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Carotid Body Paraganglioma A Rare Disease

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Abstract

This study was aimed to evaluate the management of carotid body paraganglioma, the best approach to have the diagnosis and to classify them on the basis of Shamblin's classification. A series of 5 cases reporting to the department in 4 years was taken and retrospectively analysed. One of them was having large bilateral pulsatile mass with bruit and few neurological symptoms like hoarseness of voice. One of the patient was diagnosed during routine investigation as she was having small nodular swelling right side of neck but otherwise asymptomatic. FNAC was diagnostic which was advised as in any other nodal mass. Rest three cases were also in type II but had only localized symptoms. All cases were investigated with CT scan and colour Doppler studies. Three cases were operated, one was referred and another one being asymptomatic was not ready for any surgical intervention. The outcome was evaluated. There were no major complications or recurrences seen.

Keywords - ***CBT, Paraganglioma***

Introduction

Paragangliomas are slow growing neuroendocrine tumours that commonly develop in head and neck, thorax and abdomen. They are either single or multicentric. As per literature about 75% are sporadic while 25% are hereditary which are likely to be multicentric and develop at younger age. They are highly vascular, deep red, rarely malignant and biochemically active.[1]

Paraganglionic cells are neural crest derived cells that can be found from base of skull to the pelvis. These cells are part of diffuse endocrine system and are associated with sympathetic nerves. They are usually found along the major vessels and vagus nerve in head and neck.[2] Specific sites are the carotid body, jugular bulb, tympanic plexus, vagus nerve, supra and infraglottic regions and they are named accordingly like carotid paraganglioma, jugular paraganglioma etc.[3]

The paragangliomas are more prevalent at high altitude regions, which are >2000 m from sea level. The most common paraganglioma is carotid paraganglioma followed by jugulotympanic ones. Head and Neck paraganglioma presents as a slow growing painless neck mass. Diagnosis is usually late because they are relatively rare and asymptomatic.[4] Age of onset of the CBT may be helpful in determining its familial origin. Occurrence of multiple tumours also strongly suggest that it is familial.

Carotid body paraganglioma are rare neoplasms. They comprise 65% of head and neck paragangliomas, which develop within adventitia of medial aspect of carotid bifurcation. Carotid body originates from the neural crest and has an important role during the body's acute adaptation to fluctuating concentration of oxygen, carbon dioxide and PH. Carotid body protects the organs from hypoxic damage by releasing neuro transmitter, that increases the respiratory rate when gets stimulated.[5,6]

It is a middle age disease and mean age of onset is 45years though it may occur in children also. 10-50% are inherited cases where age of onset is 20-40 years and disease may be bilateral (5% cases). Familial tumours are more common in CBT as compared to other paragangliomas. It is the disease of high altitude due to chronic hypoxia.

CBT is of 3 types[4,7]

1) Familial - 10-50% of total cases, common in younger age (20-40yrs), >5% are bilateral
2) Sporadic - Most common type, 85% of CBT
3) Hyperplastic - Common in patient with chronic hypoxia including

a) Patients living at high altitude at about 5000 feet above the sea level

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b) Patients suffering from COPD
c) In cases of cyanotic heart disease

Shamblin's classification –Based on relationship of tumour and carotid wall infiltration they are classified into Type I, Type II and Type III.[8]

Type I - Small sized tumour <5cm without encasement of vessel wall. There is no widening of carotid bifurcation. This can be excised easily.
Type II -Tumour attached to the wall of carotid, involving adventitia but no encasement of vessel.
Type III - These tumours are located inside the blood vessel with encasement of vessel wall, these are larger in size >5cm and cause widening of carotid bifurcation.

Pathophysiology - There are 2 main etiological factors responsible[9] -

1) Familial which has genetic predisposition
2) Hypoxic stimulation.

Familial cases are genetically heterogenous. Currently 4 genes are identified, the first 3 genes encode the subunits of enzymes Succinate dehydrogenase complex which is the part of Kreb's cycle. Defective succinate dehydrogenase has been postulated to cause increase in intracellular concentration of molecular hypoxia mediator and vascular endothelial growth factor (VEGF), resulting in hyperplastic angiogenesis and neoplasia.[10]

Chronic hypoxic condition such as high altitude, COPD or cyanotic heart disease can over burden the carotid bodies and subsequently leads to hypertrophy, hyperplasia and neoplasia of chief cells. This condition is seen in hyperplastic type of CBT.

CBT can be occasionally coupled with nonparaganglionic tumours and can be a part of syndromes like MEN type II, Von Hippel Lindau syndrome and neurofibromatosis type I.

Methods

Retrospective study of series of 5 patients presented in the department during the period of 3 years was performed and analysed on the basis of presenting symptoms, examination findings and investigations. Shamblin's classification was followed to classify them for deciding the line of treatment. All the patient were investigated for proper family history and relevant previous disease specially for any chronic hypoxic situation. Colour Doppler study and CT scan was done in all cases apart from the routine basic blood and radiological tests to confirm the diagnosis. Doppler study also helps in embolization of vessel, and reduces the intra-operative bleeding by occluding the feeding vessel.

All the cases were presented with swelling in the neck. 4 out of 5 were females with 1 male who had bilateral tumour which was pulsatile and bruit was also present. Palpitation, hypertension, hoarseness of voice and sweating of forehead were other features found in the same patient. One patient was asymptomatic with a small nodular mass and it was an incidental finding on routine FNAC examination. Rest three cases were having typical pulsatile nodular mass in upper neck at carotid bifurcation with no other neurological signs.

On radiological examination one patient was showing complete blockage of vessel and no blood flow could be seen beyond the tumour on MR Angiography. (Fig1)

Three cases were operated, one with bilateral mass was referred as he had multiple complications and other one refused for surgery as it was asymptomatic and patient had a small child to look after.

Surgical Procedure

Under general anaesthesia with the preparation of all vascular safety and clamps, excision of mass was done .Horizontal upper neck incision was given, skin and fascial flap raised. Sternomastoid muscle and nerves were separated, external jugular vein ligated. Tumour was exposed, all the vessels were identified and it was dissected from the vessels. Vascular clamps were applied and tumour was excised separating from the vessels. In one case where there was complete block beyond the tumour mass with the development of very good collaterals, all the vessels were ligated to remove the tumour completely. In this case the tumour was quite firm with lot of fibrous element. Superficial laryngeal branch of vagus was also found to be trapped in the mass which caused difficulty in separating and produced post operative cough and hoarseness because of nerve handling. It took 3 weeks for cough to disappear, till then RT feeding was given. Bleeding was very less in all the cases due to hypotensive anaesthesia. Wound closure done in two layers after keeping the vaccum drain which was removed after 48 hrs. Post operative histopathology was showing features of paraganglioma. (Surgical photographs are shown in Fig 2, 3, 4 and 5)

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FIG 1

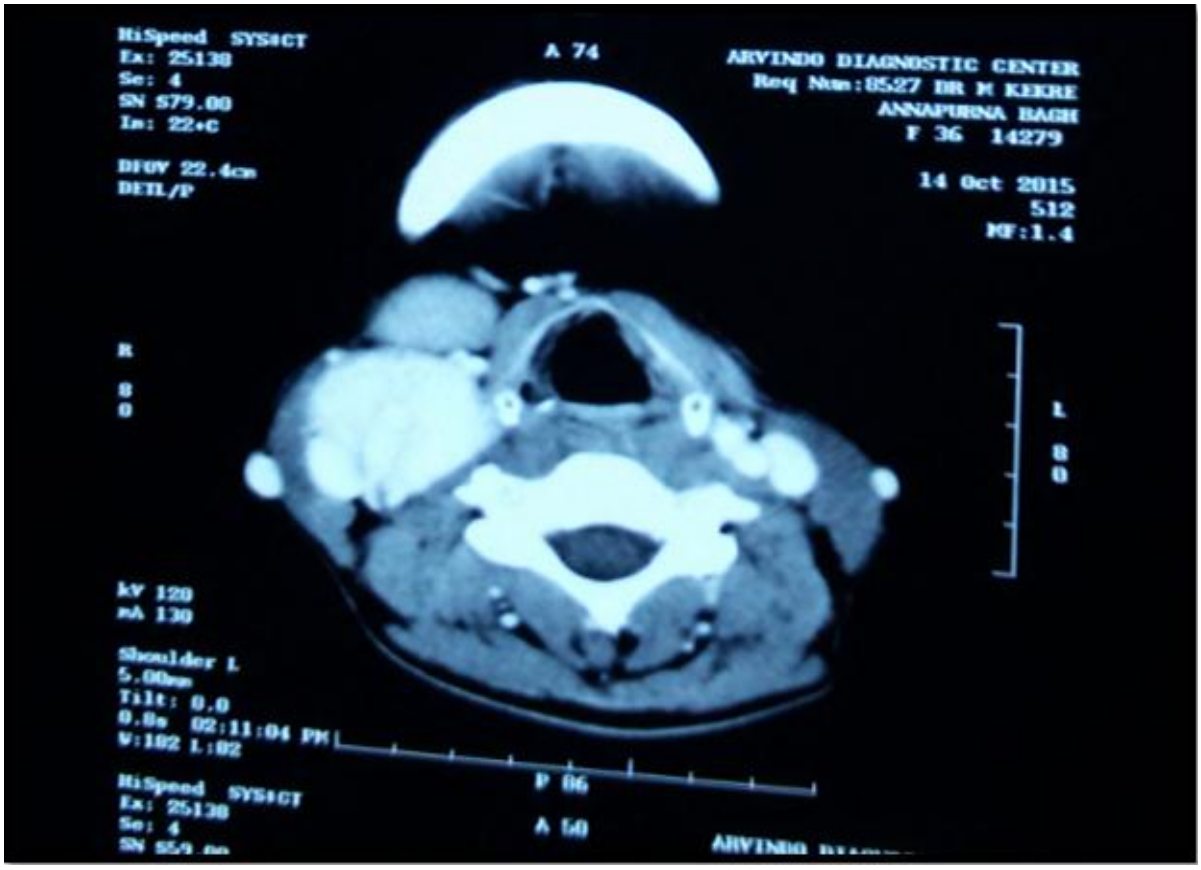

FIG: 2 and 3

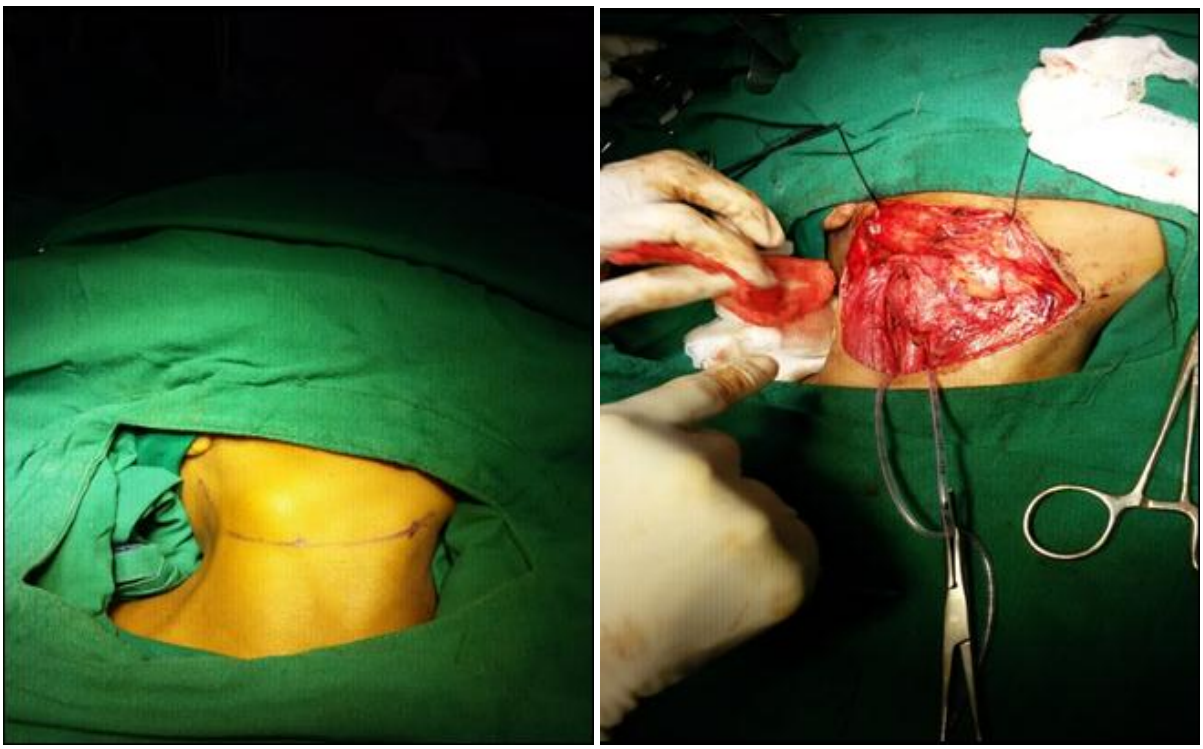

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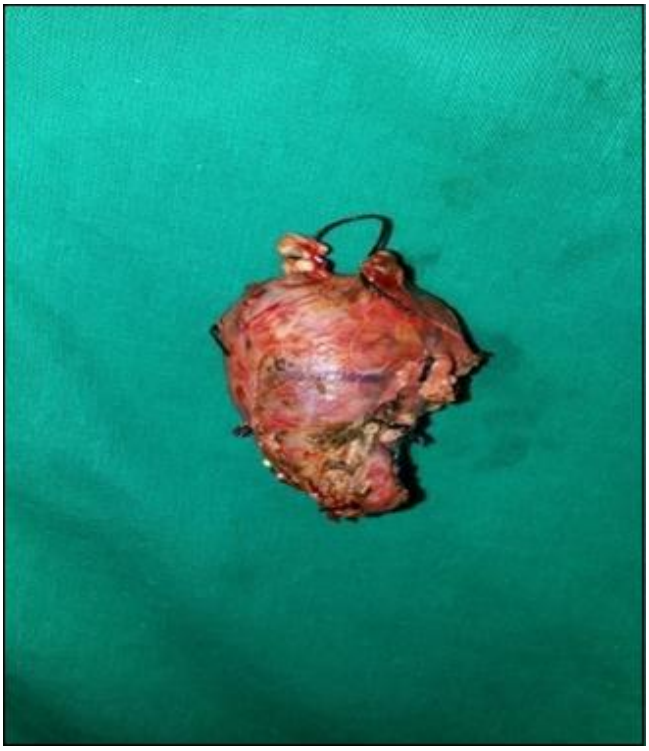
FIG: 4 and 5




Results

Post operative period was smooth in all cases, except in one case where superficial laryngeal nerve was found to be entrapped in the tumour during surgery causing difficulty in dissection. The excessive nerve handling caused oedema and hoarseness of voice and cough in the immediate post operative period. Patient was kept on RT feeding for three weeks till the nerve recovered and the patient started taking food without any difficulty. In the same case both the carotids had to be ligated as tumour was infiltrating the vessel wall causing complete obstruction of the lumen and fixity to the mass so they had to be removed along the tumour (fig 3 and 4). There was no immediate and late intracranial neurological deficit as blood flow was already compromised for long time with the development of good collateral circulation. The patients were discharged in 7 days without any other complication. They all came for follow up with no delayed complication.

Discussion

Carotid body is a small reddish brown oval structure located in postero medial aspect of carotid artery bifurcation measuring 3-5 mm (rice grain) in diameter, weighs <15mg. It is highly vascular receiving its blood supply from external carotid artery and its innervations by the Hering nerve originating from gloss pharyngeal nerve.[11]

Carotid body is comprised of small cluster of chemo receptors and supporting cells which detects changes in composition of arterial blood flow through it, mainly partial pressure of arterial oxygen and also that of carbon dioxide. It is also sensitive to the changes in PH and temperature. In addition to hypoxia carotid body has been found to respond to a variety of blood borne stimuli like glucose and immune related cytokines.

The mechanism of carotid body paraganglioma remains unknown, but hypoxia and genetic factors are thought to be involved in pathogenesis. Low pressure of oxygen and chronic hypoxia in people living at high altitude or chronic intermittent hypoxia due to sleep apnoea syndrome and COPD may stimulate the hyperplasia and hypertrophy of carotid body.[12,13] The patient with genetic mutation if experience chronic hypoxia develop these tumour at early age but under the situation of no such positive family history predisposing genes can spontaneously cause this tumour at the age 40-65 years.

These patients usually present with a neck mass, initially treated as nodal mass conservatively. Some of them report to the physician for the symptoms similar to cardiological problems like palpitation, increased pulsations, hypertension or even bruit in the neck while few others come for neurological advice as they present with cranial nerve palsies eg hypoglossal, glossopharyngeal, vagus or sympathetic chain involvement. There are patient reporting with swelling and pressure symptoms over trachea and pharynx. Fever is uncommon presentation seen along with paroxysmal hypertension in cases of tumour having functional element like pheochromocytoma.

Classification and staging is done after the colour doppler or CT /MRI studies which decide the surgical planning. DSA not only helps to confirm the diagnosis and extent but also

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provides information regarding the extra or intracranial blood circulation and helps in embolizing the feeding vessel .Balloon occlusion test can also be conducted to know whether patient will tolerate blood vessel ligation.[14-16]

If patient has positive family history or multiple foci of disease, pre operative CT scan of mediastinum and retroperitoneal sonography should be done.[17] Prior to surgical resection important measures to control the bleeding such as ligation bands or atraumatic vascular clamps should be kept ready.

The final diagnosis of malignant CBT is made depending upon recurrence or metastasis, partial recurrence with pains and lesions in the involved nerves, distant metastasis in the lungs, liver and bones. The incidence of malignancy is 5-7% with higher occurrence in younger patients and in those with family history.[18] The emergence of postoperative metastasis is reported even after 20 years therefore long term follow up should be insisted. Surgical excision is still the optimal choice for the treatment but in certain conditions where serious operative injuries are expected such as in preoperative involvement of cranial nerves, recurrence or lesions classified as Shamblin type III, in elderly patient or patient with chronic diseases, radiotherapy can effectively prolong the survival

Conclusion

Carotid body tumour or paraganglioma is one of the rare tumour, may present as a simple nodular mass or in a most severe form with multiple symptoms. It may be associated with other paraganglioma anywhere in the body or as a single mass in the neck. Both familial and hypoxic factors are found to be responsible for the occurrence. The familial are seen in younger age group and are with high risk of recurrence and malignant change where as the cases with hypoxic etiology are usually elderly with rare recurrence. Surgery is the treatment of choice depending upon the type of tumour based on Shamblin's classification type I, II, III unless there is any contraindication like extensive involvement of cranial nerves, recurrence or any other chronic medical problem. CT scan, MRI, Colour Doppler study is required for appropriate diagnosis.

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