Case Report

Carotid Body Tumour a Challenging Management: Rare Case Report in Baghdad Radiation Oncology Center, Medical City, Baghdad, Iraq

ABSTRACT

Background: Paragangliomas are rare asymptomatic painless tumors, originating from paraganglionic bodies of autonomous nerve system. The carotid body tumors are relatively rare tumor but constitute majority of head and neck paragangliomas about 70%. These tumours are generally benign but possess aggressive local growth potential.

Aim of study: The purpose of this review article is to simplify understanding the basic and clinical aspects of this challenging tumour, which is the first time study in Iraq.

Case presentation: We reported a case of 27 year-old female patient with a left neck swelling that had persisted for 5 years. She was first diagnosed and treated surgically by local excision in 2012. Histopathology revealed the histology of carotid body tumor. She was complaining of hypertension, headache, palpitation, and low grade fever. She was observed gradual onset of growing and pulsated lump on previous surgical scar site. At October 2017, the mass growing faster and she felt pulsation on lying on left side resulting in discomfort on sleep and deglutition. On physical examination a pulsating firm painless mass measuring about 6 x 5 x 4 cm in size was found on the left side of her neck. Pulsations were felt on deep palpation and a faint bruit was heard on auscultation. Work up done for her included laboratory tests, neck US, neck CT scan, carotid angiography and slid review of histopathology. All suggested recurrent carotid body tumor.

Conclusion: Throughout more than four decades working in this field, we faced 2-3 cases of CBTs, so it is very rare tumour. It is a challenging tumour whether decide to treat by surgery or radiotherapy. The main step in management of CBT is excluding of others tumour may be present in same region. Surgery is treatment of choice while radiotherapy is standard treatment for recurrent cases.

Keywords: Carotid body tumour; Paraganglioma; Chemodectoma; Glomus cells; Carotid Arteriography

1. INTRODUCTION

Carotid body tumour (CBT) or chemodectoma is a rare, highly vascular, mostly benign tumour arising from the paraganglia of carotid body. CBTs are nonchromaffin paragangliomas arising from the chemoreceptor cells found at the carotid bifurcation. The tumour is highly vascular; its blood supply is the richest per gram of tissue of any tumour [1]. The carotid body, which originates in the neural crest, is important in the body's acute adaptation to fluctuating concentrations of oxygen, carbon dioxide, and pH. The carotid body protects the organs from hypoxic damage by releasing neurotransmitters that increase the

ventilatory rate when stimulated [2]. There are three different types of CBTs have been described: familial, sporadic and hyperplastic. The sporadic form is the most common type. The familial type (10-20%) is more common in younger patients. The hyperplastic form is very common in patients with chronic hypoxia, which includes those patients living at a high altitude and may be found in patients with COPD and cyanotic heart diseases. The first anatomical description of carotid body was provided by Albrecht Von Haller in 1743 [2, 3]. Histologically, carotid body tumors have a characteristic growth pattern often referred to as a zellballen **[**6]. Carotid body tumors occur at any age but are typically diagnosed between the third and sixth decades of life [5]. The usual presentation is a slow growing mass at the angle of mandible [2, 4, 9]. It is must involve thorough evaluation for primary tumour of the thyroid, the oropharynx, and the nasopharynx is essential, since metastases to a cervical lymph node is a much more frequent cause of a neck mass than CBTs [9]. Carotid body tumors can be a diagnostic challenge for the clinician and lack of pre-operative diagnosis has been reported in up to 30% of the cases in different series [4]. The diagnostic work-up of CBT may involve one or more of the following: Duplex US scanning, CT, MRI, MRA, carotid arteriography, and serum and urinary catecholamine level assessment [2]. The treatment modalities for CBTs are surgical excision and/or radiotherapy [5, 9, 10, 11, 12, 13].

2. CASE PRESENTATION

A 27 year-old female patient presented with a left neck swelling that had persisted for 5 years. She was first treated by local excision at 29th July 2012 in a private hospital. Pathology revealed the histology of carotid body paraganglioma. She was in good health for two years before she developed her signs and symptoms of catecholamine excess such as hypertension, headache, palpitation, and weight loss. She was consulted many doctors and centers for her problem and she kept on medical treatment and follow up. In the last nine months she observed gradual onset of growing and pulsated lump on previous surgical scar site. Suddenly in two weeks exactly in October 2017, before she came to Baghdad Radiation Oncology Center, the mass growing faster and she felt pulsation on is lying on left side resulting in discomfort on sleep and deglutition.

1. Physical examination

A pulsating firm painless and well circumscribed mass measuring about 6 x 5 x 4 cm in size was found on the left side of her neck, near the angle of the mandible. There were pressure symptom & more mobile transversely than vertically. Pulsations were felt on deep palpation and a faint bruit was heard on auscultation. She was complain of headache, tachycardia and hypertension.

2. Work up

2.1. Laboratory tests

CBC, ESR, RFT, LFT, electrolytes balance and urine analysis were done.

2.2. Imaging

CXR and Echo study also done.

2.2.1. Neck US

It revealed highly vascular ill-defined hypoechoic solid mass seen above level of Left carotid bifurcation between ICA and ECA measured 35 x 20 mm, reaching up to lower border of parotid gland suggestive of recurrent CBT. Normal other structures.

2.2.2. CT scan of neck

It demonstrated a well-circumscribed lobulated hypervascular, hypoechogenivic mass with splaying of the carotid bifurcation. The mass was heterogeneously intense enhancing soft tissue density mass of size 38 × 24 × 25 mm. the findings suggested recurrent CBT. There were no infiltration into adjacent structures is seen. Both thyroid lobes were normal in size, echogenicity, normal isthmus, no cervical LAP, normal both submandibular salivary glands, normal neck vessels and no SOL.

2.2.3. Carotid angiography of neck

It showed a well-defined highly vascular blush mass (density is 250 HU while in the carotid artery is 340 HU), is about 33 x 27 x 25 mm, located at the lateral to both ICA and ECA of left carotid arteries and about 2 cms above the carotid bifurcation. Mass is not intrinsic to blood vessels & patency of the artery was preserved. This finding was considered to be consistent with a diagnosis of recurrent CBT. In compares with previous carotid angiography done in June 2012, there was irregular solid oval soft tissue density mass 63 x 39 x 27 mm in size seen in left upper the neck in left retro mandibular region beneath left sternomastoid muscle, extending from level of below left parotid gland down to below mandibular angle. It was vascular mass and protruding between ICA and ECA. There was no cervical LAP. The features were CBT.

2.3. Histopathology

The previous histopathology was done at July 2012. Grossly there was a globular piece of tissue measured 5 x 3 x 2 cm, with a grey brown cut section, also there were three firm grey nodes. Histologically, the sections showed well defined nests of uniform cuboidal cells (Zellballen) with abundant granular basophilic cytoplasm, separated by highly vascularized fibrous septa. There was no mitoses or necrosis seen. The picture was consistent with carotid body tumour CBT. The three LNs showed reactive follicular hyperplasia. The slide review done and give the same result.

3. Treatment

At that time, the mass was quite large with size and attached to left common carotid artery. Therefore, surgery was not suitable, because of very high risk for surgical injury to these vessels and nerves and she was a recurrent case. Finally, she was referred to our radiotherapy unit for definitive radiotherapy. The radiation technique was 3D-conformal radiotherapy (3D-CRT) with 2 Gy/daily fraction to the total dose of 50 Gray in 25 Fractions. The GTV (gross target volume), which was defined as gross tumor, was shown on contrast CT scan. The CTV (clinical target volume) was the volume including GTV and 1 cm margin around. The PTV (planning target volume) was the volume including CTV and 1 cm margin around. The normal tissue constraint was limited within tolerance dose.

3. DISCUSSION

Carotid body tumors are rare and uncommon entities may be found in unilateral or both sides of the neck and in both genders at the different rate of frequency, and they belong mostly to a benign group of tumors and their surgical management is technically challenging. The most common presentation in patients with CBTs is a slowly enlarging painless mass in the neck. Locally invasive growth of these tumors subsequently leads to cranial nerve deficits along with compression symptoms like Horner’s syndrome, syncope, hoarseness and dysphagia since the carotid body functions as a chemoreceptor organ that is stimulated

by hypoxia, hypercapnea, and acidosis, it is involved in the control of blood pressure, heart rate, and respiration [1, 3, 5, 11, 13]. CBDs are slow growing painless masses localized in the neck, anterior to the sternocleidomastoid muscle at the level of the hyoid bone. As the tumour grows, dysphagia, odynophagia, dysphonia, and symptoms due to compression of cranial nerves IX to XII may be seen. The most commonly involved cranial nerve is the vagus, up to one third of all cases will show cranial nerve palsies [3, 5, 8, 11].

US is the first non-invasive procedure which allows seeing that is vascular discrimination between the solid and cystic nature of the mass. Carotid arterial angiography is the most valuable diagnostic technique, it is the gold standard for diagnosis is carotid angiography, which serves both diagnostic and treatment purposes. MRI and CT angiography can demonstrate the extent of the tumor and its relationship to adjacent structures [2, 7, 8].

There are many choices of treatment for CBDs including observation, surgery, external beam radiotherapy, and stereotactic radiotherapy. Surgery is the treatment of choice. The local control by surgery alone is approximately 85-100%. Most of the reports demonstrated local control with radiotherapy alone is approximately 80-90% comparable to surgery [10, 11, 13]. Treatment with radiotherapy can achieve comparable local control and less morbidity than surgical resection in paraganglioma. Regarding definitive radiation treatment of CBTs. There are many techniques, protocols and radiation dose ranges of treatment. Although stereotactic radiotherapy has been increasingly used and their results have been generally accepted, conventional radiotherapy and 3D radiotherapy are still commonly used in the place where stereotactic radiotherapy is not available. Many reports used a radiation dose of 45 Gy in 25 fractions, with a daily dose of 1.8 Gy [3, 5, 6, 12]. Continued follow-up is necessary, however, as recurrence and metastasis may occur years later [12].

4. CONCLUSION

CBT is a rare paraganglionic tumour affecting both sex of reproductive age. Treatment of choice is surgery. Radiotherapy is standard treatment in recurrent and inoperable cases. Since late recurrences are known, patients should be adequately need for periodic follow up so that recurrences can be identified early and treatments offered. The limitation of this study is that Immunohistochemistry was not done to support the histological diagnosis and was because the markers for this technique are currently out of stoke in country.

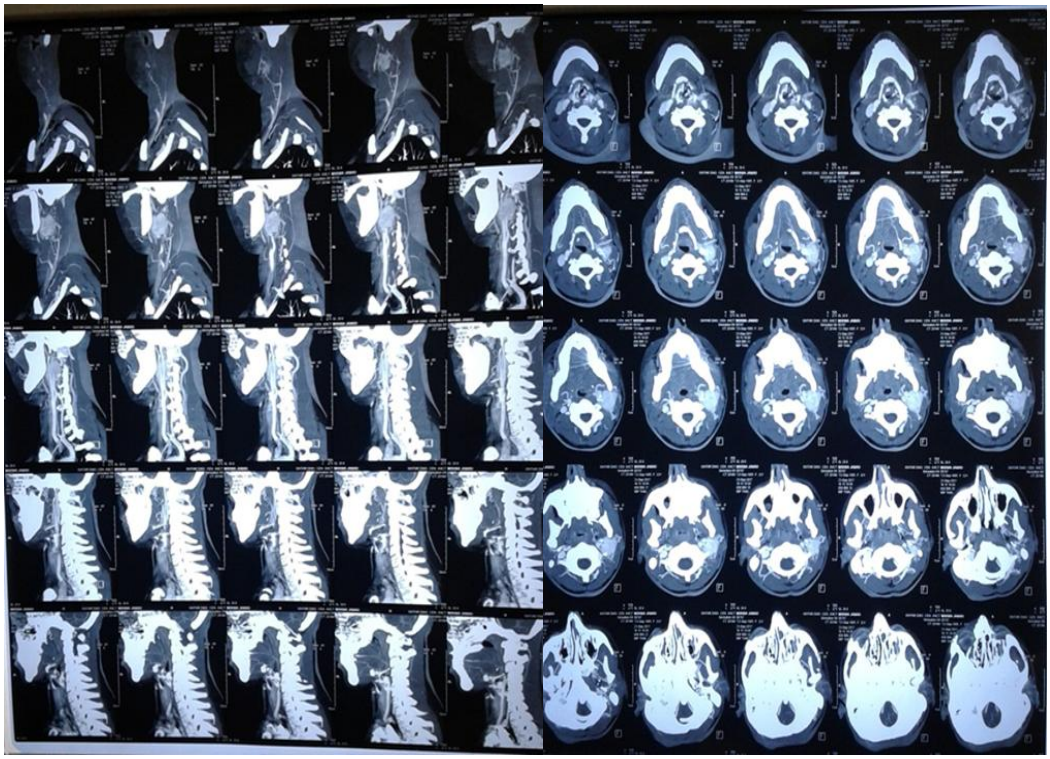

Fig. 1. Head and Neck CT scan (axial and sagittal plan).

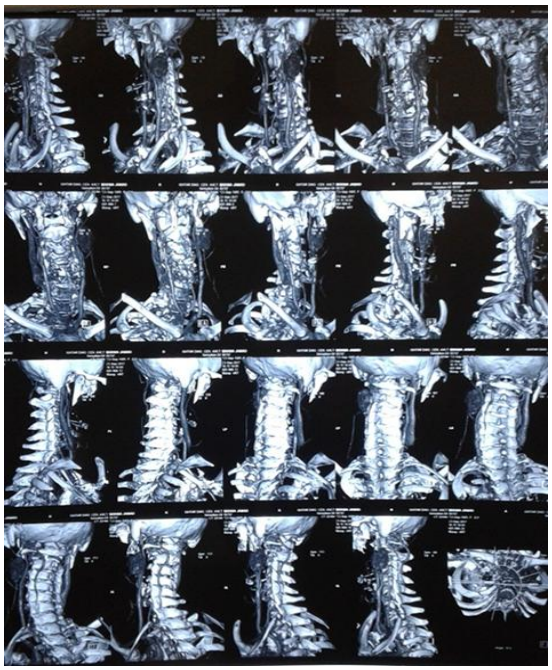

Fig. 2. Carotid angiography

COMPETING INTERESTS

Authors have declared that no competing interests exist.

CONSENT (WHERE EVER APPLICABLE)

All authors declare that 'written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images.

ETHICAL APPROVAL (WHERE EVER APPLICABLE)

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

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ABBREVIATIONS

CBT: carotid body tumour, COPD**:** chronic obstructive pulmonary diseases, CCA: common carotid artery**,** ICA: internal carotid artery, ECA: external carotid artery**,** PG: Paraganglioma, FNA: fine needle aspiration, US: ultra sound, CT: computed tomography, MRA: magnetic resonance arteriography, 3D: three-dimensional, RT: radiotherapy, CBC: complete blood pictures, RFT: renal function tests, LFT: liver function tests, Gy: gray, LAP: lymphadenopathy.