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Case Reports

Bilateral Giant Familial Carotid Body Tumors With Concomitant Skull-Base Paraganglioma and Facial Nerve Palsy

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Carotid body tumors, rare neck paragangliomas arising from the common carotid artery bifurcation, can be classified as sporadic, hyperplastic, or familial. The familial type is often bilateral and associated with germline mutation of the mitochondrial enzyme succinate dehydrogenase. We report the rare case of a 42-year-old man who presented with bilateral giant familial carotid body tumors associated with a concomitant skull-base paraganglioma, left-sided facial nerve palsy, and an incomplete circle of Willis. We describe the excision of the tumors in 2 stages (the left mass and associated paraganglioma first and the right mass second), 6 months apart, with use of general anesthesia, and we discuss other operative considerations. ***(Tex Heart Inst J 2022;49(2):e207370)***

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Carotid body tumors (CBTs) are rare neck paragangliomas of neural crest origin.[1,2] The carotid body is a chemoreceptor cluster within the carotid bifurcation that helps the body adapt to changes in oxygen and carbon dioxide concentrations and changes in pH.[3] Carotid body tumors can be classified etiologically as sporadic, hyperplastic, or familial.[3-5] They often occur in chronic hypoxic conditions and in association with a germline mutation of the mitochondrial enzyme succinate dehydrogenase.[3-5] More rarely, CBTs are associated with nonparaganglionic tumors in syndromes such as von Hippel Lindau syndrome, multiple endocrine neoplasia type 2, and neurofibromatosis (NF) type 1.[4] About 5% of CBTs are bilateral, and 5% to 10% are malignant; these rates are much higher in patients with inherited disease.[2,5] Shamblin classified CBTs into 3 types according to their involvement with the branches of the common carotid artery (CCA): type I (within the CCA bifurcation); type II (partially surrounding or adhering to the branches); and type III (encasing the branches).[6] The tumors grow slowly[7,8] and occur predominantly in women.[2,7,9,10] Accurate preoperative evaluation and diagnosis of CBTs, especially larger ones, are important because the risk of postoperative death and morbidity is high.[1,2,11] We present the rare case of bilateral giant familial CBTs associated with a concomitant skull-base paraganglioma and left-sided facial nerve palsy.

Case Report

A 42-year-old man presented at our institution with painless bilateral neck masses. The left-sided neck mass had been growing slowly for 11 years and, for the last 2 years, had been associated with intermittent nonradiating dull pain, dysphagia, left-sided hearing impairment, occasional vertigo and tinnitus, and rightward deviation of the mouth. The smaller right-sided neck mass had been growing slowly for 3 years. The patient's medical history included no other tumors, no hypertension or diabetes, and an attempted biopsy of the left-sided neck mass 10 years previously. His older brother had had a similar left-sided neck mass and had died 5 years previously.

Physical examination of the left-sided mass revealed a firm, painless, 8.0 × 6.0-cm mass in zones II and III that could be moved horizontally but not vertically (Fontaine sign); a transverse scar over the mass from the previously attempted biopsy; and left-sided facial nerve palsy of the lower motor neuron type, involving the mandibular branch. Examination of the right side of the neck revealed a 5.0 × 6.0-cm mass in zone II. A neck ultrasonogram showed bilateral masses that suggested salivary gland tumors. However, a craniocervical computed tomographic angiogram revealed highly vascular masses in the left (3.2 × 2.8 cm) and right (3.0 × 2.6 cm) CCA bifurcations and concomitant splaying (Lyre sign) (Fig. 1). The angiogram also revealed a similar mass posterosuperior to and closely related to the left-sided neck mass, producing a dumbbell-shaped image; a missing anterior communicating artery; and a fetal right posterior cerebral artery. An abdominal ultrasonogram revealed an ectopic right kidney with a simple renal cyst. The results of a 24-hour urine vanillylmandelic acid assay were normal. Together, these findings supported a diagnosis of bilateral CBTs with left-sided facial nerve palsy, incomplete circle of Willis, and ectopic kidney. We decided to excise the tumors in 2 stages: the left CBT and associated skull-base paraganglioma first, and the right CBT second.

Operative Technique

The first-stage excision was performed approximately 3 weeks after presentation. The results of preoperative evaluation, which included a renal function test, complete blood count, electrocardiogram (ECG), and chest radiograph, were normal. The operation was performed with the patient under general anesthesia and with simultaneous monitoring of central venous pressure and arterial blood pressure, capnography, ECG, and pulse oximetry.

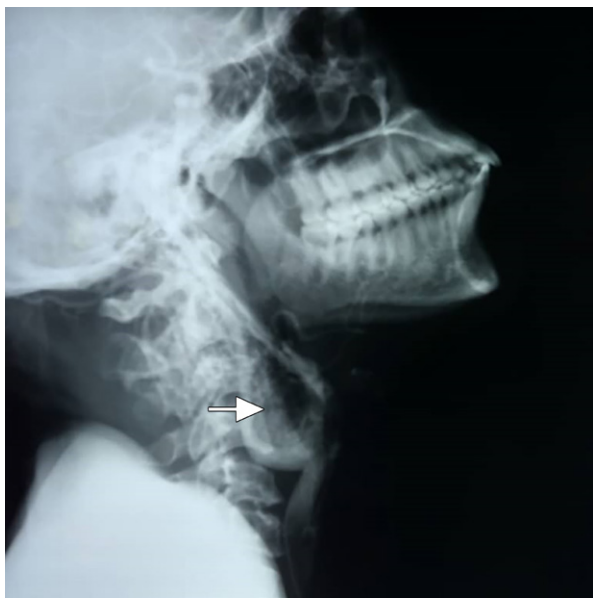

Fig. 1 *Computed tomographic angiogram shows the splayed branches of the right carotid artery (Lyre sign) (arrow).*

Anesthesia was induced by premedicating the patient with intravenous (IV) ranitidine 50 mg, glycopyrrolate 0.04 mg/kg, and midazolam 5 mg. Intravenous propofol 2 mg/kg, fentanyl 100 μg, and suxamethonium 100 mg were then administered, followed by insertion of a 7.5-mm cuffed orotracheal tube. Epinephrine 2 mg in 48 mL of normal saline was kept in a syringe pump for IV administration in case of hypotension; nitroglycerin 200 mg in 45 mL of normal saline was kept in another syringe pump for IV administration in case of hypertension. Anesthesia was maintained with IV pancuronium, 8 mg administered immediately and 4 mg intermittently in boluses, IV fentanyl intermittently in boluses of 50 μg, and inhalational isoflurane at 1 minimum alveolar concentration. Central venous pressure was maintained between 8 and 12 cm H_2O, and carbon dioxide levels were maintained between 38 and 48 mmHg. Intravenous nitroglycerin was infused at a rate of 0.02 mg/kg/min immediately after the tumor was manipulated. Four episodes of hypotension and one episode of hypertension occurred during the operation. The hypotension was controlled with intermittent boluses of IV epinephrine 0.27 μg/kg; the hypertension, with intermittent boluses of IV nitroglycerin at a rate of 1.5 mg/min.

After general anesthesia was achieved, the patient was placed in a supine position with a sandbag beneath his shoulders. His neck was then extended and rotated to the right. The skin from the earlobes to the nipples was prepared for operation; in addition, the skin on the thighs was prepared for saphenous vein harvest, if needed. To approach the left-sided neck mass, an oblique incision was made on the anterior border of the sternocleidomastoid muscle (SCM). The SCM was retracted laterally, and the superior belly of the omohyoid muscle was divided. The carotid sheath was incised to reveal an ovoid (potato-shaped), 4.0 × 2.0-cm Shamblin type II tumor and an adjoining 6.0 × 6.0-cm superior cervical tumor extending to the base of the skull (Fig. 2). The left CCA was mobilized. The left external and internal carotid arteries were similarly identified and carefully dissected from the tumor. Because the patient's incomplete circle of Willis made cross-clamping of the carotid arteries risky, hemostasis was instead achieved by a combination of vessel-looping, suture repair of injuries to the internal jugular vein, and unipolar diathermy at reduced wattage in the external carotid artery feeding branches. The CBT was excised caudiocranially from within the carotid artery bifurcation. The adjoining superior cervical tumor was dissected to its stalk from the base of the skull, divided, and excised.

After all excisions were complete, the anesthetic agents were tapered, the neuromuscular blockade was

reversed, and the patient was extubated. He was then transported to the intensive care unit while receiving IV nitroglycerin to maintain his systolic blood pressure below 140 mmHg.

The total operative time was 368 minutes, the estimated blood loss was 1,900 mL, and 1,350 mL of blood were transfused. The patient's postoperative course was uneventful, and he was discharged from the hospital on postoperative day 4.

The second-stage excision of the contralateral mass was performed 6 months later under the same anesthetic and operative conditions. The excised 4.0 × 3.0-cm tumor was a Shamblin type II. An adjoining 1.0 × 0.5-cm posterior auricular lymph node was also excised. The total operative time was 277 minutes, the estimated blood loss was 350 mL, and no blood transfusions were needed. The patient's postoperative course was uneventful, and he was discharged on postoperative day 3. He remained well thereafter.

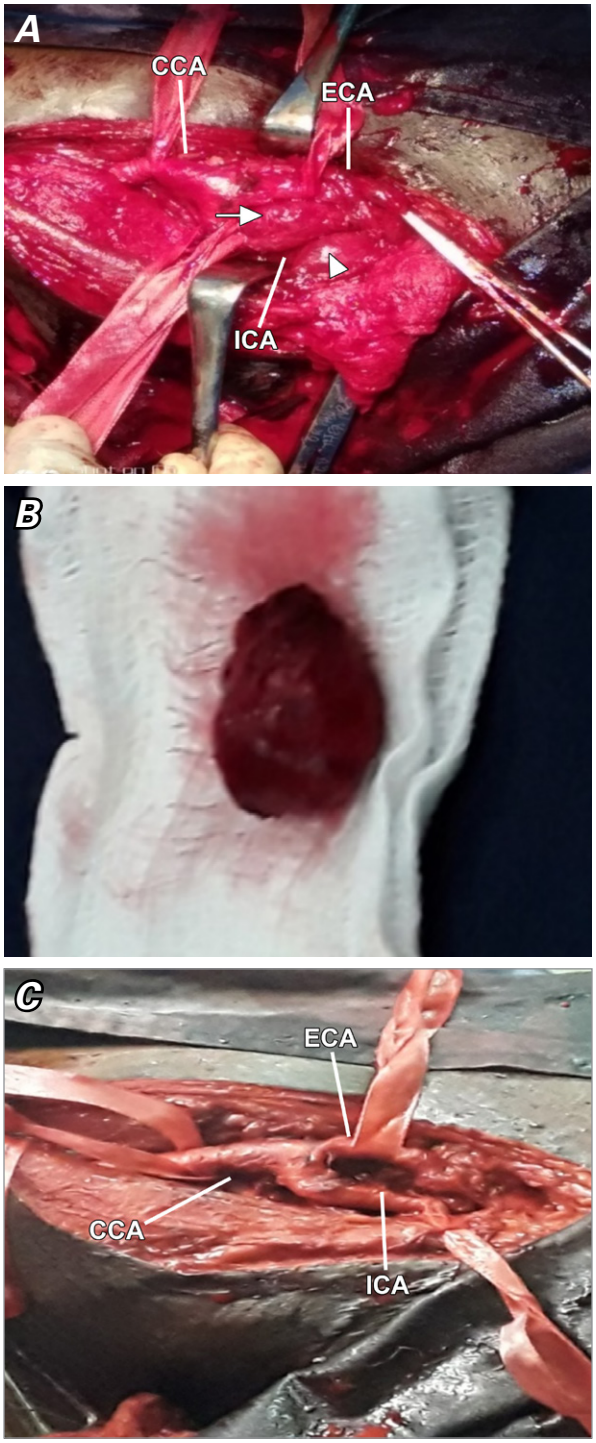

Fig. 2 *Intraoperative photographs show **A**) the left carotid body tumor (arrow) and adjoining cervical paraganglioma extending to the skull base (arrowhead) before excision, **B**) the 4.0 × 2.0-cm ovoid tumor excised from the carotid bifurcation, and **C**) the carotid artery and its branches after tumor excision.*

CCA = common carotid artery; ECA = external carotid artery; ICA = internal carotid artery

Histologic Analysis

Postoperative histologic analysis of the excised left and right CBTs and left cervical mass with skull-base extension revealed benign proliferation of nests (zellballen) of polygonal cells enclosed by sustentacular cells and fibrovascular tissue. The tumor cells contained abundant granular and eosinophilic cytoplasm and large pleomorphic nuclei. Histologic analysis of the excised lymph node revealed typical to markedly enlarged germinal centers with numerous tingible body microphages and no evidence of malignancy. These findings were consistent with a diagnosis of benign paraganglioma.

Discussion

The standard treatment for CBTs is surgical excision. Radiotherapy and clinical monitoring are reserved for patients with unresectable tumors. The need for preoperative embolization is debatable because it often results in additional neurologic complications.[1,2,8,12,13] A complete arterial anatomy, especially in the intracranial aspect, is important. In this case, the underdeveloped circle of Willis prompted our decision to avoid cross-clamping the carotid arteries intraoperatively. Because of the ever-present risk of postoperative neural injury, detailed informed consent and thorough preoperative documentation of cranial nerve status were mandatory. For bilateral CBTs with or without concomitant vagal paragangliomas, staged excision of the smaller lesion first and the larger tumor second is recommended to minimize the risk of bilateral nerve palsies.[11,14] In this case, however, the existing left-sided facial nerve palsy posed a greater risk, so we opted to operate first on the larger lesion on the left side, to minimize the risk of inflicting bilateral cranial nerve injuries. We recommend against a minimal preoperative incision biopsy, which in rare cases can be complicated by torrential hemorrhage.[1,8,11]

Although the vast majority of cervical paragangliomas are hormonally inactive, anesthesia for surgical extirpation of these tumors must take into account hemodynamic changes and instability that may occur during tumor handling. Hypotension, hypertension, and arrhythmias in that situation have all been documented.[15,16] In this case, the hypotension and hypertension that occurred during excision of the left-sided tumor were managed by administering titrated boluses of epinephrine and nitroglycerin, respectively. Most CBTs are Shamblin types I and II and can be safely excised without the need for vascular reconstruction, even though in some instances the external carotid artery may be sacrificed.[2] The greater risks associated with excision of Shamblin type III CBTs must be thoroughly explained to affected patients before surgery.[2,14]

Conclusion

In treating bilateral CBTs associated with unilateral cranial nerve palsy and an incomplete circle of Willis, the side with the nerve lesion should be operated on first because it poses greater danger to the patient. Intraoperative cross-clamping of the carotid arteries must also be avoided. Patients with a familial history of CBT should be evaluated for concomitant paragangliomas elsewhere in the body and undergo lifelong monitoring because of the risk of metachronous paragangliomas.

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