DIAGNOSIS OF CAROTID BODY PARAGANGLIOMAS BY VARIOUS IMAGING TECHNIQUES

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ABSTRACT

Introduction: Carotid body paragangliomas are rare hypervascular lesions arising from neural crest paraganglia cells, with a tendency of slow, but progressive growth, giving rise to external compression, or/and involvement of the carotid arteries, cranio-facial nerves and other neighbouring structures.
Patients and methods: We analysed demographics, mode of presentation, imaging features, Shamblin classification, treatment modalities, and neurological complications (stroke, cranial nerve injuries), of carotid body paragangliomas, in order to find specific signs and to elaborate a strategy for diagnosis and treatment.
Results: One patient had two localisations (the second was a glomus tumor of the left prelacrimal sac) and another one had a family history for carotid body tumor. All lesions were paragangliomas of the carotid bifurcation, represented by painless lateral neck mass. There was no evidence of functional tumor. The tumors were confirmed during ultrasonography (4 patients), MRI with MRA (4 patients) and DSA (2 patients). No preoperative embolisation was performed in our patients before complete resection of paragangliomas. Postoperatively, 2 patients had a transient twelve cranial nerve deficit. No stroke occured.
Conclusions: Early diagnosis of carotid body paragangliomas is possible now with color Doppler sonography and MRI+MRA. Early surgery for paragangliomas minimized the risk of complications associated with large tumours.
Key words: carotid body paragangliomas, painless lateral neck mass, color Doppler sonography, MRI+MRA.

INTRODUCTION

Carotid body tumors are primary tumors of vascular structures, which are unusual (they represent only 0.04% of cervical tumors) (Zizi A et al,2000). They arise from mesoblastic (chemodectomas) or neural crest paraganglion cells. (Padberg FT et al, 1983) Paragangliomas are hypervascular masses which are, sometimes familial. (Merino MJ et al, 1981) Only 5% are bilateral, metastases being found in 2-8% of cases (Win T et al, 1995). Local recurrence is uncommon and is usually delayed for many years. They have a tendency of slow, but progressive growth, giving rise to external compression, and/or involvement of the carotid arteries, cranio-facial nerves and other neighbouring structures, and manifestating as dysphagia and hoarness, althrough dyspnea, Horner's syndrome and facial pain may also occur (Harrington HJ et al, 1983).

We analysed demographics data, clinical characteristics, imaging features and neurological complications of patients (pts) with histologically confirmed paragangliomas, in order to find specific clinical signs, to compare the different imaging techniques, and to define a better standardized proceeding in the management of these tumors.

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PATIENTS AND METHODS

The study included between February 2003 and March 2008 four patients (3 females, 1 male), mean age 49.75 years, range 42 to 54 years, who underwent surgery for unilateral carotid body tumors arising from paraganglion cells. Patients diagnosed with neck paragangliomas were given a medical history questionnaire. Familial disease was initially determined by pedigree analysis (Isik AC et al, 2006). All patients underwent a complete head and neck examination, through clinical neurological, otorinolaryngological, and ophtalmological practices. The clinical features of paragangliomas were represented by: site, size, consistency, and pulsatility of the neck masse, the possible neurological complications (stroke and/or local cranial nerve involvement), and Shamblin classification (Matticari S et al, 1995) (Westerband A et al, 1998). The initial evaluation included colour Doppler sonography (Esaote MyLab 50), magnetic resonance imaging (Signa Horizon Lx 1.0T), and 2 D time of flight (TOF) magnetic resonance angiography. The characteristics findings on MRI+MRA were analysed for lesion shape, margin, signal intensity, angle of common carotid bifurcation, and the relationship between the great vessels and the carotid space mass. Digital substraction angiography was performed in 2 patients to define the vascular anatomy of the lesion. Intraoperative somatosensorial Evoked Potentials (SEP) monitoring has not been used (Matticari S et al, 1995).

RESULTS

A) Clinical features

The tumors were discovered on clinical or self examination. Pathologically, there were four unilateral typical paragangliomas of the carotid bifurcation. One patient had two localizations (the second was a glomus tumor of the left prelacrimal sac) and another one had a family history for carotid body tumor.

The demographics and clinical features are presented in table I.

Table I. *Pts demographics and clinical features*

Pt	Age	Sex	Site/ size of the neck mass	Consistency of the neck mass	Pulsatility of the neck mass	Postoperative local cranial nerve involvement	Postoperative stroke
1	54	M	Left 4.7/3.2	Non tender	Yes	Yes	No
2	53	F	Right 3.5/2.1	Non tender	Yes	No	No
3	50	F	Left 3.8/2.5	Non tender	Yes	No	No
4	42	F	Left 4.4/2.7	Non tender	Yes	Yes	No

A painless lateral neck mass was the main finding in all patients. There was no evidence of bilateral involvement of carotid bifurcation, and/or clinically functioning tumors in any case. All patients underwent successful excision of the tumor. Postoperative complications were transient cranial nerve deficits (patients 1 and 4) and a permanent Horner's syndrome (patient 1). No stroke occurred. During the follow-up, none of the patients developed local recurrences or metastases.

Patient 1

A 54 old man was detected with a left carotid body tumor (stage II Shambling). Surgical technique was based on resection performed from the medial surface of the carotid bifurcation which has been partially absorbed into the mass of the tumor. Postoperatively, this patient had a transient left XII cranial nerve injury, and a permanent Horner's syndrome. No stroke occurred. The follow-up period was 2 years and 6 months, without signs of local recidivation or metastatic disease (figure 1) (figure 2).

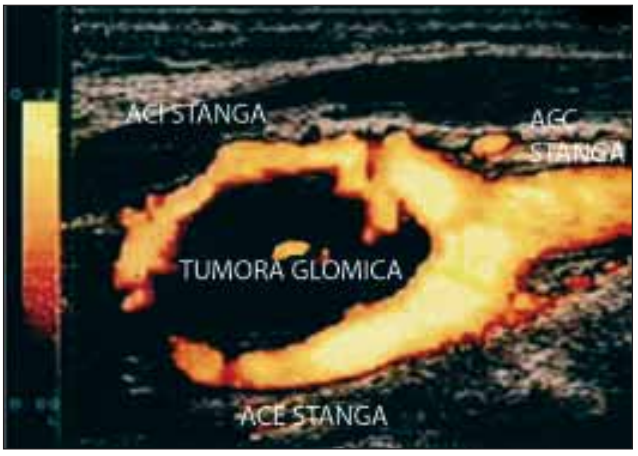

Figure 1. (first patient): *glomus tumor of the left carotid body. Colour Doppler sonography: left cervical mass widening the carotid bifurcation*

Patient 2

A 53 old woman. The suspicion of right carotid artery aneurysm was the indication for urgent operation. The intraoperative finding showed a right carotid body tumor (stage II Shambling), with a slight compression of the carotid arteries. The

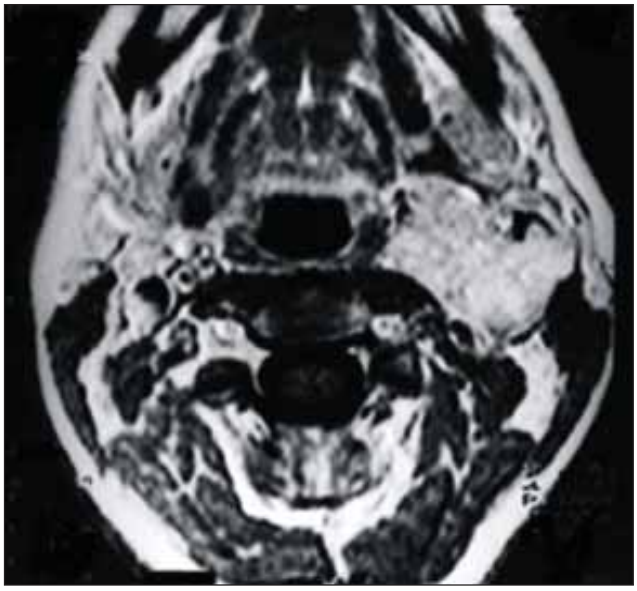

Figure 2 (first patient). *glomus tumor of the left carotid body. MRI features: axial T1 after Gadolinium; cervical mass widening the carotid bifurcation, with intense enhancement. Typical "salt and pepper" aspect due to the serpentine areas of signal-void*

Figure 3 (patient 2). *glomus tumor of the right carotid body. Microscopic examination*

subadventicial removal of the tumor was done. No neurological complications occurred. She presented an associated glomus tumor of the left prelacrimal sac and extended to the lower and temporal side in front of the medial palpebral ligament. This second tumor was whole extirpated (fig 3) (fig 4). The patient was followed-up for 2 years without signs of local recidivation.

Patient 3

A 50 old woman. Familial disease was initially determined by pedigree analysis. After subadventicial surgical removal of the tumor (stage II Shambling), without clamping or injuring the carotid arteries, the patient was followed-up for 2 years, and was without signs of local recidivation or metastatic disease (fig 5) (fig 6).

Patient 4

A 42 old woman. The intraoperative finding showed a left carotid body tumor (stage II Shambling). After subadventicial surgical removal of the tumor, a temporary left XII cranial injury occurred, with slight deviation of the tongue and

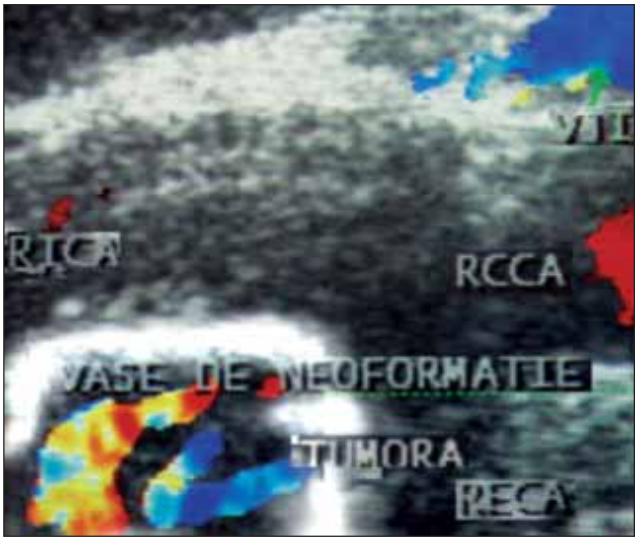

Figure 4 (patient 2). *glomus tumor of the right carotid body. Color Doppler sonography: right cervical mass widening the carotid bifurcation*

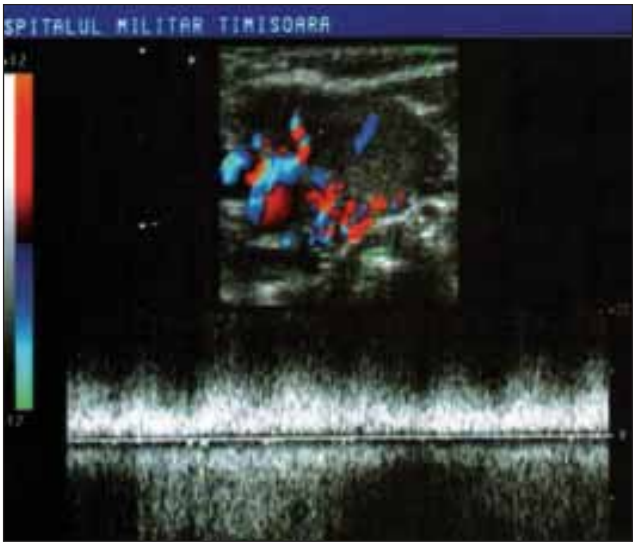

Figure 5 (patient 3). *glomus tumor of the left carotid body. Color Duplex ultrasonography: left cervical mass widening the carotid bifurcation. Accelerated and turbulent flow in the vessels of the tumor*

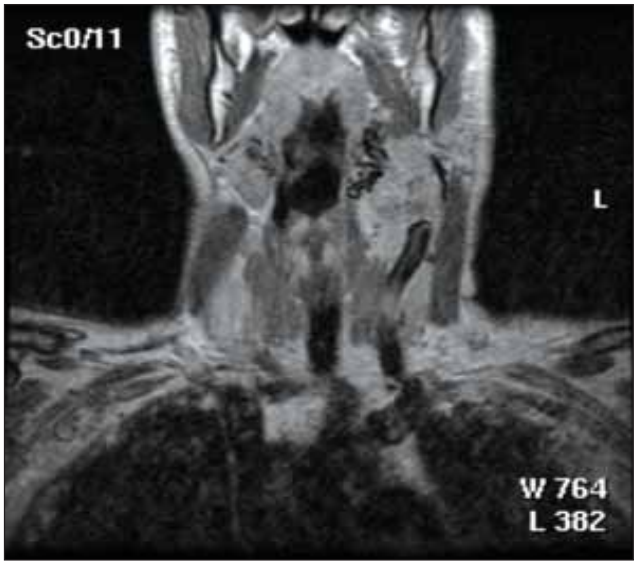

Figure 6 (patient 3). *glomus tumor of the left carotid body. MRI features: coronal T1 with enhancement. Large mass of the left carotid space*

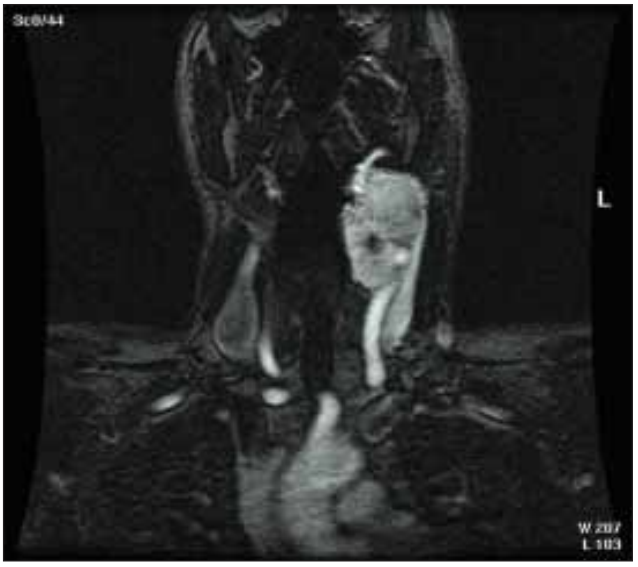

Figure 7 (patient 4). *glomus tumor of the left carotid body. MRA with dynamic enhancement. Original image with good visibility of the enhancement and the hypervascularization of the tumor*

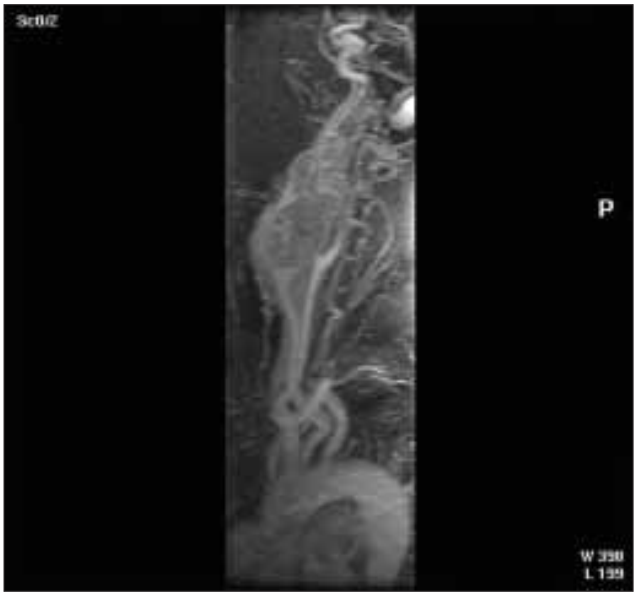

Figure 8 (patient 4). *glomus tumor of the left carotid body. MRA with dynamic enhancement.. Reconstruction MIP. Note the hypervascularization of the tumor and the good visibility of the enhancement of the tumor*

scurred speech. On 3 years of followed-up, the pt was free of local recurrences and metastases. (fig 7) (fig 8).

B) Imaging techniques

The diagnosis, prior to surgery, was achieved through ultrasonographic and MRI-MRA studies, as well as selective angiography (digital substraction).

I-Doppler ultrasonography features:

1) B-scan ultrasonography revealed well-defined, solid, even weakly hypoechoic masses, commonly broadening the carotid bifurcation.
2) An arterio-venous shunt could be seen using the Doppler technique (low IR, with accelerate and turbulent flow in the vessels of the tumor) (Koch T et al, 1990) (fig 5).
3) The diagnostic value of color Doppler sonography in the evaluation of these tumors resisted in a characteristic wide splaning of the carotid bifurcation by a hypervascular mass (the highly vascularized tumor), with shifting of the internal carotid artery posteriorly and laterally and of the external carotid artery anteriorly and medially (fig. 1) (fig 4) (fig 5).
4) Power Doppler sonography showed abundant flow, characterized as an intense blush, throughout the entire tumor in all patients. (Arslan H et al, 2000)

II- MRI-MRA features and DSA features:

1) MRI indicated the full extend of the tumors and provided confirmation about infiltration of adiacent structures (fig 2) (fig 6).
2) MRA confirmed the diagnosis, demonstrating a vascularized tumor in the carotid bifurcation. The carotid body tumors of our patients derived their blood supply from the external carotid artery. (fig 7) (fig 8).
3) DSA was performed in patients 1 and 4 to define the exact vascular anatomy of the lesions.

DISCUSSIONS

Carotid body tumors present a diagnostic challenge. Despite technologic diagnostic advances,

misdiagnosis resulting from a blind biopsy or exploration through a limited incision still occur (Worsey MJ et al,1992).

The present study reviews our experience in the diagnosis and treatment of the paragangliomas of the carotid bifurcation, which are rare hypervascularised tumors with slowly growth (Win T et al, 1995). They are more frequent than, in order, vague, thymphanic and jugular paragangliomas (Zizi A et al,2000). Sometimes, they present multicentricity (4%) (Zizi A et al, 2000), like our patient number 2 (with a glomus tumor of the left prelacrimal sac), being more common in patients with a familial history (10%) (Zizi A et al, 2000) (like our patient number 3). In the literature (Zizi A et al, 2000), the mean age of patients with paragangliomas is 40 years, with a high frequency between 30 and 60 years (Win T et al, 1995); our patients fitted as well between this range of age. The increasing use of sophisticated imaging modalities may allow earlier discovery of carotid body tumors, before they cause clinically manifestations (Westerband A et al, 1998). Generally, carotid body paragangliomas are manifestated as asymptomatic lateral neck masses (all our patients), like vagal paragangliomas. (Isik A.C et al, 2006) Sometimes, they determine disphagia, hoarseness, etc, due to their external compression and/or involvement of the neighbouring structures (Harrington HJ et al, 1983). In exchange, pheochromocytomas typically determine uncontrolled hypertension, and jugular paragangliomas present skull base extension, with symptomatic relief (Antonitsis P et al, 2006). No early or late recurrence occurred and no metastases were detected for our patients at follow-up, like in the great majority of published studies (Zizi A. et al, 2000).

Taking into consideration the practicability and invasiveness of the investigations, as well as the risks and costs involved, and last, but not least, the international literature, we propose that the imaging techniques be performed in the following sequence: 1-Doppler ultrasonography, 2-MRI, 3-MRA.(Koch T et al, 1990) DSA has to be used only in selected cases, for detect the vascular anatomy of the tumors, with eventually preoperative embolisation, for reducing tumor vascularity (Koch T, 1990).

Doppler ultrasonography is adequate to establish the diagnosis of a glomus carotidian tumor. A characteristic feature of these tumors found with Doppler ultrasonography is wide splaning of the carotid bifurcation by a hypervascular mass (Worsey MJ et al, 1992), (Zizi A et al, 2000). B-scan ultrasonography reveals an inhomogenous mass that pushes the carotid arteries apart and an arteriovenous shunt may be seen using the Doppler technique (Arslan H et al, 2000) (patient no. 3). Colour Doppler sonography permits differentiation of vascular malformations in the neck from solid and chystic tumors. (Delcker A et al, 1991) (schwanoms have central necrosis and peripheral hypervascularization) (Zizi A et al, 1995). However, there are difficulties with other entities, like ganglion's metastasis of thyroid or breast cancer. This combination of ultrasound findings is absent in cases of sympathetic neuromas (Schreiber J et al, 1996). Color Duplex imaging proved to be as accurate as DSA in the imaging of the great arteries and their relationship to the tumor, as well as of the small tumor's feeding vessels (Schreiber J et al, 1996). If the tumor is considered by the surgeon to be too small to warrant immediate exclusion, the patient is followed by periodic duplex scanning (Worsey MJ et al, 1992). Sonographic examination of the carotid body tumors is highly accurate. It is safe and reliable (Schreiber J et al, 1996).

MRI-MRA contribute to additional information about the tumor extension and to definitive treatment-plan, in patients with familial paragangliomas, like patient number 3. These techniques lead, sometimes, to the identification of an afferent vessel of the tumor, which could be selective embolised. (Zizi A et al, 2000). Carotid body tumors derive their blood supply, primarily from the external carotid artery, but may recive branchies from the internal carotid bulb (Zizi A et al, 1995). Usually, they displace the ICA, but do not cause obstruction of this vessel (all four cases).

If DSA leads to identification of an afferent vessel, selective embolisation of the tumor may be performed, thus enabling a better operation, with less blood loss (Koch T et al, 1990), (Isik AC, et al, 2006), (Schreiber J et al, 1996).

Because the natural history of carotid body tumors is believed to be unpredictable, immediate surgical removal is recommended (Matticari S et al, 1995). The surgical treatment of carotid body tumors is also a challenging complex problem. Combined therapeutic approach, (with eventually preoperative selective embolisation) followed by surgical resection by an experienced team offer a safe and effective method for complete excision of the tumors with a reduced morbidity rate. No preoperative embolisation was performed in our patients. Depending on multiple factors, different surgical technique are indicated (Matticari S et al, 1995):

a)-subadventicial resection of the tumor (all 4 patients);

b)-ligation of external carotid artery and internal carotid artery reconstruction with resection of the tumor were never necessary in our cases. Resection of carotid body tumors can be difficult to perform because of it's site, vascularity, arterial adherence and local cranial nerve involvement (Matticari S et al, 1995). Thus, the larger the tumor is, the more difficult the resection is, and more injuries are caused to the surrounding nerves and vessels (Kiris M, et al, 2003). Intraoperative meticulous dissection may decrease the incidence of postoperative complications (stroke, death, nerves involvement) (Antonitsis P et al, 2006).

CONCLUSIONS

1) Paragangliomas are hypervascularized carotid body tumors, that should be considered in the evaluation of all lateral neck mass.
2) Their early diagnosis is possible now with Color Doppler sonography and MRI+MRA.
3) Doppler ultrasonography has an important role in the detection of the vascular nature of these tumors (identifications of cervical arteries and their relationship to the tumor, as well as of the small tumor's feeding vessels; these tumors are hypervascularized masses with a characteristic wide splanning of the carotid bifurcation).
4) Color Doppler scanning is the noninvasive modality of choice for the primary diagnosis of carotid body tumors. Additionally, it may be useful in screening for familial carotid body tumors and sequential follow-up of nonoperatively managed tumors.
5) MRI-MRA is considered as the gold standard imaging technique for the evaluation of carotid space tumors, as it allows a multiplan approach which is important in the preoperative study.
6) Early surgery for paragangliomas minimizes the risk of complications associated with large tumors.

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