vascular structures and intralesional blood flow signals; Angio-CT scan and angio-RMN provide further details about regional extension of the tumor and eventual vascular encasement. Selective carotid angiography can identify the feeding vessels of the tumor, usually rising from pharyngeal and external carotid artery, but such an invasive procedure

should be used only to perform superselective embolization with the aim of preoperative tumor shrinking, reducing vascularisation and intraoperative bleeding (11, 13, 14). Preoperative embolization is suggested in high risk tumors (group 2 and 3) (11, 12) and also in our series 2 patients (1 from group 2 and 1 from group 3) underwent the procedure because of CT evidence of vagus nerve encasement. We perform embolization the day before the surgical resection to avoid any perilesional inflammatory reaction and to achieve the best reduction of intraoperative bleeding. CBTs are usually non-functioning; we did not found clinical or laboratory findings suggestive of endocrine activity in any of our patients. However, the use of a second level diagnostic imaging technique such as CT, MRI or MIBG for total body study in familial cases or multifocal disease is highly recommended.

We did not use the classification proposed by Shamblin (15), because this classification is useful for predicting vascular morbidity but not neurological morbidity (16). The location of CBTs implies the risk of intraoperative cranial nerves injuries (8, 9), with higher rates in bilateral and/or large tumors (8, 10); thus it is very important to assess the preoperative status of cranial nerves potentially involved (VII, X, XI and XII). Surgical excision must include lymph nodes with suspicious morphology or closely adjacent to the tumor predominance. Visualization and careful dissection of the principal regional nerves (vagus nerve, hypoglossal nerve and the superior laryngeal nerve) is mandatory. Careful and accurate dissection of the tumor should be performed along the sub adventitial plane or "White Line" suggested by Gordon Taylor (17); in this way it is possible to separate the tumor from the artery through a relatively avascular plane (18, 19).

Conclusions

The evolution of imaging techniques has improved the diagnostic opportunities of CBTs in the last years, and integrated multidisciplinary approach seems indispensable. On the basis of our personal experience and of the reported experiences we can draw some conclusions:

- Surgical removal of the carotid paragangliomas is the only curative treatment and should be considered as the first therapeutic option (4, 20).
- Excision should be as conservative as possible preserving vascular and adjacent nerves; demolitive interventions should be limited to cases of actual locoregional invasion.
- Family screening in patients with hereditary paraganglioma carotid is very important, if we consider the high incidence of these tumors associated with specific genetic alterations; moreover, these patients show more aggressive tumors so a early diagnosis should be obtained in relatives (7, 21).
- In our opinion, a careful planning of the intervention by integrated imaging and a full mastery of the surgical technique are necessary to minimize the risk of complications, with particular reference to neurologic ones (transitory and/or permanent).
- The usual histological criteria of malignancy do not apply to CBTs; for this reason a lifelong follow-up is crucial, particularly in patients with familial disease or sporadic lesions in an advanced stage.

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