Carotid body tumors: a subject review and suggested surgical approach

FREDRIC B. MEYER, M.D., THORALF M. SUNDT, JR., M.D., AND BRUCE W. PEARSON, M.D.

Departments of Neurologic Surgery and Otolaryngology, Mayo Clinic and Mayo Graduate School of Medicine, Rochester, Minnesota

Carotid body tumors are a rare but potentially difficult surgical entity. Their pathology, physiology, and natural history are reviewed along with surgical results reported in the literature. A surgical approach for removal of these tumors is presented which differs significantly from the recommended techniques in that emphasis is placed on intraoperative monitoring of cerebral blood flow, the selective use of shunts, a tumor-adventitial plane of dissection, preservation of the carotid artery complex, and mobilization of the parotid gland. Thirteen cases using these techniques are reviewed. The mortality rate and the incidence of cerebrovascular sequelae were both 0%. The major morbidity consisted of injury to the lower cranial nerves in five patients (39%) with tumors larger than 5 cm in length.

KEY WORDS · carotid body · brain tumor · operative approach

Basic Considerations

Embryology

The carotid body is derived from both mesodermal elements of the third branchial arch and neural elements originating from the neural crest ectoderm. These neural crest cells further differentiate into sympathogonia, which are forerunners of paraganglionic cells. Therefore, the original descriptive term "chemodectoma" is questionable since the carotid body is derived from paraganglionic cells and not chemoreceptor cells. Paraganglionic cells are also located adjacent to the aorta, with the greatest collection in the adrenal medulla. Cells within the adrenal medulla are chromaffin-positive, indicative of their ability to produce catecholamines.

Initially, carotid body tumors were thought to be non-chromaffin paragangliomas. However, recent studies have demonstrated chromaffin-positive secretory granules, suggesting that the carotid body is capable of secreting catecholamines. A few cases have been reported of hypertension due to catecholamine-producing carotid body tumors.

Physiology

The carotid body, by weight, has a greater blood flow and oxygen consumption than the brain. Its proposed
primary function is regulation of pulmonary ventilation through afferent input via the glossopharyngeal nerve to the medullary reticular formation. It is responsive primarily to hypoxia and to a lesser degree to hypercapnia and acidosis. Stimulation of the carotid body produces an increase in respiratory rate and tidal volume, and an increase in sympathetic tone as evidenced by an increase in heart rate, blood pressure, vasoconstriction, and circulating catecholamines.

Pathology

The carotid body measures approximately $5 \times 3 \times 2$ mm, and is located in the adventitia of the posterior medial surface of the common carotid artery bifurcation. Its fibrous capsule is attached to the bifurcation by Mayer's ligament through which its blood supply traverses. Grossly, carotid body tumors are rubbery, red-brown in color, and well circumscribed.

Microscopically, the carotid body tumor is composed of nests of epithelioid cells ("Zellballen") with granular eosinophilic cytoplasm. Separating these nests in a trabeculated fashion is a vascularized connective tissue. Each nest is composed of 10 to 20 neoplastic chief cells (Fig. 1). Neural elements are located in the tumor capsule. The tumor is highly vascular and draws its blood supply from the vasa vasorum. Although these tumors are well circumscribed, they have no capsule.

The carotid system is progressively distorted and extended as the tumor grows (Fig. 2). Shamblin, et al., divided these tumors into three anatomical groups. Group I consisted of relatively small tumors with minimal attachment to the carotid vessels. Surgical excision of these could be performed with minimal difficulty. Group II tumors were larger and had moderate arterial attachment. These tumors could be resected with precise surgical dissection; however, the need for a shunt was likely. Group III tumors were large neoplasms encasing the carotid arteries. Tumor removal in this group would require arterial resection and grafting.

Since the tumor originates from the adventitia, many surgeons advocate the use of a subadventitial plane for dissection. However, with larger tumors, compression of the vessel makes identification of this adventitia-media plane difficult. Furthermore, this technique severely weakens the arterial wall. In addition, there is an increased likelihood of inadvertent arterial wall tears requiring unplanned occlusion of the common carotid artery for hemostasis. We would therefore recommend against the use of a subadventitial plane of dissection.

With growth of the tumor, the vagus, hypoglossal, and occasionally the sympathetic chain are either displaced or encased by the tumor. The tumor may extend medially, producing a pharyngeal mass on oral examination, and may reach up to the skull base.

Biological Behavior

The malignancy potential of carotid body tumors is controversial; reported figures range from 2.6% to 50%. Pathological criteria for malignancy are based on the standard criteria of cellular atypia and mitoses, local invasion, and dissemination. Microscopically, it is rare for these tumors to demonstrate mitoses. However, most authorities agree that in this tumor the histological appearance does not correlate well with growth behavior. Currently, the metastatic rate is considered to be approximately
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Metastatic spread occurs most commonly to the regional lymph nodes, but metastases have been documented to the brachial plexus, cerebellum, lungs, bone, abdomen, pancreas, thyroid, kidney, and breast. Metastases should not be confused with multicentricity of paragangliomas at other sites in the body.

Carotid body tumors will grow relentlessly if not resected. Some reviews note a mortality rate of about 8% in untreated cases. Although some authors have reported palliation with radiation therapy alone, it is generally agreed that this is not acceptable primary treatment. Reported complications from radiation therapy include necrosis of the mandible, carotid artery, and larynx. Some authors recommend radiation therapy for incomplete resections, although this has not convincingly been shown to be of value.

Epidemiological studies suggest that there are two forms of this disease. First, in the more common sporadic form there is a 5% incidence of bilateral carotid body tumors. The second, rarer, form is the familial occurrence of these tumors, which is transmitted in an autosomal-dominant pattern. Within this second group, there is a 32% incidence of bilateral tumors. Therefore, if a positive family history is obtained in the initial evaluation of a patient, early examination of other family members should be performed since the ease of resection is based on the size of the tumor.

There appears to be greater incidence of carotid body tumors in people who live at high altitudes or in patients subjected to chronic hypoxia.

Clinical Presentation

The most frequent presenting symptom in a patient with a carotid body tumor is a palpable neck mass that either the patient or the local physician has noticed routinely. Other symptoms include a recently increasing neck mass, neck discomfort, hoarseness, dysphagia, stridor, tongue weakness, and dizziness.

On examination, the neck mass is located below the angle of the jaw and is often laterally mobile but vertically fixed because of its adventitial attachments. The incidence of cranial nerve involvement has been estimated at 20% and includes the vagus and hypoglossal nerves. Rarely, a patient may present with a Horner’s syndrome. Although most of these masses have a transmittable pulse, a bruit is infrequent. The stridor and dysphagia may be secondary to either vagus involvement or compression of the pharynx by the adjacent tumor. In either situation, these specific findings are suggestive of a large tumor. Although dizziness is a frequent complaint, there are only a few documented cases of a carotid body tumor presenting with cerebral ischemia. This is probably due to the fact that the tumor tends to encase the carotid arteries and grow cephalically rather than compress the arteries. In the review by Shamblin, et al., of 90 cases, six presented with hypertension. Evaluation for catecholamine production was negative in these cases. However, the reported cases of catecholamine-secreting tumors and the potential for multicentricity, including pheochromocytoma, must be emphasized. The preoperative identification of excess catecholamine production will alter anesthetic management. Therefore, preoperative measurement of urinary metanephrines and vanillylmandelic acid is justified in all these patients.

Diagnosis

Clinically suspicious neck masses should be evaluated by complete transfemoral bilateral cerebral angiography. The presence of an enhancing oval mass widening the angle of the bifurcation with displacement of the internal and external carotid arteries is essentially pathognomonic of a carotid body tumor (Fig. 3). Although blood supply is primarily from the bifurcation and external carotid artery, contribution from the internal carotid artery, vertebral artery, and thyrocervical trunk does occur (Fig. 4). Knowledge of the sources of blood supply aids in exposure and hemostasis. The extent of tumor blush will also indicate the cephalic
extent of the tumor and help in planning the surgical exposure (Figs. 4 and 5). Bilateral angiography is critical for the evaluation of concurrent atherosclerosis and collateral flow, and for identification of multicentric paragangliomas of the contralateral carotid body and glomus jugulare. Although some reports suggest the use of digital subtraction angiography,13 in our opinion the current resolution is not sufficient to supply detailed surgical anatomy.

Contrast-enhanced computerized tomography (CT) scanning of the neck is useful in demonstrating the lateral and medial extent of the tumor. However, differentiation between an aneurysm and a neoplasm is difficult. Dynamic or rapid-sequencing CT improves delineation between vascular structures59 and may emerge as the primary screening examination. Angiography would still be mandated by the need to identify collateral flow, atherosclerosis, and blood supply to the tumor. Ultrasonography has been advocated as a noninvasive diagnostic test.19 Currently at our institution it is used as a means of sequentially following these tumors in specific situations. Oculoplethsmography is a useful noninvasive technique to evaluate the hemodynamic status of both carotid arteries.

Treatment
The mainstay of therapy for carotid body tumors is complete surgical excision. Early removal of an asymptomatic tumor minimizes potential cranial nerve and carotid artery injury. Unresected tumors grow relentlessly and tend to encase the adjacent neurovascular structures, making delayed surgery more hazardous. Large tumors extend into the skull base, necessitating a combined suboccipital-cervical neck dissection. In addition, there is a 5% incidence of metastases. Specific indications for a conservative approach include the patient's age and medical condition. A patient with bilateral carotid artery tumors who has undergone a previous resection with resulting vagus nerve injury should be treated conservatively; in that specific situation, surgical removal of the contralateral tumor carries an inherent risk of injury to the remaining vagus nerve. Since bilateral vagus nerve palsies have a high risk of morbidity and mortality, the appropriate approach would be noninvasive serial evaluation by CT or ultrasound. As previously mentioned, radiation therapy has not been demonstrated to be effective in control of these tumors.14,34,48 Although there are some reports of preoperative embolization to decrease intraoperative blood loss,3,28 we have not found this necessary.

Summary of Cases

Case Material
From the years 1931 to 1984, approximately 140 patients have undergone resection of a carotid body tumor at the Mayo Clinic by various surgical specialists. The results up to 1975 have been reported previously.25,35,38,48 We analyze here the 13 patients who were operated on between 1975 and 1984 by the neurovascular service with the aid of intraoperative electroencephalography and CBF monitoring.

There were 10 females and three males with an average age of 48 years. Four patients had bilateral carotid body tumors; one of these patients also had a glomus jugulare tumor, and one had a familial pattern.
Carotid body tumors with autosomal dominant inheritance. The most frequent presenting symptom was a neck mass. Two patients had preoperative neurological deficits: one had a Horner’s syndrome and the other suffered palsy of the seventh, 10th, and 12th cranial nerves. The mean follow-up period was 3.5 years (range 1 to 7.5 years).

Surgical Technique

The current surgical approach used by the neurovascular service emphasizes six fundamental concepts. 1) The preservation of CBF during and after the operation is critical. Therefore, all patients are monitored with intraoperative electroencephalography. Furthermore, patients with large tumors in whom temporary carotid artery occlusion may be required have baseline preocclusion and occlusion xenon-133 (133Xe) CBF studies. 2) Distal exposure of large tumors is obtained by mobilization of the parotid gland. This approach facilitates identification of the lower cranial nerves cephalic to the tumor and aids in their preservation. 3) These tumors are dissected in the capsular-adventitial plane as opposed to the subadventitial plane as advocated by some surgeons. 4) Great effort is taken to maintain the integrity of the external carotid artery as it is a potential source of collateral flow. This is contrary to opinions at other institutions. 5) Although some authors recommend the routine use of a shunt in large tumors, shunts are used only when the electroencephalogram and CBF studies demonstrate insufficient perfusion during carotid artery occlusion. This minimizes the risks associated with a shunt and avoids an unnecessary arteriotomy. In most cases, meticulous dissection eliminates the need for either temporary carotid artery occlusion or shunt placement. 6) Since exposure is critical in successful removal of these tumors, a longitudinal incision is used extending from the ear to the suprasternal notch along the anterior sternocleidomastoid muscle. Although cosmetically less appealing than the horizontal incision advocated by some, it permits excellent exposure of both the distal and proximal carotid arteries. (In all patients, the ipsilateral lower section of the leg is prepared and draped for surgery in case a saphenous vein graft is needed.) The lower part of the incision is similar to that used in a routine carotid endarterectomy and parallels the anterior border of the sternocleidomastoid muscle, ending several centimeters superior and lateral to the suprasternal notch. The cervical segment of the incision runs along the anterior border of the sternocleidomastoid muscle and ascends to a point just behind the lobe of the ear. In the lower quarter of the postauricular sulcus, it drops to the bottom of the ear, skirts the ear lobe, and then ascends in a pretragal skin crease to the superior border of the zygoma. We then prefer to mobilize the parotid gland first to gain exposure of the distal internal carotid artery.

The superficial cervical fascia is incised and the posterior border of the parotid gland is exposed and elevated. The anterior-inferior surface of the auricular cartilage is followed deep to its “pointer,” the triangular projection of cartilage at its medial limit. The temporo-parotid fascia is incised between the mastoid process and the posterior margin of the parotid gland, and the facial nerve is found subjacent to the fascia. A finger placed on the mastoid tip and directed anteriorly forward, the cartilaginous “pointer,” and the palpable junction between the external auditory meatus (the tympanomastoid suture) all point to the main trunk of the seventh nerve. Once the main trunk is identified, the lower division and marginal mandibular nerve which form the upper limit of the deep dissection can be traced forward by sharp dissection and elevated safely using mobilized parotid tissues as a “bundle.” The posterior belly of the digastric muscle is now followed to its point of insertion in the mastoid groove and divided there. The stylohyoid muscle lies superior and parallel to the digastric muscle and should also be divided, exposing the deeper stylomandibular ligament which must be resected for adequate distal exposure to the internal carotid artery.

Although this relationship between nerves and muscles is referred to by the anatomists as a “retroparotid fossa,” the tissue is in fact densely bound by deep thick cervical fascia; until this fascia is divided, it is not possible to elevate or mobilize the parotid gland superiorly and thus not possible to expose and isolate the

![Fig. 5.](https://example.com/fig5.png)
distal internal carotid artery. For high exposure, it is usually necessary to identify the origin of the seventh nerve but seldom is it necessary to trace this nerve distally into the parotid gland itself. Using fishhooks as retractors rather than heavy self-restraining retractors often avoids damage to the marginal mandibular branch of the facial nerve.

After mobilization of the parotid gland and exposure of the distal internal carotid artery, the proximal common carotid artery is then exposed by dissection of the deep fascia anterior to the sternocleidomastoid muscle. This dissection is carefully extended cephalically to the bifurcation, where the causal limits of the tumor are encountered. At this point, vascular tapes are placed around the common, internal, and external carotid arteries prior to dissection of the tumor. Before dissection of the tumor, baseline preocclusion and occlusion $^{133}$Xe CBF studies are performed. It is important to know the status of the collateral blood supply ahead of time in case the carotid artery must be quickly occluded for hemostasis.

As depicted in Fig. 6 upper left, the common facial vein is often incorporated into the tumor capsule and must be ligated along with feeding veins from the surrounding tissue. The tumor is then isolated along its medial and lateral borders. The proper plane of dissection is identified between the lower pole of the tumor and the common carotid artery, using the bipolar forceps under magnification. Since the tumor's main blood supply is from the carotid bifurcation and external carotid artery, the dissection delineates this attachment first (Fig. 6 upper right). Usually there is an areolar plane between the tumor and artery except for its subadventitial attachment at the posterior wall of the bifurcation. With the use of bipolar cautery, the multiple perforating arteries arising from the vasa vasmorum are coagulated and divided (Fig. 6 center left). The tumor is usually fed by large proximal branches of the external carotid artery, which must be ligated individually. The same is true for feeding arteries from the vertebral artery and thyrocervical trunk, which develop in very large lesions. The tumor is grasped with forceps and rotated superolaterally to expose the tumor-carotid body interface. By dissecting in the periadventitial layer close to the arteries, the risks of injuring the superior and recurrent laryngeal nerves can be minimized.

It should be noted that we have encountered an anteriorly located aberrant vagus nerve on several occasions during elective endarterectomy. Furthermore, in two cases of carotid body tumors, the vagus nerve was incorporated within the tumor bed and had to be carefully dissected out. Identification of the vagus nerve in reference to the tumor is greatly facilitated by mobilization of the carotid gland. The other cranial nerve that can be injured at this point in the dissection is the hypoglossal nerve. Usually the tumor displaces the hypoglossal nerve posteriorly and superiorly. Again, identification of the nerve in the submandibular region is important for its preservation. The mandibular branch of the facial nerve can be injured by excessive retraction under the angle of the jaw, but prior parotid mobilization usually provides sufficient room. In tumors with a large lateral extension, the accessory spinal nerve should be identified.

After dissection and ligation of the feeding arteries from the common and external carotid arteries, the lateral and superior poles of the tumor are further mobilized (Fig. 6 center right). Laterally and somewhat posteriorly, it is common for the tumor to derive a very major share of its blood supply from the carotid sheath. These vessels are often quite large but can be well controlled with bipolar coagulation techniques. This approach leaves the medial posterior attachment between the internal and external carotid arteries for last. In this manner better visualization of the vagus nerve and its very important superior laryngeal branch is achieved. As the tumor is elevated from its bed, the superior laryngeal nerve is dissected away from the tumor capsule (Fig. 6 lower).

After excision of the tumor, the arteries are carefully inspected for any arterial wall injury and CBF is measured. If this flow is low, or if a segment of artery appears suspicious, the artery is occluded and a local arteriotomy is made. The appropriate arterial repair and, if necessary, endarterectomy are performed with a saphenous vein patch graft. On one occasion a soft clot was found in the artery at the site of external injury.

The rare massive carotid body tumor may extend up to and erode the foramen lacerum and petrous bone. Tumors that extend into the posterior fossa may be approached through a suboccipital craniectomy. However, tumors within the foramen lacerum should be considered unresectable.

**Operative Results**

There was no surgical mortality or any cerebrovascular sequelae, including stroke, transient ischemic attacks, or carotid occlusion. The major morbidity was cranial nerve injury, which occurred in five patients (39%). The superior laryngeal nerve was injured five times, the recurrent laryngeal nerve four times, the hypoglossal nerve twice, and the sympathetic chain once. All cranial nerve injuries occurred in five of the seven tumors that were at least 5 cm in length. No injuries were noted in tumors less than 5 cm in length. In two cases, the vagus nerve was engulfed by the tumor and had to be dissected free. Although anatomical continuity was maintained, function of the superior and recurrent laryngeal nerves was lost. The two patients with preoperative nerve deficits did not have return of function after tumor removal.

Documentation of damage to the superior laryngeal nerve is difficult unless electromyography of the cricothyroid muscle is performed. Clinically, these patients present with difficulty in swallowing and possibly aspiration. The proximity of the superior laryngeal nerve
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Fig. 6. Operative sequence. **Upper Left:** The parotid gland has already been mobilized and the rostral extent of the tumor delineated. The common facial vein is often incorporated into the tumor capsule and must be ligated. **Upper Right:** After dissection of the medial and lateral borders of the tumor from the surrounding tissue, its attachments to the common carotid and external carotid arteries are attacked with bipolar cautery. **Center Left:** The tumor is dissected from the arteries in an areolar plane between the capsule and adventitia. Only its origin from the adventitia is removed in the subadventitial plane. Bipolar cautery is used to coagulate the multiple perforating branches from the vasa vasorum. **Center Right:** The superior and lateral poles of the tumor are further mobilized after the main blood supply to the tumor has been ligated. **Lower:** As a last step, the posterior wall of the tumor is dissected from the carotid artery complex and the cranial nerves. Rotation of the tumor improves exposure at this interface.
to the recurrent laryngeal nerve means that, if there is injury to one, both are usually involved. However, hoarseness alone implies recurrent laryngeal nerve injury, which can be documented on pharyngeal examination. Injury to the superior laryngeal nerve in this series was based on clinical criteria. Treatment consisted of a soft diet until normal swallowing returned. No patient suffered from aspiration, although three had persistent complaints of dysphagia in the follow-up period.

Injury of the recurrent laryngeal nerve was treated in all four patients by Teflon injection of the paralyzed vocal cord, with good palliative effect. In one of these patients, injury of the recurrent laryngeal nerve has required a conservative approach to the contralateral tumor. Injury of the hypoglossal nerve was bothersome but not disabling to the patients.

Intraoperative shunting and ligation of the external carotid artery were each used once in the same patient. In the remaining 12 operations, tumor removal was complete without the use of these techniques. One patient had repair of a common carotid bifurcation tear. None of the patients required an interposition graft.

In all 13 cases the histological diagnosis was benign. No patient had evidence of metastatic disease. One patient had only a 95% resection as the tumor extended up to the foramen lacerum; the other 12 patients had complete resection, including one patient who underwent a simultaneous suboccipital craniectomy for tumor in the cerebellopontine angle (Fig. 5).

Discussion

This series of 13 patients demonstrates that carotid body tumors can usually be resected while maintaining the integrity of the carotid artery complex. Intraoperative electroencephalography and CBF monitoring are important adjuncts to protect the cerebral hemisphere during dissection when temporary arterial occlusion may be required. This is preferable to injudicious shunting, in our judgment.

Division of these tumors into Groups I, II, and III according to Shamblin, et al., was not of predictive values in terms of shunting or en bloc resection of the tumor-carotid complex. With precise dissection and the use of bipolar cautery, these techniques were not necessary. All of these tumors were resected in the capsular-adventitial plane. In fact, the one arterial tear in the group occurred during a subadventitial dissection when the appropriate plane had been lost. Therefore, we favor tumor removal that does not violate the adventitia except at the point where the tumor originates from the subadventitia.

The major morbidity consisted of cranial nerve injury in five patients with very large tumors. Although rarely mentioned in the literature on carotid body tumors, in our cases the superior laryngeal nerve was most frequently injured. Furthermore, in large tumors, motor branches from the vagus nerve to the pharynx may also be injured. Presumably, in other case reports dysphagia was attributed solely to recurrent laryngeal nerve palsy. Mobilization of the parotid gland aids in identifying the proximal location of the ninth, 10th, 11th, and 12th cranial nerves. However, when large tumors extend up to the submandibular region this advantage is gradually lost. Nevertheless, parotid mobilization still protects the seventh nerve and aids in rostral dissection even in large tumors.

We usually recommend removal of large tumors that have grown under observation. Small asymptomatic tumors may be followed by CT scanning or ultrasound. If growth is observed, these tumors should be resected. The 5% incidence of metastases may justify the early removal of even small tumors. Currently, we reserve radiation therapy only for large tumors in a patient who is medically unfit for surgery or for the rare tumor that is incompletely resected at the skull base.

Acknowledgments

The authors are indebted to Drs. David Piepgras and Michael Ebersold for the inclusion of their cases and to Ms. Bernita Bruns for preparation of the manuscript.

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Manuscript received May 29, 1985. Accepted in final form August 15, 1985. Address reprint requests to: Thoralf M. Sundt, Jr., M.D., Department of Neurologic Surgery, Mayo Clinic, Rochester, Minnesota 55905.