

Case Report

Carotid body tumour: an enigma that remains

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ABSTRACT

Carotid body tumour is a form of paraganglioma which arises from the carotid body. This tumour is known for its rich vascular supply mainly contributed by the ascending pharyngeal artery, a branch from external carotid artery. Surgical excision preceded by pre-operative embolization remains the definitive treatment despite the countless feared complications. We present our experience with a case of carotid body tumour which was embolised prior to surgical excision, unfortunately complicated with a thromboembolic event. This patient was found to have an anatomical variation in the cerebrovascular anatomy known as a fetal posterior cerebral artery which led to a paradoxical PCA infarction post-embolization. We would like to highlight this rare presentation along with its management as there were scarce evidence of this case in the literature.

Keywords: Carotid body tumors, Paraganglioma, Carotid artery, Cerebral infarction

INTRODUCTION

Carotid body tumour (CBT) are rare tumours of the head and neck which arises from neural crest progenitor cells.¹ CBT is a type of paraganglioma, which are highly vascular tumours owing to its blood supply from the external carotid artery, in particular ascending pharyngeal artery and are found typically at the carotid bifurcation.² Surgical excision preceded by pre-operative angioembolization is considered to be the gold standard of treatment by many surgeons. Nevertheless, both surgical excision and angioembolization carries high risk of thromboembolic events. Thus, it is imperative that preoperative imaging is done and meticulously reviewed to stratify the risk and identify any possible anatomic variants which may affect the outcome for the patient. We report a case of carotid body tumour which was managed with angioembolization followed by surgical excision, complicated with a thromboembolic event. This patient was found to have an anatomical variation in the cerebrovascular anatomy known as a fetal posterior

cerebral artery (PCA) which led to a paradoxical PCA territory infarction as reported previously.³

CASE REPORT

A 55-year-old Indian lady with underlying type II diabetes mellitus presented with a one year history of left-sided neck swelling which progressively enlarged for the past 2 months. She also complained of intermittent pain over the swelling and dysphagia during the recent 2 months. There was however no hoarseness, shortness of breath, palpitations, syncopal attacks or constitutional symptoms. Patient denies any fever or recent trauma or fall. She also had no family members with similar condition.

Upon examination, patient appeared comfortable with stable vital signs. Neck examination revealed a pulsatile swelling over left level II measuring 2×2 cms, which was firm, non-tender with no signs on inflammation. The swelling was fixed vertically but was found to be mobile in the horizontal axis. Cranial nerves were grossly intact.

We proceeded with flexible nasopharyngolaryngoscopy which was unremarkable.

Ultrasound neck done revealed a lobulated solid lesion splaying the left internal carotid artery and external carotid artery which is suggestive of carotid body tumour. Further assessment was done with contrast enhanced computed tomography, supported the diagnosis and demonstrated an enhancing soft tissue lesion within the carotid sheath measuring 2.4×1.9×3.3 cms. Magnetic resonance angiography of the carotid artery revealed a mass occupying at the left carotid bifurcation causing the characteristic splaying of the internal and external carotid arteries (Lyre sign) as well as presence of flow void signal was seen within the mass (Figure 1). Following this, a diagnostic cerebral angiogram was done which displayed tumour blush at the left common carotid bifurcation with its arterial supply from the left external carotid artery, likely ascending pharyngeal branch. Incidentally, presence of a variant was unveiled in the posterior cerebral circulation whereby the left fetal posterior cerebral artery (fetal PCA) supplied the P2 segment of left posterior cerebral artery (Figure 2).



Figure 1: MRA reveal mass occupying at the left carotid bifurcation.

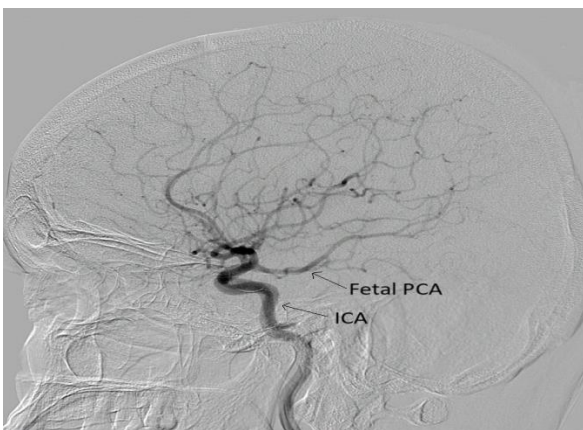


Figure 2: Cerebral angiogram with variant in posterior cerebral circulation.

Patient and her family were counseled on the diagnosis and options of management and complications were discussed thoroughly. They were keen for surgical intervention and were aware of the possible risks and complications that could possibly arise. A multidisciplinary meeting was conducted between the vascular team, interventional radiology team, otorhinolaryngology team and patient.

Patient subsequently underwent embolization of the left ascending pharyngeal artery whereby 10% glue was used under general anaesthesia. Post embolization angiogram showed minimal tumour blush. Patient remained intubated till the day for operation. Intraoperatively, a 2×3 cms smooth mass was discerned at the carotid bifurcation, posterior to the internal and external arteries (Figure 3). Excision of the tumour in its entirety was done successfully with preservation of the carotid artery without clamping. The vagus, hypoglossal, glossopharyngeal nerves were identified and preserved during the excision. In addition to that, there was minimal blood loss and patient's blood pressure was fairly stable throughout the procedure.

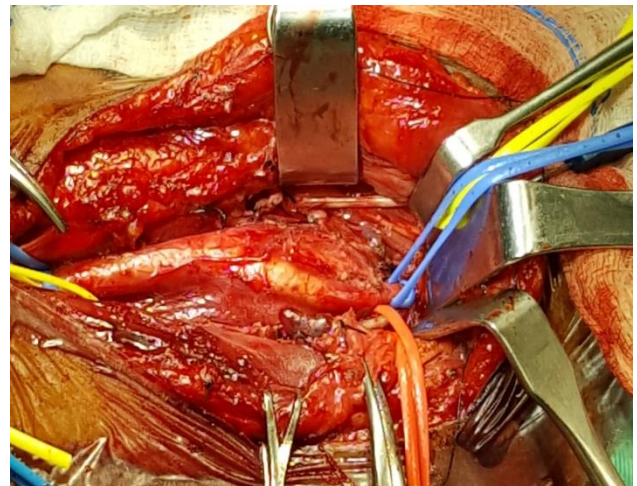


Figure 3: A smooth mass at carotid bifurcation.

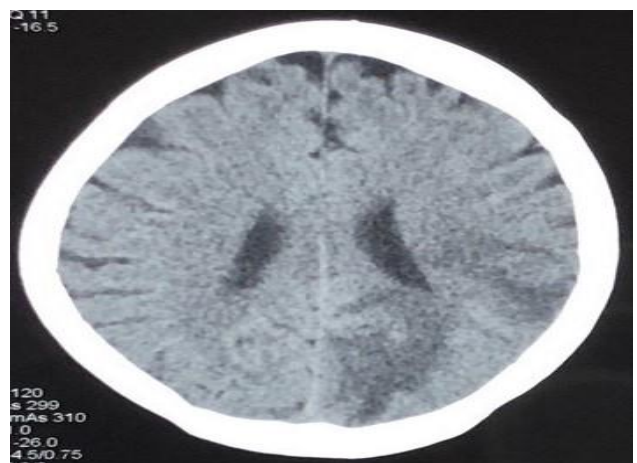


Figure 4: CT brain with left parieto-occipital infarct.

Postoperatively, patient was noted to have no movement of her right limbs. Hence, an urgent computed tomography of the brain was done which revealed a right basal ganglia and left parieto-occipital infarct (Figure 4). Patient was started on aspirin at once and subsequent repeated scans showed no evolving changes. Following intensive physiotherapy and rehabilitation, the patient successfully regained power and function in the affected limbs with marked improvement of power from 1/5 to 4/5. Subsequent follow-up revealed no evidence of recurrence with gross improvement of the motor functions of the affected limb.

DISCUSSION

CBT a rare, slow-growing tumour is the most common paraganglioma with an incidence of 1:30000.⁴ External carotid artery is the main blood supply to the CBT via multiple feeder vessel with largest being the ascending pharyngeal artery as in our case. Gold standard management has metamorphose over the past decade to complete resection with a preoperative embolization as an adjunct owing to the rich vascular nature of CBT.⁵ In addition to that, surgical resection is advocated due to not only the effects of local complications but also the risk of malignancy.⁶ Having said that, surgical excision carries high risk of major blood loss as well as neurological morbidity. Risk of stroke following excision of CBT are highly variable have been reported to range between 0% to 20%.⁷

Angioembolisation is an adjunct procedure done prior to surgery to devascularise CBT.⁸ First described by Schick et al, preoperative embolization was done to aid in reducing blood loss and operative time.⁹ Nonetheless, angioembolization has its fair share of complications mainly risk of neurological complications even in the experienced operators.^{7,10} Certain authors have reported that the risk of stroke following post-embolization may even exceed 10%.¹¹ Although the claimed benefits remain debatable, pre-operative angioembolization techniques are still employed to reduce the risk of intraoperative bleeding which allow a bloodless surgical field to ensure safe and proper excision of the tumour. Our patient described above underwent a preoperative embolization followed by a surgical excision.

Unfortunately, our patient developed significant neurological deficits resulting from ischemia in the left parieto-occipital lobe which are territories supplied by both the internal carotid artery (ICA) and posterior cerebral artery (PCA). Concurrent infarction in the left ICA-PCA territories indicated that this was not a straightforward stroke. Looking back retrospectively, preoperative cerebral angiogram did reveal an incidental finding of a left fetal PCOM supplying the P2 segment of the left posterior cerebral artery. We postulate that this was likely the reason for the paradoxical PCA infarction. The posterior cerebral circulation receive blood supply from the ICA via the fetal PCOM hence, emboli from the

ICA may enter the PCA P2 segment causing a cross embolization.³

In our patient, angioembolization was done 20 hours prior to surgery under general anesthesia and patient remained ventilated and sedated till the surgery. The neurological deficit was only detected post-operatively once the patient was weaned off general anaesthesia. It was also not possible to ascertain if the thromboembolic event had occurred as a complication of embolization or following surgical excision as patient was not assessed after angioembolization. The cause of stroke remains a conundrum till date. Nevertheless, treatment was instituted at once for the stroke with satisfactory neurological function recovery noted within two weeks. This complication has shown that it is imperative to assess the patient's neurological function following embolization. Furthermore, intraoperative electroencephalographic monitoring can also be helpful in lowering the risk of neurological morbidity during surgical manipulation.⁷ We also highlight the importance of vascular imaging preoperatively to identify possible anatomic variants which can contribute to brain ischemia.

CONCLUSION

Carotid body tumor is managed widely with surgical excision with a prior embolization albeit the notable complications. Reports of post-procedural stroke indicate that the risks of angioembolization should be weighed carefully. Our patient suffered from stroke which is a known complication of both embolization and surgery. We would like to strongly highlight that it is imperative that all patients are extubated after angioembolization to assess for possible thromboembolic events. In addition to that, possibility of atypical pattern of stroke can be due to an anatomical variant in the cerebral vasculature as in our case.

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