Operative management of symptomatic, metachronous carotid body tumors involving the skull base and its neurological sequelae

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ABSTRACT
A 44-year-old morbidly obese woman with a history of right carotid body tumor (CBT) resection presented with a symptomatic, nonfunctional, left Shamblin-III CBT. Abutment of the skull base precluded distal internal carotid artery control for arterial reconstruction, favoring parent vessel sacrifice after an asymptomatic provocative test. She underwent CBT resection with anticipated sacrifice of neural nerves X and XII and the common carotid artery and its branches, developing baroreceptor failure syndrome and sequelae of cranial nerve sacrifice. When facing a symptomatic, metachronous CBT abutting the skull base, upfront operative intervention with adjuvant radiation for residual tumor optimizes curative resection. (J Vasc Surg Cases Innov Tech 2021;7:378-81.)

Keywords: Carotid body tumor; Metachronous carotid body tumor; Cranial nerves deficit; Baroreceptor failure syndrome; Multidisciplinary care

CASE REPORT
A morbidly obese 44-year-old woman presented with painful swelling in her left upper neck. She denied recent trauma, tobacco abuse, or family history of paragangliomas. She underwent excision of a right Shamblin-III CBT. Imaging, succinate dehydrogenase levels, or genetic evaluation all remained negative for further work-up. She was lost to follow-up and did not have surveillance duplex imaging, succinate dehydrogenase levels, or genetic evaluation. On physical examination, she had a tender left neck mass; cervical swelling in her left upper neck. She denied recent trauma, to her left neck mass. Cerebrospinal fluid was negative for neoplastic cells. The lesion was nonfunctional with a normal serum normetanephrine level. Imaging (Fig 1) demonstrated a 6.2 cm, left, Shamblin-III CBT abutting the skull base. The patient declined radiation therapy because of claustrophobia and elective surgery despite the risks. A multidisciplinary discussion among Vascular Surgery, Otolaryngology, and Neurosurgery established that balloon test occlusion would determine the need for internal carotid artery (ICA)-external carotid artery (ECA) bypass, with anticipation of parent vessel sacrifice if successful because distal ICA control was prohibitive for arterial reconstruction. After successful balloon test occlusion (Fig 2), the ICA and ECA branches were embolized with Amplatzer plugs (St. Jude Medical, St. Paul, Minn) and Target 360 soft and ultra coils (Stryker, Kalamazoo, Mich), respectively. The patient was discharged home with continued oral treatment for BFS. She has been recovering slowly but remains gastrostomy-tube dependent, and she is still undecided regarding adjuvant radiation therapy and genetic consultation despite recommendations.

The patient provided written informed consent for patient information and images to be published.

DISCUSSION
CBTs are the most common head and neck paragangliomas. This rare pathology arises from the periaortic plexus of the carotid bifurcation. The carotid body maintains homeostasis of carbon dioxide, oxygen, and pH, whereas carotid baroreceptors in the carotid sinus regulate blood
Symptomatic CBTs mandate operative resection that may be associated with significant morbidity. Bilateral synchronous CBTs occur in 7% to 11% of CBTs that can require staged bilateral resection, predisposing to BFS.1,2 Our patient had a symptomatic metachronous, benign, Shamblin-III CBT abutting the skull base. She declined radiation therapy because of claustrophobia and elected for surgical intervention despite the risks, especially related to nerve sacrifice. She underwent preoperative embolization with subsequent extensive resection and carotid ligation without reconstruction, resulting in anticipated, permanent, and unilateral vagus and hypoglossal nerve deficits as well as transient hypertensive crisis due to BFS. We approach CBTs with a multidisciplinary approach in order to optimize outcomes as described by Mohebali et al.3 Herein, we discuss our complications in this complex case as well as preoperative decision-making.

Cranial nerve injury (CNI) is a common complication after surgical resection of CBTs, resulting in significant morbidity in up to 40% of patients.5,6 Patients most commonly experience deficits of the vagus (6%), hypoglossal (6%), and accessory (3%) nerves as well as the sympathetic chain (3%).7 Clinical implications include hoarseness, dysphagia, dysgeusia, unilateral neck weakness, and Horner’s syndrome. The Shamblin classification classically stratifies the degree of tumor encasement of the carotid arteries in order to predict the need for carotid reconstruction; a higher Shamblin classification is associated with greater risk of CNI.8 Other risk factors include bilateral resection, number of lymph nodes removed, distance to base of the skull, tumor volume, and estimated blood loss.2,8,9 In our patient, her Shamblin-III CBT abutting the skull base encased the vagus and hypoglossal nerves, prohibiting preservation despite initial identification and preservation during surgery. Regarding dysphagia and dysphonia after vagus nerve sacrifice, young patients recover well with minimal dysfunction after vocal cord augmentation and therapy by the speech and swallow team. Furthermore, unilateral hypoglossal nerve paralysis has minimal effect on speech and swallowing as the contralateral nerve is enough for this function, despite the expected hemitongue atrophy.

BFS is a well-documented sequela of bilateral CBT resection due to unopposed sympathetic brainstem signals from bilateral baroreceptor denervation.10,11 Headache, anxiety, hypertension, and tachycardia classically characterize this morbidity.2,6,8,10-12 and its spectrum ranges from hypertensive crisis to volatile hypertension and orthostatic tachycardia.3 Hypertensive crisis develops acutely after bilateral resection with significant risk of a cerebrovascular event; volatile hypertension and orthostatic tachycardia are exceedingly more common presentations of BFS with a more gradual onset and more mild and permanent symptoms. A staged approach to resection of synchronous, bilateral tumors minimizes the incidence of this pathology, ranging from 0% to 16%.2,12 Management typically includes clonidine and beta-blockers, counteracting the uninhibited sympathetic outflow.3,10 In our case, an antihypertensive infusion acutely controlled the perioperative hypertensive crisis with gradual postoperative transition to oral antihypertensive and beta-blockade.

Management of metachronous, Shamblin-III CBTs abutting the skull base is an individualized decision as no standardized treatment algorithm exists. Treatment modalities include definitive radiation (XRT), neoadjuvant XRT with resection, and resection with adjuvant XRT for symptomatic CBTs, while observation is appropriate in asymptomatic lesions.10 We discussed XRT for symptomatic CBTs, while observation is appropriate for additional irradiation. Given the patient’s claustrophobia in the setting of a large and symptomatic lesion, we felt that resection with adjuvant XRT for anticipated, residual tumor was the best treatment modality. This rationale guided our multidisciplinary decision for surgical resection in a young patient with a symptomatic CBT.

Tumor involvement of the ICA at the jugular foramen complicates CBT resection as it precludes ICA...
reconstruction. Its incidence is rare in the setting of bilateral CBTs; two case series (n = 306) describe this pathology only twice (<0.01%). A described preoperative evaluation of neurovascular insufficiency involves manual compression of the common carotid artery for 30 minutes. In our patient, a multidisciplinary approach assessed adequate cerebrovascular perfusion with ICA balloon occlusion via a published and standard technique. Imaging confirmed patency of the contralateral autologous interposition graft. The patient underwent uncomplicated ICA and ECA coil embolization with subsequent carotid ligation without reconstruction. In addition, preoperative embolization in CBTs is an adjunct to decrease intraoperative blood loss and CNI, although its efficacy is controversial.

CONCLUSIONS

Bilateral CBTs are a complex and rare pathology with high risk of operative morbidity. A symptomatic, metachronous, Shamblin-III CBT abutting the skull base in a young patient is a rare clinical scenario. Operative intervention requires thorough preoperative planning and a multidisciplinary approach, and surgical complications must be thoroughly discussed preoperatively.
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