



## SINGLE CENTRE EXPERIENCE WITH CAROTID BODY TUMOUR

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**ABSTRACT** **Background:** Carotid Body tumours (CBTs) are rare neuroendocrine tumours. Due to their proximity to vital structures including major vessels and cranial nerves, Surgical excision requires meticulous dissection and any injury to major vessels requires prompt repair. This study aimed to review our experience with the surgical management of Carotid Body Tumours. **Methods:** A retrospective study was performed on 20 patients who underwent excision of carotid body tumours at Vardhman Mahavir Medical college and Safdarjung hospital, New Delhi between January 2003 to June 2022. Demographic data describing the presentation, preoperative and intraoperative details, and postoperative complications reported were tabulated and descriptive statistical analysis was done. **Results:** A total of 20 patients were operated on. Complete resection was possible in all the patients. Vascular injury occurred in only 2 patients which were repaired promptly. Cranial nerve injury was reported in only 1 patient which resolved on follow-up. There was no incidence of stroke or recurrence. **Conclusions:** Periadventitial Excision is the treatment of choice for carotid body tumours and is a safe and effective procedure with minimal intraoperative and postoperative complications.

**KEYWORDS :** Carotid body tumour, Periadventitial Excision

### INTRODUCTION

Carotid body tumors (CBTs) are rare neuroendocrine neoplasms which arise near the carotid bifurcation within glomus cells derived from the embryonic neural crest and account for 0.6% of the head and neck tumors in humans.<sup>1</sup> Despite their rarity, CBTs are the most frequent neuro-ectodermal tumor of the head and neck. CBTs are usually benign with the incidence of malignant tumors below 10%, with most malignant tumors diagnosed after the detection of metastases.<sup>2</sup>

The majority of these tumors are asymptomatic and initially noticed by inspection and palpation of swelling in the neck during the physical examination or as an incidental finding on radiological imaging studies. The most common symptoms include swelling on the right side of neck, pain, dysphagia, and autonomic dysfunction. They can also produce symptoms due to pressure and local invasion of the surrounding tissue. The most commonly involved nerves are the hypoglossal nerve, glossopharyngeal nerve, vagus nerve and sympathetic chain.<sup>3</sup>

The most common and clinically relevant anatomic classification used is the Shamblyn et al.,<sup>4</sup> which was designed as a predictor of intraoperative technical difficulty.<sup>5</sup> It describes the extent of the common carotid, internal carotid, and external carotid artery envelopment by the carotid body tumor. The operative approach must be planned according to the tumor classification and preoperative imaging details.

Due to their proximity to great vessels, these patients present to the Cardiothoracic and Vascular surgery department for surgical excision.

Meticulous dissection is required during their surgical excision to prevent injury to common carotid artery, external and internal carotid artery, internal jugular vein and various cranial nerves. Thus, there are high chances of vascular injury requiring vascular repair or reconstruction, depending upon the size and extent of these tumors. The present retrospective study was conducted to review and assess the surgical outcomes of carotid body tumor excision in our patients.

### MATERIAL AND METHODS

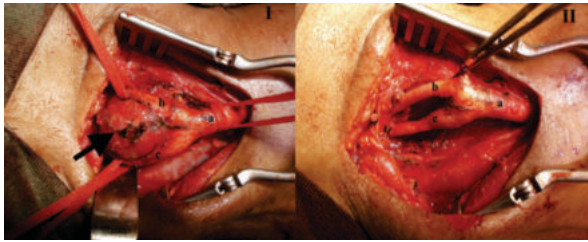
In the present retrospective study, data from the medical records of all the patients who underwent surgical excision for carotid body tumor at the Department of Cardiothoracic and Vascular Surgery, Vardhman Mahavir Medical College & Safdarjung Hospital, Delhi, between January 2003 to April 2022 were collected. Medical records were reviewed for demographics, operative details, pathology, postoperative complications, survival, and tumor recurrence. Shamblyn's grade was determined from the surgeon's operative report and preoperative radiological evaluation. Confirmation of diagnosis was made on the basis of histopathological report.

Demographic data describing the presentation, preoperative and intraoperative details, and postoperative complications reported was tabulated and descriptive statistical analysis was done.

### Surgical Technique :

Incision was made at the anterior border of sternocleidomastoid muscle. The sternocleidomastoid muscle was gently retracted. The carotid sheath was opened. Tumor was identified along with carotid arteries. Tumor dissection was done in the periadventitial plane taking care that any pressure on the tumor may lead to catecholamines release

into the bloodstream and subsequent hypertension. As these are highly vascular tumors all the feeder vessels from vertebral and ascending cervical artery were identified, doubly looped and ligated. Periadventitial tumor dissection is carried out carefully to avoid injury to the vessel and adjoining nerves. (Figure 1)



**Figure 1:** [I] Intraoperative image showing Carotid body tumour (bold arrow), at the bifurcation of common carotid Artery .[II] Final intraoperative image post excision of tumour. { a – Common carotid, b – external carotid, c – internal carotid}

**RESULTS**

In this study, the mean age was 48 9.1 years(range 21-60 years). The male-to-female ratio was 14:6. The most common presentation was slow growing neck mass (20), followed by headache (5) and pain (3). None of the patients presented with hypertension or other signs of active neuroendocrine tumor. (Table 1)

**Table 1: Demographics and presentation.**

Sl. No	Preoperative characteristics	N(n=20)
I	Gender	
1.	Male	14
2.	Female	6
II	Side	
1.	Right	12
2.	Left	7
3.	Bilateral	1
III	Presenting Symptoms	
1.	Neck Mass	20
2.	Headache	5
3.	Pain	3
4.	Dysphagia	1
5.	Hoarsness of voice	1
6.	Vertigo	1

Complete resection was achieved in all the patients. The majority of the patients 11 (55%) were Shamblin class 1, and only 2 (10%) patients were Shamblin class 3. 1 patient was found to have enlarged lymph node which was sent for biopsy and was found to be negative for metastasis. Intraoperative vascular injury occurred out of which re-anastomosis of external carotid artery was done in 1 patient and rent repair of common carotid artery was done in other. Post-operatively, 1 patient developed cranial nerve deficit which resulted in hoarseness in voice however it was transient and improved on follow up. (Table 2)

**Table 2: Intraoperative characteristics.**

Sl. No	Intraoperative characteristics	N(n=20)
I	Tumor Size	
1.	<5cm	14
2.	5cm	6
II	Shamblin Grade	
1.	I	11
2.	II	7
3.	III	3
III	Intraoperative complication	
1.	Common carotid artery injury	1
2.	External carotid artery injury	1
3.	Internal carotid artery injury	0
4.	Nerve injury	0

All patients were confirmed to have carotid body tumor on post-operative histopathological examination. Minimum follow up period was 6 months, and 14 patients were followed up upto 5 years. None of the patients presented with recurrences of tumor on follow up. (Table 3)

**Table 3: Postoperative complications and follow up.**

Sl. No		N(n=20)
I	Postoperative complications	
1.	Bleeding	0
2.	Reexploration	0
3.	Vocal cord palsy	1
4.	SSI	2
5.	Wound dehiscence	0
6.	Stroke	0
II	Recurrence	0

**DISCUSSION**

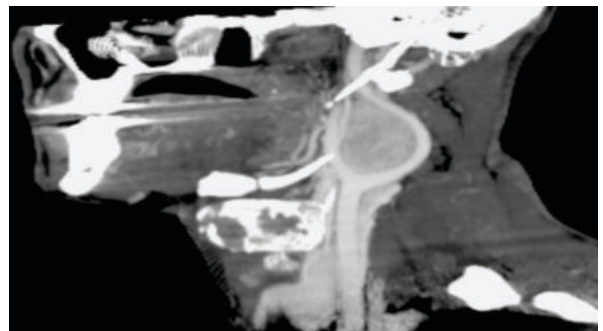
The most common site of paragangliomas in head and neck is the carotid bifurcation. Carotid body tumors usually present as a painless, rubber-like, slowly enlarging mass, mobile in the horizontal direction with limited mobility in the vertical direction (Fontaine sign).<sup>6</sup> They are located at the anterior border of the sternocleidomastoid muscle at the level of the hyoid bone. As the tumor grows, dysphagia, odynophagia, dysphonia and symptoms due to compression of cranial nerves 9 to 12 may be observed.

Carotid body tumors usually present in 3<sup>rd</sup> to 6<sup>th</sup> decade of life and are more common in females as reported in the literature,<sup>7,8</sup> however in this study, majority of patients were male. Most common presenting complaints of our patients were neck mass, headache and pain. Bilaterality has been observed in 5- 10% for sporadic tumors,<sup>9</sup> in this study only 1 patient presented with bilateral tumors. Inhabitants of high-altitude areas have been reported to have higher incidence of CBTs.<sup>9</sup> This has been attributed to hyperplasia of carotid body due to chronic hypoxia. In this study, 6 (30%) patients were residents of high-altitude areas.

CBTs can be classified on the basis of aetiology, genetic mutations and anatomy. Etiological types include sporadic (most common), familial and hyperplastic. Three genes (succinate dehydrogenase [SDH] complex subunits “B”, “C” and “D”) have been linked to an increased incidence of paraganglioma.<sup>10</sup> However, tumor function, age of onset, risk of malignant diseases, and transmission vary by genotype. However, tumour function, age of onset, risk of malignant diseases, and transmission vary by genotype<sup>10-11</sup>. Genetic testing is considered for patients with multiple paragangliomas or a positive family history. In this study none of the patients had a positive family history.

Shamblin et.al, classified CBTs according to the extent and encasement of carotid arteries and bifurcation.<sup>4</sup> This classification thus correlates well with the surgical complexity. In this study, we classified the tumors using Shamblin classification both preoperatively using imaging studies and postoperatively based on intra-operative findings.

Ultrasound duplex imaging is usually considered the 1<sup>st</sup> choice investigation and is an excellent screening modality for CBT. It reveals highly vascularized hypoechoic mass at the carotid bifurcation. Major disadvantage with ultrasound imaging is that the study is operator dependent. Computed tomography or Magnetic resonance angiography is considered the most important preoperative imaging technique as it provides complete anatomical details regarding tumor extent, relationship to vessels and feeding vessels. (Figure 2) All our patients underwent ultrasonography as a primary investigation and CT angiography for confirmation of diagnosis and to know the extent to plan for surgical resection. Three patients also underwent Magnetic resonance imaging neck for confirmation of diagnosis and to look for local invasion.



**Figure 2:** CT angiography showing carotid body tumour on the right side (a), Common carotid artery (b), internal carotid artery (c), external carotid artery (d).

The blood supply is primarily from the bifurcation and external carotid artery, but feeding vessels from the internal carotid artery, vertebral artery, and thyrocervical trunk are also frequently found. Information of the sources of blood supply helps in exposure and hemostasis. A specific perfusion increase (Lyre sign) at the carotid bifurcation can be seen due to the tumor and is pathognomic to CBTs.<sup>12</sup> Arteriographic imaging should be carried out bilaterally to rule out bilateral tumors. During arteriography, embolization can also be performed to the feeding vessels to reduce the vascularity of the tumor, resulting in ease of dissection and lower incidences of hypertensive episodes while handling the tumour. However several studies have failed to report any efficacy of preoperative embolization<sup>13-15</sup>.

We did not opt of preoperative embolization in any of our patients. All Patients should undergo Serum catecholamine levels as well as urinary vanilmandelic acid and metanephrine levels to rule out associated adrenal paraganglioma and if found positive should be further investigated before surgery. All our patients underwent the same, however it was found to be negative in all our patients.

Surgery is considered as the first choice of treatment and complete surgical removal is the only proven cure for CBTs. Radiotherapy has been considered as an alternative modality for an elderly patient in poor general status or those who are surgically unfit secondary to comorbidities, whereas surgical treatment is preferred in young patients. Good exposure with identification of carotid artery both proximally and distally is the key to good and adequate surgical excision. Meticulous surgical techniques and timely operations in the initial stages of CBTs are the optimal way to minimize complications<sup>16</sup>. Every effort should be made to identify and preserve the vagus, hypoglossal, and spinal accessory nerves during the operation because injury of these cranial nerves contributes significantly to postoperative morbidity. Tumor removal requires periadventitial dissection. The most important principle is to preserve the integrity of the internal carotid artery. If necessary, the external carotid artery can be sacrificed. Any injury to common or internal carotid artery must be identified and repaired promptly. Graft replacement can be done if necessary.

Cranial nerve injury is the most commonly reported complication after CBT resection<sup>17</sup>. In our patients, we found cranial nerve injuries in only 1 patient which was transient and improved on follow up, which is less than the results reported in literature. The incidence of stroke and recurrence are less than 5% and less than 6% respectively as reported in the literature<sup>18</sup>. However none of the patients in this study developed stroke in the postoperative period. No incidence of recurrence was reported in the follow up.

## CONCLUSION

CBT can be treated safely with surgical excision with minimal post-operative complication. Tumour handling should be minimal during surgical excision to prevent catecholamine release by pressure. A periadventitial excision is recommended as it allows complete resection with preservation of neurovascular structures surrounding the tumor. Continuous invasive blood pressure monitoring and facility for vascular repair should be readily available.

## DECLARATIONS

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Conflict of interest: none declared

Ethical approval: Not required

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