



Carotid Body Paraganglioma: Study of 5 Cases Submitted For Embolization and Surgery

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Abstract

Objectives: Report the treatment of 5 cases of patients with paraganglionic carotid body tumors who underwent preoperative embolization and surgery.

Case details and treatments performed: Five cases of patients with paraganglionic tumors were diagnosed and treated, with a mean age of 42 years. Most of them were female, with complaints of tumor in the lateral cervical region, pulsatile and painless, with no history of trauma and comorbidities. All patients underwent pre-embolization carotid angiography of the tumor, and then resection of the glomus tumor with preservation of the internal and external carotid arteries. After surgery, most patients developed dysphonia.

Conclusion: The case studies show satisfactory results in carotid body tumor treatment with the association of preoperative embolization and surgical resection.

Keywords: Carotid Body Tumor; Embolization therapeutic; Surgery; Head and neck neoplasms

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Introduction

The paraganglioma is a type of carotid body tumor located at the bifurcation of the common carotid artery at the level of its adventitial layer, which develops from neural crest-derived paraganglion cells, with an estimated incidence of 1/30,000 to 1/100,000 [1-4]. It represents about 3% of all paragangliomas that occur in the head and neck, having as anatomically most prevalent sites: the carotid body, the jugular body, the vagus nerve and along the tympanic branch of the glossopharyngeal nerve [4,5]. Of the three different types of carotid body tumors described in the literature, the sporadic form has been the most common, representing approximately 85% of carotid body tumors, appearing on average at 45 years old. The familial type may occur in 10% to 50% of cases in young individuals in the second decade of life. While the hyperplastic type is related to patients with chronic hypoxia, including patients with chronic obstructive pulmonary disease or cyanotic heart disease, and patients living at high altitudes [3,6]. Although carotid body tumors are mostly benign, about 9% of cases follow a malignant course with metastases to the regional lymph nodes, viscera and bones [5]. Thus, for early diagnosis, medical history and physical examination become essential because the carotid body tumor, in general, manifests as a firm, pulsatile, painless lateral cervical mass, located below the mandible angle, movable in the lateral direction (Fontaine sign) with slow evolution. It may cause localized pain, hoarseness due to involvement of the vagus nerve (upper laryngeal branch), involvement of the hypoglossal or glossopharyngeal nerves, Horner syndrome with invasion or compression of the cervical sympathetic chain and syncope, possibly due to compression of the carotid sinus or the internal carotid artery. Other symptoms such as tachycardia, palpitation, hypertension, and tremors may indicate possible endocrine activity [7]. Imaging tests are of great relevance in the diagnosis of carotid body tumors. Direct biopsy is considered an inappropriate procedure due to the vascular nature of this type of tumor [6]. Thus, and as a consequence of the possibility of malignant transformation, peritumoral invasion and metastasis, surgical resection with or without concomitant preoperative endovascular embolization or radiotherapy has been the most used technique of choice for treatment [3,6]. The aim of the present study was to report the treatment of 05 cases of patients with paraganglionic carotid body

tumors who underwent preoperative embolization and surgery.

Case Series

Case 1

A 34-year-old male patient sought medical attention complaining of severe sporadic pain in the cervical region with no history of trauma, and denied comorbidities. On physical examination, a palpable mass was observed in the pulsatile and hardened cervical region. The patient underwent preoperative embolization and the surgical procedure was performed with resection of the glomus tumor with preservation of the internal and external carotid arteries. After surgery, the patient presented transient ischemic accident during arteriography and developed sporadic pain on exertion and yawning.

Case 2

A 62-year-old female patient sought medical attention complaining of tumors in the left cervical region, denying neurological symptoms, hypertension, diabetes mellitus, smoking habits and trauma in the region. On physical examination, a palpable, hardened, pulsatile left cervical mass was observed. The patient underwent pre-embolization carotid angiography of the tumor (Figure 1a), and after surgical resection of the paraganglioma, we can observe the preserved internal and external carotid arteries (Figure 1b). After surgery, the patient evolved well and without complications.

Case 3

A 48-year-old female patient sought medical attention complaining of bulging in the left cervical region, denying other associated symptoms without comorbidities. On physical examination, a palpable mass was observed in the pulsatile and hardened cervical region. The patient underwent preoperative embolization and the surgical procedure was performed with resection of the carotid body tumor with preservation of the internal and external carotid arteries. After surgery, the patient developed dysphonia.

Case 4

A 43-year-old female patient sought medical attention complaining of cervical mass, denying other associated symptoms, no previous pathologies and no history of trauma. On physical examination, a palpable mass was observed in the pulsatile and hardened cervical region. The patient underwent preoperative embolization and the surgical procedure was performed with resection of the glomus tumor with preservation of the internal and external carotid arteries. After surgery, the patient evolved without complications.

Case 5

A 23-year-old male patient, with brown skin, sought medical attention complaining of a cervical tumor associated with local pain and headache. He had no comorbidities and denied local trauma. On physical examination, a palpable, pulsatile and hardened left cervical mass was observed. Preoperatively, the patient underwent carotid angiography (Figure 2a) and then embolization (Figure 2b). As a surgical procedure, paraganglioma resection with preservation of the internal and external carotid arteries was performed. After surgery, the patient developed dysphonia and refractory dysphagia. In all five cases of paragangliomas, pathological examinations were performed.

Macroscopic examination

The surgical specimens showed, in all cases, nodular formations, brown to brownish, solid, elastic, sometimes encapsulated, presenting



Figure 1: (a) Selective carotid angiography showing paraganglioma of the pre-embolization carotid body and (b) After total tumor excision with typical separation of the internal and external carotid arteries.



Figure 2: (a) Carotid angiography showing preoperative carotid body tumor and (b) After tumor embolization.

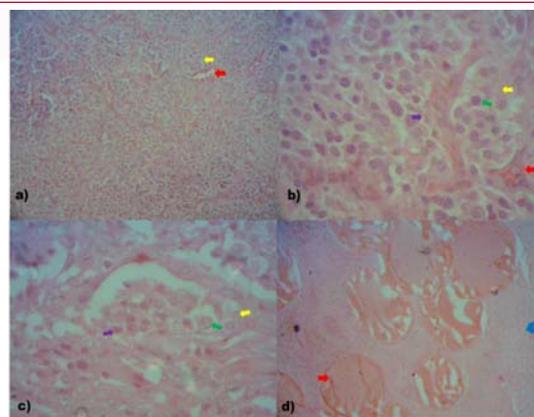


Figure 3: Histopathological examination of carotid body paragangliomas.

cystic areas filled with colloidal material, with and without foci of hemorrhage. The nodules had in their smallest dimensions 1.5 cm × 1.2 cm × 0.8 cm and the largest of 5.5 cm × 3.2 cm × 2.5 cm.

Microscopic examination

Hematoxylin-Eosin (HE) stained histological sections, where it can be observed a solid nest-like architectural pattern (Zell ballen or cell ball pattern-yellow arrow) surrounded by capillaries (red arrow) with regular morphology, with small, slightly hyper chromatic nuclei, an abundant eosinophilic cytoplasm, showing neoplasia adjacent to blood vessels (Figure 3a). The Zell ballen pattern of neoplastic cells consists of two cell types: the main cells (green arrow) with abundant

clear cytoplasm and hyper chromatic nucleus and the sustentacular cells (purple arrow), which are thinner, spindle-shaped and located peripherally around the nests (Figure 3b). We can still observe cells with small nuclei, abundant and slightly eosinophilic cytoplasm, showing neoplastic cells with dilated and congested blood vessels (Figure 3c and 3d).

Discussion

The literature reports that carotid body tumors can appear at any age, but their incidence begins to increase from the fourth decade of life, peaking from the fifth to the seventh decades [8]. Recent studies have shown a higher incidence of these tumors in females, present in the sporadic form of paraganglioma, which has a unilateral presentation in 90% of cases [9-14]. According to some authors, the relation between women/men is quite variable, from 1.4:1 to 7.3:1 [8,15]. The five patients reported in the present study with carotid body tumors ranged from 23 to 62 years old, with an average of 43.2 years, three were female and two male (3:2). All five paraganglionic tumors were unilaterally located and the patients had no family history of this type of cancer. The paraganglioma has a slow growth and may remain asymptomatic for several years. The most common form of presentation is a painless cervical mass along the anterior border of the sternocleidomastoid muscle, between the internal and external carotid arteries (Kocher's Sign I), movable in the horizontal plane and fixed vertically (Fontaine's sign). Occasionally, the tumor mass may transmit or be noticeable on the carotid pulse, like a murmur. Bidigital palpation (external and intraoral) may reveal a localized mass in the tonsillar region (Kocher's Sign II) [9,12,13]. When symptomatic, the most common manifestation of the paraganglioma is cervical pain and in about 10% of cases they present paralysis involving a cranial nerve [6,16]. In the cases described in this study, Kocher's Sign I was seen in all patients and local pain associated with tumors was reported in 2 cases. To better predict surgical morbidity, Shamblin et al., [17] proposed a classification based on the experience of 90 patients treated over a 30-year period at the Mayo Clinic. Through the variable relation between neoplasia and carotid artery wall, the authors classified carotid body tumors into three groups: the first, Shamblin I, tumors are small, localized and can be resected without significant trauma to the vessel wall or capsule of the tumor; second, Shamblin II, the tumor partially involves the adventitia of the carotid vessels; and the third, Shamblin III, the paraganglioma has a close adherence relation with the entire circumference of the carotid bifurcation, making surgical dissection impossible [17,18]. In the latter, biopsy is contraindicated due to the high risk of bleeding [19]. Surgery has been the only curative treatment. Other therapeutic approaches, such as radiotherapy, are reserved for patients with unrespectable tumors. Techniques, such as percutaneous embolization or endovascular exclusion, have been used as adjunctive therapy to resect the neoplasm to prevent bleeding and reduce tumor size before surgery [18]. These techniques should be performed within 48 h prior to surgery to prevent the inflammatory reaction from hindering tumor dissection. Embolization is not free of complications, including cranial nerve injury by embolization of perineural vessels or neurological sequelae due to material leakage by anastomotic vessels [19-21]. In our study, all patients underwent preoperative embolization, and after, simple surgical resection. Regarding postoperative complications, the cranial nerve palsy rate varies between 32 and 44%, the hypoglossal, vagus and superior laryngeal nerves are the most injured, often causing definitive paralysis, while the rates of cerebral vascular insufficiency vary between 8% to 20% and the mortality rate is reported between

5 and 13% [1,22,23]. In the present study, in particular, two patients had partial lesion of cranial pairs: hypoglossal, vagus and superior laryngeal (40%), evolving with dysphonia, and one case being associated with refractory dysphagia. A case of transient stroke after arteriography and sporadic pain on exertion and yawning (20%) was reported. Two patients followed without complications (40%). Postoperative mortality was zero. The diagnosis is mainly made by imaging techniques. The carotid duplex ultrasound may locate a tumor adjacent to the carotid bifurcation, but computed tomography or magnetic resonance imaging is usually required to further delineate the tumor's relation to adjacent structures [6]. In the present study, carotid ultrasonography was performed in all patients as preoperative evaluation, and carotid angiography for preoperative embolization and magnetic resonance imaging in only one patient. Preoperative differential diagnosis includes possibilities of bronchial cleft cyst, enlarged cervical lymph node, salivary gland tumor, neurofibroma, and internal carotid artery aneurysm [17]. The pathological examination of these tumors revealed sites of round-shaped cells (Zell ballen) and polygonal cells (chief cells), surrounded by elongated sustainable cell trabeculae [24]. Tumor cells had an abundant eosinophilic cytoplasm and uniform round or oval nuclei, sporadically vesicular with or without granules. There was little cellular pleomorphism and mitoses were not frequent. Electron microscopy reveals well-defined neuroendocrine granules, which are rare in non-functioning tumors. In most cases, individual tumor cells are relatively homogeneous in appearance. However, in a minority of paragangliomas, nuclear pleomorphism, necrosis and mitotic activity can be found, which together raise the possibility of a more aggressive lesion [25].

Conclusion

Carotid body tumors are rare neuroendocrine neoplasms that require early diagnosis and appropriate multidisciplinary treatment and should be suspected in cases involving the presence of pulsatile cervical mass. Surgery is the treatment of choice despite its risks, especially in large tumors, as it provides good cure rates with minimal recurrence and morbidity. Despite advanced surgical techniques, the prevalence of postoperative cranial nerve palsy is still quite high. Therefore, the patient should be clearly informed about their illness and operational risks during the preoperative period.

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