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OTHER ARTICLES

Carotid Body Tumors: Institutional Experience of 10 Cases and a Review of Literature

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Abstract Carotid body tumors are slow growing neck masses that arise from the neural crest cells at the carotid bifurcation. Majority are asymptomatic and are diagnosed incidentally. Surgical excision is accepted as the treatment of choice to reduce complications. In the present series, we report 10 cases of carotid body tumors and our institutional experience. All patients underwent radiological evaluation with an ultrasonography with Doppler, contrast enhanced computed tomography and MR angiography. 6 cases were operated by a transcervical excision. The tumor was excised in tototranscervically. One of the cases required saphenous vein graft intraoperatively due to vascular injury and also had postoperative vocal cord palsy. The rest had an uneventful recovery. Carotid body tumors although rare and seemingly indolent can cause substantial symptoms if left untreated. A prompt multi modality approach is needed for both diagnosis and treatment to avoid major complications.

Keywords Carotid body tumor · Neck mass · Carotid bifurcation · Angiography · Transcervical excision

Introduction

Carotid body tumors are slow growing neck masses that were originally described by Von Haller in 1743 [1]. They arise from the neural crest cells which is located at the carotid bifurcation. The reported incidence of CBTs is 1–2 per 100,000 [2]. They account for about 0.6% of the head and neck tumors in humans [3].

The tumor is histologically composed of 2 types of cells: type I paraganglionic cell also known as chief cell and type II supporting cell also known assustentacular cell.

Type 1 cells are arranged characteristically in a pseudoalveolar pattern known as zellballen. Type 2 cells have chemoreceptor activity. Grossly, they are well circumscribed and have a pseudocapsule. The cut surface is typically solid with a smooth, rubbery. Special stains used for diagnosis include neuron-specific enolase (NSE) for paraganglionic cell and S100 for sustentacularcells [4].

Unfortunately, it is not possible to predict how aggressive a paraganglioma will be based on histology. Presence of lymph nodes and distant metastasis are the only indicators of malignant CBT [5].

Majority of these tumors are asymptomatic and are often diagnosed incidentally on clinical examination or radiological imaging. However, large sized tumors may cause pressure symptoms over adjacent neurovascular structures. The commonly affected structures include the hypoglossal nerve, vagus nerve, glossopharyngeal nerve and the sympathetic chain.

Most cases are diagnosed by clinical examination and radiological investigations. Although USG, CT and MRI are all helpful modalities, an angiography is essential to comment on the status of the carotid vasculature. Surgical excision is widely accepted as the treatment of choice to reduce complications in the form of local invasion and neurological deficit. Due to the extreme vascularity and close adherence to the carotid vessels, surgery is technically challenging and requires proper preoperative planning.The incidence of surgical complications increases with large tumors and in those that are adherent to the carotid arteries. The Shamblin classification helps in predicting the feasibility of surgery

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by taking these factors into account [6]. A type I tumor is small, localized, and can be easily separated from the adjacent artery wall. Type II lesions are larger, adherent to the artery, and partially encapsulate the vessel. Type III lesions are densely adherent to and intimately surround the carotid arteries. Whereas type I lesions are easily dissected from the artery, type II lesions usually require subadventitial dissection, and most type III lesions will require arterial excision and reconstruction.

Case Series

In the present series, we report 10 cases of carotid body tumors and our institutional protocol and management. All subjects were included in the study after getting their written informed consent. Ethical approval was waived by the local Ethics Committee of All India Institute of Medical Sciences—Bhubaneshwar, in view of the retrospective nature of the study and all the procedures being performed were part of the routine care. All 10 cases presented to the opd with long standing history of neck masses. Of them 2 patients had pressure symptoms in the form of dysphonia and mild dysphagia. A total of 3 cases were bilateral of which one had postoperative left vocal cord palsy. 2 patients denied surgery.

Demographics

In our series, the male: female ratio is 8:2. Review of literature has shown that there is no marked gender preponderance for these tumors. The average of presentation in the present study was 40.1 years. Carotid body tumors commonly present in the 4th–5th decade of life [7]. We had one paediatric patient aged 11 years with bilateral tumors (type III on right and type II on left) with no symptoms (Table 1).

Investigations

All patients underwent radiological evaluation in the form of an ultrasonography with Doppler, contrast enhanced computed tomography and MR angiography (Figs. 1, 2, 3). It has been widely accepted that preoperative vascular assessment is mandatory before planning any intervention in these tumors due to their close proximity to the carotid vessels. In addition to routine laboratory investigations, all patients were tested for serum catecholamines and 24 h urinary Vanillylmandelic acid (VMA) levels. Laryngeal endoscopy was performed in all cases to rule out any vocal cord palsy preoperatively.

Types and Laterality

7 out of a total of 13 tumors were Shamblin type II. 4 were Shamblin type III. The sole postoperative complication was encountered in a case of type III tumor. The increased vascular contact of the tumor with major vessels places them at a greater risk for both neural and vascular injury.

7 out of 13 tumors were present on the left side of neck with 3 being bilateral. We encountered one patient who had bilateral carotid body tumors in addition to a glomus

Table 1 Details of the cases

Serial no	Age,sex	Unilateral / bilateral	Shamblin type		Symptoms	Investigations	treatment	Complications	Outcomes
1	66/F	Bilateral	III	II	Swelling in neck	CECT,CEMRI,MRA	Surgical excision	Nil	Asymptomatic
2	33/M	Unilateral (right)	II		Swelling in neck	CECT,CEMRI,MRA	Surgical excision	Nil	Asymptomatic
3	52/M	Unilateral (right)	I		Swelling in neck	CECT,CEMRI,MRA	Surgical excision	Nil	Asymptomatic
4	21/M	Unilateral (left)		III	Swelling in neck	CECT,CEMRI,MRA	Surgical excision with grafting of left ICA with saphenous vein graft	Left vocal cord palsy	Asymptomatic
5	45/M	Unilateral (right)	III		Swelling in neck	CECT,CEMRI,MRA	Surgery denied	Nil	Asymptomatic
6	11/M	Bilateral	III	II	Swelling in neck	CECT,CEMRI,MRA	Surgery denied	Nil	Asymptomatic
7	40/M	Unilateral (left)		II	Swelling in neck	CECT,CEMRI,MRA	Surgical excision	Nil	Asymptomatic
8	43/M	Unilateral (left)		II	Swelling in neck	CECT,CEMRI,MRA	Surgery denied	Nil	Asymptomatic
9	42/F	Unilateral (left)		II	Swelling in neck	CECT,CEMRI,MRA	Surgical excision	Nil	Asymptomatic
10	48/M	Bilateral with right glomus jugulare	II	I	Swelling in neck	CECT,CEMRI,MRA	Surgical excision of right glomus followed by left carotid body tumor excision	Nil	Asymptomatic

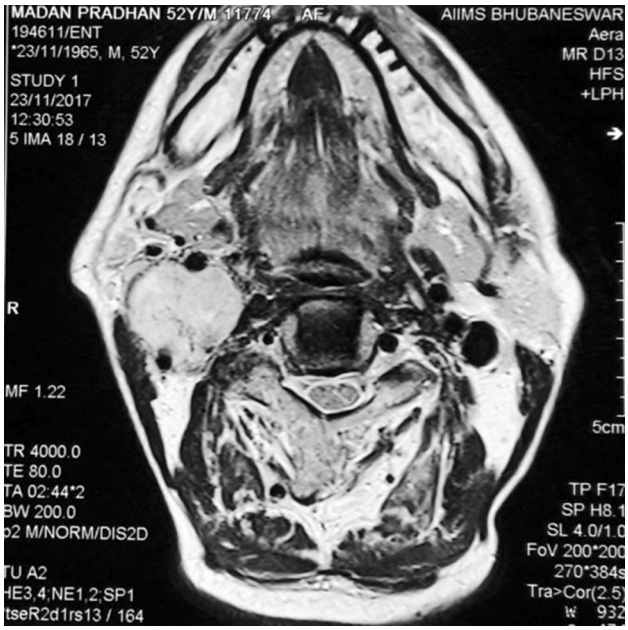

Fig. 1 CEMRI showing right carotid body tumor

jugularetumor. Excision of glomus tumor in right side was done followed by carotid body tumor on left side in a staged manner.

Surgery

6 cases were operated. All underwent transcervical excision with a cervical incision along the anterior border of the sternocleidomastoid muscle. After careful dissection, the carotid sheath was opened and control of major vessels was taken proximally as well as distally. The tumor was then excised along the subadventitial layer away from the major vessels. In all cases the tumor was excised completely. Grafting of ICA with saphenous vein graft was done in one case (Figs. 4, 5). The excised tumors were sent for histopathological examination which confirmed the diagnosis of carotid body tumor (Figs. 6, 7, 8, 9).

Complications

Fortunately most of our patients had an uneventful postoperative recovery with the exception of a 21 year old male with left sided type III tumor. The patient required saphenous vein grafting of the ICA intraoperatively. He complained of voice change on the first postoperative day and was subsequently found to have left vocal cord palsy. He was advised speech therapy and was managed conservatively. On last follow up, he did not have any significant symptoms apart from change in voice and the opposite side cord was seen to adequately compensating on laryngeal endoscopy.

Discussion

Carotid body tumors are rare head-neck neoplasms accounting for < 0.5% of all tumors; and they are the most common extra-adrenal paraganglioma [1]. They develop from epithelioid cells derived from the neural crest. The normal carotid body is located in the carotid artery bifurcation. The blood supply is usually from branches of the external carotid. Neural innervationisby afferent branches of the glossopharyngeal and vagus nerves and the cervical sympathetic ganglia. It has extremely high blood flow and oxygenconsumption.

CBTs are mostly benign with the incidence of malignant tumors being less than 10% of the total. These tumors may be functionally active (i.e., catecholamine secreting) or inactive. Carotid body tumors may be familial (10–50%) or nonfamilial i.e. sporadic and hyperplastic. The incidence of bilateral carotid body tumors is 32% in the familial group and 5% in the nonfamilial group [8]. The succinatedehydrogenase (SDH) gene subunits, succinate dehydrogenase

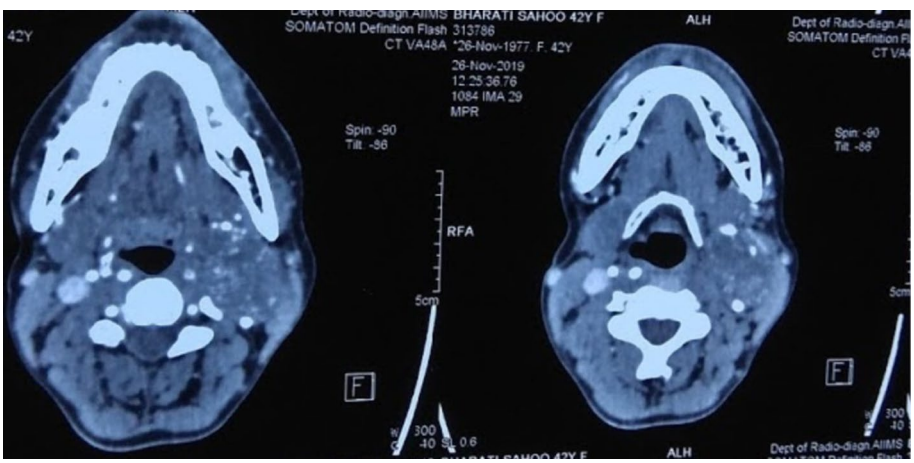

Fig. 2 CECT showing left side carotid body tumor

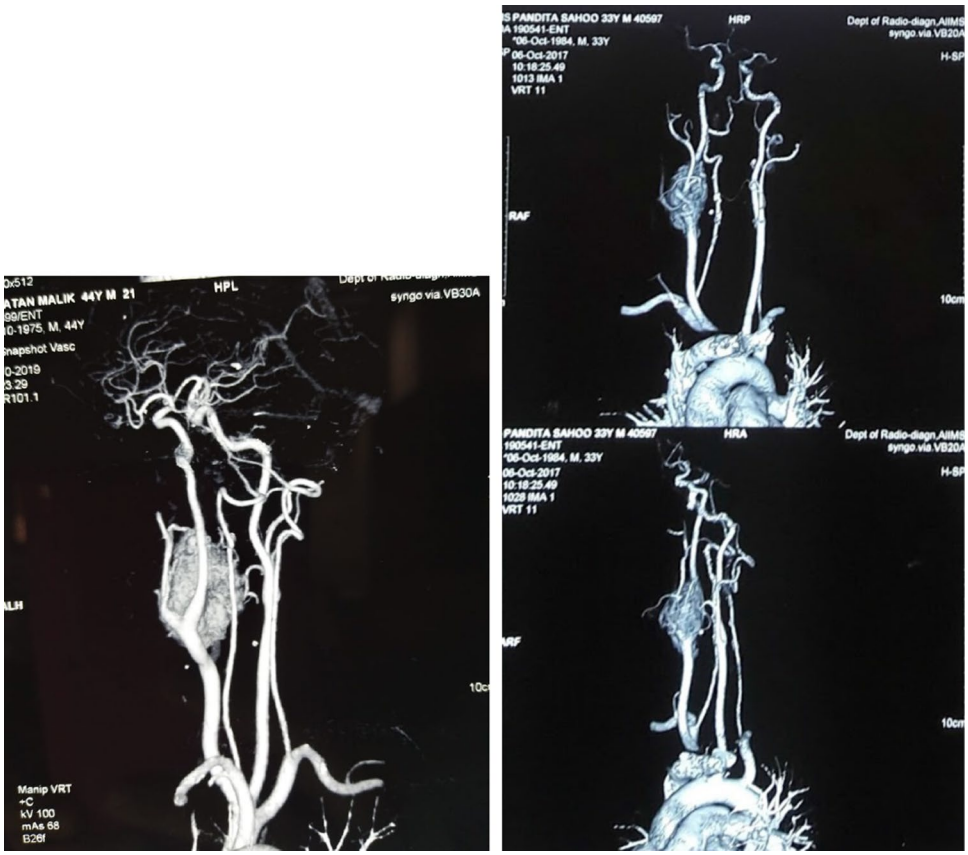

Fig. 3 DSA showing carotid body tumor and lyres sign—splaying of bifurcation of common carotid artery

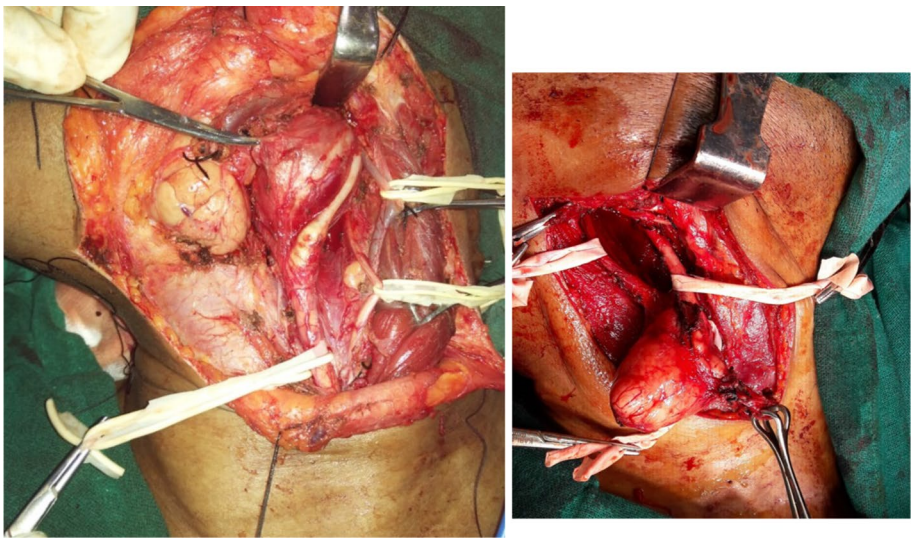

Fig. 4 intraoperative image showing right carotid body tumor exposed along withcommon carotid artery and external and internal carotid artery

complex subunit D(SDHD) and succinate dehydrogenase complex iron-sulfur subunit B (SDHB) are suspected to be responsiblefor a significant percentage of familial type paragangliomasof the head and neck [4, 8]. Familial paragangliomas can also beassociated with genetic syndromes such as Von Hippel-Lindau syndrome, neurofibromatosis type 1, multiple endocrine neoplasia(MEN) type 2A and type 2B [9].

The patient usually presents with a long duration slow growing neck swelling with or without pressure symptoms. Dysphagia, hoarseness are some common symptoms. The tumor usually causes compression of various cranial nerves leading to these symptoms. The hypoglossal, glossopharyngeal, vagus nerve and the sympathetic chain are commonly affected. Functional catecholamine secreting tumors may have symptoms like tachycardia, hypertension, palpitations,

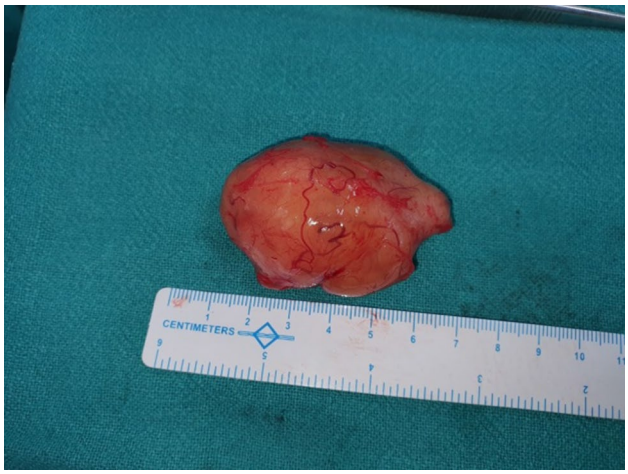

Fig. 5 Surgical specimen showing completely excised carotid body tumor

facial flushing and headaches. In such cases, serum catecholamine levels are urinary VMA needs to be assessed.

The presence of a slow growing, painless neck mass presenting in the lateral side neck anterior to sternocleidomastoid at the angle of mandible at the level of the hyoid,which is pulsatile on clinical examination raises the suspicion of a CBT. Differentials include branchial cyst,parotid gland tumor, carotid artery aneurysm, lateral aberrant thyroid gland, lymphoma,neurofibroma, cervical lymphadenitis, and metastatic carcinoma.

Biopsy is contraindicated because of the extremely vascular nature of the disease.

Radiological investigations usually help in confirming the diagnosis. Doppler ultrasonography, contrast enhanced CT, MRI scan and MR angiography are the investigations routinely done.

On CECT, the tumor appears as an avidly enhancing hypervascular mass located at the carotid bifurcation. The ECA is splayed anteriorly and the ICA posteriorly. This characteristic splaying is termed "Lyre's sign". MRI shows serpentine or punctate vascular high and low flow voids ("salt and pepper") corresponding to the vascular structures on T2 sequences. Angiography done shows the ascending pharyngeal artery to be the most common arterial feeder [10]. It may also show arterio-veneous shunting.

The role of preoperative embolization is controversial. If done it should be 24–28 h prior to surgical intervention to avoid formation of collaterals [11]. Tumor volume and vascularity can be reduced by effective embolization. The risk of ICA thrombosis, operator dependence and lack of widespread availability are some of its demerits. We did not preoperatively embolize any tumor.

Surgical excision of the tumor has been widely accepted as the primary treatment. The first successful excision of a carotid body tumor was performed by Albert J Van Der Bogt in 1889 [12]. In our series too all consenting patients were planned for surgery. Although the surgical approaches may differ according to surgeon choice, expertise, size of tumor-etc the most critical aspect is careful dissection and preservation of adjacent neurovascular structures.For this, accurate dissection with excision of the tumor should be performed along the subadventitial plane or "white line" as suggested by Gordon-Taylor [13]. All of our cases were operated by a transcervical incision.(fig) along the anterior border of the sternocleidomastoid muscle. After careful dissection, the carotid sheath was opened and control of major vessels was taken proximally as well as distally. The tumor was then

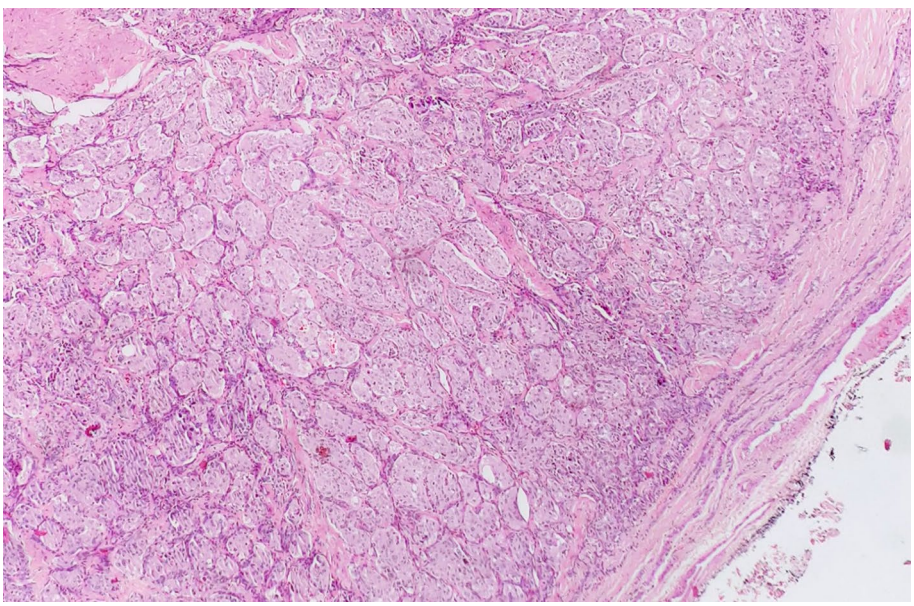

Fig. 6 The tumor is completely encapsulated. It is composed of cells arranged in nests and separated by fibrovascular septae (H & E 40x).

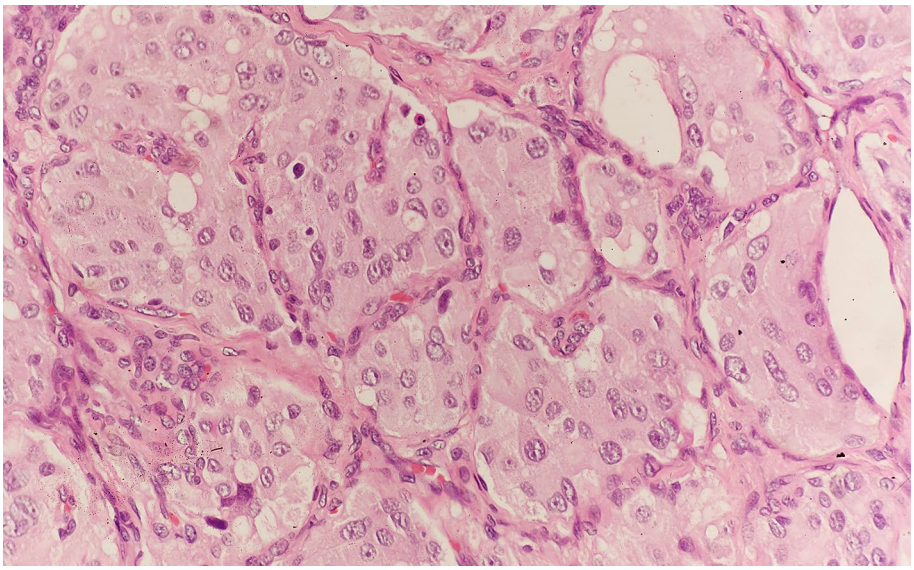

Fig. 7 Nests of tumor cells separated by fibrovascular separate (Zell ballen pattern). The cells have abundant eosinophilic granular cytoplasm. The nuclei are round, centrally located and show granular chromatin. occasional cell show anisonucleosis (H & E 400x).

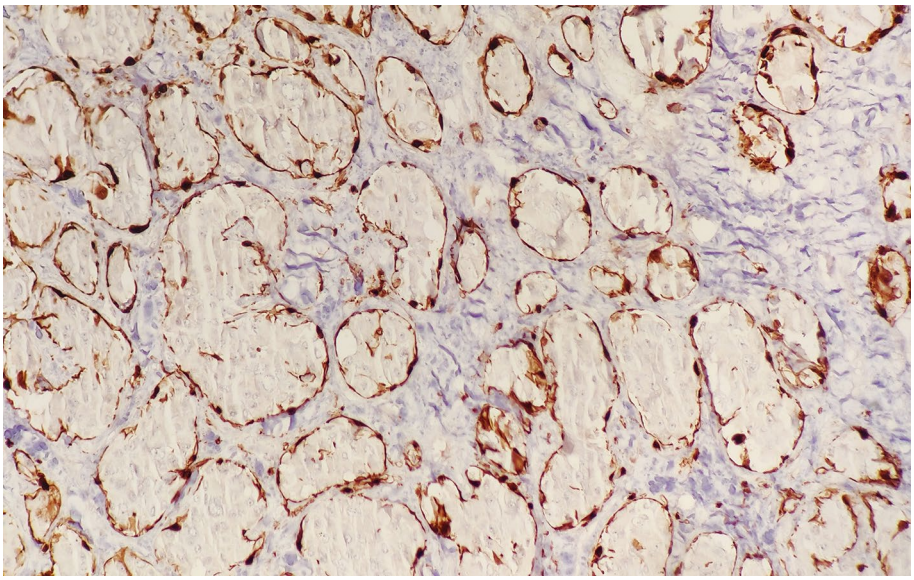

Fig. 8 Immunohistochemistry for chromogranin shows diffuse strong cytoplasmic positivity in the tumor cells. IHC stain (400x)

excised along the subadventitial layer away from the major vessels. In all cases the tumor was excised completely.

Postoperative complications like stroke, cranial nerve injury, and bleeding have been reported in literature.However recent studies have shown a less than 5% risk of stroke post CBT excision [14]. The risk of cranial nerve injury in the postoperative period remains high ranging from 20 to 50% in reported literature [15]. Dysphagia and change in voice are common. In the current case series also we had one case of post operative vagal palsy. While around80% of complications resolve, 20% may be permanent [16].

Intraoperative bleeding is a potentially life threating risk and someauthors have recommended standardizedICA shunting during excision in order to cut off the vascular supply of ECA, reduce the size ofthe tumor and to ease resection in difficult cases [17]. One of the cases in our study had intraoperative ICA injury for which a saphenous vein grafting was done.

Rate of recurrence in literature is about 6% [18]. None of our operated cases have had recurrences till last follow up.

Rate of complications can be predicted to some extent by theShamblin typing due to the increasing proximity of major neuro vascular structures.

Conclusion

Carotid body tumors although rare and seemingly indolent can cause substantial symptoms if left untreated. As seen in literature the risk of major neurovascular injury increases

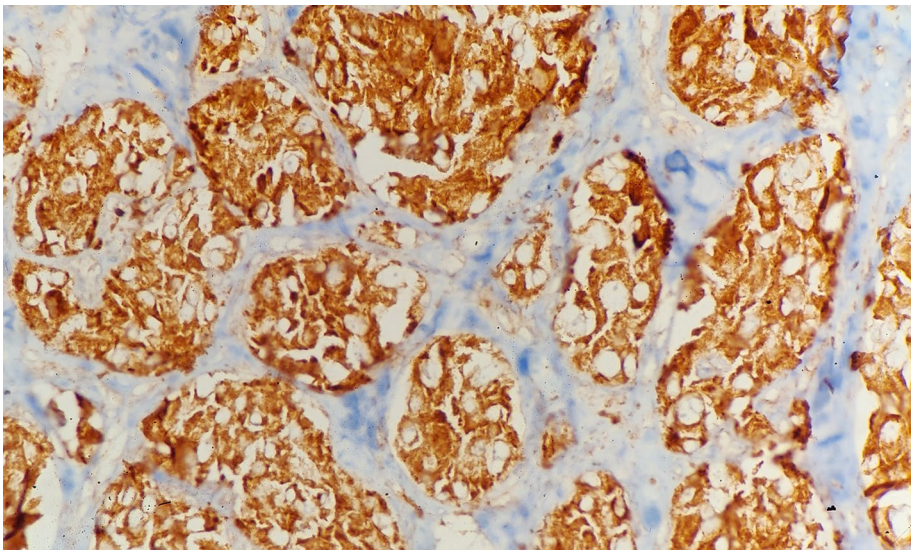

Fig. 9 Immunohistochemistry for S-100 shows positivity in the sustentacular cells surrounding the tumor cells. The tumor cells are negative for S-100. IHC stain (200x)

as the size of the tumor increases, it is therefore imperative to diagnose them early. A multimodality approach with the radiologist and vascular surgeon may be needed in some cases. Most patients will require a long follow up.

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Declarations

Conflict of interest All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

Ethics Approval Our study was conducted on human participants in accordance with ethical standards as laid down in the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards. Ethical approval was waived by the local Ethics Committee of All India Institute of Medical Sciences—Bhubaneshwar, in view of the retrospective nature of the study and all the procedures being performed were part of the routine care.

Patient Consent Prior written consent was obtained from all the patients.

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