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CASE REPORT

Bilateral Carotid Body Tumor – A Rare Case Report

Nanda Patil, Neha Ghadge, Dhara Dodia

Abstract

Carotid body tumor is an uncommon tumor seen near bifurcation of carotid artery which arises from chemoreceptor cells of carotid body. They are slow growing and often asymptomatic and can be functioning or non-functioning tumors. Bilateral carotid tumors are rare. We present a case of bilateral carotid body tumor in 55 year male patient presented with painless, pulsatile, gradually increasing bilateral neck swelling. The diagnosis was done on the basis of history, clinical examination and radiological investigation. The tumor was excised and histopathological examination confirmed the diagnosis.

Key Words

Carotid Body Tumor, Chemodectoma

Introduction

Carotid body is a sensory organ which is located at the bifurcation of common carotid artery bilaterally[1]. It is the largest collection of paraganglia in the head and neck region and it can be a site for carotid body tumor which is also known as chemodectoma. They are extra adrenal neoplasms having neuroendocrine origin, slow growing and highly vascular tumors comprising of 0.3% of all paragangliomas[2,3]. Majority of these tumors are benign and non-functioning. The reported incidence is 1 to 2 per 1 lakh individual[4].

Case report

A 55 year old male patient presented with bilateral neck swelling since 5 to 6 months. On clinical examination it was observed that swelling was pulsatile, firm, slightly mobile and was seen near both sides of angle of mandible. There was no history of headache, sweating or palpitations. There was no lymphadenopathy.CT angiogram of head and neck revealed well defined oval soft tissue lesions noted at both sides of carotid bifurcation, splaying internal and external carotid artery and was diagnosed as bilateral carotid body tumor (Fig.1).

Right side of carotid body tumor was excised and sent for histopathological examination. We received a well circumscribed, encapsulated, globular, grey brown tumor measuring 3.5 x 3.2 x 0.8 cm. Cut section was homogenous and dusky brown to red in color. (Fig.2 and Fig. 3)

Microscopic examination revealed neoplastic cells arranged in nests separated by fibrovascularstroma creating zellballen pattern. Neoplastic cells were round to polygonal with abundant eosinophilic cytoplasm without nuclear atypia (Fig.3 and Fig.4). Considering these features diagnosis was given as carotid body tumor. Postoperative followup was uneventfull.

Discussion

Carotid body tumor presents as palpable, painless mass in the anterolateral aspect of neck which is pulsatile. Most of the tumor are unilateral. Bilateral occurance is very rare as seen in our case which is seen in only 5% of affected patients[5]. Most of the tumors are benign while 5 to 10% being malignant. Carotid body tumor is frequently seen in adults at the age group of 45 to 50 years with

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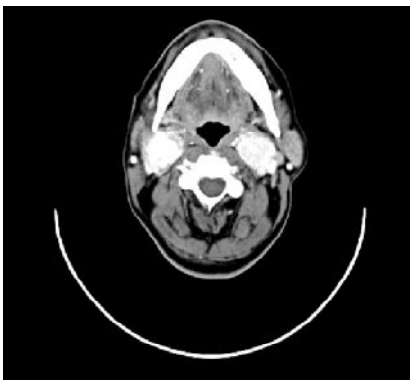
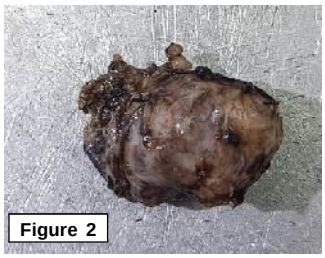


Fig 1 : CT angiogram of head and neck revealed well defined oval soft tissue lesions noted at both sides of carotid bifurcation, splaying internal and external carotid artery



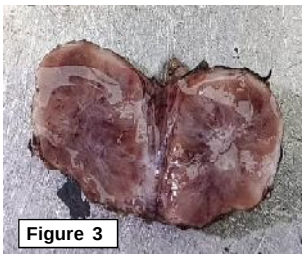


Figure 2, 3: Gross – A well circumscribed encapsulated globular grey brown tumor
Cutsection - Homogenous, dusky brown.

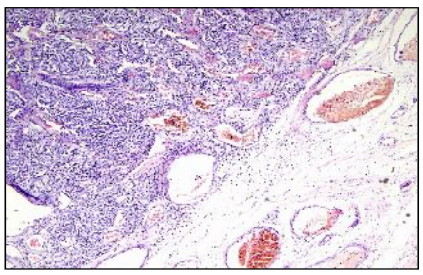


Figure 4 : Microscopy – Neoplastic cells arranged in nests separated by fibrovascular stroma (Zellballen pattern. 100X H and E).

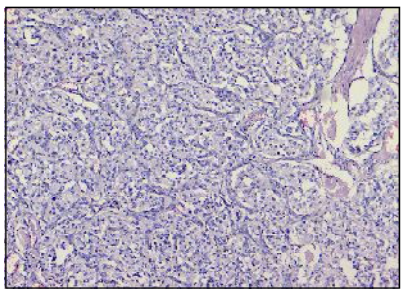


Figure 5 : Microscopy - round to polygonal neoplastic cells with abundant eosinophilic cytoplasm without nucler atypia (400 X H and E).

female preponderance[6]. Howeverour patient was male patient. Majority of tumors are non-functioning. Functioning tumors produce symptoms of headache, sweating, palpitation, hypertension, flushing, arrhythmia and transient ischemic attack and stroke[7]. The most frequent cause of carotid body tumor is sporadic 30 to 50% have hereditary predisposition. It is thought that genetic factor and hypoxia play significant role in the etiology of carotid body tumor. Commonest mutation observed is succinatedehydrogenase mutation which is enhanced with hypoxia. Hyperfunctioning carotid body tumor are evaluated with 24 hour collection of urine for catecholamine, vanilylmandelic acid and metanephrins. Malignant carotid body tumor can invade local cranial nerve and cause Horner's syndrome[8]. The most important differential diagnoses include aneurysm, hematoma, vagal schwannoma or carotid body hyperplasia and head and neck carcinoma[9].

Digital subtraction angiography helps in the diagnosis of carotid body tumors which shows splaying of internal and external carotid artery which is known as Lyre's sign. Similar observation was noted in our case. In 1971, Shamblin et al. classified carotid body tumors to help in the prediction of the prognosis and difficulty level of surgery excision. Group 1 tumors are small having minimal attachment to carotid vessels making surgical excision easier. Group 2 tumors are larger and show moderate attachment to carotid artery and no encasement, while group 3 tumors are large and fully encase carotid arteries. Surgical excision in group 3 tumor is associated with most complications and replacement of vessels should be considered[10].

In 2005, Lunar-Ortiz et al. added group 3b category encompassing tumors which encase circumferentially as well as infiltrate the vessels making the prognosis worse[11].

Surgical excision is the treatment of choice. Postoperative complications are nerve damage, stroke, hemorrhage and death[14]. Today however enhancement in imaging technology and selective embolization, surgical excision has become safe[12].

Conclusion

Bilateral carotid body tumors are very rare. Due to potential of these tumors for malignancy, early diagnosis is essential.

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