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ORIGINAL ARTICLE

The Outcome of Multidisciplinary Management of Carotid Body Tumors: Retrospective Cohort Study

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Abstract
Background Carotid body tumor (CBT) is a rare paraganglionic hyper-vascular tumor of the carotid body. The standard treatment for CBTs is surgery, but it involves risk. The study is aimed to assess the CBTs and evaluate the outcome of multidisciplinary management.
Materials and Methods A retrospective cohort study included patients with CBTs who were managed by surgical excision between May 2006 and April 2018. A multidisciplinary team was established to excise the tumor completely with minimal neurovascular compromise.
Results The study comprised of 32 patients in the age group of 23–65 years. The main presentation was a unilateral painless neck mass. Six cases (18.75%) were Shamblin I, 10 (31.25%) Shamblin II and 16 (50%) Shamblin III. Complete excision was performed for all cases. Ligation of the external carotid artery was done in 15 cases (46.88%) and repair of the internal carotid artery in 6 (18.75%). Postoperative cranial nerve complications occurred in six patients (18.75%): four transient hypoglossal pareses and two hoarseness of voice. Histopathologically, one case (3.125%) was malignant. No recurrence was detected through the follow-up period.
Conclusion A multidisciplinary approach is essential for management of CBTs. Early diagnosis and surgical resection minimize morbidity and carry good surgical outcome.
Trial Registration Number ChiCTR1800018722 (*Agency*: Chinese Clinical Trial Registry).

Keywords Carotid body tumors · Shamblin classification · Excision · Ligation of external carotid artery

Introduction

Carotid body tumor (CBT) is a rare paraganglionic hyper-vascular tumor of the carotid body. The incidence of paraganglionic tumors in head and neck is about 1/30,000, among which CBT accounts for 65% [1]. It is often unilateral. Bilateral disease occurs with a family history [2]. The sporadic form of CBT is common than the inherited variety and tends to occur slightly more often in women. It is seen more frequently in people living at high altitudes and is multicentric in approximately among 10% of cases [3]. Malignancy occurs in 6–12.5% of cases, which ranks CBT as the most frequently occurring malignant head and neck paraganglioma [4].

The main presentation of CBT is a slowly growing painless mass in the neck, which may enlarge and compress the surrounding structures and invade the skull base, the cranial nerves and the sympathetic chain with correlated symptoms, like dysphagia, shocking, hoarseness, tongue muscle atrophy and Horner's syndrome [5].

Due to the proximity to the blood vessels and nerves, CBT is mainly diagnosed by radiographic examination,

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fine-needle biopsy is contraindicated, and it may cause uncontrolled bleeding and false aneurysms. The characteristic manifestation of CBT in digital subtraction angiography (DSA) is the displacement of internal carotid artery (ICA) and external carotid artery (ECA), and the goblet-like or "Lyre sign." DSA can also be used for preoperative assessment of the extent of tumor involvement in the blood vessels; through the temporary balloon occlusion test to assess the collateral circulation of the brain, predicting the need to ligation of ECA or sacrifice and reconstruction of the ICA can also help preoperative embolization and stent implantation [2].

Shamblin [9] classified the CBT into three types according to the degree of the tumor involving the carotid artery. In recent years, it has been suggested that Shamblin classification cannot fully assess the complications of the nervous system and the difficulty of operation, so Luna-Ortiz [13] proposed the improved Shamblin classification. Ma et al. [14] used the texture of the tumor to classify the tumor and could better predict the ease of complete resection.

The standard treatment for CBTs is surgery. The key to the surgical treatment is to deal with the relationship between the vessels and nerves surrounding the tumor. Gordon-Taylor [6] described a safe, sub-adventitial dissection in 1940. Since the 1980s, devascularization of CBTs by ligation of feeding branches of the ECA during operation has been a common practical technique [7]. Later, a craniocaudal dissection technique was proposed to minimize postoperative morbidity [8].

The aim of this work was to study the CBTs in our institution and to evaluate the outcome of their surgical treatment by a multidisciplinary approach.

Patient and Methods

Study Design, Study Setting, Study Population

A retrospective cohort study included patients with CBTs that had been presented to the Department of Surgery in the period from May 2006 to April 2018 and treated by surgical excision.

Ethical Clearance

This study was approved by the Institutional Review Board and Ethics Committee of our institution and was conducted in accordance with the principles of the 1975 Declaration of Helsinki. It was registered at the Chinese Clinical Trial Registry (ChiCTR180001872). All patients provided an informed consent.

Inclusion Criteria

Patients diagnosed with CBTs and who were treated by surgical excision voluntarily agreed to participate in the study.

Exclusion Criteria

Exclusion criteria are the recurrent cases, cases that refused surgery and/or received radiotherapy as a primary treatment, and patients who were not willing to participate in the study and unable to give informed consent.

Data Collection

The collected data included demographic details, radiologic findings, surgical details and outcome during the follow-up period.

Methodology

All patients were subjected to complete clinical evaluation, duplex ultrasonography, CT angiography and digital subtraction angiography (DSA) (Fig. 1a–e). All cases were diagnosed clinically and radiologically. We avoided the need for preoperative biopsy. Shamblin classification [9] was used to group our patients into three categories:

Type I: The tumor is small and easy to separate from blood vessel, and the blood vessels are not injured.

Type II: The tumor surrounds the blood vessels and needs careful stripping, the tumor can be resected completely, and the blood vessels are retained. A small number of patients need to establish bypass and vascular reconstruction.

Type III: The tumor is very large and encases the carotid artery and its bifurcation, there are obvious symptoms and signs, and surgical resection of the blood vessels at the same time is required.

Complete surgical excision was done for all cases. Clinical diagnosis was confirmed by postoperative histopathology. Postoperative follow-up was done clinically and by duplex scan every 3 months for the first year and each 6 months later to evaluate the outcome and detect recurrence.

Surgical Procedure

Our goal was to excise the tumor completely with minimal neurovascular compromise. A multidisciplinary team composed of maxillofacial surgeons, vascular surgeon and neurosurgeon was established to achieve that goal. The surgical procedures are briefly described as follows: The patient is under general anesthesia with endotracheal

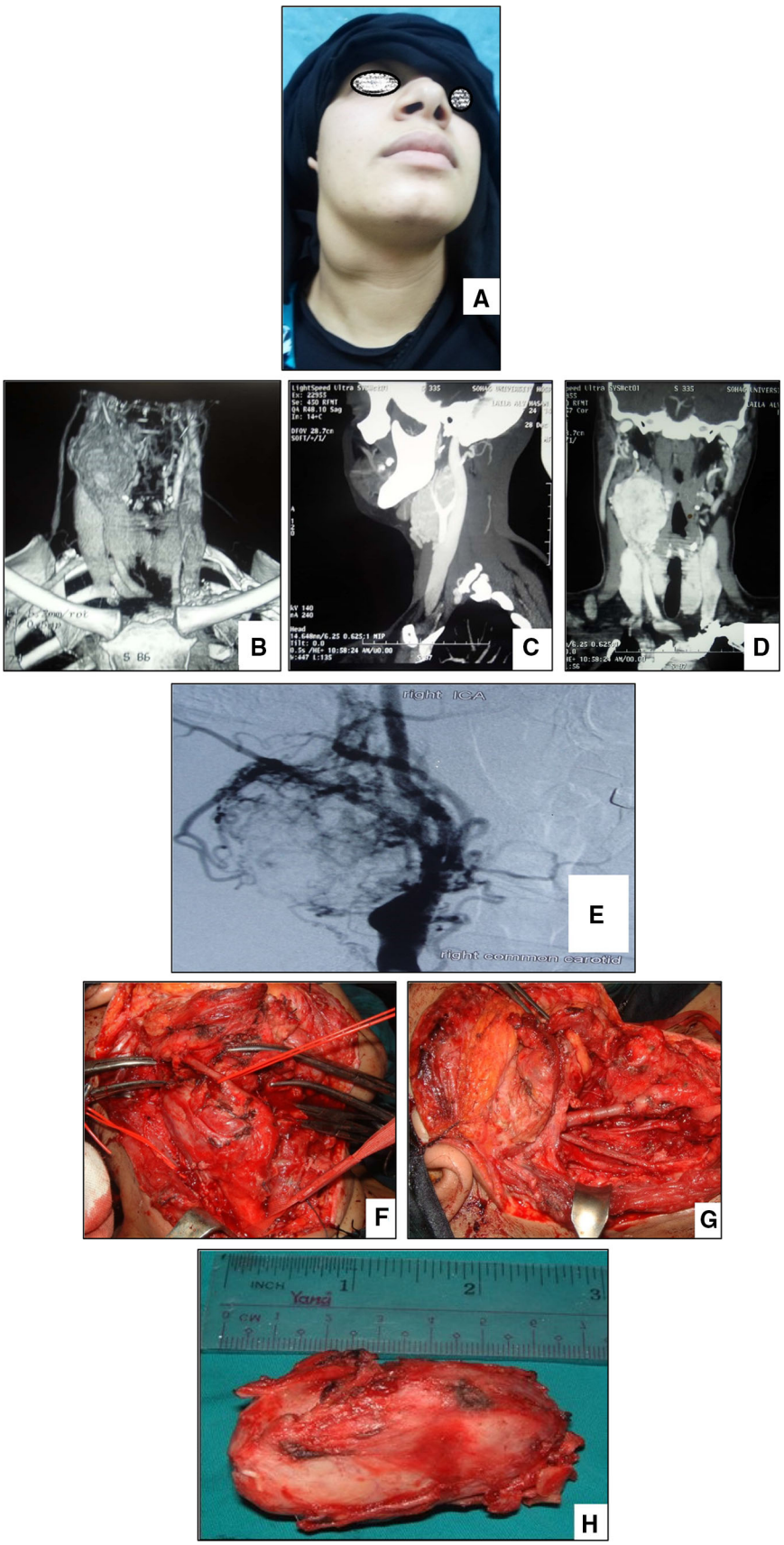

Fig. 1 25-year-old female patient presented with right side CBT and treated by surgical excision: **a** preoperative photograph showing right side CBT, **b** three-dimensional view of CT angiography showing the extent of CBT in the neck, **c** sagittal view of CT angiography showing the relations of CBT to the neck vessels, **d** coronal view of CT angiography showing the relations of CBT to the neck vessels, **e** DSA of the right carotid artery showing a huge hyper-vascular CBT, **f** intraoperative photograph showing the tumor mass and controlling the ECA and the ICA, **g** intraoperative photograph after complete resection of the CBT and primary repair of the ICA, **h** excised CBT measuring about 7 cm, Shamblin III

intubation. Wide transverse cervical incision was achieved to provide good exposure of the tumor, and the carotid vessels were controlled proximally and distally. Careful dissection is done to preserve the adjacent cranial nerves (vagus, hypoglossal, superior laryngeal and mandibular branch of the facial nerve). Certain precautions were taken to reduce the bleeding before tumor resection as using gentle blunt dissection in the adventitial plane, ligation of the feeding vessels supplying the tumor, ligation of ECA and use of electrocautery. Any undesirable injuries of the vessel wall are immediately managed by vascular surgeon via primary vascular repair or vascular reconstruction (Fig. 1f–h). After complete hemostasis, wound was closed with suction drain application.

Statistical Analyses

The data for each case were recorded in patient's file. Data were collected in the Microsoft Excel spreadsheet, processed using the Statistical Package for the Social Sciences version 20 (Inc. Chicago, Illinois, USA) and analyzed.

Results

Demographic and Clinical Characteristics

A total of 32 patients were included in the study period and there were 20 (62.5%) females and 12 (37.5%) males and their age ranged from 23 to 65 years (mean 42 ± 13). All patients presented with unilateral painless lump in the neck.

In 14 cases (43.75%), the CBT was in the right side and in 18 cases (56.25%) it is in the left side. None of the patients had a functioning CBT. According to Shamblin classification, it was found that Shamblin group I included six (18.75%) patients, group II included 10 (31.25%) cases, and group III included 16 (50%) patients.

Intraoperative Data

Complete surgical excision of the tumor was performed in all cases. In sixteen patients (50%) with Shamblin group I and II, sub-adventitial tumor excision was performed after ligation of the feeding vessels of the tumor and six (18.75%) of them were in need of ligation of the ECA and primary vascular repair of the ICA was needed in two patients (6.25%). In the remaining 16 (50%) patients with Shamblin group III, ECA ligation was done in nine (28.13%) patients and primary vascular repair of the ICA was needed in four (12.5%) patients.

Postoperative Complications

Postoperative hemiparesis occurred in five (15.63%) patients with Shamblin III: Four of them improved in 3 months after surgery, while the other one was permanently affected. Postoperative cranial nerve complications occurred in six patients (18.75%): Four of them with transient hypoglossal paresis and the remaining two developed hoarseness of voice due to permanent vocal cord paralysis (Table 1).

Histopathological Diagnosis

All postoperative specimens were diagnosed histopathologically to be benign paraganglioma except one which was malignant CBT. This patient was referred to oncology department for adjuvant postoperative radiotherapy.

Follow-Up

All patients were followed up from 6 to 72 months (mean 22 ± 12) with no local recurrence.

Discussion

CBTs in the head and neck represent a dilemma and their treatment is challenging. Most CBTs are benign and slowly growing, and surgical excision is the treatment of choice when the patient is young and fit.

In accordance with other studies [3, 9], female predominance was noticed in our study where 20 cases (62.5%) were females and 12 cases (37.5%) were males. It is believed that the occurrence of CBT is related to genetic susceptibility and estrogen is considered to be another important factor that can accelerate the formation of CBT [9].

In our study, we depended on the clinical and radiologic diagnosis. No preoperative open biopsy or even percutaneous needle biopsy was done as they may cause intractable bleeding, pseudoaneurysm formation and carotid artery thrombosis [10].

Color Doppler ultrasonography is simple, reproducible and noninvasive, and can be used as the preferred noninvasive method for the diagnosis of CBT. However, due to mandibular obstruction and poor development of small blood vessels, the application range is limited. DSA is still considered the gold standard for preoperative diagnosis of CBT [11]. It was reported that CT angiography can replace DSA because it can provide accurate angiography and show the size, shape and growth mode of the tumor and the relationship with the adjacent tissue structures, especially when there is tumor invasion of skull base where it can

Table 1 Clinical characteristics, operative data and postoperative complications

Shamblin classification	Frequency (%)	Vascular injury		Postoperative complications		
		Ligation of ECA	Repair of ICA	Cerebral ischemia	Hypoglossal nerve paresis	Hoarseness of voice
I	6 (18.75%)	–	–	–	–	–
II	10 (31.25%)	6 (18.75%)	2 (6.25%)	–	2 (6.25%)	–
III	16 (50%)	9 (28.13%)	4 (12.5%)	5 (15.63%)	2 (6.25%)	2 (6.25%)
Total no.	32 (100%)	15 (46.88%)	6 (18.75%)	5 (15.63%)	4 (12.5%)	2 (6.25%)

show bone destruction [12]. In our study, we found that CT angiography and DSA are complementary to each other for the diagnosis of CBTs.

All cases in our study were unilateral tumors that were treated by surgical excision. We think that surgical resection is still the preferred treatment because CBT is slowly growing and not sensitive to radiotherapy and chemotherapy. Some studies reported that the unilateral CBT regardless of the size of the tumor should be surgically treated. The operation can not only remove the tumor and release the tumor compression to the surrounding tissues, but also provide tissue specimen for pathological diagnosis [2].

The literature reported that CBTs are mostly benign, while malignancy was found in 2–8% [9]. In our study, postoperative histopathological examination revealed that 31 cases (96.88%) were benign and only one case (3.13%) was malignant. This patient was referred to oncology department for adjuvant postoperative radiotherapy. Some authors have found that radiotherapy is effective in inhibiting further growth of CBTs, However, it is often considered to be an alternative treatment modality for patients who cannot undergo surgery due to extensive involvement [13].

In our study, the main purpose of surgery was to remove all neoplastic tissue, saving the nearby important neurovascular structures. Surgical resection of CBTs was performed in our cases with special maneuvers to facilitate and make the surgical procedure safer via wide transverse cervical incision, dissection and controlling the carotid vessels proximally and distally, and then sub-adventitial dissection of the carotid vessels and early ligation of the feeding vessels.

For intraoperative sharp separation of tumors and blood vessels, bipolar electric knife is used which can reduce blood vessel injury and intraoperative bleeding, and research has shown that blunt separation can lead to the occurrence of false aneurysm. For severe intraoperative bleeding, blood transfusion can be considered [2, 14].

The CBT is closely related to the vessels and nerves, which makes the operation more difficult and has a high complication rate due to the particularity of its anatomical structure. Bleeding is a common intraoperative complication, and the amount of bleeding is related to the size of the tumor [2] and the distance from the skull base [14]. Although it was reported that for large or high tumor, preoperative embolization of external carotid artery branches and tumor-supplying vessels can reduce intraoperative bleeding and reduce the proportion of carotid reconstruction [15]. In our study, the preoperative embolization of the external carotid artery branches or the feeding vessels was not performed because this may induce embolic reverse flow embolization of the brain or eye vessels and other risks.

As CBTs are close to important vessels and nerves in the neck, there is a risk of morbidity and mortality, which is variable. Incidence of ICA injury has been reported to be 2% to 23% with or without reconstruction. The sacrifice of ECA and ICA has been reported to be needed from 13–39 and 11%, respectively [7, 16]. In our study, ligation of the ECA was done in 15 patients (46.88%), while primary vascular repair of ICA was done in six patients (18.75%). It was noticed that the incidence of vascular complications in our study was higher than the previous studies because this study included cases with more advanced tumors.

Ischemic stroke is the most serious complication after CBT resection. It can occur with or without manipulation of major neck vessels, and its incidence has been reported to be 0–11% [16, 17]. In our study, five (15.63%) patients developed postoperative cerebral ischemia: Four (12.5%) of them were transient and improved by conservative measures, while permanent left sided hemiparesis persisted in one patient (3.13%).

It was reported that the occurrence of ischemic stroke is more with cases of preoperative carotid artery compression symptoms and Shamblin grade II, III type, the internal carotid artery occlusion time is too long, the internal carotid artery reconstruction, the side carotid artery asymptomatic plaque and the intraoperative or postoperative systolic pressure difference [18]. Therefore, preoperative need to fully assess the situation of circle of Willis, accurate measurement of brain compensation for blood supply,

Matas pressure neck test [19] training, meticulous surgery to minimize vascular injury, shortening the carotid artery occlusion time, low-temperature, controlled-pressure (systolic pressure control around 130 mmHg) anesthesia, perioperative attention to maintain the stability of hemodynamics and appropriate use of anticoagulant drugs should be considered [12].

Postoperative neurological complications as cranial nerve deficits can occur, and the incidence of postoperative affection ranges from 6.9 to 42%. The hypoglossal and vagus nerve appeared to be the most vulnerable to injury from the sacrifice or retraction [17]. In our study, four cases (12.5%) showed temporary tongue deviation that may be due to retraction of the hypoglossal nerve and both cases improved spontaneously. Another two patients (6.25%) developed permanent postoperative hoarseness due to vocal cord paralysis. These findings are parallel to the results of other studies [8, 17, 20, 21]. Postoperative mortality was not encountered in our cases. However, it is not unusual hazard after surgery especially in Shamblin III [7].

In this study, it has been noticed that all patients with permanent postoperative neurovascular complications and most of patients who had carotid artery reconstruction were referred to Shamblin group III. These results also support the reports that the Shamblin classification can be used to predict the possible postoperative neurovascular complications [12, 20, 22].

There is a study to show that high risk factors for cranial nerve injury are the deficient preoperative evaluation and Shamblin II, III type. Therefore, early diagnosis, adequate evaluation and early resection are the primary measures to prevent cranial nerve injury, and full exposure of the surgical field is the key to reduce cranial nerve injury. Also, dexamethasone and mannitol can be used postoperatively to reduce the edema of nerves and brain after cerebral reperfusion [23].

Conclusions

Patients with CBT in our locality are usually presented in an advanced stage. Early diagnosis and surgical resection of these tumors will minimize the morbidity and carry good surgical outcome. A team approach including maxillofacial surgery, vascular surgery and neurosurgery is essential to develop reasonable treatment plan, achieve good surgical results and reduce the occurrence of intraoperative and postoperative complications.

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Compliance with Ethical Standards

Conflict of interest The authors report no conflicts of interest associated with this work.

Ethical approval This study was approved by the Institutional Review Board and Ethics Committee of our institution (Permit Number: 21/2018) and was conducted in accordance with the principles of the 1975 Declaration of Helsinki. All patients provided informed consent.

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