BILATERAL CAROTID BODY TUMORS: A CASE REPORT AND REVIEW

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Abstract

Carotid body tumors (CBTS) also known as chemodectomas are a type of paragangliomas which originate from the carotid body. Carotid body tumors present as a palpable mass in anterolateral aspect of the neck causing splaying of the external carotid artery (ECA) and internal carotid artery (ICA). Bilateral carotid body tumors are uncommon with a reported incidence of 5 - 10%. CBTs are commonly seen in 4th to 5th decades with female predilection. We report a case of 16 years old boy with bilateral carotid body tumors.

Key Words:

carotid body tumor, paraganglioma, bilateral.

Introduction:

Carotid body tumor is an extra adrenal paraganglioma that may also be termed as chemodectoma. CBT originate from the neural crest tissue in the carotid bifurcation. Usually, CBT is a solitary occurrence. These tumors release catecholamines although less common than with adrenal paraganglioma1.Carotid body tumors occur frequently in adults averaging 45-50 years of age and are uncommon in young age. The majority of cases are considered to be benign, with only 10-20% demonstrating malignant potential. Familial forms accounts for 10% with bilateral tumors in 32% of cases. In sporadic cases bilateral tumors are detected in only 5% of patient2.These are slow growing tumors, presenting as asymptomatic palpable masses in the anterior neck. However untreated cases may surround the ICA and ECA and subsequently involve the cranial nerves, skull base and show intra cranial extension. Functional carotid body tumors are rare.

Case Report:

A16 years old boy was admitted to the department of ENT of Bolan Medical Complex hospital (BMCH) with bilateral palpable masses in upper neck since two years. There was no history of cardiac arrythmias, hypertension, tremors or features of lower cranial nerve paralysis. On examination there was palpable masses of approximately 3cm on right and 3.4cm on left. The masses were near the angle of mandible and were firm, non tender, pulsatile and could be moved from side to side (positive fontaine's sign). No bruit was present over these masses. Tough large in size dysphagia / hoarseness were absent. Vocal cord paralysis was excluded after laryngeal examination. No sign of hypoglossal neuropathy was seen. Few nodular swellings were also palpated in bilateral cervical regions and were suspected as being lymph nodes. Patient was advised Ultrasound neck and FNAC of these masses. Plan was of surgical resection after proper radiological investigations. USG done showed heterogeneously hypoechoic well defined lesion approximately measuring 2.9 x 3.2cm and 3.1 x 3.9 cm on right and left sides respectively. These were seen at the carotid bifurcation causing splaying of the ECA and ICA however no encasement or evidence of infiltration was noted. On color flow imaging these lesions were hypervascular with flow direction being predominately cephaled. Both tumors had low resistance flow. These masses were displacing the external carotid artery anteriorly and internal carotid artery posteriorly (fig 1). The left sided mass was relatively large, however the upper margin was not detected clearly due to the deep location of areas of the lesion. Few mildly enlarged cervical lymph nodes were seen. Based on sonographic findings likely

diagnosis of paraganglioma with differential of lymphoid masses were given.. The histopathologist did FNAC of the right sided enlarged lymph node which revealed chronic nonspecific lymphadenitis. . Contrast CT neck was advised by the ENT specialist. CT was done by primeaquilion 128 slice Toshiba which demonstrated well defined solid masses that showed avid homogenous enhancement on right side with peripheral enhancement and central non enhancing necrotic areas on left (fig 2). These masses were seen at carotid bifurcation on both sides causing splaying of internal and external carotid artery giving a positive lyre sign (fig 3). Few prominent bilateral cervical lymph nodes were also seen. Final diagnosis of bilateral carotid body tumor was given and patient was further advised workup for multiple endocrine neoplasia (MEN II). ENT department referred the patient to Karachi for multi disciplinary approach and further management by the vascular surgery. 1 month later we got in contact with the vascular surgeon who told us that the patient underwent CE MRI neck in which cranial nerves (namely glossopharyngeal, vagus and hypoglossal nerves) couldnot be separately demarcated from the CBTs and involvement of these nerves by masses couldnt be commented upon with certainity. The vascular surgeon was of the opinion that treatment of bilateral CBT should focus on preservation of the quality of life rather than on cure of the disease. Patient was given two options either to perform a two staged surgery to remove the tumor however it wasnt possible without sacrificing the vagal nerve. In one step surgery there was risk of bilateral cranial nerve palsies and cerebrovascular complications. Second option was to observe tumor growth with serial imaging studies. Cases has been reported of 5 years of disease free progression. The patient opted for the observation and follow up of the tumors.

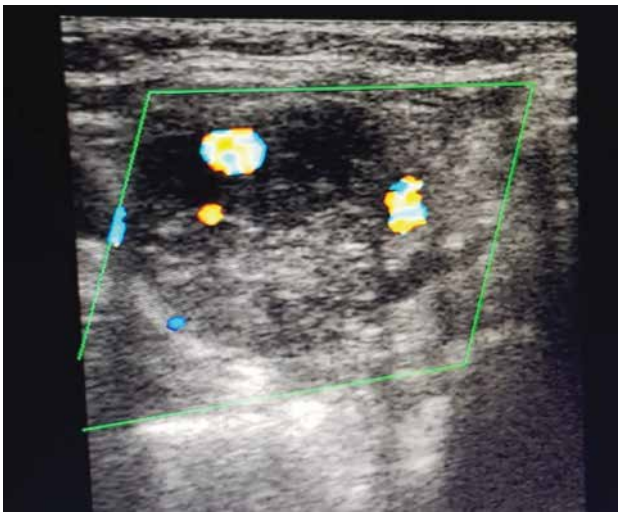

Fig1. Color doppler showing splaying of ICA andECA

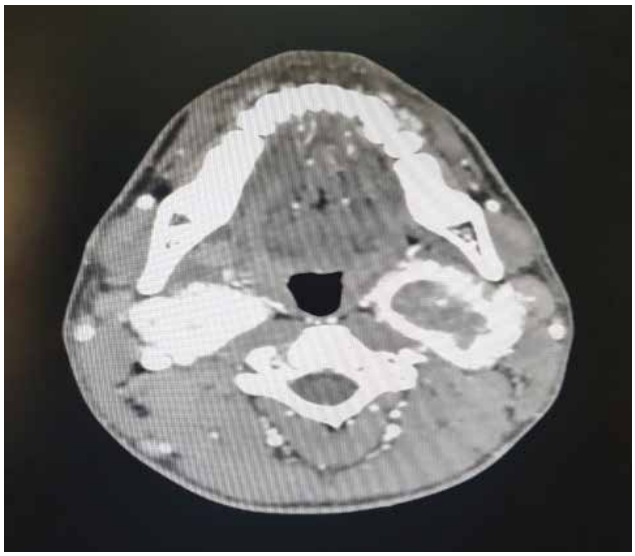

Fig2. axial CE CT showing bilateral carotid body tumors showing avid enhancement with central necrotic area in left sided tumor

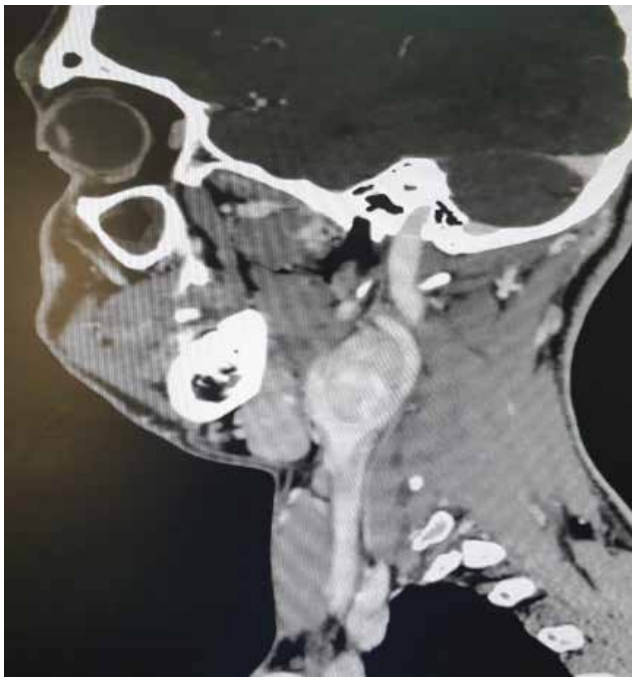

Fig3: CECT showing positive lyre sign

Discussion:

Carotid body tumors are type of extra adrenal paraganglioma. CBT are often diagnosed using the location, clinical symptoms and imaging findings of the tumor. The majority of CBTs are benign, however certain

lesions may demonstrate malignant inclinations.3

Radiological findings are important in diagnosing CBT. Usually conventional grey scale, color doppler ultrasound, CT and computed tomography angiography (CTA), MR and MRA, carotid conventional angiography (CA) are used 4.In our case grey scale USG, doppler USG and CT scan were used for the diagnosis of the tumors. An ultrasound is a rapid, convenient and non invasive measure that may be used to detect the margin, vascularity and invasion of a mass, and any regional lymph node5 .Ultrasounds are more useful for screening familial cases and follow up procedures. The possible diagnosis of CBT may be anticipated when a solid mass is detected at the carotid bifurcation.

A Doppler analysis of the mass is useful to evaluate intratumor blood flow and is valuable in differentiating chemodectomas from other solid non hypervascular masses2.Doppler analysis may reveal the association between the tumors and carotid artery clearly. Doppler imaging is also sufficient for the primary diagnosis of CBT as it may reveal abundant blood flow, which is characterized as intense blush of the tumor5. Contrast ultrasonography may also aid the evaluation of the blood supply to the tumor6 .Therefore ultrasound is a suitable technique for the identification of the CBT. However ultrasound is unable to differentiate whether the CBT is benign or malignant. The possibility of the malignant CBT may only be considered if significant vascular infiltration or regional lymph node invasion or distal metastasis are present. Ultrasound is also limited due to an inability to identify deeply located lesions7.

Cross sectional study best reveals the shape, size, margins, enhancement and extension of the tumor in the present case. In our case CE CT was done. MRI was suggested but was not done in BMCH due to the non availability of closed MRI and poor resolution of the open 0.3 T MRI which was the available option in our hospital.

On CT images, a carotid tumor is identified as a well defined solid mass with homogenous enhancement that is located within the carotid sheath. Larger tumors are frequently inhomogenous due to necrotic and hemorrhagic regions8. In our case the left sided carotid body tumor was large in size and showed internal non enhancing necrotic centre however no evidence of any hemorrhagic component was noted on plain CT scan. The ECA is usually displaced anteromedially and the ICA is typically displaced posterolaterally9.In our case similar displacement of the ECA and ICA were also noted which strongly suggested bilateral CTB. These features were characteristically identified in the images of the present study however vascular invasion was not identified.

CT demonstrates a well defined solid mass that shows homogenous enhancement on intravenous contrast administration in the regions of carotid bifurcation on both side causing splaying of internal carotid artery (ICA) and external carotid artery(ECA)2.The presence of uniform contrast enhancement and large feeding vessels helps it to differentiate it from lymph nodes and shwannomas. It is differentiated from the glomus vagal tumors,which is the paragangliomaof the vagus nerve by its relative location in the neck, with glomus vagal tumors arising slightly more cephalad and presenting as an intensely vascular, enhancing mass on CECT displacing the ICA and ECA anteriorly and internal jugular vein posteriorly. So after ruling out other differentials diagnosis of carotid body tumors was made in our case.

Among 89 patients with CBT, Shamblin et al report only one case in 12 years old child 1. shamblin et al mentioned three groups for future surgical cases based on attachment to the carotid vessels. Group one was minimally attached to the carotid vessels. Group 2 was moderately attached whereas group 3 are usually large and incarcerate the vessels.In our case shamblin type 2 was expected on left and shamblin type 1/2 on right. Dickinson et al reported only one case of young 14 years old girl10, complicated by XII nerve permanent paralysis out of 37 CBTs. Takutz et al found only one 12 years old boy having CBT in a large population of patients younger than 21 years. Regional prominent nodes were noted in the current case which however on FNAC revealed nonspecific lymphadenitis.

In conclusion bilateral carotid body tumors in young age is rare. Radiological investigations are non invasive techniques that may be used to diagnose and evaluate the extent of lesions.

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Author's contribution:
Dr Asma; data collection, data analysis, manuscript writing, manuscript review
Pari Gul; data collection, data analysis, manuscript writing, manuscript review
Palwasha Gul; concept,data collection, data analysis, manuscript writing, manuscript review