



REVIEW

Paraganglioma of the carotid body: Report of 26 patients and review of the literature



Zainine Rim, Bechraoui Rim *, Chahed Houda, Jbali Souheil, Beltaief Najeh, Besbes Ghazi

Ear, Nose and Throat Department, La Rabta Hospital, Tunis, Tunisia

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KEYWORDS

Paragangliomas;
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Abstract *Introduction:* Paragangliomas are extra-adrenal tumors originating from the neuroectoderm, 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.

Objectives: The aim of the present study was to review clinical profile, treatment outcomes and long-term follow-up in patients with paragangliomas of the carotid body.

Materials and methods: It is about a retrospective study of 26 patients (28 paragangliomas) followed and treated in Ear, Nose and Throat Department of La Rabta Hospital. Pre-, intra- and postoperative findings were analyzed.

Results: The present study included 6 men and 20 women. Bilateral involvement of the carotid glomus was noted in 2 cases and tympano-jugular location was associated in 2 other cases. Ultrasound of the neck, computed tomography (CT) and magnetic resonance (MR) tomography were performed in 13, 17 and 10 cases, respectively. 2 patients have had preoperative embolization and 22 patients, in total, were operated. External radiation was an exclusive therapeutic option in 2 patients and adjuvant to incomplete surgery in one patient. Only one case of malignant paraganglioma was noted that evolution was, in fact, rapidly fatal.

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* Tel.: +216 98255815.

E-mail address: rbechraoui@yahoo.fr (B. Rim).

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1. Introduction

Carotid paragangliomas are tumors developed from chromaffin cells of the extra-adrenal tissue. Clinical presentation of these tumors may be misleading and the treatment, often, arduous. Hence, specific pretreatment assessment is the only guarantee of adequate management. The development of diagnostic and interventional radiology techniques has, obviously, improved the prognosis of these tumors.

2. Material and methods

Over a 27-year period (1981–2007), 26 patients were treated for 28 carotid paragangliomas in the Ear, Nose and Throat Department of La Rabta Hospital. Their clinical and operative data were collected and analyzed retrospectively. Preoperative work-up in patients with carotid paraganglioma consisted of careful family history, clinical examination, ultrasound of the neck, computed tomography (CT) or magnetic resonance (MR) tomography. The Shamblin classification was used to grade the tumor size. Treatment modality, including the necessity of preoperative interventional embolization, was assessed by the operating surgeons and vascular radiologist. Postoperative follow-up consisted of clinical evaluation 1 month after surgery, ultrasound 1 year after and CT or magnetic resonance imaging (MRI) if new pathological findings occurred in ultrasound.

3. Results

Our study includes 26 patients. Mean age was 38 years, ranging from 18 to 75 years. There was a female predominance with 20 women and 6 men (sex ratio = 0.3). Carotid paragangliomas were bilateral in 2 cases (10%) and associated with tympano-jugular location in 2 cases.

A family history of paraganglioma was found in 3 cases who were a patient and his 2 nieces.

- This patient and one of his nieces had a double localization. Bilateral carotid localization in one case and a carotid paraganglioma associated with contralateral jugular location in the other case.
- The last patient had a carotid body paraganglioma on a single side.

Consultation average delay was 3 years (6 months–15 years). The chief complaint was a slowly growing, painless lateral neck mass associated with high dysphagia in one case. The carotid paraganglioma was an incidental finding on the occasion of an MRI angiography performed as part of explorations of tympano-jugular paraganglioma in 1 case.

The paraganglioma was discovered intraoperatively during a cervicotomy performed for a supposed chronic lymphadenopathy in 3 cases.

Symptoms related to catecholamines release, giving place to hypertensive crises and arrhythmias, were not reported in our patients.

On physical examination, the mass was located at the high or middle jugulo-carotid region. Its average size was 4 cm (range from 2 to 6 cm). Limited mobility in transverse direction has been reported in 14 cases (54%). Pulse-like sensation and palpated thrill, testifying the vascular character, were noted in 14 (54%) and 2 cases (9%) respectively. Examination of the oropharynx allowed objectifying a parapharyngeal bulge in 1 case.

Otosopic examination revealed the existence of a bleeding tumor filling the external auditory canal in 2 cases.

Ultrasound of the neck was performed in 13 patients. It was suggestive of the diagnosis in 7 cases showing a well limited hypoechoic solid mass, located in the carotid bifurcation and vascularized in Doppler ultrasound exam. In the other cases, it evoked a cervical lymphadenopathy except in one case where it was in favor of a parotid tumor.

CT was performed in 17 patients. It helped to advance the diagnosis in 15 cases. The suggestive aspect was that of a carotid space mass which splay the internal and external carotid arteries at the level of the bifurcation and demonstrates intense homogenous enhancement following the intravenous administration of iodinated contrast (Fig. 1).

In one case, the evoked diagnosis was metastatic lymphadenopathy due to the presence of a thyroid suspicious nodule in both ultrasound and cytology.

The MRI was performed in 10 patients. The carotid paraganglioma took the appearance of a lesion sitting at the carotid bifurcation with intermediate and heterogeneous signal on T1 and T2 sequences. Hypointense flow voids of punctuate, serpiginous or channel-like structures on the set of sequences, achieving an aspect of “salt and pepper” is a distinctive feature in MRI. Intense and homogenous enhancement following the intravenous administration of gadolinium is the last character noted in our patients (Fig. 2).

The MR angiography, performed in 2 patients, showed the spacing of the carotid bifurcation and confirmed the permeability of vessels. It also showed the absence of the posterior communicating artery in 1 case. Nevertheless MR angiography was not able to identify the feeding pedicles in our study.

Angiography was performed in 16 patients allowing to confirm the diagnosis in all cases (Fig. 3).

Biologically, measurement of urinary VMA (vanillylmandelic acid) was performed in 12 patients. It was normal in all cases.

A fine needle aspiration was performed in 10 patients without incident, fortunately. It was inconclusive in 9 cases and in favor of metastatic lymphadenopathy in 1 case.

Concerning treatment, 22 patients were operated. Two patients had both large tympano-jugular and carotid paragangliomas. One of them received radiation therapy to the first location and was operated for the second one. The other was lost.

Three patients have already had a cervicotomy in another department for the diagnosis of chronic cervical

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