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Five-year outcome of a staged giant Shamblin type III carotid body tumor excision

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ABSTRACT

Giant carotid body tumors, defined as those >8 cm in size, are extremely rare. Definitive surgical management is a complex undertaking because these large tumors tend to have grown to envelop cranial nerves and the carotid artery, and few data exist regarding the long-term outcomes for these patients. We present the case of a patient with bilateral giant carotid body tumors who underwent staged embolization and excision of a >10-cm carotid body tumor. After 5 years of follow-up, we demonstrated that elective open repair can provide long-term symptomatic relief. We describe and illustrate the crucial steps and considerations regarding the excision of complex Shamblin type III carotid body tumors. (J Vasc Surg Cases Innov Tech 2023;9:101320.)

Keywords: Carotid body tumor; Carotid tumor; Shamblin type III; Vascular mass excision; Vascular tumor

Carotid body tumors (CBTs) are rare neuroendocrine neoplasms that arise from around the carotid body at the carotid bifurcation. Almost one quarter infiltrate the carotid vessels and require vascular surgical expertise for removal.[1] Only seven cases of giant CBTs >8 cm in size have been reported in the literature to date, providing few details regarding the exact methods by which excision is performed. The gold standard for the management of CBTs is to surgically resect these tumors as early as possible, which can present an especially complex undertaking for Shamblin type III tumors in which the tumor has grown to encase the cranial nerves, with the carotid artery within the tumor.[2] A thorough description of the methods by which a giant Shamblin type III CBT can be excised to minimize blood loss and the risk of cranial nerve damage will assist surgeons undertaking excision.

A 38-year-old woman presented to our clinic 1 year after completing chemotherapy for mediastinal diffuse large B-cell lymphoma with bilateral large >10-cm right and 8-cm left pulsating neck masses with bruits that were tender to palpation and had been growing slowly for 15 years (Fig 1, *A and B*). Her symptoms were notable for dysphagia, intermittent stridor with exertion, a raspy voice, significant neck pain that was greater on the right, and aural fullness with mild hearing loss. No neurologic symptoms, such as persistent headache, amaurosis fugax, or focal weakness, were present.

The imaging findings were consistent with bilateral Shamblin type III CBTs (Fig 1, *C and D*). The serum metanephrine and normetanephrine test results were within normal limits, and the patient had no signs of a hyperadrenergic state. Genetic testing was performed and was positive for a heterozygous pathogenic p.P81 L variant in succinate dehydrogenase B. The patient provided written informed consent for the report of her case details and imaging studies.

OPERATIVE TECHNIQUE AND OUTCOMES

The operative objectives included preservation of cranial nerve function (cranial nerves X-XII), avoidance of stroke, complete tumor resection, relief of compressive aerodigestive symptoms, and minimization of bleeding during surgery. The larger and more symptomatic right-sided lesion was planned to be addressed first in two stages. The first stage was preoperative and involved endovascular embolization of the blood supply to the right-sided CBT using Onyx glue (Medtronic) by interventional neuroradiology (Fig 2). The second stage was intraoperative, with neck dissection and tumor excision performed jointly by the vascular and otolaryngology teams 48 hours after preoperative embolization had been achieved.

During preoperative embolization, the Onyx 34L liquid embolization system was used to embolize the arterial supply feeding the tumor, including the branches arising from the external carotid artery, mainly the ascending

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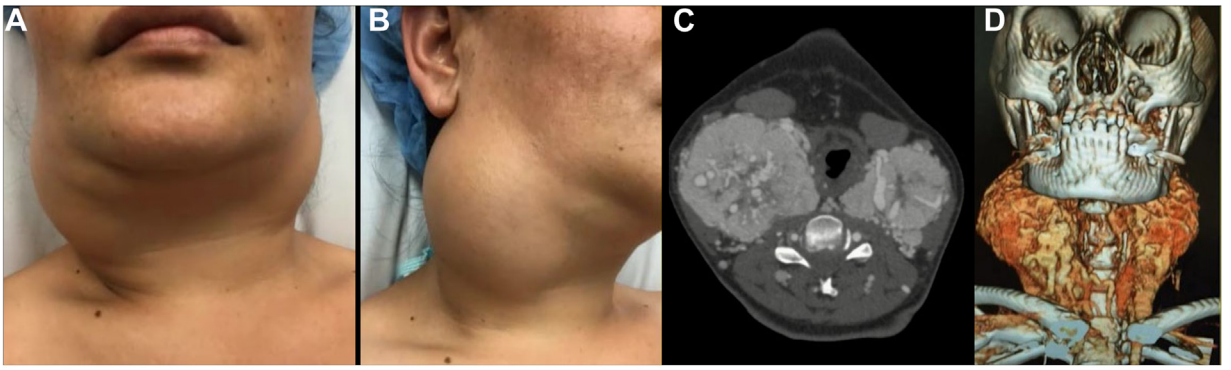

Fig 1. A. Anterior external appearance of bilateral carotid body tumors (CBTs) at presentation. **B.** Right external view of giant CBT on presentation. **C.** Computed tomography angiography of the head and neck showing bilateral Shamblin type III CBTs. **D.** Three-dimensional reconstruction of computed tomography angiography of the head and neck.

Fig 2. Preoperative embolization of the blood supply to the carotid body tumor (CBT). **A.** Pretreatment angiogram. **B.** Angiogram after Onyx administration.

pharyngeal artery, facial artery, occipital artery, and superior thyroid artery. Careful selection of feeder branches using microcatheters was used to direct Onyx into the respective feeder branches to avoid the risk of extension of the embolic material into the external carotid artery. Angiographic balloon test occlusion of the treated side was performed with a second catheter in the contralateral internal carotid artery.

For the neck dissection and tumor excision, the patient was positioned supine with preparation that included the right neck from above the mandible to the clavicle and the right leg for potential saphenous vein recovery (Fig 3, *A*). A generous transverse oblique incision was made that was centered on the tumor. The platysma muscle was traversed, and wide subplatysmal flaps were elevated both superiorly and inferiorly. The sternocleidomastoid muscle was then mobilized laterally, away from the tumor (Fig 3, *B*). The flaps were secured to the drape using skin hooks. The eleventh cranial nerve was identified first (Fig 3, *C*), followed by the twelfth cranial nerve in its medial orientation (Fig 3, *D*). The cranial nerves were encased in tumor. Bipolar electrocautery was used to dissect the tumor free from the nerve. Multiple draining veins were identified, ligated, and divided. The tumor was then mobilized.

Proximal control was obtained of the common carotid artery with an umbilical tape (Fig 3, *E*). Further dissection of cranial nerve XII was completed, and distal control of

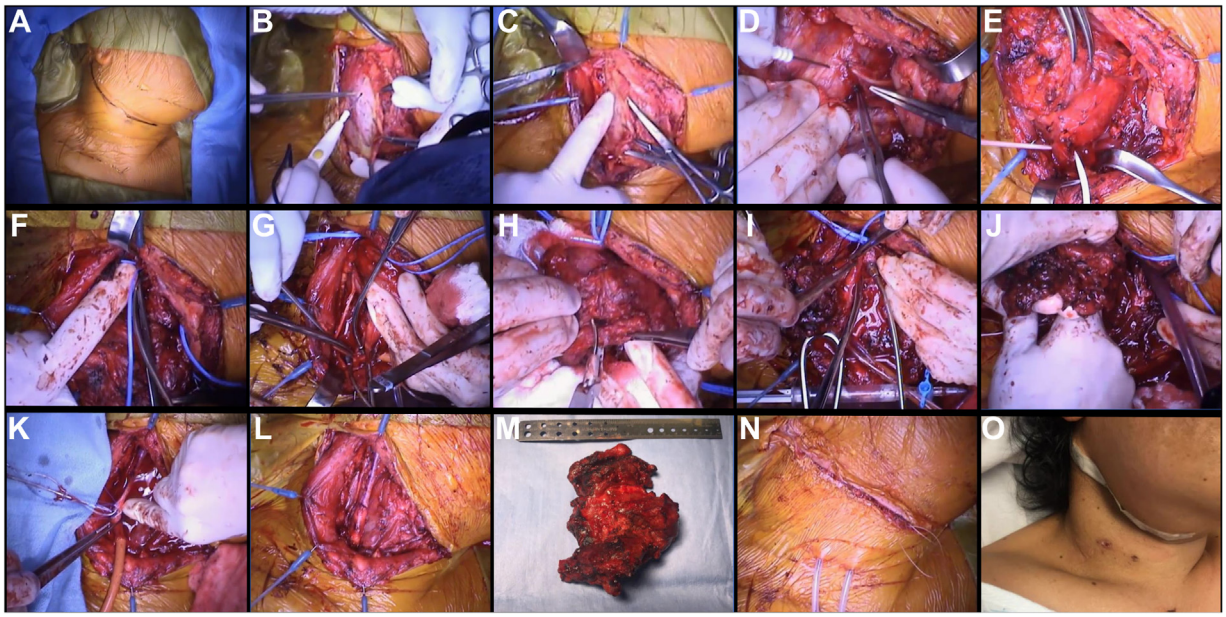

Fig 3. Giant Shamblin type III carotid body tumor (CBT) excision, step-by-step. **A.** Preoperative preparation. **B.** After dissection of platysma, the sternocleidomastoid muscle is mobilized. **C.** Cranial nerve XI is identified and protected. **D.** Cranial nerve XII is mobilized from the tumor. **E.** Proximal control of the common carotid artery is obtained. **F.** Distal control of the internal carotid artery is obtained. **G.** Cranial nerve X is mobilized from the tumor. **H.** Electrocautery is used to divide the tumor. **I.** The carotid artery is mobilized from the tumor. **J.** With protection of the nerves, the tumor is removed. **K-L.** Carotid artery reanastomosis is performed if needed. **M.** Tumor inspection. **N.** Wound closure with drain placement. **O.** Postoperative drain removal.

the internal carotid artery was obtained with a single vessel loop (Fig 3, *F*). The tenth cranial nerve was identified running between the internal jugular vein and tumor laterally and was dissected free throughout its entire length (Fig 3, *C*). This was facilitated with the use of a nerve hook. The entire tumor was mobilized further with a combination of a harmonic scalpel and bipolar electrocautery (Fig 3, *H*). Unplanned injury at the carotid artery bifurcation occurred, requiring intraoperative shunting with a Pruitt-Inahara shunt.

Meticulous periadventitial dissection directly around the artery with bipolar electrocautery under loupe magnification was performed. Because of the injury at the carotid artery bifurcation, the external carotid artery was suture ligated at its origin and was resected with the tumor en bloc. Once the carotid vessels were free, the remainder of the tumor was dissected free and removed in its entirety, ensuring protection of cranial nerves X, XI, and XII (Fig 3, *I and J*).

Resection and end-to-end anastomosis of the internal carotid artery was required to prevent kinking due to overt laxity of the vessel resulting from the large tumor burden (Fig 3, *K* and *L*). Over the Pruitt-Inahara shunt, the ends of the internal carotid artery were brought together to complete an end-to-end anastomosis. The shunt, which was surrounded with vessel loops, was pulled out just before completing the anastomosis. The tumor was inspected in its entirety (Fig 3, *M*). The wound was closed over two suction drains in standard fashion (Fig 3, *N*).

Immediately after surgery the patient's symptoms were limited to mild transient hoarseness, and she was discharged on postoperative day 3 after drain removal (Fig 3, *O*). Flexible laryngoscopy was performed within 1 week of surgery, which revealed right vocal fold paresis, for which she received bilateral vocal cord injection with hyaluronic acid gel (Restylane; Galderma), which resulted in resolution of the paresis. Her postoperative course was notable for dysphagia requiring percutaneous endoscopic gastrostomy (PEG) tube placement. She could only tolerate small amounts of oral food initially. She required the PEG tube for 9 months before being able to fully tolerate an oral diet and having the PEG tube removed. The surgical pathology findings indicated paraganglioma with virtually absent mitoses.

At her 5-year follow-up visit, the patient had no symptoms on the side from which her tumor had been removed, and she declined operative intervention for the contralateral tumor. At 5 years, she had no evidence of recurrence on the operative side.

DISCUSSION

We present a specific operative technique for a staged approach to open surgical excision of a giant Shamblin type III CBT. The most important steps for successful resection are the achievement of proximal and distal

control of the common carotid artery at the base of the neck and the internal carotid artery at the base of the skull and the identification of the cranial nerves.

One of the greatest challenges is the avoidance of nerve injury, because the distance from the tumor to the skull base has been shown to be highly predictive for the occurrence of cranial nerve injury during CBT resection.[3] Cranial nerve XII is the most often injured during CBT resection, and the best method to prevent such injury is to protect the nerves during tumor mobilization.[4] Inadvertent injury to the carotid artery or bifurcation is an inherent risk of this surgery, because dissection along the carotid artery occurs in the periadventitial plane. Although not preventable, inadvertent entry into the carotid artery can be managed with any shunt the operator is most familiar with using. We used the Pruitt-Inahara shunt in the present patient owing to the length and laxity of the carotid artery. When injury involves the carotid bifurcation, the external carotid artery can be ligated, such as was performed in the present patient.

The operative approach for removing Shamblin type III CBT relies on individual patient anatomy and surgeon preference. Although one case study suggested that a Shamblin type III tumor can be removed in a bloodless plane using a finger dissection method, this can be challenging when the tumor is adherent to structures and warrants the use of a combination of blunt and sharp techniques and electrocautery.[5] Although in the present case, the exposure was adequate to achieve both proximal and distal control, approaches for extension of giant CBTs to the skull base have been described, including covered stent application before CBT resection with the arterial wall en bloc and midline mandibulotomy.[6,7] Transmural tumor invasion of the carotid artery can also require partial vessel sacrifice with internal carotid artery reconstruction using great saphenous vein grafts; thus, we included the leg in our operative preparation.[8]

Although much debate has centered on whether preoperative embolization is useful in CBT resection in general, a recent large propensity-matched analysis suggested that doing so significantly reduces operative blood loss.[9] In addition, studies suggest that as the time between preoperative tumor embolization and excision increases, both the blood loss and the operative time increase.[10] Regarding excision of giant CBTs specifically, using Onyx for preoperative embolization at a 48-hour time point before excision has previously been described with good results.[11] In the present case, we used Onyx 34 because the injection is slow, controlled, and less likely to cause catheter entrapment. Care should be given to the avoidance of standard electrocautery after the use of Onyx, because the compound is highly flammable and quite literally causes microexplosions on ignition, increasing the chance of adjacent nerve injury. Alternative approaches to embolization include platinum-based, fully detachable packing coils.[12] We prefer intra-arterial Onyx injection for embolization when the number of feeders is limited (fewer than five to six). If too many feeders are present, we also use direct stick embolization with preference for an *N*-butyl cyanoacrylate with ethiodol 25% mixture, because Onyx can cause breakage of the hub of the needle used to puncture the lesion due to a reaction with dimethyl sulfoxide.

Mutations of the SDHB gene are known to be responsible for most hereditary head and neck paragangliomas, and patients with CBT who are positive for SDHB mutations have a higher risk of metastatic disease.[13,14] In general, these patients benefit from a multispecialty approach that should include vascular surgeons, otolaryngologists, neurointerventional radiologists, and genetic counselors to reduce the operative times and blood loss and allow for identification of tumors in related asymptomatic patients.[15] The identification of genetic components of the disease are especially important because mortality for these patients has been found to be influenced by recurrence and metastasis rather than the tumor size or Shamblin type.[16]

We advised our patient to undergo resection of the contralateral tumor after she had recovered from her procedure; however, the stridor that had motivated her to pursue intervention had resolved. At 5 years after surgery, the patient no longer had symptoms on the right side. Because the mass effect from her remaining contralateral CBT has caused a return of dysphasia and pain, we plan to use our multidisciplinary approach and excision in the coming year. In addition to counseling her as before regarding the risks of cranial nerve injury, bleeding, and infection, it will be crucial to counsel our patient on the specific risks of contralateral surgery in the context of her flexible laryngoscopy findings consistent with ongoing right-sided vocal cord paresis, which could possibly be related to sacrifice of the right-sided superior laryngeal nerve despite preservation of the vagus. Our approach to excise the contralateral tumor would be the same as has described in the present report. It is important to recognize that these challenging procedures can be accomplished safely with good results and symptomatic relief to the patients in the long term.

CONCLUSIONS

Excision of giant CBTs is a complex undertaking that benefits from the expertise provided by a multidisciplinary perioperative team and a staged approach involving preoperative embolization of the blood supply to the tumor before neck dissection and tumor excision.

DISCLOSURES

None.

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