# Carotid body tumor surgery: challenges and management

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**Purpose**
To assess the challenges and management of carotid body tumors (CBTs). Adequate planning by the surgeon can greatly reduce adverse events of such rare and difficult tumors.

**Background**
CBTs are rare. Despite over a century of describing surgery, controversy remains surrounding details of their treatment. Most complications are considered technically preventable.

**Patients and methods**
This prospective study included 13 cases diagnosed as CBT. Patients were subjected to clinical evaluation, carotid duplex ultrasound, computed tomography angiography scan, and sometimes MR angiography. All patients underwent CBT resection and when needed vascular reconstruction. Patients were classified into three groups depending on Shamblin classification.

**Results**
Mean operative time was longer in patients of Shamblin class 3 ($5.4\pm0.77$ h; range, 4.5–6 h; $P<0.001$). Regarding vascular reconstruction, all patients of Shamblin class 3 were reconstructed by interposition vein graft and two patients of Shamblin class 1 were reconstructed through primary repair, but patients of Shamblin class 2 were reconstructed by all means. In postoperative follow-up complications, bleeding was more significantly in Shamblin class 3 ($P=0.032$). Cranial nerve deficit was observed in four (33.3%) patients, and all of them were transient. The mean size of excised tumor was $5.17\pm1.4$ cm, with range of 3–7.5 cm.

**Conclusion**
Early surgical excision by a vascular surgeon is the only proven cure for CBTs, because they are small and easy to remove. However, if neglected, there will be many challenges, as most tumors can become locally invasive. Meticulous periadventitial (and sometimes subadventitial) dissection can greatly reduce the rates of complications.

**Keywords:**
carotid body tumors, challenges, management, technique

## Introduction

The carotid body is the largest chemoreceptor tissue mass located within the periadventitia of posterior surface of carotid bifurcation. Normal carotid body is ovoid in shape and ~5 mm in its longest dimension. It usually receives arterial supply from external carotid artery and derives sensory innervation from glossopharyngeal nerve [1,2].

Carotid body tumors (CBTs) have many names, such as chemodectomas, paragangliomas, and glomus tumors. They represent neoplasia of chemoreceptive tissue. These tumors belong to paragangliomas, which occur along autonomic ganglion chain. In head and neck, CBTs are the most common paragangliomas [3].

Macroscopically, they are reddish brown and rubbery. Although they lack a true capsule, they are highly vascular and can invade adventitia of carotid vessels. As the tumors enlarge, they splay carotid artery branches, giving 'lyre sign' on angiography [4].

Most carotid chemodectomas are benign slowly growing tumors. Patients usually present with pulsatile neck masses. The exact incidence of malignant transformation is unknown. Malignancy here is based mainly on clinical behavior more than histology. Even benign tumors are capable of aggressive invasion into adjacent structures. If carotid body cells are found in lymph nodes biopsied during surgery, this is diagnostic for malignancy. Some cases have been reported with hypertension, resolved after tumor resection, which may be caused by tumor-related catecholamine release [1,5–7].

Conventional angiography was traditionally the mainstay and provides excellent details especially about vascularity of the lesion. Duplex ultrasound

has been considered the most important, noninvasive method to examine carotid lesions with no discomfort or risk. Recently, computed tomography angiography and MR angiography (MRA) have been used instead of conventional angiography. These methods provide high levels of anatomic and vascular details [1,8,9].

Shamblin and colleagues at Mayo Clinic developed a classification system for carotid chemodectomas that depends on tumor extent and neurovascular involvement. Group I tumors are smaller, so can be dissected easily. Group II tumors are larger, adherent to adventitia and partially surround vessels. Group III tumors are more adherent and encase internal and external carotid arteries, so dissection is difficult in the periadventitial plane [10,11].

Surgical resection is the gold standard treatment for carotid chemodectomas. Although these lesions tend to be small, they are best treated early. The first successful carotid chemodectoma resection was done by Scudder in the United States. He was able to preserve carotid system, and there were no significant complications [8,12,13].

Surgeons must plan their operative approach according to the tumor characteristics, and attempts must be made to reduce blood loss and preserve vital structures as these tumors are highly vascular and carry high risk of hemorrhage during surgery. The lesions may also adhere to or envelop neurovascular structures [1,14].

Radiation therapy is another treatment modality for CBTs, but it remains controversial owing to conflicting studies and lack of long-term follow-up. There is currently no effective chemotherapy for these tumors [1,12].

## Patients and methods

After approval by the local ethical committee of Zagazig and Benha universities and Benha Insurance Hospitals and obtaining written fully informed consent from the patients, this prospective study was conducted on patients diagnosed with CBT at the Vascular Surgery Department of Zagazig and Benha universities and Benha Insurance Hospitals from August 2015 till January 2020; the enrollment period was 42 months, and the patient follow-up period was 12 months.

All patients were subjected to detailed clinical evaluation, laboratory assessment, carotid duplex ultrasound, computed tomography angiography scan, and sometimes MRA examination. All patients

underwent CBT resection followed by vascular reconstruction if needed. Patients were classified depending on Shamblin classification into three groups.

Patients included in this study were fit for general anesthesia with resectable tumor or if tumors demonstrated airway compression. However, patients excluded from this study were patients with significant comorbidities, with limited life expectancy, with contralateral cranial nerve dysfunction, or with irresectable tumor.

### Operative procedure

The procedure was performed general anesthesia. Cerebral monitoring and protection were done with internal carotid artery occlusion or reconstruction using routine shunting by Pruitt-Inahara shunt (Figs 1–3).

Patient was kept supine with head rotated to the contralateral side. Shoulder roll was placed to extend the neck, and table head was elevated 10–15° to reduce venous pressures and incisional blood loss. A longitudinal skin incision was made along anterior border of sternomastoid muscle. If needed, the incision was extended proximally toward sternal notch and distally toward mastoid process.

The dissection continued until carotid sheath appears. Then we opened this sheath along anterior border of internal jugular vein. Common facial vein typically joins internal jugular vein near the carotid bifurcation and was a useful landmark; we divided this vein. Here, laryngeal nerves were protected. Proximal common carotid artery was circumferentially mobilized in the periadventitial plane, with sufficient length for bypass shunt.

We proceeded dissection distally on the lateral aspect of carotid system until beyond the upper end of the mass where we circumferentially mobilized internal carotid artery. Here hypoglossal nerve was carefully separated and protected. Then dissection was continued down along medial aspect of internal carotid artery till bifurcation. At this point, we circumferentially mobilized the external carotid artery.

For small tumors (Shamblin group I), we conserved the external carotid artery; branches of external carotid artery supplying the tumor were divided and ligated. Once the posterior, lateral, and medial aspects of the tumor were mobilized, the superior extent was divided between the ligature and the mass removed. Primary repair was done for small tears (Fig. 1a–c).
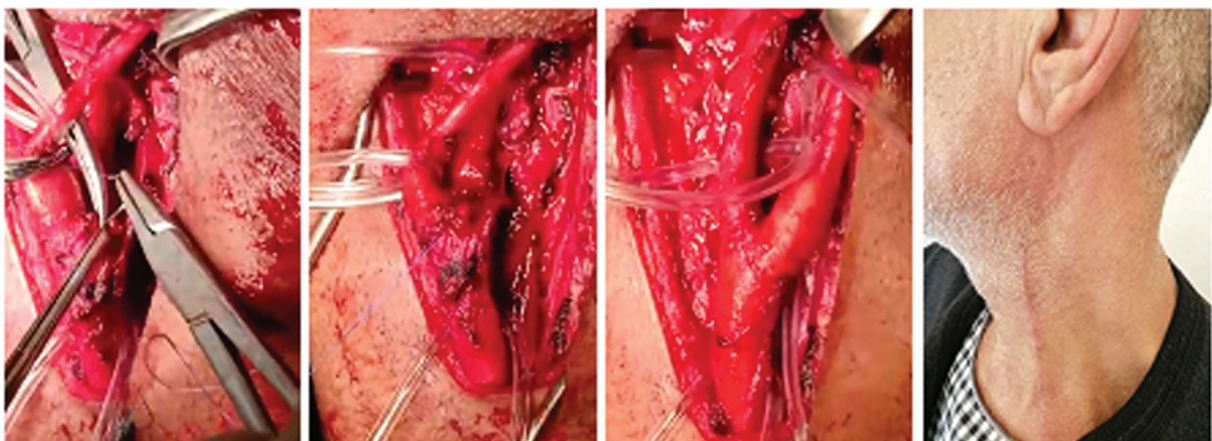
**Figure 1**
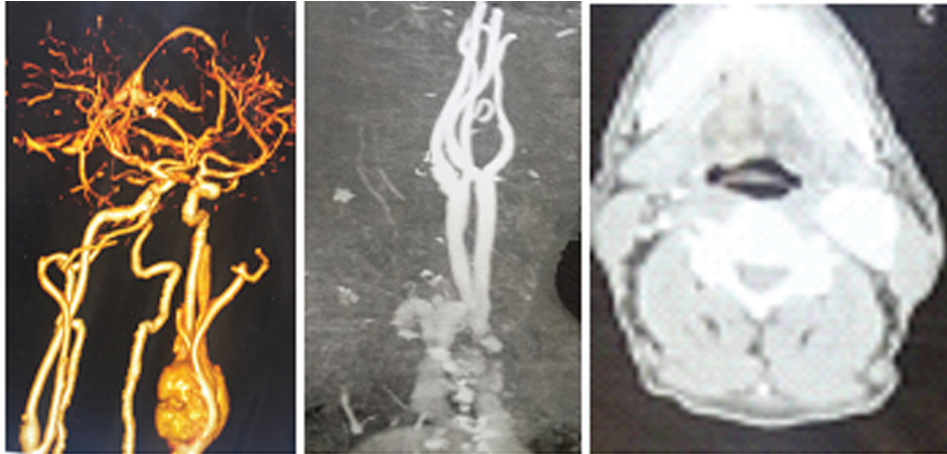


(A): CTA of Lt CBT splaying internal & external carotid arteries "lyre sign".
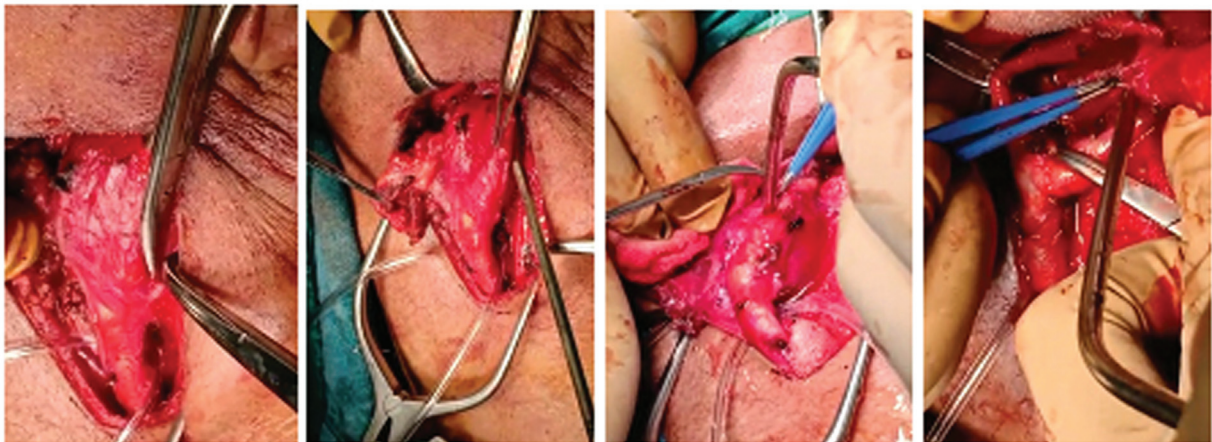
(B): Steps of tumor dissection in the periadventitial plane.

(C): Primary repair of the carotid bifurcation and incision postoperative.

Shamblin class 2 carotid body tumor with primary repair. (a) Computed tomography angiography of left CBT splaying internal and external carotid arteries 'lyre sign'. (b) Steps of tumor dissection in the periadventitial plane. (c) Primary repair of the carotid bifurcation and incision postoperative. CBT, carotid body tumor.

For larger tumors that were adherent to the external carotid artery (Shamblin group II), we tried to conserve the External Carotid Artery (ECA) first, but if tedious, it was divided at its origin, and proximal stump was oversewn to reduce blood loss and provide a handle to rotate the tumor helping dissection. Patients of Shamblin class 2 were reconstructed by all means (Figs 1a–c and 2a–c).

**Figure 2**
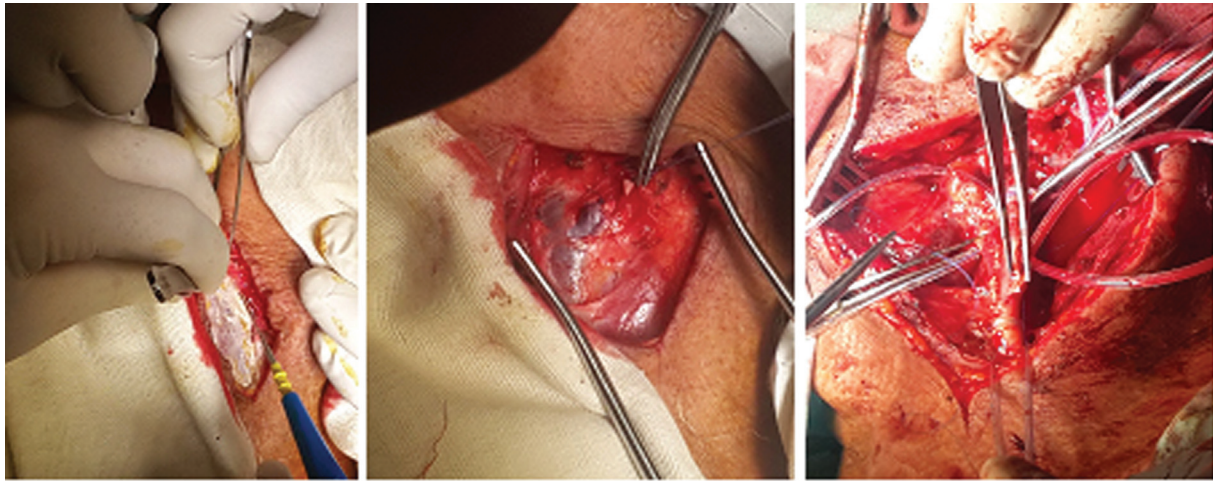


(A): Skin incision     (B): Internal Jugular Vein     (C): Patch repair over shunt.

Shamblin class 2 carotid body tumor with patch repair over shunt. (a) Skin incision. (b) Internal jugular vein. (c) Patch repair overshunt.

**Figure 3**
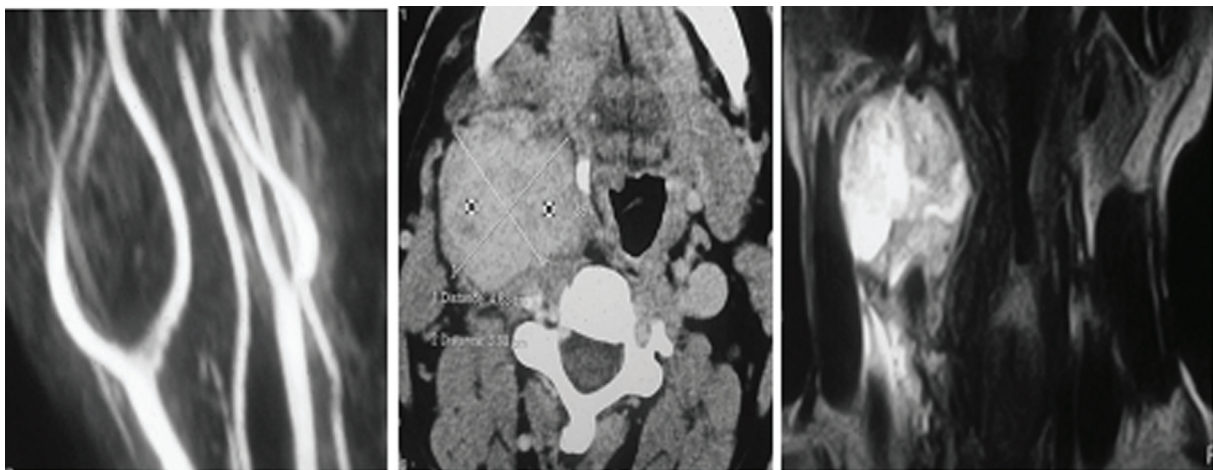


(A): MRA of Rt CBT
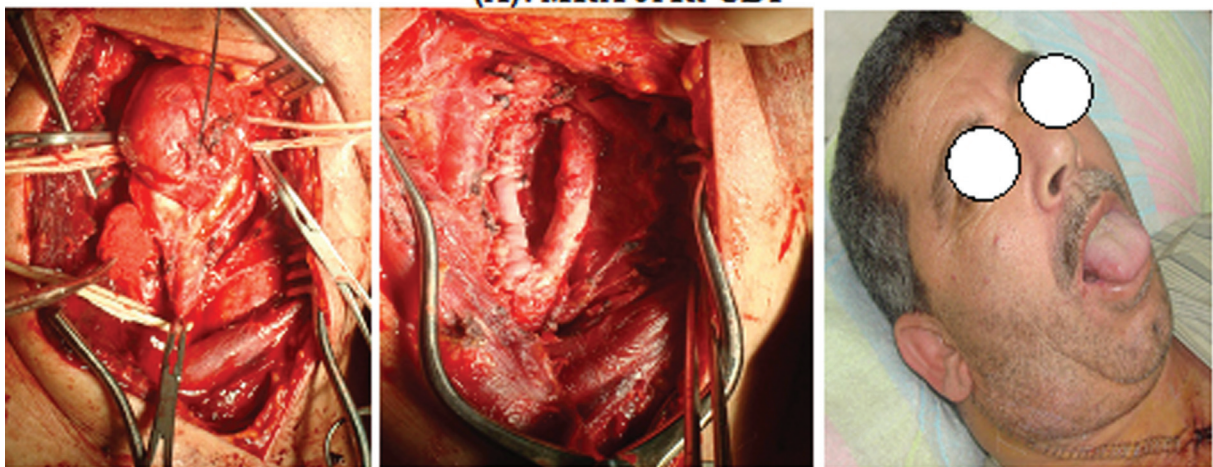
(B): Dissection     (C): Vein interposition graft     (D): Intact hypoglossal N.

Shamblin class 3 carotid body tumor with vein interposition graft. (a) MRA of right CBT. (b) Dissection. (c) Vein interposition graft. (d) Intact hypoglossal nerve. CBT, carotid body tumor; MRA, MR angiography.

For tumors that could not be dissected from the bifurcation (Shamblin group III), both internal and external carotid arteries were removed with the tumor. Vascular reconstruction for the internal carotid was performed with autologous vein interposition graft using shunt through the graft and removing the shunt before the second anastomosis was completed. In addition, if clamping of carotid arteries was performed, the patient received systemic heparin (Fig. 3a–d).

### Follow-up
A preliminary neurologic assessment was performed when patient awakens from anesthesia. Patient was brought to recovery room where blood pressure and neurologic function could be carefully monitored. Care was done for neck hematomas. Drain was removed after 24 h. Patients were followed closely for 12 months.

### Statistical analysis
The study data were tabulated and analyzed using SPSS, version 16 soft ware (SPSS Inc., Chicago, Illinois, USA). Categorical data were presented as number and percentages, and analyzed using Fisher's exact test. Continuous data were expressed as mean ±SD and range. Differences between groups were tested using analysis of variance ($F$ test) for variables or Kruskal–Wallis test for nonparametric ones ($P<0.05$ was considered significant).

## Results
No mortality related to surgery was reported. One patient missed follow-up, and this patient was excluded from the study data analysis. So at completion of the study, clinical data were available for 12 patients. Their age range was 35–69 years old.

Overall, seven (58.3%) patients were females and five (41.7%) patients were males. All patients were fit for general anesthesia depending on American Society of Anesthesiologists ($ASA_I$ and $ASA_{II}$). Patients were classified depending on Shamblin classification into three groups: seven (58.3%) patients were class 1, three (25%) patients were class 2, and two (16.7%) patients were class 3 (Table 1, Fig. 4).

Neck mass was the main presenting complaint in 11 (91.7%) patients; only one (8.3%) patient was incidental finding. Sensory changes (pulsatile tinnitus, numbness, and face or neck pain) were observed in five (41.7%) patients. Duration of symptoms was 1.25±0.46 years. Bilateral CBT was reported only in one (8.3%) patient (Table 2, Fig. 5).

All patients passed uneventful intraoperative course without complications. Mean operative time was

**Table 1 Patients' demographic data**

| Data | Findings ($N=12$) |
| --- | --- |
| Age groups (years) | |
| <40 | 2 (16.7) |
| 40–60 | 6 (50) |
| >60 | 4 (33.3) |
| Sex | |
| Female | 7 (58.3) |
| Male | 5 (41.7) |
| American Society of Anesthesiologists grade | |
| $ASA_I$ | 8 (66.7) |
| $ASA_{II}$ | 4 (33.3) |
| Patients classification depending on Shamblin classification | |
| Class 1 | 7 (58.3) |
| Class 2 | 3 (25) |
| Class 3 | 2 (16.7) |

Data are presented as $n$ (%). ASA, American Society of Anesthesiologists.

**Figure 4**
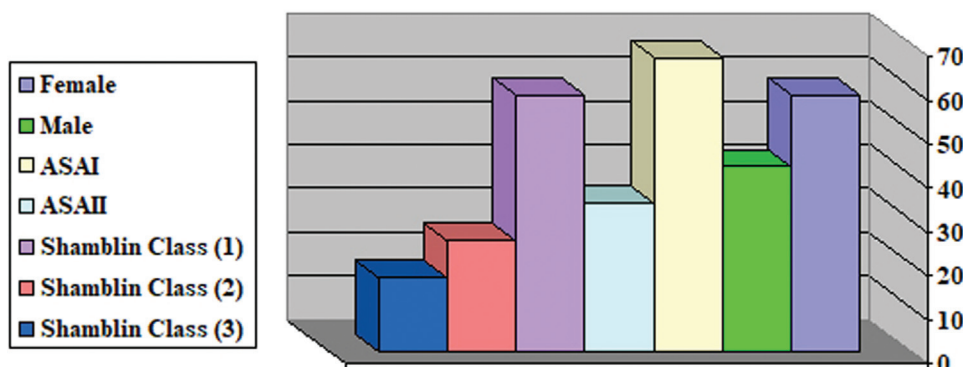


Patients' demographic data.

longer in patients of Shamblin class 3 (mean, 5.4 ±0.77 h; range, 4.5–6 h; $P<0.001$). Intraoperative blood loss was also more in patients of Shamblin class 3. Mean duration of postoperative hospital stay (in days) was longer in the patients of Shamblin class 3 (mean, 9.3±4.1; range, 6–14 days; $P=0.003$) (Table 3).

Regarding vascular reconstruction, all patients of Shamblin class 3 were reconstructed by interposition vein graft and two patients of Shamblin class 1 were repaired primarily, but three patients of Shamblin class 2 were reconstructed by all means (Table 4).

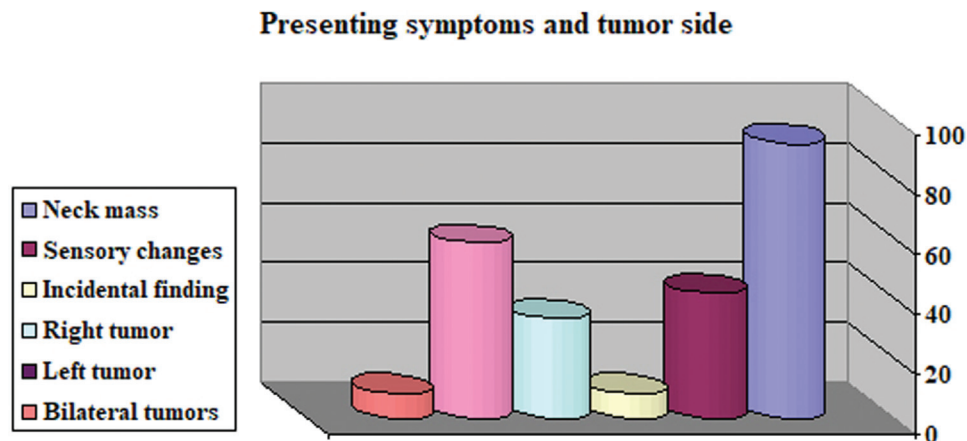The mean size of excised tumor was 5.17±1.4, with range of 3–7.5 cm. Most tumors were benign paraganglioma [10 (83.4%)], and only one (8.3%) tumor was malignant, with positive surgical margin and lymph node invasion. There was a histological surprise in one (8.3%) patient, having lymph node hyperplasia (Table 5).

Challenges in the postoperative follow-up were as follows: bleeding was observed in three (25%) patients in the form of hematoma or ecchymosis; only one (8.3%) patient needed re-exploration to ligate slipped ligature of common facial vein, and others were managed conservatively. Stroke was reported in two (16.7%) patients; both patients were transferred to neurological ICU for management. Cranial nerve deficit was observed in four (33.3%) patients; all of them were transient and treated by neurotonic drugs for up to 6 months and consisted of superior laryngeal nerve in one patient in the form of chocking, recurrent laryngeal nerve in one patient in the form of hoarseness of voice, hypoglossal nerve in one patient in the form of deviated tongue to the same side of injury, and marginal mandibular nerve in one patient in the form of wry lip.

**Table 2 Presenting symptoms and tumor side**

| Variables | Findings (N=12) | |
|---|---|---|
| | n (%) | Mean±SD (range) |
| Presenting symptoms | | |
| Neck mass | 11 (91.7) | |
| Sensory changes | 5 (41.7) | |
| Incidental finding | 1 (8.3) | |
| Duration of symptoms (years) | | 1.25±0.46 (0.5–2) |
| Treated side | | |
| Right | 4 (33.3) | |
| Left | 7 (58.3) | |
| Bilateral | 1 (8.3) | |

More than one symptom was observed in one patient.

**Figure 5**



Presenting symptoms and tumor side.

**Table 3 Operative and immediate postoperative data**

| Variables | Shamblin | | | ANOVA | P value |
|---|---|---|---|---|---|
| | 1 | 2 | 3 | | |
| Operative time (h) | 1.46±0.34 | 3.3±1.5 | 5.4±0.77 | 18.6 | <0.001 (HS) |
| | 1–2 | 2–5.6* | 4.5–6*† | | |
| Intraoperative blood loss (ml) | 290±90.1 | 680±256.5 | 963.3±464.7 | KW=10.98 | 0.004 (S) |
| | 150–400 | 350–1000* | 700–1500* | | |
| Duration of PO hospital stay (days) | 1.38±0.51 | 5.7±2.7 | 9.3±4.1 | KW=11.9 | 0.003 (S) |
| | 1–2 | 2–13 | 6–14* | | |

Data are presented as means±SD (range). ANOVA, analysis of variance; HS, highly significant; KW, Kruskal–Wallis; PO, postoperative; S, significant. *Sig in comparison with Shamblin 1. †Sig in comparison with Shamblin 2.

**Table 4 Vascular reconstruction of carotid system**

| Vascular reconstruction | Findings (Shamblin) | | | | Fisher's test and P value |
|---|---|---|---|---|---|
| | 1 | 2 | 3 | Total | |
| Primary repair | 2 (100) | 1 (33.3) | 0 | 3 (25) | 5.23 and 0.073 (NS) |
| Patch repair | 0 | 1 (33.3) | 0 | 1 (8.3) | |
| Interposition graft | 0 | 1 (33.3) | 2 (100) | 3 (25) | |
| Total of each class | 2 (100) | 3 (100) | 2 (100) | 12 (100) | |

Data are presented as n (%).

**Table 5 Pathological finding of excised tumors**

| Data | Findings |
|---|---|
| Size (cm) | |
| Longest diameters | 5.17±1.4 (3–7.5) |
| Histological types | |
| Benign paraganglioma | 10 (83.4) |
| Malignant | 1 (8.3) |
| Lymph node hyperplasia | 1 (8.3) |
| Surgical margin invasion | |
| Yes | 1 (8.3) |
| No | 11 (91.7) |
| Lymph node status | |
| Positive | 1 (8.3) |
| Negative | 11 (91.4) |

Data are presented as means±SD and n (%).

Infection was treated in one (8.3%) patient by proper antibiotics (Table 6).

**Table 6 Postoperative challenges**

| Variables | Shamblin | | | P value |
|---|---|---|---|---|
| | Class 1 | Class 2 | Class 3 | |
| | 7 (100) | 3 (100) | 2 (100) | |
| Bleeding | 0 | 1 (33.3) | 2 (100) | 0.032 (S) |
| Stroke | 0 | 1 (33.3) | 1 (50) | 0.56 (NS) |
| Cranial nerve deficit | 1 (14.3) | 2 (66.7) | 1 (50) | 0.61 (NS) |
| Infection | 0 | 0 | 1 (50) | 0.026 (S) |

Data are presented as means±SD and n (%). S, significant. More than one complication was observed in one patient. Fisher's exact test was used.

## Discussion

Surgery of carotid chemodectoma is the gold standard for cure. These tumors carry a high risk of massive hemorrhage during resection because they are highly vascular. The lesions may also adhere to or envelop neurovascular structures, increasing the risk of complications [8].

These tumors are best treated early for several reasons: smaller tumors are easier to remove; most tumors can become locally invasive; lesions may grow distally into the skull or involve cranial nerves, complicating resection; and these tumors may develop malignant behavior [1,12,13].

Because complications are common, these morbidities dramatically can be decreased by meticulous dissection and adequate control of carotid system [1].

In the current study, most patients' age was more than 40 years old; despite the exact age incidence of CBT being unknown, they are usually common in the age of 30–50 years old. There was near-equal sex distribution in this series despite sex predominance not being established [1]. This age and sex distribution was comparable to that mentioned by

Ma et al. [15], who reported their study on 18 patients (female: eight and male: 10, mean age: 52 years). The current study patients were more than that mentioned by Zhu et al. [16], who reported 11 cases, and they mentioned that CBT is reported at 50–70 years old, with higher female incidence.

Patients can be anesthetized either cervical block or general anesthesia; in this study, general anesthesia was preferred owing to better airway management and reduced patient movement during the operation as mentioned by Warren et al. [1].

Patients were classified depending on Shamblin classification into three groups; seven (58.3%) patients were class 1, three (25%) patients were class 2, and two (16.7%) patients were class 3. This was near similar to that mentioned by Ma et al. [15], who reported 10 (55.6%) cases could be classified into Shamblin I, five (27.8%) cases were Shamblin II, and only three (16.6%) cases were Shamblin III.

Most reported carotid chemodectomas are symptomatic mass, which is similar to the current study. A neck mass was the main symptom in 11 (91.7%) patients. Duplex ultrasound imaging, cpmputed tomography angiography, or MRA allowed for adequate operative planning [8,9,17].

Regarding treated side in this study, bilateral CBT was reported only in one (8.3%) patient, right CBTs were reported in four (33.3%) patients, and left CBTs were

reported in seven (58.3%) patients. This was comparable to that mentioned by Ma *et al.* [15], who observed unilateral mass in 17/18 (94.4%) cases, with 13 masses in the left and five masses in the right side and bilateral in 1/18 (5.6%) case .

By reviewing operative events, mean operative time was longer in patients of Shamblin class 3 (5.4 ±0.77 h; range, 4.5–6 h; $P<0.001$). Mean intraoperative blood loss was more also in patients of Shamblin class 3 (963.3±464.7; range, 700–1500 ml; $P=0.004$). Mean duration of postoperative hospital stay (in days) was longer in the patients of Shamblin class 3 (9.3±4.1; range, 6–14 days; $P=0.003$). This was similar to that mentioned by Warren *et al.* [1] and Power and Hallet [18] but less than that mentioned by Davila *et al.* [8], where the mean time for resected carotid chemodectoma was 224.5 min (52–696 min), and the mean blood loss was 143.9 ml (10–2000 ml).

Regarding vascular reconstruction, all patients of Shamblin class 3 were reconstructed by interposition vein graft and two patients of Shamblin class 1 were repaired primarily, but the three patients of Shamblin class 2 were reconstructed by all means. This result was comparable to that observed by Davila *et al.* [8], where 20 (10.9%) patients underwent reconstruction: primary closure in eight cases, saphenous interposition graft in six cases, and patch closure in six cases.

The mean size of excised tumor was 5.17±1.4, with range of 3–7.5 cm. Most histological types were benign paraganglioma in 32 (94.2%) and only one type was malignant one (2.9%) and another case one (2.9%) was histological surprise, that is, lymph node hyperplasia. This tumor size was comparable to that reported by Ma *et al.* [15], where the resected tumor was larger than 3.5 cm and smaller than 8 cm. Risk of malignancy of carotid chemodectoma is less than 5% [19]. This was reported also by Davila *et al.* [8]. One case developed pulmonary secondaries after resection by 2 years. However, Ma *et al.* [15] found neither recurrence nor metastasis in 11-year follow-up.

This study confirms that patients can safely undergo excision, and most complications are considered technically preventable, and adequate planning by the surgeon can greatly reduce the rates of adverse events. The current study complication percentage was more as compared with that observed by Davila *et al.* [8] Postoperative complications were observed in 36 (19.7%) patients: cranial nerve deficit in 28 (15.2%),

stroke in two (1.0%), and hematoma in three (1.6%). However, Ma *et al.* [15] reported no nerve injuries, except hoarse voice, which recovered later on. Although recurrence was rare, patients should be followed closely after discharge [1,18].

## Conclusion

Early surgical excision by a vascular surgeon is the only proven cure for CBTs because they are small and easy to remove. However, if neglected, there will be many challenges as most tumors can become locally invasive. Meticulous periadventitial (and sometimes subadventitial) dissection can greatly reduce the rates of complications.

### Conflicts of interest
There are no conflicts of interest.

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