ORIGINAL RESEARCH PAPER

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KEY WORDS: Carotid body, paraganglioma, parasympathetic chain.

CAROTID BODY TUMOR: A RARE CASE REPORT

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ABSTRACT

In the head and neck region, the normal paraganglia are associated with the parasympathetic nervous system and paragangliomas arising from these sites account for up to 70% of extra adrenal paragangliomas. The most common site is the carotid body. They arise at the bifurcation of internal and external carotid arteries. Flow voids from the numerous vessels are typically seen on MR imaging and this finding is part of the classic "salt and pepper appearance" seen on T2 weighted images. Histologically they have a characteristic "Zellballen" growth pattern. Surgical resection is the treatment of choice. Here we present a case of 40 years, Female presenting with a swelling in the right side of neck for last 5 years. It was firm in consistency, non-tender and non-pulsatile. CEMRI showed well defined lesion in right carotid space extending from C2 to C5 vertebra splaying the carotid bifurcation- suggestive of paraganglioma – carotid body tumor. FNAC of swelling was suggestive of- soft tissue spindle cell neoplasm. Patient was operated, excision of tumor mass done. Intraoperative and postoperative period turned out to be uneventful. Patient is now on follow up.

INTRODUCTION

In the head and neck region, the normal paraganglia are associated with the parasympathetic nervous system and paragangliomas arising from these sites account for up to 70% of extra-adrenal paragangliomas(1) Carotid paragangliomas are the most common type of head and neck paragangliomas.

History- Von Luschka first described a tumor of the carotid body in 1862. The first successful removal of a carotid body tumor in the United States was reported by Scudder in 1903(2).

Anatomy and physiology- The carotid body is located in the adventitia of the posteromedial aspect of the bifurcation of the common carotid artery. The normal carotid body measures 3 to 5 mm in diameter. The average weight of the normal adult gland is 12 mg. The carotid body has a chemoreceptor role by modulating respiratory and cardiovascular function in response to fluctuations in arterial pH, oxygen and carbon dioxide tension.

Etiology – It appears to be multifactorial, but most are solitary. Genetic mutations responsible for the hereditary form have been identified in SDHD, SDHB and SDHC genes(3). Hereditary paraganglioma syndrome has been classified genetically into 4 entities: PGL1, PGL2, PGL3 and PGL4. PGL1 is associated most commonly with head and neck tumors. It is estimated that 86% of individuals with a gene mutation will develop a tumor by age 50 years(4).

Clinical presentation and diagnosis- Majority of them are slow growing, benign lesions. It has a median increase in dimension of 0.83mm/yr and a median tumor doubling time of 10 years. Many tumors are pulsatile by transmission from the carotid vessels. The consistency varies from soft and elastic to firm and are generally nontender. Some 10% patients present with neurological symptoms, most frequently affecting the X nerve. Although all of them have neurosecretory granules, only 1% to 3% are considered functional. Malignancy is rare in head and neck paragangliomas. National cancer database report shows that the rate of malignancy in paragangliomas is approximately 10%(6).

Classification: In 1971, Shamblin et al. described a classification system used to grade the difficulty of resection of carotid body tumors(10). It is a radiologic classification into 3 types based on the maximum degree of circumferential contact of the tumor with the Internal carotid artery.

Investigations:

Endocrine investigations: β blockade should be ceased for at least 7 days. Combined 24 Hr urinary collection of metanephrine, norepinephrine, epinephrine and dopamine can be done.

Radiological investigations: CT scan with intravenous contrast will demonstrate a hypervascular mass with avid contrast enhancement, at carotid bifurcation, which splays the internal and external carotid arteries. MRI with gadolinium contrast may be the most useful study in evaluation. It is sensitive for tumors as small as 0.8 cm. Tumors larger than 2cm in diameter demonstrate "salt and pepper pattern" seen on T2 weighted MR images(not always present in carotid paragangliomas). A characteristic "Lyre sign" is seen as a bowing and displacing of the internal and the external carotid arteries(9). Radionuclide techniques help in whole body screening for occult disease and post operative assessment.

MIBG scanning depends upon the uptake of radiolabelled material by the APUD system. F-DOPA-PET identifies neuroendocrine cells due to their uptake of dopamine and is the most sensitive test.

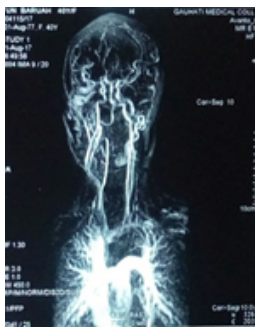

Fig 2. MRA showing splaying of right Internal and external carotid arteries.

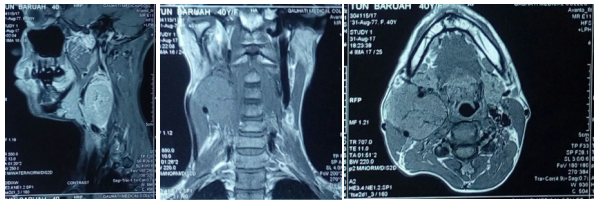

Fig 3,4,5- Showing MRI Sagittal, Coronal and Axial views of tumor mass respectively.

HISTOPATHOLOGY:

Tumors are encapsulated by a dense fibrous pseudocapsule. They demonstrate same cell types as normal paraganglia, type I- which have the capacity for synthesis and storage of catecholamines and type-II which are sustentacular cells. They have a rich density of somatostatin receptors and tissue is rich vascularized. It is characterized by "Zellballen appearance" a nested arrangement of type I cells, surrounded by fibrovascular stromal tissue(5)

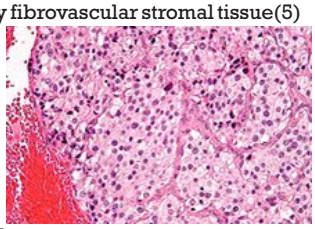

Fig 6. Zellballen appearance

Management: In 1970's radiotherapy was mainstay of treatment. Surgery is the treatment of choice at present. Structures most at risk in the procedure are Hypoglossal nerve and Superior laryngeal nerve. Vagus nerve and External carotid artery may need to be ligated to facilitate resection. Post operative results are generally good.

Preoperative Photos

CASE REPORT

CHIEF COMPLAINTS

A 40 years old female came with swelling in the right side of neck for last 5 years. It was gradual in onset, size increased progressively. No history of tenderness, no difficulty in swallowing or breathing, no history of change in voice. No history of sweating or palpitations.

PHYSICAL EXAMINATION

The swelling was oval shaped, size 4x4x3 cm(approx), firm in consistency, non tender, it was mobile horizontally but less mobile vertically. Swelling was non pulsatile. Cervical lymph nodes were not palpaple. Ear, nose, throat examination was found to be normal. No other systemic examination findings were present.

DIAGNOSIS

Patient underwent CEMRI of the neck swelling which was suggestive of carotid body tumor. FNAC of swelling showed possibility of spindle cell neoplasm. Rest routine blood investigations were found to be normal.

TREATMENT

Patient underwent excision of the tumor mass, intraoperative findings showed tumor belonging to shamblin II classification. Intraoperative and postoperative period turned out to be uneventful.

INTRAOPERATIVE PHOTOS

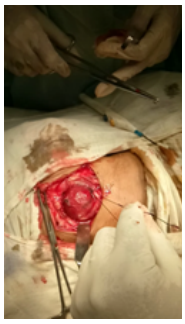

Fig 6- Common Facial vein being ligated

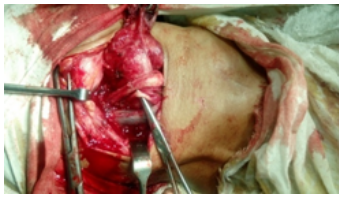

Fig 7 Vagus nerve identified (above internal carotid artery and below internal jugular vein).

Fig 8 Tumor mass being removed (size- 4x4x5 cm)

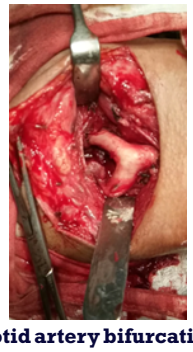

Fig 9 Common carotid artery bifurcation

POST OPERATIVE PHOTOS

DISCUSSION

- The term paraganglion was first used by histologist Kohn in 1903 to describe the carotid body. This term was most appropriate because cells of the carotid body originate from the neural crest crest and migrate in close association with autonomic ganglion cells, hence the name "paraganglionic"(7).
- It usually presents as a lateral cervical mass, which is mobile laterally but less mobile in the craniocaudal direction because of its adherence to the carotid arteries. This physical finding has been called a positive Fontaine sign(1). Here in the present case this finding was found to be present.
- In a study done by Choakchai et al. in 2016, 40 cases were taken, Carotid Angiography was done in 29 cases and CT scan, MRI and MRA was done to detect the widening of carotid bifurcation(8). In this case, CEMRI was done and splaying of carotids was identified.
- A study done by Arya S et al in 2008 evaluated a study on 10 cases to establish a criteria to accurately predict the Shamblin group on preoperative MR imaging for a uniform reporting system(9).
- In a study done in 90 cases of carotid body tumors by Shamblin et al. in 1971, classification of tumors was done and the overall surgical mortality rate was found to be 5.7%. In this case, intraoperatively it was found to be shambling classification type II. Postoperative period was uneventful(10).
- In a Review literature of Patlola R. et al in 2010, mentioned that surgical advances have greatly decreased the mortality rates, but the morbidity rates secondary to cranial nerve injuries remain high. In the present case no postoperative cranial nerve palsy was seen(11).

CONCLUSION

Our above case report shows us that carotid body tumors are a slowly progressive benign lesion which can be excised based on intraoperative findings. However, an extensive preoperative investigation and histopathological assessment is necessary prior to excision of mass.

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