

# High Altitude Bilateral Carotid Body Tumor: A Surgical Management

Sudesh Kumar<sup>1</sup>, Narender Kumar<sup>1</sup>, Umesh Kumar<sup>1</sup>, Niraj Gupta<sup>2\*</sup>, Puneet Sharma<sup>3</sup> and RS Kanwar<sup>3</sup>

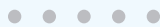
<sup>1</sup>Department of Otolaryngology and Head Neck Surgery, Rajendra Prashad Government Medical College, Himachal Pradesh, India

<sup>2</sup>Department of Surgery, Rajendra Prashad Government Medical College, Himachal Pradesh, India

<sup>3</sup>Cardiothoracic and Vascular Surgery, Rajendra Prashad Government Medical College, Himachal Pradesh, India



**\*Corresponding author:** Niraj Gupta, Department of Surgery, Rajendra Prashad Government Medical College, Tanda Hospital Rd, Pushp Vihar Colony, Kangra, Himachal Pradesh 176001, India. E-mail: drdudds@gmail.com



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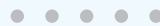
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## Abstract

Carotid body tumor arise from the paraganglionic cells located at the medial surface of the carotid bifurcation. Bilateral tumors occur in only 5% of these cases. Our case report is about a 29-year-old female resident of high altitude (> 3500 meters above sea levels), she developed bilateral neck swelling and her radiological investigations determined she had bilateral carotid body tumors. The larger tumor (Shamblin III) was successfully excised and the smaller tumor was kept under follow-up.

## Introduction

Para-Ganglion cells (PG) or neuro-endocrine cells are derived from neural crest and based on location, they are classified as abdominal and extra-abdominal. Tumor arising from these cells are known as Para Gangliomas (PGS). The adrenal gland is the largest abdominal organ containing PG cells, they also contain chromaffin cells, which is pivotal in its functioning as an endocrine organ. The extra-abdominal PGS are located in the Head and Neck (HN) and mediastinum. They contain mostly non-chromaffin cells and may function as chemo-receptors e.g. carotid body tumor. In the HN region, these are found along the parasympathetic nerve, cranial nerve IX and X, and the superior and inferior laryngeal nerves [1]. PGS are benign and highly vascular tumors that arise from paraganglionic cells [2]. About 0.5% of HN tumors are PGS [3]. Around 65% of all head and neck PGS are Carotid Body Tumors (CBT) [4]. In the beginning, it appears as a painless mass in the neck, but it can grow to a considerable size over time, making it life threatening [5]. They most often occur unilaterally, while approximately 5% are bilateral in HN region [6]. The radiological appearances of PG are characteristic and are very useful in the diagnosis. Not only do they assist in staging, but they also provide guidance on how to manage them [7]. After a diagnosis is made, surgery is the only definitive treatment, although it has risks and complications [8]. In bilateral tumors, either a staged operation should be performed or a smaller tumor should be observed after resection of the larger tumor [4]. When bilateral resections are done simultaneously, a high rate of cranial nerve injuries and cerebrovascular complications can result, which can have severe consequences [9]. We describe the case of a young female, who was a native of high altitude and had bilateral CBT. The larger tumor of these was surgically removed and she is in follow-up for the tumor of the contra-lateral side.

## Case Presentation

A 29-year-old married female patient native of high altitude (11400 feet), in Himalayan state of India, presented to us with complaint of right upper neck swelling for one year duration. It was painless initially but for the last two months there was intermittent pain with

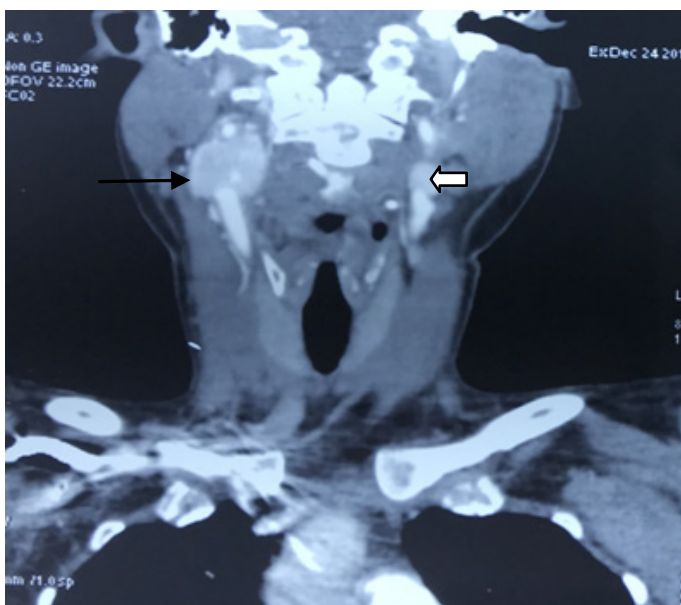
mild relief on medication. Around two months ago, she noticed similar swelling on the left side. She denied any family history of similar swelling in the head and neck region. There was no history of palpitation, flushing or fluctuating hypertension. During the physical examination, in the right upper neck, there was a 5 cm x 4 cm non-tender, pulsatile swelling in upper part of anterior triangle of neck, superiorly it was extending under the mandible and inferiorly up to the level of hyoid bone. On the left, there was a 1 cm x 2 cm similar pulsatile swelling at level of hyoid bone. The examination did not revealed any intra-oral bulges and all the cranial nerves were functioning normally. Contrast-Enhancing Computed Tomography (CECT) and CT angiography of the neck revealed a massive mass measuring 6.5 cm x 5 cm x 4.5 cm on the right side (Shamblin III) and 1 cm x 1.5 cm x 1.2 cm on the left side (Shamblin I). The larger right side tumor extended superiorly to the skull base (Figure 1, Figure 2).

CT chest and abdomen showed no similar lesions. CT head revealed a patent circle of Willis on the contralateral side. This was performed to assess the risk of hemiplegia if during the surgery ICA is temporary clamped for some time. Her vitals were stable and the 24 urinary Vanillyl mandelic acid level was 5 µ/L. After a detailed discussion with the patient and a review of the literature, informed consent for the right CBT excision was obtained. The vascular team was consulted regarding the possibility of carotid artery and subsequent need for repair due to the large size and extension of the tumor to the base of the skull. Nasal intubation was preferred because it aids in exposure by sub-luxating the mandible anteriorly. The incision was made beginning from a point two fingers below the inferior border ramus of the mandible and extended posteriorly to the mastoid process. Sub-platysmal flap was elevated and sternocleidomastoid muscle was retracted posteriorly and carotid artery sheath was opened. The internal jugular vein was secured and retracted. The vagus, hypoglossal

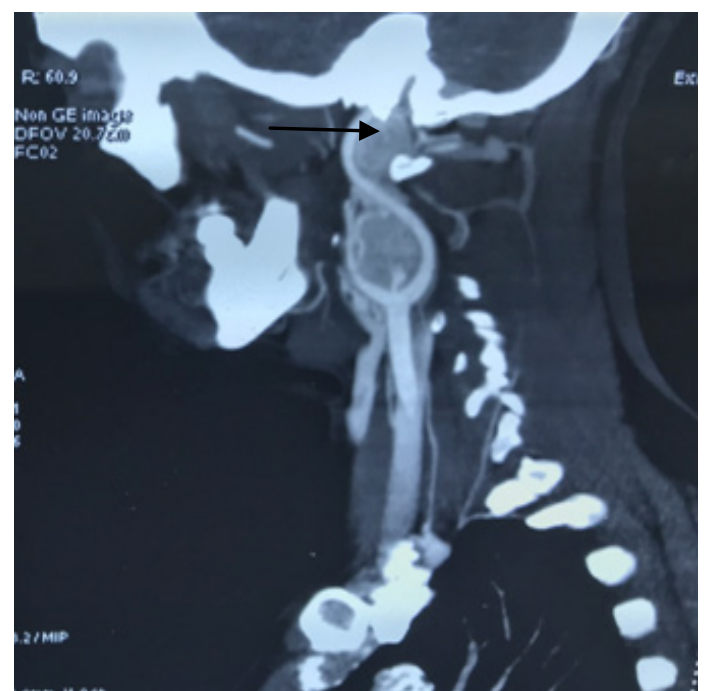
and spinal accessory cranial nerve were identified and preserved. The control of the Common Carotid Artery (CCA), Internal Carotid Artery (ICA), and External Carotid Artery (ECA) was obtained through passing vascular loops. Initial dissection of the tumor from ECA was followed by identification and ligation of the main feeder ascending pharyngeal artery. The stylo-mandibular ligament was cut and the cranial end of the ICA was secured by superior dissection of the tissue around the ICA. In the end, dissection was started on the medial side of carotid bifurcation, where the tumor was found firmly adherent to the vessel wall.

During dissection, Prolene No. 6 was immediately used to patch up two rents accidentally produced in the ICA wall. The tumor was then excised and it was around 6.5 cm x 4 cm x 2 cm in size, bilobed structure, and it was sent for the histopathological examination (Figure 3).

Intra-operative doppler ultrasound showed absence of the pulse in the ICA; and it was suspected to be thrombosed, probably due to manipulation and sutures applied to control bleeding from ICA. As a result, the vascular team opened the ICA, removed the clot that was occluding its lumen, and checked for retrograde bleeding. An end-to-end anastomosis of CCA to ICA was performed due to the risk of re-thrombosis. The anastomosis was performed with continuous sutures of Prolene 6, and vessels were clamped for approximately ten minutes. Patient was immediately heparinised with 3000 units, and application of heparin in this dose continued every 6 hourly for one week. In the intensive care unit, that was brought after surgery, she was kept intubated overnight and extubated after 24 hours. There were no neurological deficits or cranial nerve injuries after surgery. After one week on low dose aspirin, she was discharged and kept on follow-up.



**Figure 1:** Coronal CT scan the neck region showing a right Shamblin III (Black Arrow) and left Shamblin I (White Arrow) carotid body tumor.



**Figure 2:** Sagittal CT of the right neck showing a large bilobed tumor with splaying of carotids and superior extension up to the base of the skull (Black Arrow).



**Figure 3:** Excised surgical specimen of 6.5 cm x 4 cm x 2 cm size with bilobed tumor (larger arrow main tumor, smaller arrow the tumor part extending towards the base of skull).

On microscopic examination there were well defined nests of cuboidal cells (Zellballen pattern) separated by highly vascular fibrous septa suggestive of carotid body PG and excised carotid vessel wall did not show infiltration. CT scan after six months showed right normal patent ICA and stable tumor on left side and she was kept on follow-up.

### Discussion

CBT was first described in 1973 by Albrecht Von Haller as ganglion minimum [10]. The three variations are described in the literature as familial, hyperplastic, sporadic (85%), and sporadic usually occurs in the 4<sup>th</sup> decade of life [11]. Associated with an early presentation and a bilateral CBT rate of 30%–40%, is the familial variant [12]. As seen in our patient, the hyperplastic variant is associated with chronic hypoxia, including chronic pulmonary obstructive disease, cyanotic heart disease, and high altitude (> 5000 feet) [13].

Shamblin in 1971 was first to suggest a grading system for CBT depending upon the size of tumor and encasement of carotid vessels [14]. Grade I refers to small tumor, that show no encasement of carotid vessels, and can be easily dissected. Grade II tumors are adherent or partially encase the carotid vessels and compress them but they can be dissected. Grade III are encasing and adherent requiring vessel resection and reconstruction. Our patient had a Shamblin III tumor on the right side, which is associated with a 30%–46% risk of neurovascular injury [15]. During surgical excision of a CBT tumor, the vagus and hypoglossal nerves are most vulnerable to injury [16]. If both sides of these nerves are injured in bilateral CBT surgery, severe morbidity can result. Consequently, a conservative treatment of smaller tumor either by staging or observing is a better treatment option [4]. Whenever a patient develops postoperative cranial nerve palsy after the removal of larger tumor, it is always recommended to observe the other side tumor by physical examination or radiologically. There is another school of thought, based on their experience, that the

smaller tumor should be removed first, followed by the larger ones [9]. The surgical resection of bilateral CBT tumors, however, may result in baroreflex failure post-op, which is characterized by sympatho-excitation, hypertension, and tachycardia [17].

Tumors larger than 6 cm size, the exposure of tumor especially superior or cranial end is always a area of concern. The cranial exposure can be enhanced extension by nasal intubation, anterior sub-luxation of the mandible, and excision of the styloid process and mandibular osteotomy [18]. Similarly, we did naso-tracheal intubation for anaesthesia which was useful for the cranial (superior) exposure as it increases the space between tumor and mandible. It has been reported that 23.2% of ICA injuries occur during CBT surgery and that most of them occur on the medial surface of the carotid bifurcation [19]. The repair of vessel injuries can be performed though direct closure, patch plasty, or graft reconstruction [16]. In our case also the tumor was firmly adherent and accidental tear was managed with lateral suturing but it had lead to ICA thrombosis which was finally managed by end to end anastomosis of CCA and ICA.

### Conclusion

Bilateral Carotid Body Tumor presents a unique diagnostic as well therapeutic challenge. Excision of the larger tumor and observation of the smaller one is well-accepted management option. In tumors of six centimetres or larger, there is a high chances of vascular injury, so a vascular team consultation is utmost important. We also saw that naso-tracheal intubation improves the exposure of tumor by sub-luxating the mandible anteriorly.

### Conflict of Interest

The authors declare no potential conflicts of interest with respect to the research, authorship, and/or publication of this article. Informed consent was obtained for this publication.

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