

## ORIGINAL ARTICLE

# CAROTID BODY TUMOR: SURVEY OF 97 PATIENTS

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### Abstract

**Background-**Carotid body tumor is not a common disease but it should be considered in the differential diagnosis of neck masses at the mandibular angle. Without a correct diagnosis the surgeon unexpectedly encounters leading to a fruitless termination in 23.7% of cases.

**Methods-**In this study, medical records of 97 patients with carotid body tumor were reviewed retrospectively. These patients had been referred to the vascular surgery division in a teaching hospital and one of the private hospitals in Tehran from November 1985 to July 2001.

**Results-** Male to female ratio was 1: 2 and the mean age of the patients at the time of presentation was 40 years (range: 10 - 75 years). The most common complaints of the patients at first visit included: neck mass (100%), discomfort in the neck (13.4%), dizziness (4.1%), palpitation (3.1%), hypertension (2.06%), and hoarseness (3.1%). The tumor was bilateral in five patients (5.1%). The mean interval between onset of symptoms and the first visit was 4.6 years. The size of the neck mass was less than 5 cm in 60.8% of cases and greater than 5 cm in 39.2% of cases. The mass was excised in all patients, with ligation of the external carotid artery in 10.3% of cases and ligation of the internal carotid artery and graft replacement in 5.1% of cases with no postoperative consequences. The final pathologic diagnosis included paraganglioma (86.6%), schwannoma (11.3%), and mesenchymoma (2.06%). Three cases (3.1%) were malignant paraganglioma.

**Conclusion-** Many of our patients are referred in late stages with large tumors. However, following a successful resection, complete cure will ensue with no further recurrence.

**Keywords** • Paraganglioma • carotid body tumor • neck mass

### Introduction

**P**araganglioma or chemodectoma is a rare tumor of the head and neck, which is derived from the neural crest. Histopathologically, it is similar to the adrenal gland neoplasm, pheochromocytoma.<sup>1</sup> This infrequent tumor is usually benign and non-functional. It grows and expands slowly and rarely metastasizes.<sup>2,3</sup> This tumor is classified according to its location: carotid body, jugular vein, vagal body, orbital, and laryngeal.<sup>1</sup> Sometimes they arise within the abdominal cavity, usually in the retroperitoneal space.<sup>3</sup> Since these extra-adrenal

paraganglionic cells contain very small amounts of catecholamines, a clinically significant catecholamine release is rare.<sup>1</sup>

Carotid body tumor is the most common type of paraganglioma and usually presents as an asymptomatic mass in the anterolateral region of the neck. On physical examination, this tumor is laterally mobile but vertically fixed because of its attachment to the carotid bifurcation.<sup>3</sup>

The malignant form of this tumor is very uncommon and the diagnosis is made on the basis of clinical behavior such as development of recurrence or invasion rather than histopathological appearance.<sup>4,5</sup> According to some references, only 6% to 10% of these tumors are malignant<sup>1,6,7</sup> and clinical manifestations of bone metastatic tumor

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may occur up to 17 years after diagnosis.<sup>3</sup>

Paraganglioma is a highly vascularized tumor displaying a specific pattern in angiography; compact vascular network (tumor blush) with a distinct border usually derived from the external carotid artery is seen. It is located at the bifurcation of the carotid artery and due to the size of the tumor these two branches are separated from each other. In the larger tumors, these arteries pass through the bulk of the tumor.<sup>8</sup> On sonography, paraganglioma present as a well-defined, solid hypoechoic mass and on color Doppler imaging, hypervascularity with a low-resistance flow pattern is seen.<sup>9</sup> Color Doppler and angiography are crucial in showing vascularization details of the tumor.<sup>10</sup> Fine-needle aspiration (FNA) has a beneficial role in the diagnosis of carotid body tumor.<sup>11</sup> CT-scan and MRI provide good information about the extension of the tumor.

Treatment of carotid body tumor is performed by complete resection, with subadventitial dissection.<sup>1</sup> No complication is anticipated in ligation of the external carotid artery and larger tumors render themselves easier to excision. The internal carotid artery must be saved under all circumstances. Cranial nerves also should be saved even if the nerve has to be shaved away from the tumor.<sup>8</sup> If the resection is not complete, radiotherapy should be used.

In this study, we describe demographic and clinical profiles, complication and mortality rate, as well as the pathology reports of 97 patients referred to our units with carotid body tumor during a 16-year period.

### Patients and Methods

In this study, we reviewed the medical records of 97 patients with carotid body tumor retrospectively. Fifty-two patients were referred to Ayatollah Taleghani Hospital, a teaching hospital in Tehran with vascular surgery referral center, and 45 patients were referred to Iran-Mehr Hospital, Tehran, from November 1985 to July 2001. The data were collected using code sheets and a descriptive analysis was carried out to summarize data and to count frequencies.

### Results

Sixty-five patients (67%) were females and 32 (33%) were males. Mean age of the patients at the first presentation was 40 years (range 10-75 years).

The mean age of the women at the first presentation was 40.6 years (range 10- 75) and the mean age of the men at the first presentation was 39.2 years (range 21-67). The mean interval between manifestation of the symptoms and referring to the surgeon was 4.6 years (range 10 days to 22 years). The mean interval between presentation to the surgeon and the operation was 7.3 days (range 1-50). The mean postoperative duration of hospitalization was 3.7 days (range 1-12 day). Symptoms of the patients at presentation included a neck mass in all (100%) and neck pain in 13 (13.4%). Other signs and symptoms are presented in Table 1. No syncopes or tinnitus were elicited.

Five patients (5.1%) were bilaterally affected by the tumor. The first-degree relatives of 9 patients were affected by the disease comprising a total of five families (Figure 1). History of diabetes mellitus was present in 2 (2.0%), hypertension in 8 (8.2%) and cardiac disease in 2 (2.06%) patients.

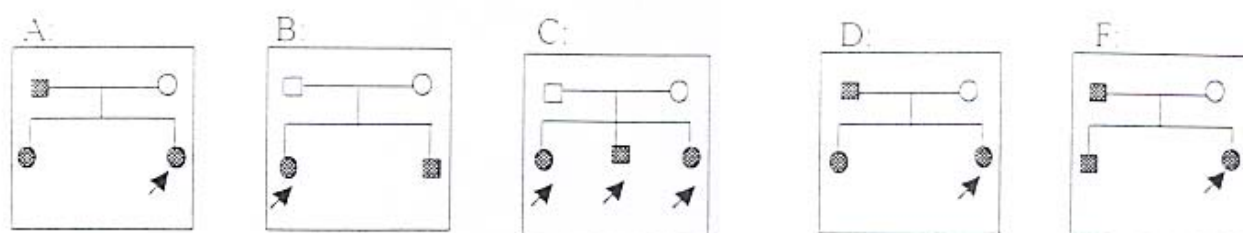
On the physical examination, blood pressure was within the range of 140/90 to 160/100 mmHg and pulse rate was within the normal limits all the patients. In 2 patients (2.06%), cardiac murmur was heard but there was no arrhythmia. Auscultation of the lungs and abdominal examination were normal in all patients.

Head and neck examination revealed that 38 patients (39.2%) had a mass larger than 5 cm and 59 patients (60.8%) had a mass smaller than 5 cm in diameter. In 13 cases (13.4%), the mass was pulsatile and in 20 patients (20.6%) a bruit was heard audible over the mass. One patient (1.03%) presented anterior chain lymphadenopathy, and in one other patient (1.03%), the neck mass was accompanied by ptosis in the same side.

The diagnostic measures before referral consisted of angiography in 25 (25.7%), open

**Table 1.** Clinical profile of patients with carotid body tumor.

Symptom	Incidence rate
Mass in the neck	100%
Painful neck	13.4%
Dizziness	4.1%
Palpitation	3.1%
Hoarseness	3.1%
Hypertension	2.06%
Headache	1.03%
Dysphagia	1.03%
Hearing loss	1.03%



**Figure 1.** Five families with carotid body tumor. Those specified with arrow underwent surgery.

biopsy in 11 (11.3%), CT-scan in 9 (9.3%), color-Doppler sonography in 7 (7.2%), ultrasonography in 6 (6.2%), FNA and sialography in one case.

The previous surgical operations performed at other centers were as follows: incisional biopsy in 14 (14.4%), exploration of the mass in 8 (8.2%), and incomplete excision in 5 others (5.1%).

At our centers, complete excision of the mass was performed for all the patients. In 10 patients (10.3%), the mass was excised following by ligation of the external carotid artery and in 5 (5.1%), excision was accompanied by ligation of the internal carotid artery with graft replacement, all of which were without untoward consequences. In 5 (5.1%), an intraluminal shunt was used during the course of the operation.

The intraoperative complications comprised perforation of the common carotid artery in one (1.03%) and perforation of the internal carotid artery in another (1.03%), which were repaired without noticeable sequelae. The postoperative general complications were hematoma and bleeding in 2 (2.06%). The postoperative neurologic complications consisted of paresis of the mandibular branch of facial nerve, hoarseness, disorder in swallowing and tongue deviation. All such complications improved within three to six weeks. One patient suffered from blindness due to central retinal artery occlusion (Table 2).

The postoperative pathology reports revealed

**Table 2.** Postoperative neurologic complications.

Postoperative neurologic complications	Frequency (%)
Facial nerve paresis	4 (4.1%)
Hoarseness	3 (3.1%)
Disorder in swallowing	3 (3.1%)
Tongue deviation	3 (3.1%)
Blindness	1 (1.03%)
<b>Total</b>	<b>14 (14.4%)</b>

that paraganglioma was the most common tumor of which three cases were malignant. Schwannoma and mesenchymoma ranked second and third, respectively (Table 3).

## Discussion

Carotid body tumor is not a common disease in general but in patients with a neck mass, near the mandibular angle, the probability rises significantly and should be placed on top of the differential diagnoses list. Performing operations, without a proper diagnosis in 23.7% of patients and the surgeon's unexpected encounter with an unfamiliar and highly hemorrhagic tumor and the termination of the operation in vein, all indicate the lack of clinical attention to this peculiar tumor. Many of our patients are referred in late stages of disease with large tumors. However, following a successful resection, complete cure will ensue with no further recurrence. In one patient with large tumor, significant adherence to the near organs and the base of skull was found during the operation. The tumor was resected and the internal carotid artery was ligated. Pathology report revealed a malignant tumor with local invasion. There is still no sign of recurrence in this patient after 16 years. With high familial incidence, a common genetic factor seems to be responsible and the tumor will be bilateral in more than 50% of cases. Irrespective of the size of the tumor, a successful resection is performed to save the internal carotid artery. However, in 5.1% of our cases, this artery was

**Table 3.** Pathology reports of carotid body tumors.

Pathology report	Percentage
Paraganglioma	86.6%
Schwannoma	11.3%
Mesenchymoma	2.06%

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ligated but no important post-operative consequences occurred.

The male to female ratio in our study was 1: 2. Rodriguez et al reported that in the areas lower than 2000 m above sea level, this ratio was 1: 2 and in areas more than 2000 m above sea level, it was up to 1: 8.3.<sup>12</sup> As the average altitude of Tehran is 1200 m above sea level, our findings are in accordance with those of Rodriguez et al.

The mean age of the patients at the first presentation was 40 years in our study. Mitchell et al have reported that the mean age of the patients at the time of initial presentation was 54.4 years<sup>13</sup>, while Defraigne et al reported a mean age of 58 ± 5 years.<sup>14</sup> Fazel reviewed 7 patients with a mean age of 37 years in 1985.<sup>8</sup>

Preoperative neurologic deficits in our patients were as follows: hoarseness in 3 cases (3.1%), neurologic deficit, dysphagia, hearing loss and ptosis of eyelid in one case (1.03%), respectively, that makes 7.2% of the cases. Defraigne et al reported that the frequency of peripheral nerve deficits defined as vagus nerve paralysis, hypoglossal nerve paralysis, and Horner's syndrome was 11%.<sup>14</sup> Rutherford has declared that a mass in the neck was found in 73%, headache or neck pain in 35%, and dysphagia in 8% of the cases.<sup>7</sup> Fazel has reported one case (14.3%) with hypoglossal nerve paralysis and tongue deviation and another case (14.3%) with the Horner's syndrome.<sup>8</sup>

The tumor was bilateral in 5 patients (5.1%). Rodriguez et al stated that in the areas less than 2000 meters above sea level, the prevalence of bilateral tumor is between 10% and 20%.<sup>12</sup> In Gardner's report, bilateral carotid body tumor was seen in 64% of the patients<sup>15</sup> while Rutherford<sup>7</sup> and Fazel<sup>8</sup> reported it to be 8% and 29%, respectively.

In our study, the familial history in the first-degree relatives was seen in 9 patients consisting of five families. While Netterville et al reported a familial pattern in 53.3%<sup>16</sup> of cases, Gardner et al believe that a positive family history is present in about 5.5% of cases.<sup>15</sup> Rodriguez et al have announced that the positive family history in those living in the areas less than 2000 meters above sea level is between 7% and 25%.<sup>12</sup>

Lavold et al believe that perioperative complications are hemorrhage, stroke and cranial nerve injury.<sup>17</sup> In Gardner's report the carotid artery was repaired in 29% and cranial nerve injury had occurred in 27.2% of the patients.<sup>15</sup> Little

reported that 4.1% of the patients had a cranial nerve deficit.<sup>18</sup> Netterville et al reported that a vascular repair was performed in 10% of the patients with a tumor less than 5 cm and in 55.5% of those with a tumor larger than 5 cm. Also, cranial nerve injury had occurred in 13.3% of the patients.<sup>16</sup> Anand et al reported that the rate of injury to the vascular walls requiring surgical intervention was 22.2%.<sup>19</sup> Guerrier et al also declared that a carotid resection was performed in 12.1% of the patients.<sup>20</sup> Fazel, in his series, noted a case (14.3%) in whom a shunt was used because of severe adherence of the tumor to the internal carotid artery.<sup>8</sup>

Mortality did not occur in any of our patients. Defraigne et al also reported the absence of perioperative mortality or vascular complication due to the operation, but the vagus nerve was sacrificed in 22.2% of their cases.<sup>14</sup> Gardner<sup>15</sup> and Fazel also reported no mortalities before.<sup>8</sup>

Regarding the postoperative neurologic complications Defraigne et al found peripheral nerve deficit in 44.4% of the cases.<sup>14</sup> Mitchell et al have reported that 12% of the patients had peripheral nerve paralysis after the operation lasting more than six months.<sup>13</sup> Muhm et al had seen cranial nerve paresis in 20.8% of their patients<sup>10</sup> and Fazel reported a case (14.3%) with paresis and dysphagia that had improved within less than six weeks after the operation.<sup>8</sup>

The diagnosis of malignant paraganglioma in three patients, which was made by a single pathologist, is noteworthy because almost all of the malignant cases were found unusually adherent to surrounding structures. However, none of these patients have shown any evidence of clinical recurrence of metastasis. Walsh and Alvarez claim that a malignant form of this disease is very rare and its malignancy is defined by observing the clinical behavior of the tumor as an invasive development.<sup>4,5</sup>

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