Case Report

A rare case of bilateral carotid body tumor with post-operative high vagal palsy

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ABSTRACT

This case report details a rare instance of bilateral carotid body tumors (CBTs) in a 32-year-old female, emphasizing the clinical presentation, diagnostic process, staged surgical intervention, and subsequent management of post-operative complications, notably high vagal palsy. CBTs, a subset of paragangliomas, are rare neuroendocrine tumors with less than 10% malignancy rate, and bilateral occurrences are particularly uncommon. The patient's treatment involved staged excisions of the tumors, with the post-operative period marked by the development of high vagal palsy, leading to significant swallowing difficulties. This case underscores the complexity of managing bilateral CBTs, highlighting the need for meticulous surgical planning and post-operative care to mitigate neurological complications. The report advocates for further research to enhance the understanding and management of such rare tumors.

Key words: Bilateral carotid body tumor, Paraganglioma, Vagal nerve palsy

Paragangliomas are rare neuroendocrine tumors that arise from the extra-adrenal autonomic paraganglia which are small organs that consist mainly of neuroendocrine (chromaffin) cells that are derived from the embryonic neural crest and have the ability to secrete catecholamines. Paragangliomas are often called extra-adrenal pheochromocytomas as two tumors are indistinguishable at the cellular level [1,2]. The carotid body was first described by Von Haller in 1743, who described it as a chemoreceptor located in the adventitia of the carotid bifurcation [3]. Carotid body tumors (CBT) account for 0.6% of the head-and-neck tumors in humans. The bilateral CBTs are usually benign with the incidence of malignant tumors below 10% [4]. The incidence of bilateral carotid tumors is even rarer and associated with familial forms and accounts for approximately 5% [5,6]. The incidence of neurological complications is rare.

In this study, we will discuss a bilateral CBT with post-surgery high vagal palsy.

CASE REPORT

A 32-year-old female presented to the outpatient department with complaints of throbbing type headaches which occurred once in 10 days and then the frequency gradually where she had to take medications to get relief along with blackouts that occurred once a month initially and then the frequency gradually increased over 1 year. There was no history of loss of vision or weakness of the upper or lower limbs. The patient was not diabetic or hypertensive.

On examination, the patient was ECOG 1 well-built and nourished with no signs of pallor. On examination of the neck region, we found bilateral pulsatile swelling of peanut size at the level II region.

Ultrasound examination of the neck revealed heteroechoic lesions in bilateral carotid spaces medial to the internal carotid artery measuring 2 × 1.4 cm on the right side and 9 × 7 mm on the left side showing significant internal vascularity.

Further computed tomography angiogram revealed a well-defined soft tissue lesion noted in the right carotid space at the level of carotid bifurcation causing splaying of both internal and external carotid arteries measuring 1.5 × 1.9 × 2.2 cm (Fig. 1). A similar lesion of size 1.36 × 1.36 × 1.75 cm was noted in the left carotid space at the bifurcation displacing both internal and external carotid arteries suggestive of bilateral CBT.

The patient was planned for bilateral carotid tumor excision in a staged manner. First, she underwent an excision of the right tumor as the tumor was bigger compared to the left side. We gave a vertical incision along the anterior border of the sternocleidomastoid, and subplatysmal flaps were raised and then retracted the sternocleidomastoid muscle laterally and approached the level 2 region. We opened the carotid fascia and identified the bifurcation of the common carotid artery along with the tumor. The hypoglossal nerve, spinal accessory nerve, and vagal nerves were identified and looped. We carefully dissected the tumor after looping the common, internal, and external carotid

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arteries by taking control. The whole tumor was removed in total. Similar steps are repeated over the left side (Fig. 2). The patient developed deviation of the tongue toward the right side due to right hypoglossal nerve palsy which was treated conservatively with steroids; the patient recovered well and ate food with no swallowing or breathing difficulties.

After 2 weeks, the patient underwent an excision of the left carotid tumor. The patient was asymptomatic for 2 days. After orals were started, the patient started developing cough and breathing difficulty. Immediate video-laryngoscopy was done which showed high vagal palsy. The patient has been kept on Ryle's tube feed for 3 weeks and sent for speech and swallowing therapy. The patient recovered well. We removed Ryle's tube after 3 weeks and started on liquids slowly. The patient was able to tolerate liquids and semisolids. Later on, she could eat solids without difficulty and was discharged.

DISCUSSION

The carotid body is a 2–6 mm, round bilateral sensory organ in the peripheral nervous system located in the adventitia of the bifurcation of the common carotid artery. They play a vital physiological role in maintaining homeostasis. Paragangliomas originate from the cells derived from the embryological neuroectoderm. There are four types of paragangliomas [1,2]: Branchiomeric group: In the region of the embryological branchiomeres (jugulotympanic ganglion, carotid body, laryngeal ganglia, subclavian ganglion, and aorticopulmonary ganglion). There is a close relationship with blood vessels. Intravagal group: In the region of the parasympathetic nerves (jugular ganglion and nodose ganglion). They have their origin within the perineurium. Aortosympathetic group: In the region of the sympathetic nerves of the aorta. Visceral autonomic group: In the nervous system of the heart, digestive tract, liver hilus, and bladder. These sensory chemoreceptors act to detect chemical changes in the body through interaction with surrounding arterial blood flow by monitoring blood gas tension and pH [7]. In a low oxygen state, the glomus cells of the carotid body become depolarized, and several excitatory and inhibitory neurotransmitters are released [8]. Located at the bifurcation of the internal and external carotid arteries, the primary functional role of the carotid body is the reflex adjustment capabilities in response to the components of arterial blood flow and physiologic oxygen sensor [8,9].

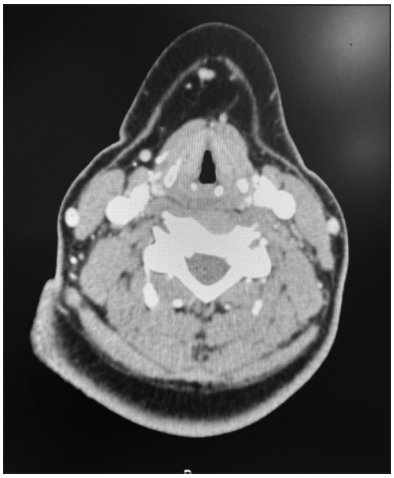

Figure 1: Contrast-enhanced angiogram showing bilateral carotid tumors

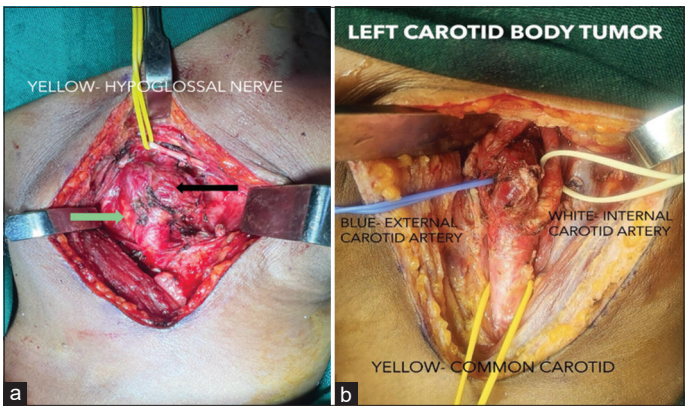

Figure 2: (a) Right carotid body tumor, green-internal carotid artery, black-external carotid artery; (b) left carotid tumor

Two types of capillary beds are responsible for the perfusion of the organ. Type I has large diameter capillaries that are fenestrated and comes in close contact with type I cells. The second type of capillary bed has narrow, non-fenestrated vessels, surrounded by pericytes, and does not make close contact with Type I or glomus cells. Blood exits the carotid body through the internal and external jugular veins [10]. It is also important to note that CBTs such as paragangliomas are frequently associated with a genetic mutation. This mutation is notable in the SDH gene. Interestingly, carotid body paragangliomas are also noted most frequently in populations who reside at higher altitudes [11].

A similar case was reported by Xu *et al*. [12] where the patient presented with a bilateral carotid tumor and was investigated and underwent surgery, but no complications were reported post-surgery. Surgery in a bilateral carotid tumor is challenging and should be done in a staged manner. In our case, there was hypoglossal nerve praxia on the right side which subsided after 1 week with steroids; on the left side, the patient developed high vagal nerve palsy which led to dysphagia and aspiration of food leading to severe cough. The mean interval between the procedures was 10.75 months [6], one had her surgery after 1 week which is the same as in our case report. In one study done by Chandra Sakaran *et al*., [13] they gave a gap of 3 months for surgery. In a study done by Johari *et al.* [14] 14.3% developed neurological abnormalities after the surgery. The size of a tumor of more than 3.2 cm is most predictive of post-operative neurological complications [14]. Another study was done by Bobadilla-Rosado *et al*. [6] showed that 3 patients had neurological complications out of those one patient had facial and two with vocal cord palsies. No neurological deficits were found after follow-ups at 2 and 8 weeks after bilateral CBT excision in the Pordal *et al*. [15] study.

High vagal nerve praxia is very rare and refers to the region of the nerve injury from the jugular foramen and could not find much data related to CBTs. It is a type of palsy where there is discoordination between the pharyngeal musculature leading

to difficulty in swallowing. Anesthesia of the larynx leads to aspiration often due to an incompetent larynx because of the disruption of sensory and motor pathways [16].

CONCLUSION

As the bilateral carotid tumor is very rare, a better understanding of the clinical characteristics of patients with bilateral CBTs may lead to more standardized and optimal management with fewer complications and a better quality of life afterward. Many more studies and further data can help prevent and anticipate neurological abnormalities.

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