

# Carotid Body Tumors: 16 Years Experience at the Jordanian Royal Medical Services

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## ABSTRACT

**Objectives:** Of all the paragangliomas in the head and neck, carotid body tumors are the most common. They are rare hyper-vascular neoplastic lesions derived from the neural crest paraganglion cells. In this study, we present our 16 years experience in dealing with this neoplasm.

**Methods:** The medical records of 33 patients with 37 surgically treated carotid body tumors between the year 1993 and 2009 at the Vascular Unit of Queen Alia Military Hospital and King Hussein Medical Center were retrospectively reviewed. Twenty patients were females and 13 were males. The mean age was 46 years. All the patients presented with a painless neck mass that was investigated by duplex ultrasound, CT angiogram and arteriography. Two patients had hoarseness of voice and dyspnea, one had dysphagia and one had vertigo. Complete surgical excision was performed in all the patients with close follow-up in respect to complications, recurrence and behavior of the tumor.

**Results:** According to Shamblin classification, 5 tumors were type I, 22 were type II and 10 were type III confirmed intraoperatively. Four patients had bilateral involvement at the time of presentation. Six patients had positive family history of the tumor. After successful surgical resection, a mean follow-up period of 24 months showed a zero mortality rate and no evidence of recurrence. Transient neuropraxia of the facial nerve was noticed in one patient, injury of recurrent laryngeal nerve in another patient while post operative localized hematoma had to be evacuated in two patients.

**Conclusion:** Although rare, carotid body tumor should be kept in mind in the differential diagnosis of anterior triangle neck mass. Complete surgical excision after adequate investigations is the treatment of choice. This is best achieved in a specialized centre with experienced vascular surgeons to get the finest outcome.

**Key words:** Carotid body tumor, Paraganglioma.

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## Introduction

Paragangliomas are benign tumors arising from brachiomeric paraganglia dispersed near or in the autonomic nervous system extending from the skull bone down to the pelvic floor.<sup>(1,2)</sup>

Paragangliomas located outside the adrenal glands are termed extra adrenal paragangliomas which in

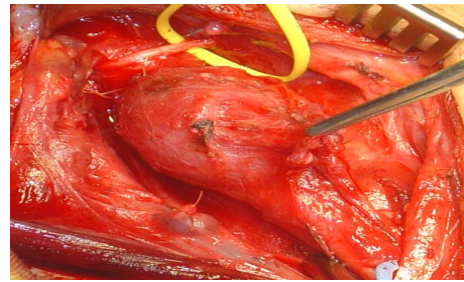
turn are divided into two groups; Group I which is associated with parasympathetic system and located in the head, neck and anterior mediastinum and thought to have chemoreceptive function, and Group II that is associated with sympathetic system and located in the posterior mediastinum and retroperitoneum and believed to have a function similar to the adrenal medulla.<sup>(1-3)</sup>

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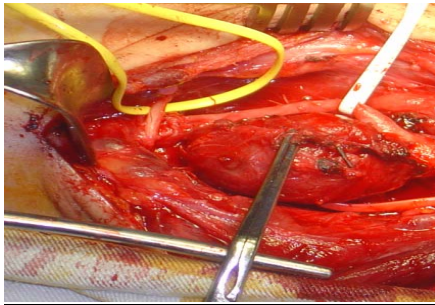
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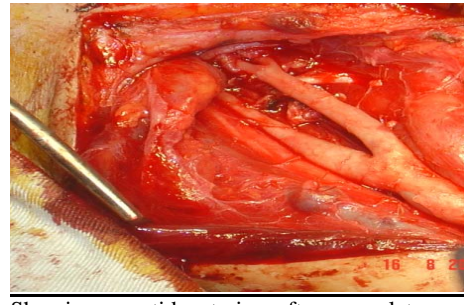
**Fig. 1.** Showing Carotid Body Tumor in the neck



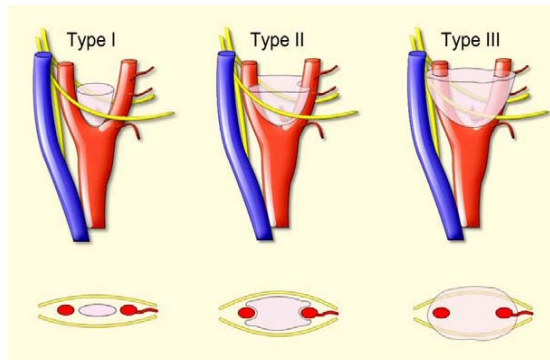
**Fig. 2.** Showing carotid body tumor



**Fig. 3.** Showing dissection of carotid body tumor from carotid arteries



**Fig. 4.** Showing carotid arteries after complete excision of carotid body tumor



**Fig. 5.** Shamblin's classification

Paragangliomas of the head and neck include: carotid body tumors, glomus jugulare, glomus tympanic and glomus vagale.<sup>(2,4)</sup> Paragangliomas arising from the carotid body are uncommon neoplasms accounting for 0.5% of all body tumors but comprise 60-70% of head and neck paragangliomas.<sup>(5-8)</sup>

Carotid body is a chemoreceptor found in the adventitia of the postero- medial surface of the common carotid artery bifurcation and is attached by Mayer's ligament which carries its blood supply that mostly arises from the external carotid artery.<sup>(7,9,10)</sup> Carotid body tumor has high blood flow and oxygen consumption averaging approximately 0.2 L/g/min.<sup>(10)</sup>

Macroscopically, carotid body tumor is a well circumscribed, rubbery and reddish brown tumor. Microscopically, it is characterized by cell nests

(Zellballen) composed of chief cells (type I) encircled by a thin layer of sustentacular cells (type II).<sup>(3,9,11)</sup>

Carotid body is responsive primarily to hypoxia and to a lesser extent to acidosis and hypercarbia and once stimulated it causes increase in the heart rate, blood pressure, respiratory rate and tidal volume.<sup>(1,9,10)</sup>

In this study we report our experience in dealing with surgically treated carotid body tumor with close follow-up in respect to complications, recurrences and behavior of the tumor.

## Methods

The medical records of 33 patients with 37 surgically treated carotid body tumors between the year 1993 and 2009 at the Vascular Unit of Queen Alia Military Hospital and King Hussein Medical

Center was retrospectively reviewed. Twenty patients were females (60%) and 13 were males (40%). Their mean age was 46 years (22-60 years).

All the patients presented with painless neck mass (Fig. 1). Two patients had hoarseness of voice and dyspnea, one patient had dysphagia and one had vertigo.

Besides the clinical diagnosis, duplex ultrasound, angiography, and CT angiography were used to confirm the diagnosis, to evaluate the extent of the tumor and to check for bilaterality.

Complete surgical excision was performed in all our patients with close follow-up in respect to complications, recurrence and behavior of the tumor.

### ***Surgical Technique***

Excision of the carotid body tumors is the mainstay of treatment.<sup>(3,8,10,12)</sup> Under general endotracheal anesthesia, the neck is explored through an incision made along the anterior border of sternocleidomastoid muscle. Dissection is continued to expose the tumor, carotid bifurcation and internal jugular vein (Fig. 2). The posterior belly of the digastrics muscle is separated from the tumor mass. The common and internal carotid arteries are exposed and a tape is placed around each for control (Fig. 3). Special care is to be taken to avoid injury of the cervical branches of the facial, hypoglossal and vagus nerves. A subadventitial plane is developed on the common carotid artery and the dissection is progressed cephalad (Fig. 3). A mosquito forceps are introduced and gently opened in the periadventitial plane, ligating or applying diathermy to the separated vascular tumor tissue and the adventitia. Ligation of the branches from the external carotid artery feeding the tumor may decrease tumor's vascularity and bleeding during resection.<sup>(8,12)</sup>

Occasionally, the external carotid artery may be ligated and divided to facilitate further resection and decrease intra operative blood loss. If proper plane between the tumor and the arteries cannot be established due to transmural tumor invasion of the arterial wall or in cases where the carotid arteries get damaged during resection of the tumor especially in large sized tumors encasing the vessels (Shamblin III), carotid arteries resection may be considered and replacement with interposition greater saphenous vein graft harvested from the thigh is applied.<sup>(8,12,13)</sup> The preoperative work up including vascular imaging should indicate the likelihood of vascular

involvement and give some indication as to how well the patient will tolerate temporary internal carotid compromise. Intra operative neuromonitoring and carotid shunting to prevent hemispheric ischemia are applied during clamping and reconstruction. A drain is placed and wound closed after hemostasis is achieved.

### **Results**

All patients underwent surgical resection with complete excision of their carotid body tumors achieved. A standard anterior approach to the carotid vessels and meticulous subadventitial dissection of the tumor were employed. The diagnosis of carotid body tumor was confirmed histologically in all patients. According to Shamblin classification, 5 tumors were type I, 22 were type II and 10 were type III confirmed intra operatively (Table I). Two patients required arterial reconstruction (Shamblin III tumors) where the external carotid artery was ligated and interposition vein graft was used to replace the internal carotid artery.

**Table I.** Distribution of patients according to Shamblin Classification

Shamblin	I	II	III
Patients No	5	22	10

The histological examination showed benign pathology in all but one patient. Radical neck dissection was done for her 4 years ago with no evidence of recurrence shown on follow-up.

Four patients had bilateral involvement at the time of presentation while six had positive family history of their carotid body tumor.

Transient neuropraxia of the facial nerve was noticed in one patient, injury of the recurrent laryngeal nerve in another patient while post operative localized hematoma had to be evacuated in two patients.

A mean follow-up period of 24 months (18-60) conducted periodically every 6 months for the first year and then yearly afterwards by clinical assessment, duplex scan and CT angiography showed a zero operative mortality and no evidence of recurrence.

### **Discussion**

Von Haller was the first to describe carotid body tumor in 1743.<sup>(6,8)</sup> In 1880, Reigner first attempted resection of carotid body tumor but unfortunately the patient died. Six years later, Maydl removed

successfully the tumor, nevertheless his patient developed stroke. Not until 1903, when the first successful removal of carotid body tumor was announced by Scudder.<sup>(6,8)</sup>

In 1940, Gordon-Taylor demonstrated a safe subadventitial plane for tumor resection to become the current surgical technique and the procedure that we used in our patients.<sup>(6,10,14)</sup>

As most of the other studies reported, all our patients presented with painless neck mass located at the angle of the mandible. Carotid body tumors are slow growing non tender masses that are mobile laterally but vertically fixed due to their attachment to the carotid bifurcation (Fontaine's sign).<sup>(6,7,15,16)</sup>

Because of its location in close proximity to cranial nerves (X-XII) and cervical sympathetic chain, progressive enlargement of carotid body tumors occasionally produce symptoms such as dysphagia, hoarseness of voice, stridor, dysphonia and Horner's syndrome.<sup>(1,6,8)</sup> Two of our patients were noticed to have hoarseness while one presented with dysphagia. These symptoms disappeared after surgery as reported by these patients during their post operative follow-up visits.

Patients may give history of symptoms suggestive of neuroendocrine activity in less than 5% of tumors such as dizziness, palpitation, tachycardia and hypertension.<sup>(1,8,11,17)</sup> In these patients, screening for catecholamine secretion with estimation of urinary metanephrines is indicated although some authors do routine screening as this neuroendocrine activity obviously has anesthetic implication.<sup>(7,9,11,12)</sup> In our patients only those with suggestive symptoms and signs underwent screening.

Female predominance has been reported in some studies. Likewise, we noticed that more than half of our patients with carotid body tumor were females and most of the tumors become apparent between the third and sixth decade of life which keeps our results in harmony with the other studies reports.<sup>(6,8,9,18-20)</sup>

The published literature has described the biological behavior of carotid body tumor to be either sporadic or familial with the former being the commonest.<sup>(3,6,11,19,20)</sup> In the familial type (20%), there is an incidence of bilaterality in about one third of the patients and a high incidence of multicentricity.<sup>(1,4,6,19)</sup> Transmission is by an autosomal dominant pattern inherited due to alteration in gene coding for succinate dehydrogenase subunit D (SDHD), B (SDHB) and C (SDHC). The subunits D and B account for a

significant percentage of head and neck paragangliomas.<sup>(1,3,11,17)</sup> In our review, six patients (18%) had positive family history and four patients (12%) had bilateral involvement, two of these were brothers. We believe that family member screening is strongly recommended in these cases for early detection and easier resection when discovered at a small tumor size. For those, Duplex ultrasound scanning was considered the primary diagnostic investigation in our series.

Investigations used in our study as in others included: Duplex scan, CT angiogram, magnetic resonance angiography and conventional angiography.<sup>(1,8,11,16,21,22)</sup> Duplex with color flow imaging demonstrates the highly vascularized tumor causing widening of the carotid bifurcation.<sup>(10,16)</sup> We used duplex also in all our patients for post operative follow-up. Both CT and MR scans can reveal the size, extent and relations of the tumor and help in proper surgical exposure. A slight superiority of MR noted as it demonstrates the classic "Salt and Pepper" appearance.<sup>(3,8,16)</sup> Angiogram remains the gold standard for the diagnosis and management of carotid body tumor. It shows the tumor blood supply and widening of the carotid bifurcation by the tumor blush (Lyre sign) which is a pathognomonic angiographic finding in carotid body tumors.<sup>(1,3,13,16,21)</sup> Another advantage of angiography is the possibility for concomitant embolization to decrease tumor blood supply.<sup>(6)</sup> Embolization is still debatable as some authors believe that the response of the tumor makes subadventitial plane dissection difficult besides that the procedure may be complicated by internal carotid artery thrombosis and embolization.<sup>(6,7,9,10,12)</sup> None of our patients reviewed underwent embolization for their carotid body tumor beforehand. We used Duplex scan as an effective modality in making the diagnosis, but angiography and CT or MRI are helpful to plan surgery.

It is worth mentioning that incisional biopsy and fine needle aspiration are contraindicated and should not be included in the investigations of this highly vascular tumor. They may be complicated by massive hemorrhage, false aneurysm and carotid thrombosis.<sup>(8,9,12)</sup>

A surgical classification system was developed by Shamblin *et al* in 1971 to assess the resectability of these tumor and foretell the surgical morbidity by dividing the tumors into 3 groups (Fig. 5). In Group I, tumors are relatively small with minimal attachment to the carotid vessels and can be easily

removed. Group II, tumors are larger with moderate attachment but separable from adjacent vessels with careful dissection. Group III, tumors encase carotid arteries and frequently require arterial reconstruction.<sup>(1-3)</sup> The majority of the patients that we studied were classified intra operatively as having Shamblin II (Table I). Two patients with Shamblin III required arterial reconstruction to achieve complete excision of the tumor. Some authors have stated that carotid body tumor cannot be classified preoperatively but Aryl *et al* have illustrated that pre operative MR imaging can predict Shamblin group using the degree of circumferential contact of the carotid body tumor with the internal carotid artery on axial images.<sup>(3,21)</sup>

As demonstrated by most other studies, carotid body tumor mostly exhibits benign characteristics with less than 10% being malignant.<sup>(5,17,23)</sup> Malignant potentials cannot be predicted by histological markers and can only be assumed in the presence of lymph nodes or distal metastasis.<sup>(3,6,8,11,23)</sup> So the histological appearance of a carotid body tumor is not a reliable guide to its propensity for malignant behavior. One of our patients demonstrated localized lymph node involvement. She underwent neck dissection and has been on follow-up for the last four years with no recurrence up to date.

In our study, two patients were from Yemen where they live at about 2200 meters above sea level. High altitude with chronic hypoxic states were considered as proposed risk factors for carotid body hyperplasia in some other studies.<sup>(10,19,24,25)</sup>

Although there is a general agreement that complete surgical excision is the best treatment option, a more conservative approach is occasionally justified.<sup>(12)</sup> Patients may be managed non-surgically, as advocated by some authors because of age and concomitant atheromatous disease of the contralateral carotid vessels as the risks of operation in these patients are judged to be high. Radiotherapy has been used as an alternative treatment in high-risk patients and in the occasional cases where the tumor is inoperable, large or recurrent.<sup>(5,9,26-28)</sup> In our series, all the patients were operated successfully achieving complete surgical excision since all the patients were in shape for surgery and anesthesia.

Though the current surgical techniques have reduced the post operative complications especially stroke rate, the incidence of cranial nerves injury remains relatively high ranging between 20-

40%.<sup>(8,13,23)</sup> Most of these cranial nerve injuries are temporary. The incidence of permanent cranial nerve deficit has been quoted less than 1% in international literature.<sup>(14,19)</sup> Transient neuropraxia of the facial nerve was noticed in one of our patients (3%) and another patient had injury of his recurrent laryngeal nerve (3%) which were diagnosed on clinical basis and neurological examination / laryngoscopy. To minimize the risk of complications, we advocated subadventitial dissection at an early stage when the tumor excision can be carried out with less bleeding and with a clearer operative field. No recurrences were reported in our series but we believe that continued follow-up is required as recurrences and metastasis may occur years after the initial diagnosis.<sup>(3,5,11)</sup>

## Conclusion

Carotid body tumors are rare but the diagnosis should be kept in mind in any patient presenting with a neck mass in the anterior triangle of the neck. Both sides of the neck need to be investigated to detect bilateral tumors. Misdiagnosed patients may undergo unnecessary procedures, which could be disastrous. Complete surgical excision after adequate investigations is the treatment of choice. In order to obtain good results; these tumors should be dealt with in a specialized center, where the availability of an experienced team who is familiar with carotid surgery improves the chances for a more successful outcome in these patients.

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