Glomus Caroticum - Our Experience in a Tertiary Care Center

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ABSTRACT

BACKGROUND

Carotid body tumour is most common paraganglioma in the neck with an incidence of 1:30000. We wanted to determine the optimal management of carotid body tumour to provide the best outcome in patients.

METHODS

We conducted a retrospective study of the records of 8 patients who presented with carotid body tumours at the Department of Surgical Oncology, Vydehi Institute of Medical Sciences, Bengaluru, India. Epidemiologic, aetiologic, diagnostic, and therapeutic features were evaluated in this multidisciplinary study.

RESULTS

The average age of patients was 38 years with 7 females (87.5%) & 1 male (12.5%). The location of tumour was on the left side of neck in all patients. Out of the 8 cases, 7 had only carotid body tumour, and one had history of tympanic paraganglioma. 1 case had hypoacusis due to previous surgery. A slow-growing neck mass was the main clinical presentation. One of the patients had deviated left tonsillar fossa (on ENT examination). CT angiography in 6 cases showed homogenous (5) and heterogenous (1) enhancing lesion with splaying of ECA and ICA (Lyre sign). MR angiography 3 (37.5%) T1 hypointense and T2 iso-hyperintense enhancing soft tissue mass at carotid bifurcation. The clinical presentation and imaging results strongly suggested the diagnosis of carotid paraganglioma in all cases. Treatment was surgical excision in all cases associated with a preoperative embolization in 2 case and a pre-operative radiotherapy in 1 case. Pathology confirmed the diagnosis.

CONCLUSIONS

Carotid body tumour (CBT) requires early diagnosis and an adequate multidisciplinary team. A high suspicion of index must be considered in the case of any pulsatile lateral cervical mass. Surgical resection is the treatment of choice for carotid body tumour, other neo-adjuvant therapies such as embolization and radiotherapy may only assist stabilization of the tumour, particularly with cranial extension, bilateral or multifocal tumours. Risks associated with tumour excision are considerable, especially with the Shamblin III group, hence the neo-adjuvant modalities of treatment may be considered in these tumours. The therapeutic indication should, ideally, be set in a multidisciplinary consultation.

KEY WORDS

Carotid Body Tumour, Glomus Caroticum, Paraganglioma, Multidisciplinary Approach

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BACKGROUND

Paragangliomas are vascular neoplasms, that arise from widely dispersed characteristic neural crest chromaffin cells that are associated with autonomic ganglia and may have the ability to secrete catecholamines.[1,2] They can derive from either parasympathetic or sympathetic ganglia, and the clinical features vary by its type of origin. Parasympathetic ganglia-derived paragangliomas are located almost exclusively in the head, neck, and skull base, arising most often from the carotid body and jugulotympanic paraganglia. They are non-functional.[2,3,4]

Carotid body tumour is a rare disease. Its incidence is 1:30000.[1,2] The carotid body tumour was first described by Von Haller in 1743.[1] is the only known pathologic condition of the carotid body.[1,2] Carotid body tumour is hypertrophy of the carotid body tissue, which is the most common type of paraganglioma. It is also known as, glomus caroticum.[5,6] Chemodectoma, Perithelioma, and Non-chromaffin paraganglioma.[7] Carotid body tumours may be familial in 10%.[1] They are slow-growing and benign. Approximately 5-10% CBT may progress to malignancy,[7] with local vascular and lymph node invasion and rarely metastases.[2,8]

We wanted to determine the optimal management of carotid body tumour to provide the best outcome in patients.

METHODS

All patients of carotid body tumour, diagnosed and operated at Vydehi Institute of Medical Sciences and Research Center from 2016 to 2019 were included in a CASE SERIES Descriptive type study. Written informed consent for surgery and embolization from all patients was obtained. Multidisciplinary team assessment and combined decisions were taken as per the need of individual cases.

RESULTS

The average age of patients was 38 yrs. (21 yrs. – 55 yrs.) with 7 females (87.5%) &1 male (12.5%). In a meta-analysis of 4418 patients, the mean age was 47 years, with the majority being female (65%).[1]

The location of the tumour was on the left side of the neck in all patients. Out of the 8 cases, 7 had isolated carotid body tumours and one had a history of Tympanic paraganglioma surgery. 1 case had hypoacusis due to previous surgery. On clinical examination all the cases had a firm non-tender neck swelling varying in size (2*3 cms - 5*4 cms) compressible (Sign of Recluse and Chevassu).[2,4]

All the swellings had transmitted pulsations and were horizontally mobile (Fontaine sign). One of the patients had deviated left tonsillar fossa on ENT examination. 2 out of 8 patients underwent preoperative embolization of feeding ECA with PVA particles, 7 days & 48 hrs. before surgery in 2 settings in one case & 48 hours before surgery in another.

Tumours were classified into the Shamblin groups, 6 Cases (75%) were group II & 2 cases (25%) group III. Complications as mentioned occurred in group III patients.[9,10]

Presenting Symptom	No. of Patients
Painless Neck Mass	7 (87.5%)
Transient Ischaemic Attack/Dizziness	2 (25%)
Headache	1 (12.5%)

Table 1. Tumour Presentation

Investigations	Number of Patients	Findings & Report
Doppler ultrasound	8 (100%)	Well defined hypo/hetero/ mixed echoic lesion at carotid bifurcation with significant vascularity
CT Angiography	6 (75%)	Homogenously (5) and heterogeneously (1) enhancing lesion with splaying of ECA and ICA (lyre sign)
MR Angiography	3 (37.5%)	T1 Hypointense and T2 ISO- hyperintense enhancing soft tissue mass at carotid bifurcation

Table 2. Radiological Investigations Done

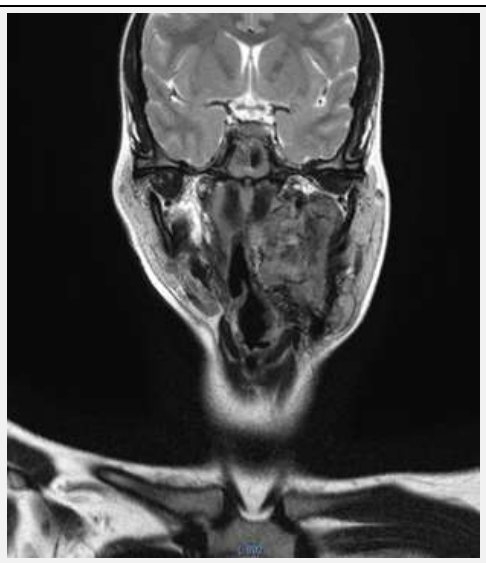

Figure 1. T1W Hypointense Lesion Abutting Left Phyrangeal Wall

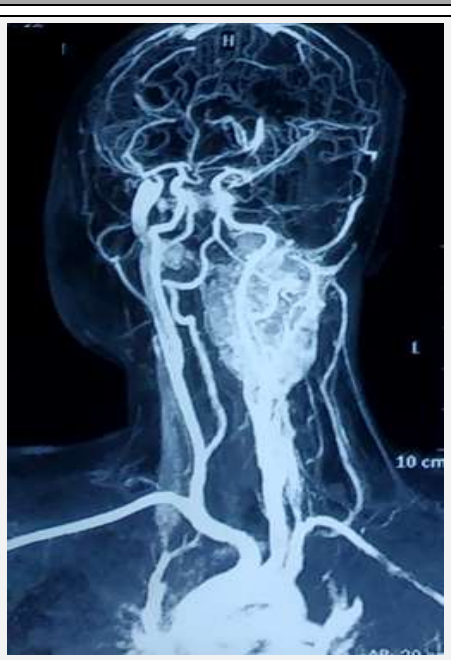

Figure 2. Lyre Sign

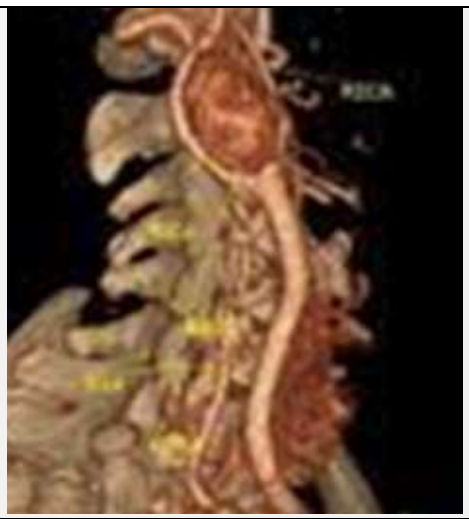

Figure 3. Carotid Body MRA Reconstruction Image

Figure 4. Peri Adventitial Plane of Dissection

Figure 5. Carotid Body Separated from Internal Carotid Artery

Figure 6. After Resection of Carotid Body Tumour

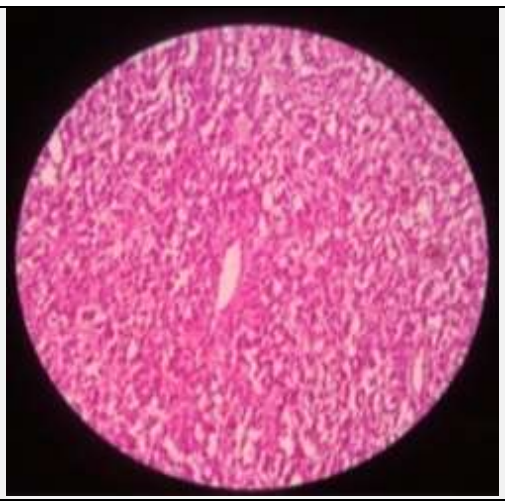

Figure 7. No Differentition between Supporting Cell and Chromaffin Cells 40X

Surgical Technique

In all 8 pts modified Aryans/modified Shobinger incision was employed to expose the carotid sheath and its structures. Meticulous dissection of the tumour was performed from the vessels. The tumour was dissected in the periadventitial plane and resected completely in all cases.

Depending on the local extent of the tumour further surgical technique was differed. All patients underwent complete surgical resection without any obvious nerve injury and with ligation of ECA in 3 patients, in 1 patient IJV was sacrificed. In one case vascular repair with Prolene 4-0 was done for a tangential tear in ECA. A drain was placed for all cases and removed on the 3rd to 5th post-op day. The average intraoperative blood loss was 300 ml (200 ml to 700 mL). One patient was transfused 1 packed RBC postoperatively.

The median time of hospital stay was 14 days (10- 24 days). Pathology confirmed the diagnosis of paraganglioma in all cases, 1 showing high-grade lesion. The postoperative period was uneventful in 6 patients, 1 case had unilateral vocal cord paralysis which was treated by medialization of the cord by fat injection, another case had dysphonia and deviation of the tongue (which was thought due to over handling of the hypoglossal nerve), Dysphonia was improved on 3 months follow up. The patient with high-grade CBT was advised for post-operative Radiation Therapy but didn't agree to adjuvant treatment. Patients were followed up on POD 10, 3 months, 6 months post-op & every 6 months for the first 2 years, thereafter annually. Out of 8 patients, 7 were on regular follow up. 1 patient visited once.

DISCUSSION

Glomus caroticum is a paraganglioma originating from paraganglionic bodies of autonomous nerve systems of embryological neural crest cells.[2,6,7] They are the largest mass of chemoreceptor tissue in the body. It is located in the periadventitia at the posterior surface of the carotid bifurcation.[1,2,5,8]

There are four types of paraganglioma as described below.[2,1]

- Branchiomeric group: In the region of the embryological branchiomeres (jugulotympanic ganglion, carotid body, laryngeal ganglia, subclavian ganglion, aorticopulmonary ganglion). There is a close relationship with blood vessels.
- Intravagal group: In the region of the parasympathetic nerves (jugular ganglion, 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. (Image 1)

Jugulotympanic paragangliomas are slow-growing lesions that usually present with pulsatile tinnitus with or without conductive hearing loss.[11,12] In one study 90 percent of patients with a jugular foramen paraganglioma had pulsatile tinnitus and 81 percent had hearing loss. In patients with a jugular paraganglioma, there may be facial nerve paralysis, vertigo, hoarseness, and paralysis of lower cranial nerves. An examination may show a bluish pulsatile mass behind the tympanic membrane.[12]

Vagal paragangliomas can occur at any point along the course of the cervical vagal nerve, but usually arise from the inferior ganglion, thus resulting in a wide variety of clinical symptoms including dizziness, blurred vision, facial droop, dysphagia, neck mass, pain, cranial nerve deficits, or Horner syndrome.[13] Rarely, cervical paragangliomas can also arise from or in proximity to the thyroid gland.[1,13]

Paragangliomas that are located within the dura can also present with symptoms of neurologic compression.

CBT is more common in women than in men.[3,14] In our study it was similar. (F:M::7:1) CBT are classified as sporadic (most common), familial (10%) and hyperplastic.[2]

Hyperplastic variety occurs in patients exposed to prolonged hypoxia, people living in high altitudes, and COPD.[2,6] In the literature, it is mentioned that CBT may occur at any age with a high frequency between 30-40 years[15] and was 38 years in our study series. CBT usually presents as a painless, slow-growing neck swelling lateral to the tip of the hyoid bone.[3]

In later stages, dysphagia, deficits of cranial nerves VII, IX, X, XI and XII, and hoarseness or a Horner syndrome may result from pressure on the vagus or sympathetic nerves.[1,2,11] Physical examination discloses a rubbery non-tender mass in the lateral neck that is more freely movable in the horizontal plane than vertically, referred to as a positive Fontaine's sign.[1] There may be a carotid bruit or the tumour may be pulsatile.

Transient ischaemic attack/ dizziness & coughing are due to pressure on the vagus or sympathetic nerves.[4] In our series, most of the patients presented with a painless neck mass and few with other symptoms like TIA/dizziness and headache.

However, some patients may present with hypertension, sweating & headache (possibly due to vasoactive catecholamine production), although the secreting paraganglioma is rare (< 3%), these signs must be kept in mind.[14]

A thorough ENT examination is necessary,[15,5] to look for features like para-pharyngeal extension, laryngeal paralysis, eustachian tube malfunction, jugular tympanic location.[9,3,5]

For the skull base and neck paragangliomas, the initial evaluation may include ultrasound or cross-sectional imaging (CT or MRI). On ultrasound, a carotid body tumour typically presents as a solid, well-defined, hypoechoic tumour with a splaying of the carotid bifurcation.[15,16] Duplex sonography typically indicates the mass to be hypervascular, although the absence of hypervascularity does not exclude the diagnosis.[3,14]

Although duplex ultrasound is the first line of imaging modality,[1] CTA/MRA can have added advantage since they have supplanted conventional angiography. The classic CT findings of a paraganglioma at any site include a homogeneous mass with unenhanced Hounsfield units in the 40 to 50 range.[17] There is intense enhancement following the administration of intravenous contrast and delayed washout. Cystic changes, necrosis, and internal calcifications are commonly described.[18,17]

In our series 6 pts underwent Computed Tomographic Angiography (CTA) that showed homogenously solid mass, fast and avid enhancement of contrast except one case showing heterogeneity. MRA shows features that are characteristic of paraganglioma. The typical smooth contour, signal characteristics, and location of paraganglioma confirms the radiological diagnosis. On T1-weighted MRI, paragangliomas typically have a background tumour matrix of intermediate signal density, with scattered areas of a signal void, reflecting high-flow blood vessels.[13] On T2-weighted images, an intense hypervascular appearance is present with classic "salt and pepper" appearance in most lesions larger than 1.5 cm, reflecting signal voids intermixed with regions of focally intense signal intensity.[13,19]

In our study, MRA was done in 3 patients, one of which showed 1 the classic “salt and pepper” appearance, salt is high signal foci of hemorrhage or slow void, and pepper low signal flow void.[20,7,8]

Surgical resection is the treatment of choice for carotid body tumour. Advanced cases with intra-cranial extensions or difficult surgeries may be assisted by Embolisation of feeding vessels and or radiation therapy prior to surgery for achieving R0 resection with minimal compiications. Definitive Radiation Therapy may be considered in cases that are in-operable, with a very high risk of cranial nerve injury or bilateral CBT where chances of vagal nerve injury may be detrimental to patients.

In our series one case had undergone excision of glomus tympanicum 6 yrs back with 20 dB of hearing loss. When multiple paragangliomas are present, a whole-body scan should be considered if the lesion is a high grade on histopathology and metastasis should be checked. In a retrospective study,[21,19] ninety percent of the body regions that were falsely negative on MIBG and metastatic lesions were detected by FDG-PET. 30 patients with *SDHB*-related paragangliomas (29 with metastatic disease) reported 100 percent sensitivity (per patient) for detection of metastases with FDG-PET, versus 80 percent for iobenguane I-123 scan and 88 percent for 18F(FDOPA)-PET. [21,18,22] If a patient has a skull-base or neck paraganglioma, tests for mutations in

the following genes should be ordered*:* SDHD*,* SDHC*,* SDHB, SDHAF2*,* and SDHA.[18,22] If genetic testing reveals inherited gene is only active from the father (maternal imprinting) then SDHD and SDHAF2 should be ordered first. If a mutation is identified at any point in the investigation, no further testing should be performed, all first-degree relatives should be offered germline mutation testing for the known mutation.[10,22] Additional evaluation for patients with an identified mutation or syndromes like VHL syndrome, MEN2, and NF1 must be considered.[1,2]

In the study by Youssef et al of 10 patients, 2 pts underwent partial resection due to cranial extension.[15]

In our study, all patients underwent complete resection, while one with cranial extension was subjected to embolization followed by surgery. None of the patients in our study had a stroke or post-operative deficits. In a study, surgical resection before expansion toward the base of the skull reduces complications as every 1-cm decrease in the distance to the skull base results in 1.8 times increase in >250 mL of blood loss and 1.5 times increased risk of cranial nerve injury.[23]

CONCLUSIONS

Carotid body tumour must be considered as a possible diagnosis in patients presenting with progressive painless pulsatile lateral neck swelling. A complete pre-operative evaluation and multidisciplinary team involvement are important for optimal surgical outcomes. Surgical resection is the treatment of choice for carotid body tumour. Other therapies prior to surgery, such as embolization and radiotherapy (45 Gy) may only assist stabilization of the tumour, particularly with cranial extension, bilateral or multifocal tumours. Risks associated with tumour excision are considerable, especially with the Shamblin III group; hence, the neo-adjuvant modalities of treatment may be considered in these tumours.

Definitive Radiation Therapy may be considered in cases that are inoperable.

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