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Surgical resection of carotid body paragangliomas: 10 years of experience

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Abstract

BACKGROUND: Carotid body tumors (CBTs) are relatively rare neoplasms, and even if they are considered predominantly benign, there is an indication for early surgical removal. The objective of this study was to conduct a review of the surgical management of CBTs.

METHODS: A retrospective study identified 34 cases (12 men and 19 women) of tumors in patients who had undergone surgical resection of pathologically confirmed CBTs over a period of 10 years from 2001 to 2011 in 2 academic departments of general surgery in Italy.

RESULTS: In our series, 10 CBTs (31%) were Shamblin class I, 13 (41%) were class II, and 9 tumors (27%) were class III. Two patients (6%) had transient cerebral ischemia immediately after operation. One patient (3%) died of postoperative cerebral ischemia after surgery for internal carotid artery thrombosis.

CONCLUSIONS: The experience of this casuistry shows that the procedure is relatively low risk for Shamblin I and II classes, whereas there is an increasing risk of neurovascular complications for Shamblin III class.

Carotid body tumors (CBTs) belong to the classification of paragangliomas because they originate from paraganglia in chromaffin-negative glomus cells derived from the embryonic neural crest, functioning as part of the sympathetic nervous system (a branch of the autonomic nervous system). These cells normally act as special chemoreceptors located along blood vessels, particularly in the carotid bodies (at the bifurcation of the common carotid artery in the neck) and in aortic bodies (near the aortic arch).

Accordingly, CBT and other paragangliomas are categorized as originating from a neural cell line in the World Health Organization classification of neuroendocrine tumors. In the categorization proposed by Wick,[1] paragangliomas belong to group II. Given the fact that they originate from cells of the orthosympathetic system, paragangliomas

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are closely related to pheochromocytomas, which are chromaffin positive. The most well-known tumor of the glomus caroticum is the CBT.

Carotid body paragangliomas are more common in women.[2–9] The incidence of bilateral carotid body lesions is approximately 10%. Most of these lesions are benign; however, malignant behavior is often encountered. For the diagnosis of malignant carotid body paragangliomas, there are no clear histological characteristics that differentiate it from benign lesions. This diagnosis is reserved for tumors with local, regional, and distant metastasis. The rate of malignancy is reported to be 6% to 12.5% of all cases[2–4,10–18]; 7% to 9% of the cases are hereditary.[1,3,5–9,19]

In 1742, von Haller described the carotid body as a glomus body-like structure situated just inferior to the carotid bifurcation.[20] Reigner first attempted resection of a carotid body paraganglioma in 1880, but the patients did not survive. Maydel was the first to remove a carotid body paraganglioma successfully in 1886, but the patient became aphasic and hemiplegic because of internal carotid artery (ICA) ligature.[21] In 1903, Scudder performed the first successful removal of a carotid body paraganglioma.[22]

CBT surgery remains a challenge for surgeons because of the tumor's location in the vicinity of important blood vessels and cranial nerves. Because of preoperative impairment of nerve function, surgery may result in deficits of cranial nerves VII, VIII, IX, X, XI, and XII.[23–25] The aim of this study was to provide a brief updated review of CBTs and to present our experience with 34 tumors that were operatively explored.

Methods

After approval from the institutional review board, a retrospective study identified 34 tumors in patients who underwent surgical resection of pathologically confirmed CBTs over a period of 10 years from 2001 to 2011 in 2 academic departments of general surgery in Italy. Patients consisted of 12 men (1 bilateral case) and 19 women (2 bilateral cases), and their ages ranged from 39 to 67 years (median = 48 years) (Table 1).

All tumors were completely resected according to the following technique. The transcervical approach was used to approach tumors with a postaural extension in 1 of the cases. After flap elevation, the proximal and distal controls of the vessels in the carotid sheath were achieved by adequate exposure by placing threads. The ascending pharyngeal artery, which is the most common feeding vessel, and other feeding vessels were ligated. The tumors were then circumscribed and were carefully dissected in a periadventitial plane using bipolar cautery and a radiofrequency scalpel, avoiding injury to the vessel wall (Fig. 1). The tumors were then released from their attachment to the surrounding tissues. The carotid was covered by temporalis fascia in all cases.

We performed a review of the medical records focused on the preoperative assessment, including a complete history and physical examination with a cranial nerve examination, and intraoperative and postoperative assessment, which focused on arterial bleedings and cranial nerve deficits. Outcome data were collected at 1 month and 1 year postoperatively. A review of surgical reports focused on cranial nerve sacrifice and carotid involvement defined as sacrifice or bypass. Pathology reports were analyzed for the presence of metastatic disease and locally infiltrative characteristics defined as the involvement of surrounding bone, connective tissue, blood vessels, or nerves.

The primary end point was functional outcome as defined by the incidence of cerebral vascular accidents. Secondary end points included the incidence of postoperative cranial neuropathies, pathological findings such as potential prognostic factors for morbidity, and the impact of preoperative cranial neuropathies on long-term functional outcome.

Neurologic assessment and functional outcome were evaluated using the National Institutes of Health Stroke Scale[26] for all patients in the first week of the postoperative period even if they did not have a detectable neurologic deficit. In this study, health-related quality of life was determined with the Sickness Impact Profile.[27] Descriptive statistics are reported as numbers and percentages.

Results

Twelve tumors (35%) were located on the right side and 21 (64%) on the left side. A multicentric paraganglioma not from the head and neck was discovered in 2 patients (7%) who had a thoracic and a paravesical paraganglioma. No patient had a family history of paragangliomas. The most common symptom was a pulsatile neck mass without cranial nerve deficit. No patient had a functioning CBT who presented with palpitations, tachycardia, and hypertension accompanying an elevated catecholamine level. Preoperative cranial nerve X dysfunction was observed in 1 patient (3%).

Table 1 Clinical presentation of 34 cases of carotid body tumors

Shamblin class	No. of patients	Side (right:left)	Sex (female:male)	Median age (y)	Preoperative cranial nerve deficit
I	10 (1 bilateral) (31%)	6:5	6:4	44 (39–58)	None
II	12 (2 bilateral) (41%)	4:10	7:5	51 (44–59)	None
III	9 (27%)	2:7	6:3	59 (39–67)	1:9 (12%)
Total	31 (3 bilateral)	12:22	19:12	48 (36–67)	1:34 (3%)