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Institutional report - Carotid and imaging

Management for carotid body paragangliomas☆

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Abstract

The carotid body tumor is a rare neoplasm that has generated much literature over the last century, and for which continued controversy exists regarding natural history, biologic behavior, proper technique of excision, and the risk of morbidity and mortality. The present study reviewed a 16-year experience of managing carotid body paraganglioma (CBP) between 1988 and 2004. There were 10 consecutive patients aged between 18–42 years with tumors and median follow-up was 10 years. Preoperative information was derived from spiral CT scanning, magnetic resonance imaging (MRI), color Doppler imaging (CDI), and four-vessel digital subtraction arteriography. In five patients the tumor excision was attempted before they were referred to our tertiary care hospital. Two patients had bilateral tumors. Four patients had preoperative embolization, and blood loss was minimal, and excision was relatively easier in them. There was difficulty in deglutition (nasal and laryngeal regurgitation) in three patients with large tumors and who required nasogastric tube feeding (1 to 3 weeks). Surgical planning and prediction of peri-operative complications can be obtained by digital subtraction angiography, spiral CT angiography and color Doppler imaging. The peri-operative blood loss can be reduced by preoperative embolization.

Keywords: Carotid body tumor; Paraganglioma; Embolization; Radical excision

1. Introduction

The carotid body tumor is the most common paraganglioma in the head and neck, and the most frequent combination of multiple tumors is bilateral carotid body tumors [1]. The overall incidence of multiple tumors is about 10%, and constituting less than 0.5% of all the body tumors [1,2]. The sporadic form of carotid body paraganglioma is more common than the inherited variety and tends to occur slightly more often in women [3]. It is seen more frequently in people living at high altitudes and is multi centric in approximately 10% of cases with bilateral carotid body lesions being the most common combination. Malignancy occurs in 6–12.5% of cases, which ranks carotid body paragangliomas as the most frequently occurring malignant head and neck paraganglioma [4,5].

2. Patients and methods

Twenty-four patients with carotid body tumors were seen in the vascular surgery outpatient department during the past 10 years and only 10 patients out of them were admitted for excision treatment. Six patients were men and four women (Fig. 1). They were aged between 18–42 years. These patients were referred to us with a diagnosis of carotid body tumor after an initial biopsy, duplex scan, CT scan or partial excision of carotid body tumor. In two patients there was infection at the site of the previous biopsy and they required antibiotic therapy for one week prior to the major surgical excision therapy. The small tumors (<4 cm) were asymptomatic. Tumors larger than 4 cm, sometimes produce difficulty in deglutition. Three out of four of our patients with a tumor size of more than 4 cm, presented with difficulty in deglutition. None of the patients had symptoms suggestive of catecholamine production such as hypertension, blushing or palpitations. Some patients were referred after aspiration cytology (3) from the tumor. In two patients (out of 10) there were small carotid body tumors on the contra lateral side. Hoarseness of voice was noted in two patients preoperatively who had partial excision of the tumor (M1, F1).

2.1. Diagnosis of the tumors

Diagnosis of the carotid body in the patients referred to us was not difficult as there are classical clinical features (site, size, and consistency) and non-invasive tests such as duplex scan or CT scanning (Fig. 2). In all these patients a preoperative angiogram was done to see the vascularity of the tumor. Tumor blush and splaying the carotid bifurcation is classical in these patients. In the recent cases we have done preoperative embolization to reduce the intraoperative bleeding to facilitate dissection. We still feel that the angiogram is a better investigation procedure in these patients though the other investigations such as spiral CT angiogram and MR angiogram are able to give good information because during angiography there is an additional benefit of considering the preoperative embolization in

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Fig. 1. Patient with left carotid body tumor.

these patients. In four patients we have given preoperative embolization. We are not routinely performing screening for the catacholamines secretion (estimation of urinary metanephrines or VMA). In one patient (elderly with mild hypertension) we checked for the VMA but they were within normal limits. MRI with gad (tumors as small as 5 mm) and

Fig. 2. CT scan showing left carotid body tumor.

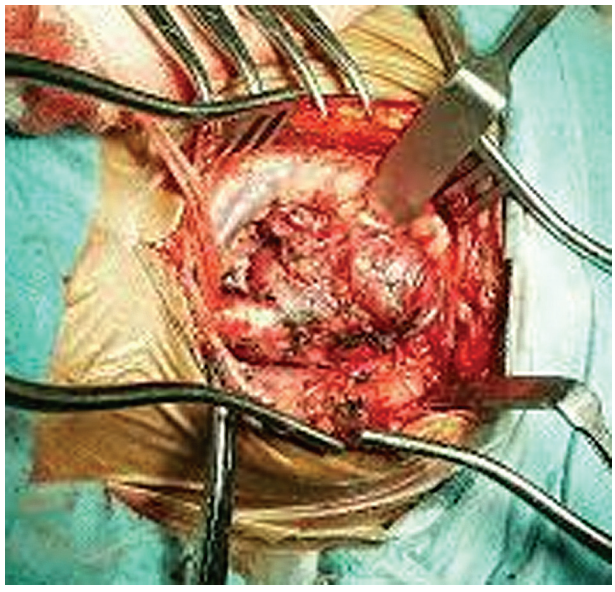

Fig. 3. Operative picture of carotid body tumor engulfing carotid vessels.

contrast CT are also effective imaging modalities in this area and are non-invasive especially in diagnosing the bilateral lesions. If the carotid angiogram is not on both the sides there is a chance that one may miss the CBP on the contra lateral side. Biopsy, including fine needle aspiration is unnecessary, dangerous, and contra-indicated in the evaluation of paragangliomas.

2.2. Surgical treatment

The main stay of treatment for carotid body tumors is surgical excision (Figs. 3, 4). Our primary aim was to excise the tumors completely without neuromuscular compromise. Neck exploration was done through an incision along the anterior border of the sternocleidomastoid muscle. There was difficulty in planning the incision when earlier transverse incisions were made in the neck with partial excision of tumor. Special care was taken to avoid injury to the

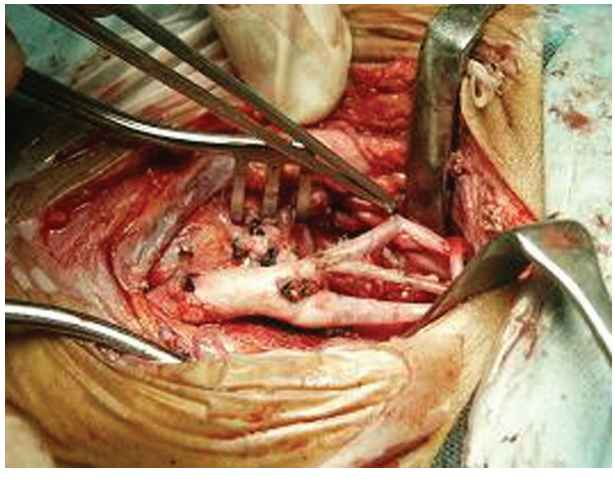

Fig. 4. Operative picture showing excision of carotid body tumor, sparing carotid vessels.

cervical branch of the facial nerve, Hypoglossal nerve, Vagus nerve. It was difficult to identify all the branches of the Glossopharyngeal nerve. The dissection plane between the tumor and internal jugular vein was helpful laterally in mobilizing the tumor. The tumor tissue around the common carotid artery is carefully dissected to get plane. Dissection of plane between the tumor and the carotid artery is done by magnifying loops. Dissection of the plane between the tumor and the carotid bifurcation is considered crucial as there are always short neck (wide mouth) branches from the carotid which will be entering the tumor immediately. Except in patient (Shamblin III), but in all the other patients, the internal carotid artery above the tumor, was dissected and taped safely. In this patient the tumor was excised with the carotid artery (intra luminal shunt) and later the carotid artery was reconstructed with a vein graft. In two patients the external carotid artery was ligated close to the carotid bifurcation during the excision to reduce the bleeding from the tumor. All of the excised specimen was sent for histopathological examination (Figs. 5, 6).

A drain was placed and the wound was closed after securing the hemostasis in all the patients. Sternocleidomastoid muscle cover was provided to the carotid artery in all the patients to prevent secondary hemorrhage from the carotid artery. Postoperative recovery wound drain persisted for 2–6 days depending on the size of the tumor and the extent of dissection.

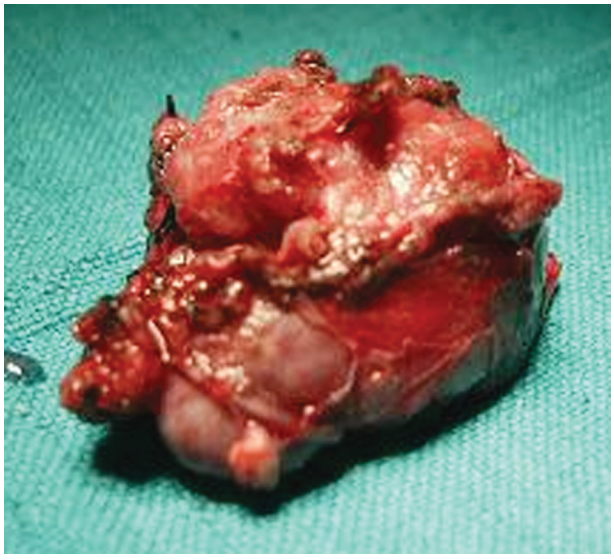

Fig. 5. Operative specimen of carotid body tumor.

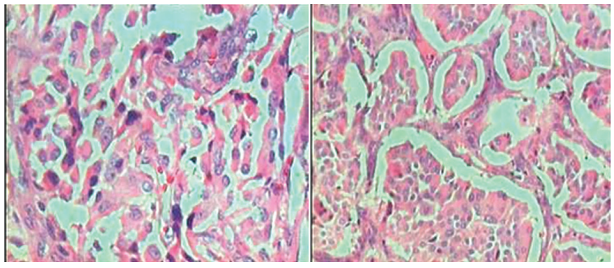

Fig. 6. Histopathological specimen of carotid body tumor.

3. Results

Only 10 out of 24 patients came forward for surgical excision. The remaining 14 patient were lost to follow up, either they were asymptomatic or it was due to financial constraint. The tumor was completely excised in all the ten patients. The operative time was 4–8 h. Intraoperatively on exploration of the neck, nine out of ten were intimately associated and compressing carotid vessels (Shamblin II), and the remaining was involving the carotid vessels (Shamblin III). In nine cases of Shamblin II carotid body paragangliomas, a subadventitial tumor excision was performed. Two patients in this group underwent ligation of the external carotid artery as this decreases the tumor vascularity and local bleeding, thus facilitating complete removal and tumor dissection away from the internal carotid artery. In patients with Shamblin III, part of the internal carotid was also excised along with the tumor, and then repaired with interposition vein graft group. There was difficulty in swallowing in the early postoperative period due to pain. In three out of four patients with large tumors there was nasal and laryngeal regurgitation in the early postoperative period. These patients were fed with a nasogastric tube (1–3 weeks) until they recovered their throat reflexes. In four patients there was temporary tongue deviation for 24 to 48 h due to stretching of the hypoglossal nerve during surgery. We did not take any special precautions regarding the catecholamine excess as none of the tumors in our study were secreting catacholamines. Radiotherapy was not considered in our patients as we thought the excision was complete. The histopathological examination showed no signs of malignancy in any of the tumors. The patients were followed up from 6 months to 16 years and no local, regional or distant metastasis was noticed. There was no early or late mortality related to the carotid body tumor surgery.

4. Discussion

The treatment of choice for paragangliomas is surgical excision [6–9]. Because these are close to the important vessels and nerves there is risk of morbidity and mortality (3–9%) [10]. The risk seems to be significant when the tumor size is more than 5 cm (67% in >5 cm and 15% when <5 cm). Shamblin developed a classification system for the carotid body tumors in 1971 [11]. Group 1 tumors are those which can be easily dissected from the adjacent vessels, group II tumors include those which are moderate in size adherent but separable from the adjacent vessels with careful dissection, and grade III tumors are usually large and engulf the carotid vessels necessitating partial or complete resection and replacement of the carotid vessels. A complete preoperative evaluation is necessary for safe resection of the tumor and reconstruction of the artery [12]. Preoperative embolization is going to reduce blood supply to the tumor but it may elicit inflammatory response in the tumor and some feel that it can make the subadventitial plane dissection more difficult [13]. Wide exposure is

helpful in achieving the adequate hemostasis. Radiotherapy, either alone or in conjunction with surgery, is a second consideration and an area of some controversy [14].

Historically, paragangliomas were considered radioresistant. Some authors believe this is false and is based on past experience where only large, recurrent or inoperable tumors were treated with this modality. Several more recent studies indicate good responses to super-voltage radiation including some complete responses [3,15]. They report only minimal acute complications (skin changes) and no long-term complications. However, other studies have demonstrated persistence of disease in lesions whose growth was stabilized by radiotherapy. Most authors still recommend radiotherapy only for very large tumors, recurrent tumors or for those patients who are poor surgical candidates. As it is a very slow growing tumor with a growth rate of less than 0.5 cm per year, small tumors in old patients, with significant risk factors for surgical interventions, can be managed by observation only [14].

5. Conclusion

Carotid body paraganglioma is a rare neoplasm. Its special anatomical position imposes great difficulty during surgery. Surgical planning and prediction of peri-operative complications can be obtained by digital subtraction angiography, spiral CT angiography and color Doppler imaging. Arteriography, the gold standard for diagnosing CBPs, demonstrates a pathognomonic tumor blush as well as the feeding vessels of the tumor; and is an excellent screening tool for concomitant paragangliomas. Adequate preoperative preparation and embolization of feeding arteries could reduce operative blood loss, improve tumor excision and preserve the internal carotid artery flow.

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