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Carotid Body Tumor with Skull Base Extension

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Conflict of interest: None declared

> **Patient:** Male, 59

Final Diagnosis: Benign carotid body tumor

Neck mass Symptoms:

Medication:

Clinical Procedure:

Specialty: Otolaryngology

Objective: Rare disease

Background: Carotid body tumors are rare tumors that arise from the paraganglionic cells of the carotid body. They are usu-

ally benign, requiring surgical resection as the treatment of choice.

Case Report: We present a case of a 59-year-old man with a benign left carotid body tumor that progressed to a very large

> size, compromised the patient's airway, completely encased the carotid vessels, vagus and hypoglossal nerves ipsilaterally, and reached the contralateral carotid vessels and ipsilateral skull base. Because of the cranial extension of the tumor, the patient had to undergo preoperative endovascular coiling of the carotid vessels pri-

or to total excision of the tumor.

Conclusions: Due to the critical location of carotid body tumors, their vascularity, and high risk of neurovascular complica-

> tions, surgical resection can be quite challenging, especially when the tumor is large. We propose an approach to managing large parapharyngeal tumors by endovascular occlusion of the internal carotid artery above the skull base. Further, a suggestion is made to add a category to Shamblin's classification - Shamblin IV - for pa-

tients with skull base extension requiring preoperative endovascular intervention.

MeSH Keywords: Carotid Body Tumor • Embolization, Therapeutic • Paraganglioma • Skull Base

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/905526

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Background

Carotid body tumors (CBTs) are rare tumors arising from the paraganglionic cells of the carotid body present near the bifurcation of the common carotid artery [1]. The most commonly encountered are head and neck paragangliomas, accounting for 60–78% of them [1]. The criterion standard treatment for CBTs is surgery [2]. Shamblin et al. classified CBTs in the year 1971 into 3 classes: in Class I the tumor is small and easily

resectable, in Class II the tumor is adherent, partially surrounding the carotid vessels, and its resection is technically more difficult in comparison to Class I, and in Class III the tumor is large (usually >5 cm), completely encasing the carotid vessels, its resection is very difficult, and interrupting the cerebral circulation is almost always inevitable [3,4]. Patients with Shamblin III tumors have a higher risk of developing postoperative neurovascular complications compared to those with Shamblin I or II.



Figure 1. (A) An axial CT image showing the complete engulfment of the internal and external carotid arteries by the tumor. (B) A reconstructed coronal image showing the tumor's extension to the skull base. (C) An angiogram showing splaying of the internal and external carotid vessels (Lyre's sign), and tumor blush. (D) A post-embolization picture showing the coils in the left common carotid artery (star), and in the left internal carotid artery above the skull base (arrow), and the vascular plug in the left external carotid artery (circle).



Figure 2. The carotid body tumor and internal and external carotid vessels after their complete resection.

In this case report, we present a challenging case of CBT, in which the tumor was large, completely encasing the carotid artery ipsilaterally and extending up to the skull base.

Case Report

A 59-year-old male was referred to our hospital with a large, left-sided neck mass and a tracheostomy tube which was inserted at an outside hospital due to an impending airway obstruction. The mass has been progressively growing for 17 years, but got worse 2 months prior to his presentation. The patient started suffering from progressive shortness of breath and changes in his voice 1 year ago, but there had been no change in his ability to swallow.

On examination, the patient looked well, was not in respiratory distress, and the tracheostomy tube was in place and patent. On palpation, there was a large, left-sided neck mass measuring 7×5 cm, crossing levels 2, 3, and 5. The mass was firm and non-tender, with no palpable lymph nodes. Fiber-optic examination showed a significant bulge on the left side of the pharynx, which was almost completely obstructing the airway.

A computed tomography (CT) scan showed a large, well-defined, intensely enhancing mass on the left side of the neck, surrounding the distal portion of the left common carotid artery and encasing the left internal carotid and external carotid arteries. The mass was seen extending medially and bulging into the oropharynx and compressing the airway, reaching the carotid artery on the contralateral right side. Moreover, the mass was extending superiorly to touch the skull base. It measured $9.1 \times 9 \times 6.8$ cm. There were no significantly enlarged cervical lymph nodes. CT angiogram showed a clear Lyre's sign (Figure 1).

To confirm the diagnosis, given the unusual large size of the mass and its recent rapid growth, a small incisional biopsy

was performed. The biopsy result was consistent with paragangliomas. Carotid balloon occlusion test was done and the patient showed no neurological deficits during the procedure. Due to the tumor's skull base extension, and anticipating the potential challenges of achieving adequate intraoperative distal control of the internal carotid artery, we opted to occlude the common, external, and internal carotid arteries to allow safe excision of the mass. The internal carotid artery was occluded intracranially using coiling technique to facilitate safe resection of the tumor off the skull base bone. Occlusion of the external carotid artery was done by using a vascular plug device (Figure 1).

Two days after the embolization, excision of the left carotid body tumor was performed. The vagus and hypoglossal nerves and internal jugular vein were found to be encased by the tumor and were sacrificed. We were able to preserve the spinal accessory nerve, sympathetic chain, phrenic nerve, and brachial plexus. The common carotid artery was ligated low in the neck just above the coiled segment of the artery. The parapharyngeal space and skull base parts of the tumor were completely excised. The internal carotid artery was ligated at the level of the skull base. In addition, the tumor was dissected off the contralateral carotid artery to fully mobilize it and allow enbloc excision (Figure 2).

The patient's recovery was uneventful and he was discharged in a good condition, tolerating oral diet well.

Discussion

Carotid body tumors often require surgical excision as a definitive treatment. But considering their location, high vascularity, and close relation to the carotid vessels and cranial nerves, these tumors can be technically challenging to excise. Patients with Shamblin III tumors were found to have higher risks of developing neurovascular injuries (30-46%) in comparison to those with class 1 or 2 due to the nerve involvement [2,5,6]. After finding an association between the tumor's proximity to the skull base and increased incidences of cranial nerve injury and intraoperative blood loss, Kim et al. suggested adding the tumor's distance to the base of skull (DTBOS) and its volume, along with Shamblin's classification, as additional predictors of patient risk for cranial nerve injuries and intraoperative bleeding [7]. As demonstrated by our case, temporary balloon occlusion (TBO) testing of the carotid artery is essential in patients who are at a risk of requiring internal carotid artery ligation to facilitate the tumor's excision.

In our case, we were able to successfully coil the internal carotid artery above the skull base to allow easy distal control of the artery below the skull base.

Conclusions

Endovascular occlusion should be considered when managing large, vascular parapharyngeal tumors. We suggest adding a further category to Shamblin's classification: Shamblin IV. This new category should include tumors with cranial direction growth up to the skull base, restricting access to the

distal internal carotid artery in the neck and necessitating endovascular blockage.

Conflict of interest

None.

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