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CASE REPORT

Giant carotid body paraganglioma: our approach to surgical excision

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ABSTRACT

BACKGROUND: Carotid body tumours typically present as an asymptomatic anterior neck mass. However, larger tumours can become symptomatic. Surgical excision is the primary treatment, often preceded by embolisation to reduce tumour haemorrhage and perioperative risks. This case report presents a case of a successful excision of a giant Shamblin III carotid body tumour.

CASE REPORT: We present a case of a 37-year-old woman, originally from Mozambique, with a decade-long painless, gradually growing pulsatile left cervical swelling and progressive dysphonia and dysphagia to solids. Cervical computed tomography angiography revealed an expansive hypervascular lesion centred in the left carotid space spanning from the left carotid bifurcation to the base of the skull, with approximate dimensions of 11 x 8.1 x 5.5 cm. She had no impairment of cranial nerves but had a tumour-compressed upper airway. Further diagnostic investigations were carried out, and the diagnosis of dopaminergic-producing left Shamblin III carotid body tumour was made.

The patient underwent tumour embolisation followed by surgical excision the following day. Under general anaesthesia to safely protect the upper airway until tumour removal, the authors deployed detachable hydrocoils to occlude several ECA branches supplying the tumour. After 24 hours, a multidisciplinary team undertook surgical excision. During the procedure, a peripheral neuromonitoring system was used to minimise the risk of injury to the lower cranial nerves. A regular longitudinal exposure of the carotid vessels was conducted. After proximal control of the common carotid artery, an easily bleeding tumour was medially mobilised to enable internal carotid artery exposure and preservation. Due to the high adhesion of the tumour and complete encasement of the external carotid artery, the authors ligated this artery and reconstructed the CCA bifurcation with an end-to-end common-to-internal carotid anastomosis. She was extubated on postoperative day one and was discharged home after ten days, without neurological deficits.

CONCLUSIONS: The management of carotid paragangliomas, particularly those classified as Shamblin III, requires a multidisciplinary approach and refined surgical technique to ensure complete tumor removal without compromising essential structures. The combination of preoperative embolization and a well-planned surgical procedure resulted in an excellent outcome for the patient, with no neurological complications.

Keywords: carotid body tumour; carotid paraganglioma; Shamblin; surgical excision; embolisation

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BACKGROUND

Carotid body tumours (CBTs) are rare neuroendocrine neoplasms accounting for 0.6% of all head and neck tumours.[1] Diagnosis is frequently based on the discovery of a pulsatile and painless lateral anterior neck mass, confirmed by imaging finding.[2] Larger tumours can become symptomatic, with discomfort, dysphagia, and autonomic dysfunction being the most commonly reported symptoms.[2] Surgical excision is usually the primary treatment, often preceded by embolisation to reduce tumour haemorrhage and perioperative risks, especially in larger tumours adherent to carotid arteries.[3]

Surgical resection of CBT is technically challenging due to its hypervascularity and proximity to the carotid vessels and cranial nerves.[4] In highly risk and selected cases, radiotherapy may be chosen as the first therapeutic option. Postoperative peripheral neurological morbidity, mainly involving the cranial nerves, is between 14 and 49% of early deficit and between 6 and 23% of persistent deficit.[5]In 1971, Shamblin *et al.* developed a surgical classification for CBT based on its relationship with carotid vessels. This classification divides CBT into three distinct groups, each with a positive predictive value for surgical difficulties and postoperative complications. Carotid body paragangliomas classified as Shamblin I or II can usually be removed en bloc.[6] However, Shamblin III CBTs, which encase the carotid vessels, present a significant surgical challenge and are often difficult to remove without carotid artery ligation. WE report a case of successful excision of a giant Shamblin III CBT.

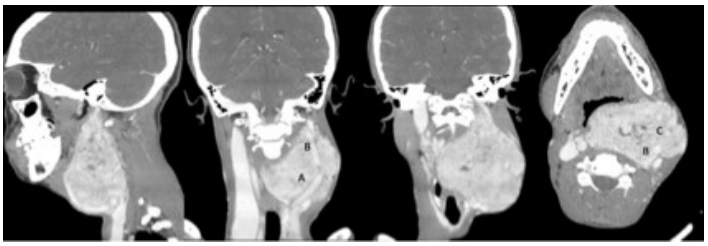
CASE REPORT

We present a case of a 37-year-old woman, originally from Mozambique, with over a decade of a painless, gradually growing pulsatile left cervical swelling and progressive dysphonia and dysphagia to solids, without dyspnea (Figure 1).

Figure 1. Pre-operative images of the giant left cervical paraganglioma

In her rural home village, the local healer made several cervical cutdowns over the years to bleed the bulge and try to reduce it without success. Upon admission to the emergency department, she had an asymptomatic moderate hypotension, and the cervical computed tomography angiography (CTA) revealed an expansive hypervascular lesion centred in the left carotid space, causing splaying and encasement of the external and internal carotid artery. The lesion extended into the parotid, parapharyngeal, retropharyngeal, prevertebral, and submandibular spaces, spanning from the left carotid bifurcation to the base of the skull, with approximate dimensions of 11 x 8.1 x 5.5 cm, Figure 2.

Figure 2. Pre-operative cervical computed tomography angiography (sagittal, coronal and axial views)



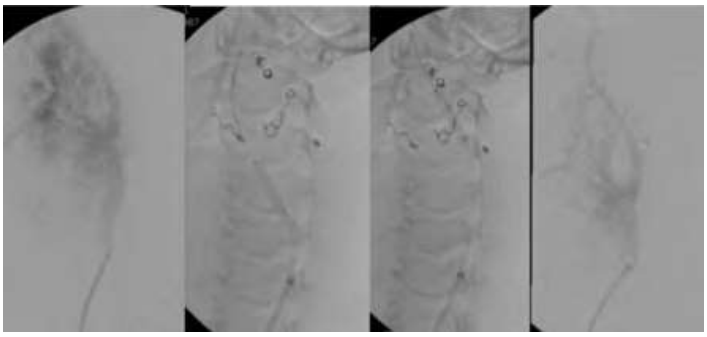
A – common carotid artery; **B** - internal carotid artery; **C** – external carotid.

A presumable diagnosis of CBT was made. The patient was admitted to the Ears, Nose and Throat (ENT) department for a 15-day observation period, during which she received corticosteroid therapy and a liquid diet with dysphagia improvement. She had no impairment of cranial nerves but had a tumour-compressed upper airway. Further diagnostic investigations were conducted, including CTA for thoracoabdominal-pelvic staging, urinary metanephrines and urinary vanillylmandelic acid, serum and urinary catecholamines, and cranial and cervical magnetic resonance imaging scans. The diagnosis of dopaminergic-producing left Shamblin III CBT without metastases or synchronous tumours was established.

In line with the diagnosis, the patient underwent tumour embolisation followed by surgical excision the following day. Under general anesthesia to safely protect the upper airway until tumor removal, the authors used a 70cm femoral 6Fr-sheath and telescopic technique to deploy detachable 0,018'' hydrocoils (AzurTM Terumo®) to occlude several left External Carotid Artery (ECA) branches supplying the tumour. The contrast enhancement of the tumour was significantly reduced and delayed after the procedure. ECA balloon occlusion was performed to evaluate the potential reduction in tumor perfusion following ECA occlusion; however, as no significant decrease in tumor filling was observed, endovascular exclusion of the ECA was not performed.

An Internal Carotid Artery (ICA) balloon occlusion test did not reveal any cerebral oximetry or EEG change, Figure 3.

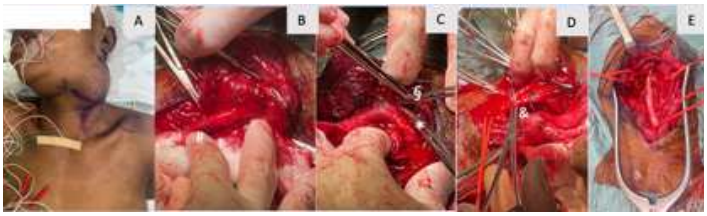
Figure 3. Intra-operative digital subtraction angiography images of the carotid body tumour embolization procedure



From left to right: initial angiography showing hypervascular tumor; external carotid artery balloon occlusion; C – internal carotid artery balloon occlusion test; D – final angiography.

After 24 hours, a multidisciplinary team of vascular and ENT surgeons undertook CBT surgical excision. During the procedure, a nerve monitoring system (NIM Medtronic®) was used to minimise the risk of injury to the lower cranial nerves (electrodes on the trachea, left tongue, soft palate and trapezius muscle would emit an audible alert if tissue dissection were near the vagus, hypoglossal, glossopharyngeal and accessory nerves, respectively). A regular longitudinal exposure of the carotid vessels was planned using a common left cervical oblique incision. After proximal control of the Common Carotid Artery (CCA), an easily bleeding CBT was medially mobilised (and ligated in some segments) to enable CCA bifurcation, ICA exposure and preservation. The tumour was firmly adherent to the vagus nerve on its postero-lateral wall, and it compressed the hypoglossal nerve crossing its anterior wall, but these nerves were released and preserved. Due to the high adhesion of the tumour and complete encasement of the ECA, ligation was decided, and reconstruction of the carotid bifurcation was performed by an end-to-end CCA-ICA anastomosis, Figure 4.

Figure 4. Intra-operative images of the carotid body tumour surgical excision



A – Positioning and monitorisation **B** - Common cartotid artery; **C** – internal carotid artery; **D** - vagus nerve; **E** - External carotid artery.

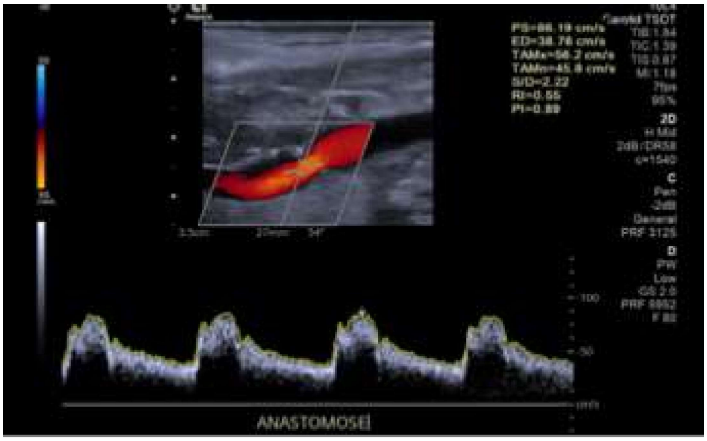
The procedure lasted 2 hours, and the patient showed a labile blood pressure profile, suggesting CBT dopamine release during tumour manipulation. Blood loss was estimated at 1000 ml.

Postoperatively, the patient was transferred to the intensive care unit, requiring temporary vasopressor support and blood transfusion. She was extubated on postoperative day one and transferred to the general ward on postoperative day three, starting speech and swallowing training. The patient was discharged 10 days postoperatively with no neurological deficits. She quickly resolved the previous dysphonia and solid dysphagia from upper airway compression. After one month, the patient presented with a healed scar and no neurological signs, and she stated that this procedure had given her a “new life”, Figure 5.

Figure 5. Clinical presentation at one month postoperatively.

Pathology confirmed it was a paraganglioma, and Doppler ultrasound showed a patent carotid axis without stenosis, Figure 6.

Figure 6. Post-operative Doppler ultrasound of the carotid axis.



DISCUSSION

The management of CBT, especially those classified as Shamblin III, presents significant clinical challenges due to their close anatomical relationship with vital neurovascular structures. This condition is associated with morbidity due to its slow, progressive growth, often leading to local invasion and long-term nerve damage. The goal of surgical treatment is to prevent such morbidity. In cases without large tumour volume, minimising intraoperative morbidity becomes even more crucial. While carotid artery involvement poses a risk for ischemic vascular injury, postoperative morbidity is more commonly related to iatrogenic nerve injury—precisely the outcome that surgical resection aims to prevent. Therefore,

utilising all available strategies to minimise surgical morbidity is essential, including intraoperative nerve monitoring and collaboration with experienced ENT surgeons specialising in skull base procedures.

In this case, the strategic use of preoperative embolisation played a crucial role in reducing intraoperative blood loss and facilitating a safer and more controlled resection of the tumour. Coupled with a carefully planned and executed surgical approach, this led to complete tumour excision without compromising cranial nerve function or critical vascular integrity. The absence of postoperative neurological deficits underscores the success of this collaborative approach and highlights the importance of individualised planning in managing such rare and high-risk cases.

Overall, this case exemplifies how advanced surgical techniques and interdisciplinary coordination can lead to favourable outcomes even in the most complex presentations of carotid body paragangliomas. It also contributes valuable insight to the limited body of literature on the successful treatment of extensive Shamblin III tumours with vascular involvement.

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