

## Bilateral Carotid Body Tumour: Stormy Physiology and Hemodynamics A Rare Case Report

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### ABSTRACT:

Carotid body tumours (CBT) are rare and highly evolved vascular neoplasm arising at the bifurcation of common carotid artery splaying the internal and external carotid arteries. Surgery for carotid body tumour may be associated with severe cardiovascular alterations due to its location, proximity to vital structures, involvement of nerves and stimulation of richly innervated chemoreceptors and carotid sinus. Here the authors describe a case of bilateral carotid body tumour in a 30 year old male patient posted for surgical removal of the tumour and sudden the event of hemodynamic instability due to injury to external carotid artery.

**Keywords:** Bilateral carotid body tumour, Chemodectoma, Paraganglioma, Hemodynamic monitoring, Lyre sign, Early vein, Salt and pepper appearance

### INTRODUCTION

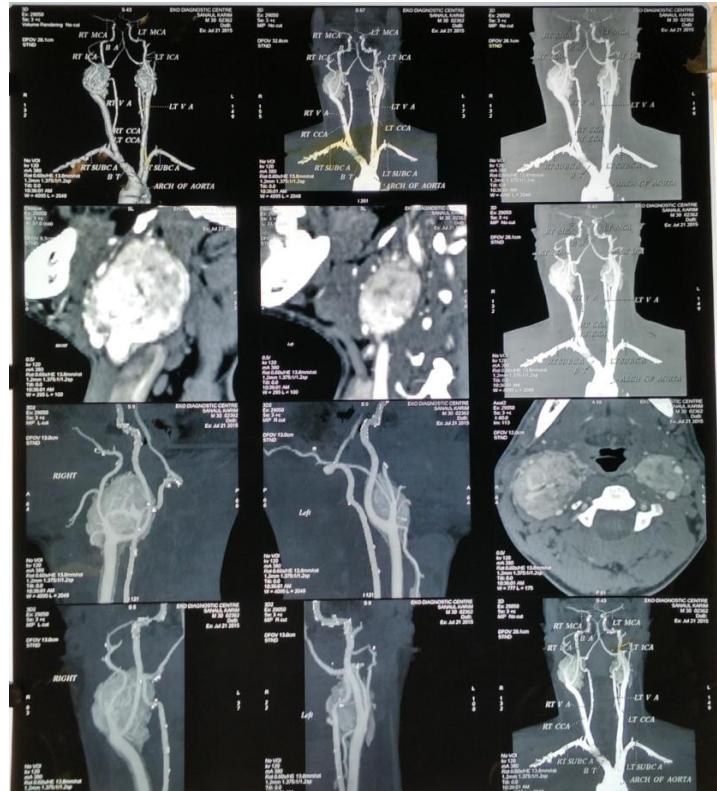
Carotid body tumours (CBT) also known as chemodectoma or carotid body paragangliomas, are rare and highly vascular neoplasm that arises from the paraganglion cells of the carotid body and develop within the adventitia of medial aspect of carotid bifurcation. Surgical removal of CBT may be associated with severe hemodynamic instability due to injury to the major vascular structure or stimulation of the highly innervated chemoreceptors. Here the authors describe a case of a middle aged male patient posted for surgical removal of carotid body tumour who developed severe hemodynamic instability due to injury to external carotid artery.

### CASE REPORT

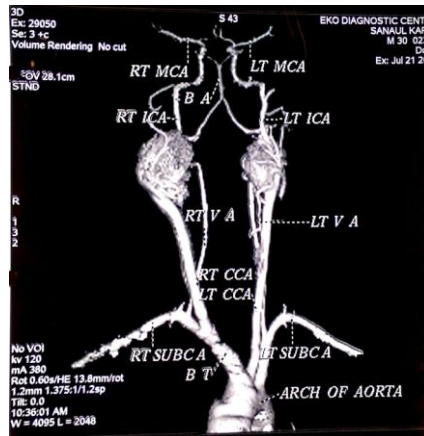
A 30 year old man presented with a bilateral swelling on the neck, near the angle of jaw since childhood. The swelling was slow growing painless, non pulsatile with dimension of 4.4x4.3x2.8 cm on right side and 3x2.7x2.5 cm on the left side. There was no history of trauma, fever, or cough. On general physical examination, the patient was 60 kg in weight, 164 cm in height with a pulse rate of 74/min, and blood pressure of 120/70 mm Hg. The airway assessment showed Mallampati grade II, thyromental distance of 6 cm and adequate mouth opening. MDCT angiogram showed well-defined hypo to isodense soft tissue mass lesion at level of carotid bifurcation bilaterally showing intensely enhancing lesion in arterial phase (Figure 1,2 and 3). Circle of Willis was normal. A diagnosis of bilateral carotid body tumour was made and resection of right side CBT was planned under general anaesthesia.

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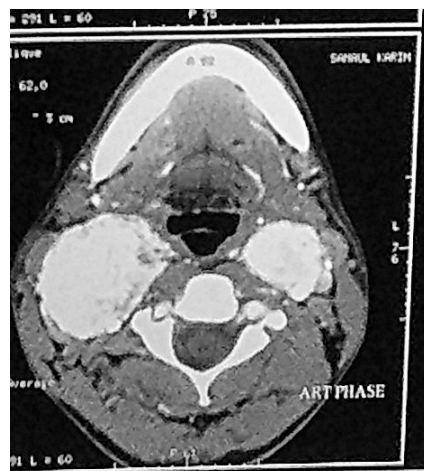
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**Figure1.** Sections of MDCT angiogram of neck showing two separate lesions at the level of carotid artery bifurcation with splaying of internal and external carotid artery



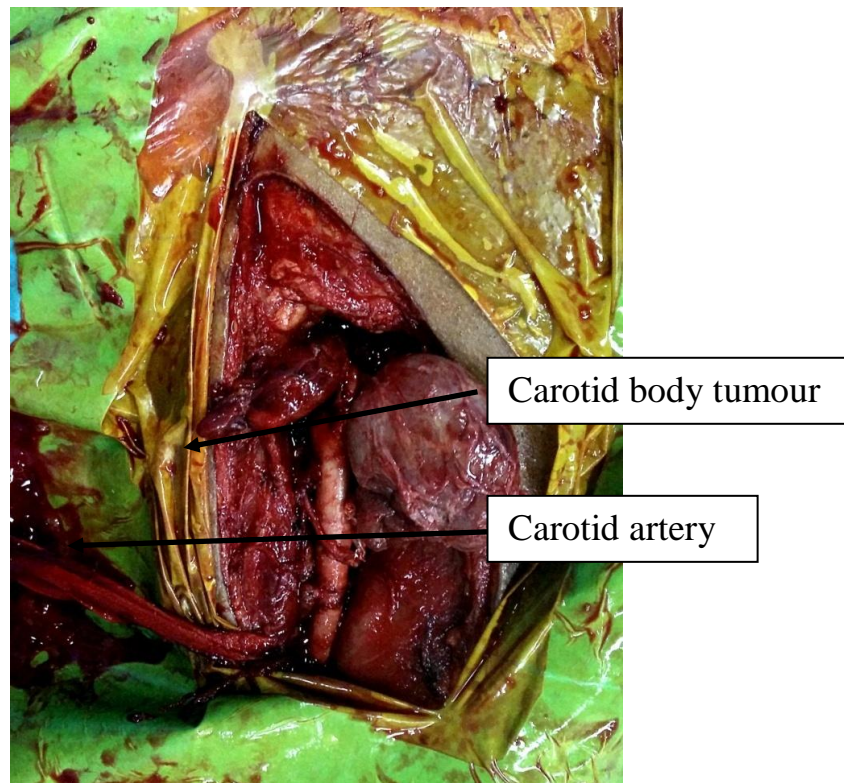
**Figure2.** Location of bilateral carotid body tumour in relation to carotid arterial system



**Figure3.** Transverse section of base of the skull showing soft tissue mass at the level of carotid bifurcation intensely enhancing lesion in the arterial phase larger in the right side

On arrival to the operating room, noninvasive blood pressure (BP) of 132/76 mmHg, heart rate of 88/min, regular, respiratory rate of 14/min, and oxygen saturation of 100% were recorded. Monitors were attached including, ECG pulse oxymetry, End tidal CO<sub>2</sub> measurement and temperature measurement. A radial arterial line was established for invasive blood pressure measurement under local anesthesia. After anesthesia induction, a central venous line was established via right subclavian vein and nitroglycerine infusion was started to keep mean BP around 80mmhg. Ambient temperature of the operation theatre was kept 21 C.

The neck was explored on the right side by a vertical incision. The tumour was dissected and found to be occupying the carotid bifurcation splaying the internal and external carotid artery. The area of carotid bifurcation was infiltrated with 3ml 0.1% lignocaine. While separating the mass from carotid artery, injury to external carotid artery occurred leading to blood loss of around 2 liters with hypotension. 1 liter of colloid was transfused. The artery was clamped and repaired. Inj Thiopentone 300 mg was given as a loading dose followed by an infusion of 250 mg/hour was started to provide neuroprotection. Patient's temperature was maintained between 32 to 35<sup>0</sup>C to provide neuroprotection. 4 units of packed red blood cells were transfused while maintain map around 80 mm hg. Total duration of surgery was 4 hour. Patient was extubated on the next morning. He was conscious, oriented with no neurological deficit.



**Figure4.** Neck dissection and the carotid body tumour.

## **DISCUSSION**

The carotid body is a small, oval, reddish brown structure located at posteromedial aspect of the carotid artery bifurcation. Normally it is 3-5 mm in diameter and around 15 mg in weight. The blood supply of carotid body arises from ascending pharyngeal artery through the Mayer ligaments and the nerve supply originates from Hering nerve, branch of the glossopharyngeal nerve<sup>1</sup>. The chemoreceptors in carotid body sense the change in partial pressure of oxygen and carbon dioxide in the blood and regulate ventilation to control hypoxia, hypercarbia and acidosis<sup>4</sup>.

Usually they are ellipsoid, red brown masses of 5 to 7 mm in height and 2.5 to 4 mm in width, located in the posterior face of the common carotid artery bifurcation<sup>7</sup>. They represent 60-70% of paraganglioma of the head and neck and bilateral in approximately 10% case<sup>9,10</sup>. CBT may be sporadic, familial or hyperplastic. 7-10% cases may be familial and 35-50% of familial cases may be multicentric<sup>6,7</sup>. Hyperplastic CBT is more common in patients with chronic hypoxia, patients living in high altitude and in patients of chronic obstructive pulmonary disease<sup>11</sup>.

The tumour is initially confined within carotid trigone and enlarges in relation to common carotid artery and its bifurcations including internal and external carotid artery, internal jugular vein and vagus nerve. Later hypoglossal nerve, the glossopharyngeal nerve, the pharyngeal and superior laryngeal rami of the vagus nerve, the mandibular ramus of the facial nerve, the accessory nerve, and the cervical sympathetic chain may also be involved and injured during surgical resection of the lesion<sup>6</sup>.

The lesion is usually asymptomatic in the initial stage. The tumours are slow growing with doubling time (T<sub>D</sub>) 7.13 years with a median growth rate of 0.83 mm/year<sup>9</sup>. Most patients present at their 4<sup>th</sup> or 5<sup>th</sup> decade of life. In 93% patients the the initial presenting symptom is painless palpable neck mass in the anterior triangle of neck<sup>8</sup>. Symptoms may appear in the later stage due to compression and involvement of the nearby structures (Table 1). On examination there is limited mobility in vertical direction because of its attachment to the bifurcation of the common carotid (Fontaine sign).Pulsation, thrill or bruit may or may not be present. The differential diagnosis includes branchial cysts, carotid aneurysms, metastatic carcinomas, intravagal tumors, lymphomas and ectopic thyroid<sup>12</sup>.

**Table1.** Symptoms of carotid body tumour<sup>6,10, 11</sup>

Symptoms	Incidence	Remarks
Accelerated growth of the mass	73%	
Headache or neck pain	35%	
Buzzing, dizziness		
Hoarseness	8%	
Syncope		
Neural involvement <ul style="list-style-type: none"> <li>• Vagus</li> <li>• Recurrent laryngeal nerve</li> <li>• Hypoglossal</li> <li>• Glossopharyngeal</li> <li>• Cervical sympathetic chain</li> </ul>	10%	May be responsible for pain, hoarseness, dysphagia, Horner syndrome, or shoulder drop.
Fever		Rarely. May cause pyrexia of unknown origin
Functional carotid body tumour		Symptoms similar to those of pheochromocytoma, such as paroxysmal hypertension, palpitations, diaphoresis, may be seen

The histological findings of the tumour does not corroborate with its behaviour. Malignancy is indicated by presence of lymph node involvement or distant metastasis<sup>10,12,13</sup>. Morbidity depends on size and compressive effect of the tumour, involvement of local structure, involvement of cranial nerves, obstruction of airways and invasion of the base of the skull. Difficulty of surgical resection may be assessed by the Shamblin classification<sup>7</sup>.

Prebiopsy imaging study is essential in a patient presenting with suspected CBT. Ultrasound may delineate a mass in relation to carotids and colour doppler may be used to assess the vascularity. Contrast enhanced computerised tomography scan is an excellent tool in depicting anatomy of these lesions and helps the analysis and detection of involvement of the cranial base, pharynx and jugular vein.. Classical findings are soft tissue density similar to muscle, bright and rapid enhancement and splaying of carotid bifurcation. Magnetic resonance imaging (MRI) scan shows iso to hypointense signal in T1 weighted image and hyperintense signal in T2 weighted image. Classical appearance is 'salt and pepper' appearance. (Table2). Magnetic resonance angiography may be helpful to identify the vascularity of the tumour and the feeding vessel. Angiography is necessary to establish the accurate diagnosis. It also detects the presence of occlusive carotid disease, the status of intracerebral blood circulation, tumor irrigation and the presence of small asymptomatic contralateral tumors, as well as allowing preoperative embolization.

**Table2.** Some specific signs and their appearances associated with carotid body tumour

Signs	Appearances
Lyre sign	Splaying of the carotid vessels
Early vein	Angiography finding, due to arteriovenous shunting
Salt and pepper appearance	Combination of punctate regions of haemorrhage or slow flow (salt) and flow voids (pepper) in T1 and T2 weighted MRI

Perioperatively carotid body tumour may pose several anesthetic challenges and perioperative morbidity approaches 20-40%<sup>14, 15</sup>. Stroke, excessive blood loss, injury to vital structures may account for perioperative mortality. Factors related to perioperative morbidity include tumour size and involvement, surgical manipulation, postoperative edema, injury to nearby nerves, compression and injury to nearby vital structures, blood loss and hemodynamic perturbation during surgery etc. Mild hypothermia (32-35°C)<sup>16</sup> and thiopentone infusion<sup>17, 18</sup> may offer neuroprotection when cross clamping exceeds 11 minutes. The patient in this discussion had an injury to external carotid which was repaired after application of cross clamps. Induced hypotension with nitroglycerine was used to reduce blood loss and hypothermia and thiopentone was used to provide neuroprotection. Postoperative airway obstruction maybe related to tumour involvement, intraoperative injury or postoperative perineural edema of the ninth, tenth or twelfth cranial nerves. Patient should be mechanically ventilated following surgery and closely observed following extubation to identify respiratory obstruction at the earliest. The patient discussed was extubated on the next morning and no symptoms of airway obstruction was noted. Bradycardia may occur due to stimulation of the carotid sinus chemoreceptors and may necessitate atropine or temporary pacing and may necessitate permanent pacemaker placement after bilateral surgery<sup>19</sup>. Local lidocaine infiltration may be used to reduce carotid sinus sensitivity<sup>20</sup>.

## CONCLUSION

Bilateral carotid body tumour is extremely rare and patients usually present at later age due to slow growth of the tumour. Symptoms may appear due to compression of the surrounding nerves or vascular involvement. Management is essentially surgical. Major vascular injury is a serious concern and should be managed immediately to prevent catastrophe. Tumour manipulation may give rise to release of catecholamines or carotid sinus syndrome. Perioperative transvenous pacing may be an attractive option. Postoperative complication may arise due to neurological injury or airway edema. If any obstructive airway symptoms develop during postoperative period the case must be reviewed for cranial nerve injury or perineural edema or airway injury and maintenance of airway is of vital importance.

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