Rural Centre Based Management of the Carotid Body Tumour

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Abstract A 40 year old male patient reported to our rural based hospital with a complaint of discomfort associated with a swelling on the left side of the neck since 8 years. A provisional diagnosis of a carotid body tumour was made based on clinical examination and ultrasound examination. Higher investigations could not be performed due to unavailability at the rural setup and referral to a specialty centre was not possible due to financial constraints of the patient. Even with advances in diagnostic and surgical techniques, surgery still presents a major threat of injury to the cranial nerves. Nevertheless, it remains the preferred method of treatment for these tumours. Our case shows that such masses can be removed successfully and that, with care, the cranial nerves and the carotid arteries can be preserved at the rural hospital.

Keywords Carotid body tumour · Chemodectoma · Potato tumour · Non-chromaffin paraganglioma · Zellballen formation

Introduction

Carotid body tumours are vascular tumours that arise from the paraganglionic cells in the outer layer of the carotid artery and develop at the bifurcation of the common carotid artery in the neck. It is also known as chemodectoma (from chemoreceptor cells) or Potato tumour (typical shape of a potato). It is a type of Non-chromaffin paraganglioma and is a non-functional tumour. Its incidence is very rare. It is

seen with a slight more frequency in females [1]. Malignancy occurs in 6–12.5% of cases, which ranks carotid body paragangliomas as the most frequently occurring malignant head and neck paraganglioma [1].

Case Report

A 40 year old male patient reported with a complaint of discomfort due to a swelling on the left side of the neck (Fig. 1) which first appeared 8 years ago. It progressively increased in size of a pea to the present size of a large lemon. The patient did not experience any symptoms of dysphagia or dyspnoea or hoarseness of voice, any syncope attacks. The patient's medical history was unremarkable. Physical examination revealed a firm, discrete, oval swelling of size 5 × 4 cm in the left carotid triangle anterior to the sternocleidomastoid muscle at the level of the upper border of the thyroid cartilage. With smooth surface the tumour was non adherent to the overlying skin and underlying muscle, with mobility in the transverse direction. Fluctuation, reducibility, compressibility, transillumination, movement on deglutition or protrusion of tongue was absent. There were no transmitted or expansile pulsations associated with the swelling. Auscultation did not reveal any bruit or murmur. No cervical or generalized lymphadenopathy was noted. Ultrasound showed a rounded, highly vascular mass of size 5.3 × 4.8 cm at the carotid artery bifurcation causing splaying of the external and internal carotid arteries on that side. Hence, a provisional diagnosis of a carotid body tumour was made. The perfusion of the contralateral carotid artery was assessed by manual occlusion of the ipsilateral common carotid artery and assessment of weakness in the extremities which was found to be absent at end of 15 min, which was found to be

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Fig. 1 Swelling in the left carotid triangle anterior to the sternocleidomastoid muscle at the level of the upper border of the thyroid cartilage

adequate. Carotid angiography and balloon occlusion test could not be performed due to unavailability at the centre.

Surgical Management

In hypotensive anaesthesia, saline and adrenaline solution was infiltrated along the incision line (the anterior border of the sternocleidomastoid muscle (SCM)) to reduce bleeding and to achieve a plane of dissection. For good exposure of tumour, a flap was raised in the sub-platysmal plane and the SCM was divided. Due to distortion of the regional anatomy identification of the great vessels was difficult. The carotid sheath was incised to expose the common carotid artery and the tumour was found to engulf the carotid artery at its bifurcation. The branches of external carotid artery supplying the tumour were ligated (Fig. 2) along with feeder vessels cauterized using bipolar diathermy. After achieving hemostasis and dissection plane, encasement of internal carotid artery is relieved by excising tumour in toto. The surgery and post-operative period was uneventful, even without neurological deficit.

Histopathological Diagnosis

Grossly 50 grams, $5 \times 4 \times 3$ cm size, pink to tan coloured, fairly circumscribed tumour covered with a very thin fibrous capsule with indentation of the internal carotid artery (Fig. 3a). Microscopically predominant

Fig. 2 Ligation of feeders to the tumour

monomorphic cell population comprising of neuroendocrinal cells forming clusters are seen. Also, areas of spindle-shaped cells (sarcomatoid foci) at places specially surrounding the cell clusters and highly vascular areas found randomly in the stroma. Nuclear pleomorphism and cellular hyperchromatism seen at places though occasional should not be considered evidence of malignancy. Features are suggestive of paraganglioma (Figure 3b).

Discussion

Carotid body tumour is a benign neoplasm, originates from the chemoreceptor cells located at the carotid bifurcation with incidence of malignancy of 5–13% [1]. It is the most common Chemodectoma (non-chromaffin paragangliomas) in the head and neck region, others being Glomus vagale, Glomus jugulare and Glomus tympanicum [1]. More prevalent in individuals who live at higher altitudes due to chronic hypoxia; hyperplasia of the carotid body resulting into carotid body tumour [3] with slight predominance in females [1, 2]. Larger tumours may cause dyspnoea, dysphagia due to pressure of the mass or may even be associated with cranial nerve palsy (X, XII) in 25% of the cases [2]. Usually manifest as a painless lump in the lateral part of the neck and grow over a period of years with few episodes of sudden syncopal attacks because of compression of the internal carotid artery and hypoxia to the brain. Multiple or metastatic tumours might secrete catecholamines which will manifest with symptoms of palpitation, sweating, headache and hypertension and raised urinary vanillyl—mandelic acid level. Non-invasive imaging for diagnosis like, CT scan shows hyperdense mass as compared to adjacent muscle on a non-enhanced scan due to large volume of blood in it. MRI scan shows mildly hypointense to isointense mass to adjoining muscle on T1

Fig. 3 **a** Pink, circumscribed tumour with thin fibrous capsule with indentation (*plastic tube*) of the internal carotid artery. **b** Areas of spindle-shaped cells surrounding the cell clusters with nuclear pleomorphism and cellular hyperchromatism in 40× zoom

weighted images and moderate hyperintense on T2 weighted images. Angiography should be used as a definitive tool for diagnosis, as it helps in identifying the tiny bleeder vessels and larger feeder vessel if any can be embolised prior to surgery. FNAC and open biopsy are contraindicated. However, benign and malignant tumours cannot be distinguished on histopathology, but only based on local spread and distant metastasis. Management options are local excision of the tumour by securing a subadvential plane.

Ideally pre-operative carotid angiography should be performed to visualize the feeder vessels and embolisation of a larger branch should be done. Preoperative evaluation of sufficiency of the contralateral internal carotid artery to supply both sides of the brain should be performed using balloon occlusion test. If perfusion is insufficient a bypass graft using a Dacron graft or a saphenous vein graft may be performed prior to excision of the tumour [1]. This would reduce the chances of neurologic deficits due to inadequate perfusion of the brain. Also if need for excision of part of the carotid artery is essential, vascular grafting should be done after excision of the tumour. The growth of neoplasms is slow but relentless, and therefore mandates an aggressive surgical approach in low-risk patients [4]. Radiation therapy appears to be of minimal help as a primary treatment for paragangliomas, although an occasional anecdotal report in the literature indicates some benefit [5]. However, even with advances in diagnostic and surgical techniques, surgery still presents a major threat of injury to the cranial nerves. Nevertheless, it remains the preferred method of treatment for these tumours. Our case shows that such masses can be removed successfully and that, with care, the cranial nerves and the carotid arteries can be preserved.

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