Review Article

What's New in the Management and Treatment of Carotid Body Tumors

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Abstract

Carotid body tumors (CBT) are a rare, hypervascular tumor involving the carotid bifurcation. CBT represent the majority of paragangliomas of the head and neck region. Despite over a century of experience, we continue to advance our knowledge in its management and treatment.

Keywords: Carotid body tumors; Neoplasm; Paragangliomas

1. Introduction

The carotid body is a specialized chemoreceptor located within the semi-adventitial tissue of the carotid bifurcation. It is stimulated by conditions that induce hypoxia, hypercapnia, and acidosis and controls the body's response to these conditions with changes in blood pressure, heart rate, and respiratory rate by release of catecholamines [1]. The carotid body receives its blood supply through glomus arteries traversing Mayer's ligament from the common carotid artery and the ascending pharyngeal arterial branch of the external carotid artery, and its innervation derives off a small branch of the glossopharyngeal nerve (Figure 1) [2-4]. CBT (paragangliomas) are a rare neoplasm originating in the neural crest cells at the carotid bifurcation. The incidence of CBT is estimated to be 1-2 per 100,000 and represent two thirds of head and neck paragangliomas of the head and neck [5]. The majority of these neoplasms are sporadic while 10-15% are familial or hyperplastic. The hyperplastic type is most common in populations living in high altitudes (>5000 feet above sea level). Most patients will present in the 4th to 5th decade of life with a slightly earlier onset in the hereditary group [6]. 10% of CBT are bilateral, and only 5% of CBT are considered malignant in nature with metastatic disease on presentation. Historically, the mortality and stroke rate associated with surgical resection was 6% and 23%, respectively [7]. Advancement of preoperative planning, multidisciplinary collaboration, and surgical technique has reduced mortality rate to less than 1% and stroke rate to 1-2%.

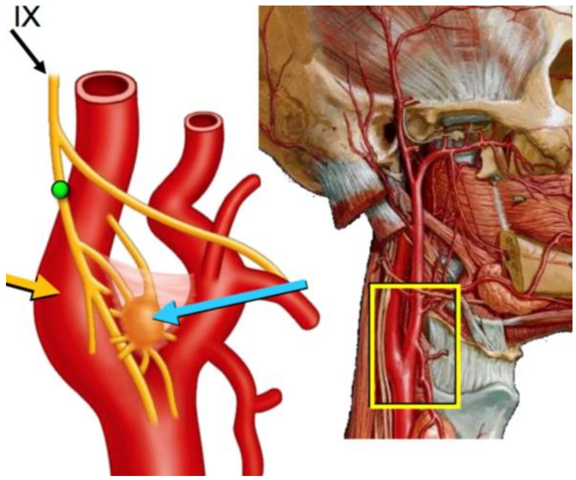


Figure 1: Location and innervation schematic of a paraganglioma (CBT) and its anatomic relationship to cranial nerve IX [4].

2. Initial Evaluation

Majority of patients with CBT present with a painless neck mass (around 65%) anterior to the sternocleidomastoid muscle. The pathognomonic sign on exam is a fixed mass in the vertical axis while mobile on the horizontal axis (Fontaine sign). Approximately 10% of patients will present with hoarseness, dysphagia, pain, winged-scapula, or Horner's syndrome consistent with nerve involvement. The remaining patients with CBT will be diagnosed incidentally during carotid duplex exam or cervical axial imaging. Fine needle aspiration should not be performed given the vascularity of the neoplasm and high risk for hemorrhage.

A careful history is required to identify patients with a genetic predisposition for CBT and other paragangliomas (e.g. pheochromocytoma). Approximately 10-30% of patients have CBT associated with at least one germline mutation in a subunit of the succinate dehydrogenase (SDH) gene which has vital role encoding mitochondria's role in the Krebs cycle. Deactivation of SDH promotes dysregulation of hypoxia-induced genes and growth factors [8]. Genetic counseling may be of benefit for patients and their family members that present at a younger age, with a positive family history, or having bilateral location or multiple paragangliomas [9].

3. Radiographic Evaluation

Duplex ultrasonography is the first-line diagnostic modality for evaluation of CBT. It permits identification of any concomitant carotid artery disease and the vascularity of the neck mass (Figure 2). Digital subtraction angiography was once considered the "gold standard" for diagnosing CBT. It has been replaced by cross-sectional imaging. CT angiography (Figure 3a and 3b) and/or MR angiography provide diagnostic as well as preoperative staging of CBT into the appropriate Shamblin classification (figure 4) [10]. Shamblin classification is an important predictor of neurovascular morbidity with Group 3a and 3b carrying the highest risk of nerve injury or requirement of vascular reconstruction. In cases of suspected metastasis or multicentric tumors, 18F-fluorodeoxyglucose positron emission tomography-computed tomography (FDG PET-CT) provides high specificity and sensitivity for detection (Figures 5a, 5b, and 5c) [11].

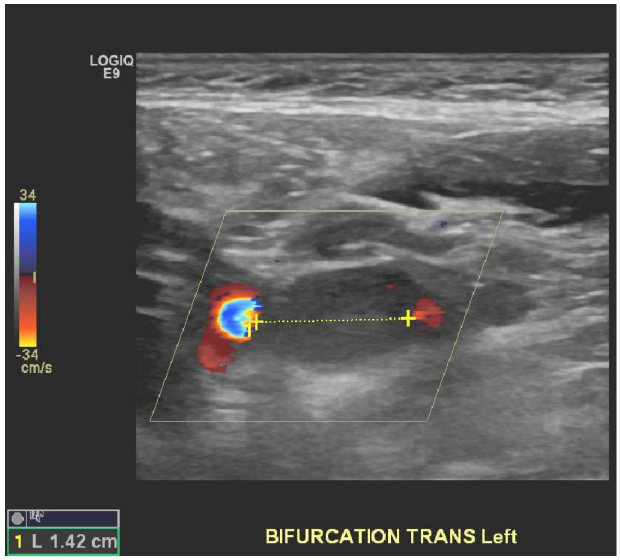
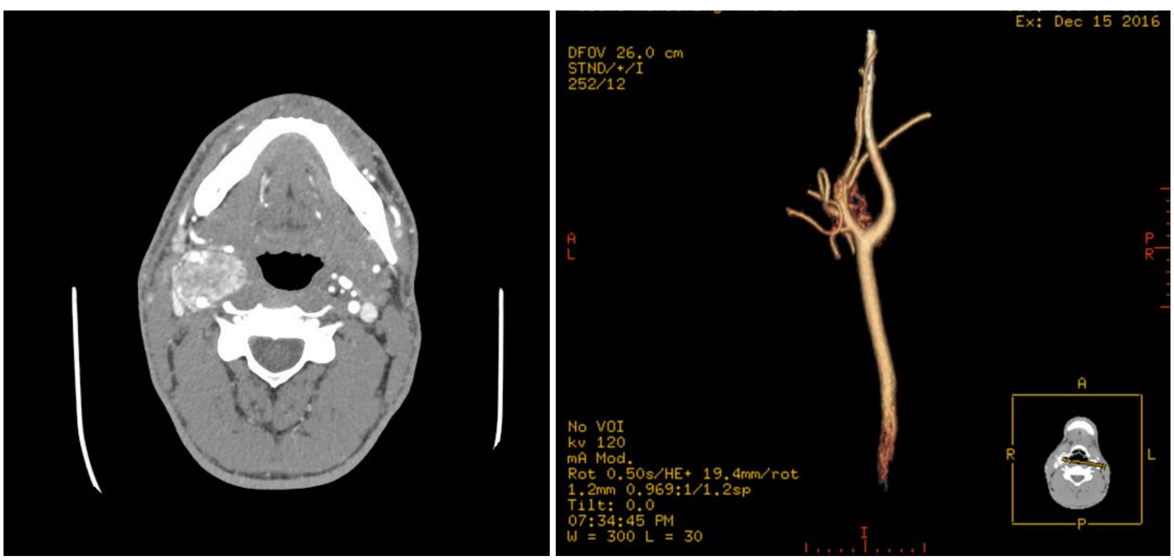


Figure 2: Carotid duplex transverse view of left carotid bifurcation demonstrating a 1.42 cm carotid body tumor splaying the internal carotid and external carotid arteries.



(a) (b)

Figure 3: (a) CT angiogram of a Shamblin II carotid body tumor of the right carotid bifurcation; (b) Volume rendering CT reconstruction demonstrating the same carotid body tumor with splaying of carotid bifurcation and neovascularization.

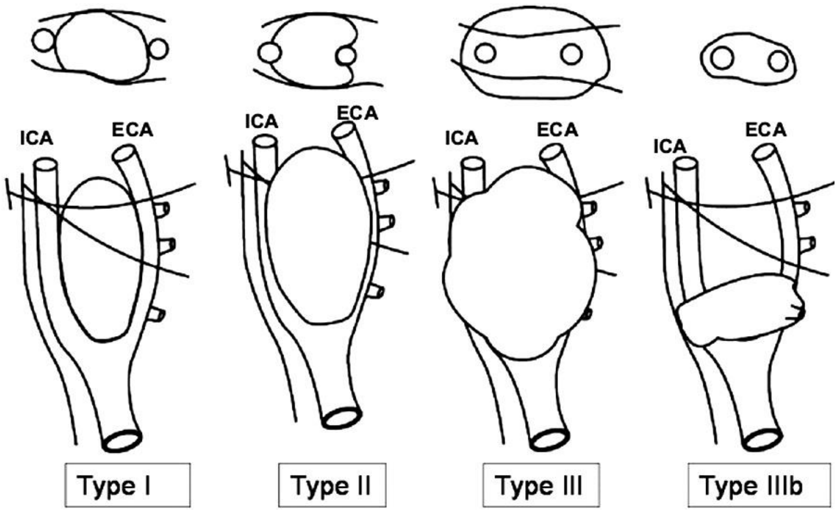


Figure 4: Shamblin classification system. The system is based on the carotid body tumors involvement of the carotid vessels. Cranial nerve XII is depicted superiorly and Cranial nerve X (vertically oriented) and its superior laryngeal branch (oblique course) are shown in relation to the CBT.

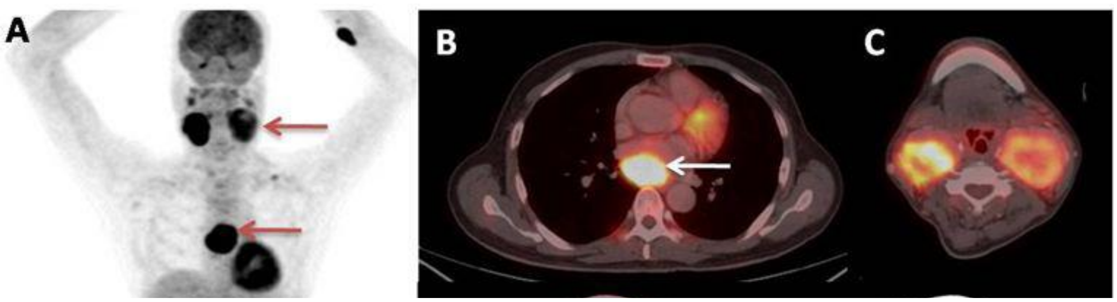


Figure 5: 18F-FDG PET CT images (A) Maximum intensity projection (MIP) demonstrating hyper-metabolic lesions (arrows) in the neck and mediastinum; (B, C) Corresponding transaxial images with tracer uptake in the CBT of the mediastinum and bilateral carotid bifurcations.

4. Preoperative Evaluation

CBT are infrequently hormonally active, but it is recommended to perform biochemical testing for plasma metanephrines or 24-hour urine collection for catecholamines and metanephrines. If the paraganglioma is functional, phenonxybenzamine 10mg two times daily should be initiated 7-10 days prior to surgery. Beta blocker is started only after achieving adequate alpha blockade with phenoxybenzamine. Patients presenting with history of previous neck surgery or new hoarseness should undergo a preoperative otolaryngology evaluation of the vocal cords. Depending on local expertise, collaboration of otolaryngologists and vascular surgeons may facilitate surgical planning and improve outcomes. Patients with bilateral CBT should have a staged surgical resection with an intermediate assessment of the vocal cords.

Preoperative embolization of the feeding vessels to CBT is controversial related to lack of evidence of improved outcomes and procedural stroke risk. Some studies have shown decreased blood loss associated with embolization while others have found no difference. If embolization is performed, surgical resection should be planned within 48 hours to avoid the secondary inflammatory reaction [12-14]. Vein mapping of the greater saphenous vein may be beneficial for larger CBT when vascular reconstruction is expected.

5. Treatment

CBT remains a surgical disease. That being said, there may be a role for observation for small CBT (<2 cm) in certain populations as demonstrated by Muno in Bogota (elevation >2000 meters above sea level) [15]. Radiation therapy is available for elderly patients that may be high risk for surgery or patients who present with local recurrence or metastasis [16]. An important but often overlooked step of the operation is patient positioning. After our anesthesiologist achieves satisfactory intravenous access and arterial line monitoring, the patient is placed in a relaxed, semi-fowler position (lawn chair) (Figure 6A) with both arms tucked. The patient’s head is slightly externally rotated to the contralateral side. Some surgeons place a small roll under the shoulder blades to accentuate the neck hyperextension. We monitor EEG waveforms during the procedure in case carotid artery cross-clamping is required, and the EEG technician will establish the patient’s baseline prior to draping and incision. During draping, anatomic landmarks are clearly visible such as the ear lobe, mastoid process, angle of mandible, clavicle and sternal notch. The use of a portable ultrasound to map and mark a segment of greater saphenous vein is beneficial prior to prepping and draping this segment of the upper thigh. We apply an IobanTM (3M, St. Paul, MN) to the neck and thigh surgical sites to prevent drape migration.

The incision may be made obliquely along the medial border of the sternocleidomastoid muscle or transverse following the natural neck lines (Langer’s line). Either incision will provide adequate exposure. Ligation of the facial vein facilitates lateral retraction of the internal jugular vein and access to the common carotid artery and its bifurcation. Early identification of the vagus nerve and the hypoglossal nerve are important steps in their preservation. The vagus nerve is located in the carotid sheath, and the hypoglossal nerve may be identified by tracing the ansa cervicalis nerve superiorly to its attachment to the hypoglossal. The ansa cervicalis may be sacrificed to simplify access to the carotid bifurcation. The next step is the precautionary use of Rummel umbilical tape tourniquets for vascular control of the carotid and its main branches as well as the additional benefit provided by them for vascular shunt stabilization, if required.

The major tenets of surgical resection of these highly vascularized neoplasm is early identification and preservation of the nerves, maintaining a bloodless field to facilitate the former, and a subadventitial plane along the carotid arteries to ensure complete excision. Bipolar diathermy (Figure 6B) provides an efficient, safe technique for maintaining hemostasis during excision and is similarly used by endocrine surgeons for thyroidectomies for smaller vessels. Van der Bogt and Hamming, et al. were one of the first to describe a cranial to caudal dissection technique which permitted early identification of vital neurovascular structures and demonstrated an associated reduction in surgical morbidity (Figure 6C and 6D) [17]. This technique simplifies the more tedious dissection on the ventral surface of the tumor. The most common nerve injury occurs during the ventral dissection to the superior laryngeal nerve. An injury to this nerve results in the alteration of pitch of the patient's voice and can make it difficult for the patient to yell or be heard in loud areas. A high vagal nerve injury is associated not only with hoarseness but high risk for aspiration and possible need for a PEG tube. In the case of Shamblin 3, with proximal extension, the accessory spinal nerve can be involved and lead to weakness of the trapezius muscle producing a drooping shoulder and a winging of the scapula [18].

A B

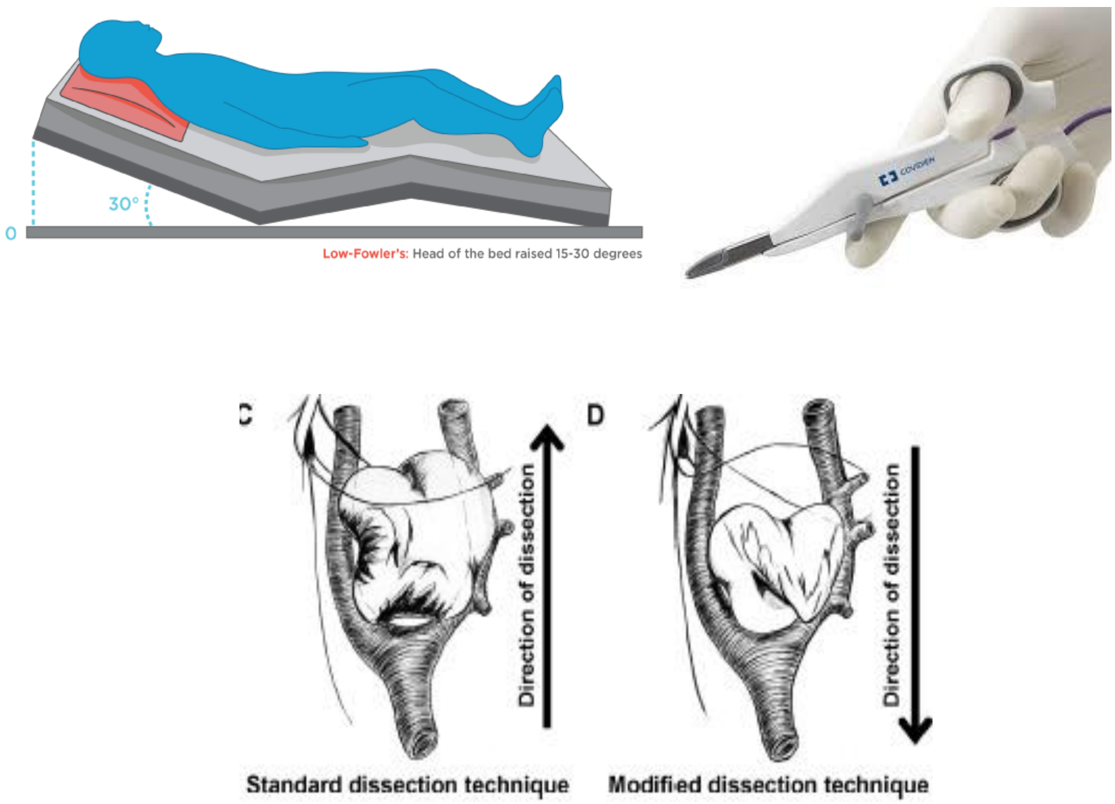


Figure 6: (A) Depiction of modified semi-fowler positioning of patient; (B) LigaSureTM Exact Dissector (Medtronic, Minneapolis, MN) is a commonly used bipolar thermal device for hemostasis; (C, D) Change from standard technique to a craniocaudal technique to reduce neurovascular injury [3, 17].

Postoperative care involves an overnight hospitalization for observation due to risk of bleeding and nerve injury or palsy. The complication risk directly correlates to the size of the neoplasm, the cranial extent of resection, and the Shamblin classification. Postoperative life-long surveillance is required for patients with hereditary paragangliomas or identified SDH genetic mutation.

6. Conclusion

CBT are a rare neoplasm but the most common paraganglioma of the head and neck. Advancement of radiographic imaging techniques has reduced the role and associated risk of preoperative angiography. Molecular genetics have assisted the identification of patients and their family members who may benefit from genetic counseling, comprehensive preoperative imaging, and postoperative surveillance. We continue to refine surgical technique to reduce neurovascular morbidity associated with resection.

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