

A Case of Bilateral Carotid Body Tumours.

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ABSTRACT

A 21 year old male patient presented to the out patient department with swelling on the right side of the neck which was gradually progressing without any other manifestations. Vitals & laboratory tests are like CBP, urinary catecholamine's are within normal limits. Ultrasound neck revealed a soft tissue mass lesion in right side of neck in the carotid triangle which is hypervascular on Doppler study & Similar lesion on left side of neck, which is relatively smaller. MRI neck revealed a 3.5x2.9x2.3cm sized, well circumscribed lesion in the right side of neck at the bifurcation of right common carotid artery which is iso to hypo intense on T1WI & hyperintense on T2WI with intense post contrast enhancement and characteristic "salt and pepper appearance". A concomitant lesion measuring 2x1.3x1.1cm noted in left side of the neck at the bifurcation of the left common carotid artery, which is iso to hypo intense on T1WI & hyperintense on T2WI with intense post contrast enhancement. MR Angiography revealed splaying of internal and external carotid arteries. FNAC findings from right side lesion revealed it as carotid body tumour.

Keywords: Carotid body tumor, Neoplasm, Paraganglioma

INTRODUCTION

Extraadrenal paragangliomas are neoplasms of the paraganglia located within the para-vertebral sympathetic and parasympathetic chains. Paragangliomas may arise anywhere along these tracts and the most common site in head and neck is the carotid body. Carotid body paragangliomas are located in the superolateral neck, inferior or deep to the mandible at the carotid bifurcation and with splaying of the internal and external carotid artery. The average age at diagnosis is usually the fifth decade of life, and the sex incidence is known to be nearly equal. Bilateral involvement is uncommon but occurs more frequently in familial cases. On ultrasound paragangliomas are demonstrated as well-defined hypoechoic masses with a heterogeneous echo pattern and numerous arterial and venous vessels inside the lesion. On MRI Flow voids from the numerous vessels are typically seen and this finding is part of the classic "salt and pepper" imaging appearance of these lesions seen on T2 weighted images. Upon contrast administration the lesions avidly enhance reflecting their vascular nature. The pathognomonic angiographic finding is widening of the carotid bifurcation by a well defined tumour blush (lyre sign). The preferred method of treatment is surgery.

manifestations. No significant family history. Vitals is within normal limits. CBP, urinary catecholamine's are within normal limits.

USG Neck: Revealed a soft tissue mass lesion in right side of neck in the carotid triangle which is hypervascular on Doppler study.

- Similar lesion is seen on left side of the neck, which is relatively smaller and is also hypervascular and causing splaying of internal and external carotid arteries.

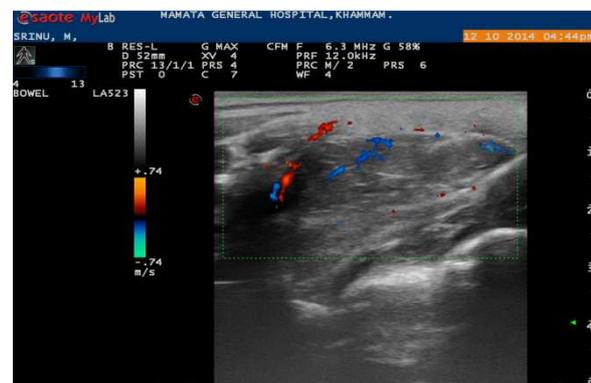


Figure 1: Right side hypervascular mass in carotid

MRI NECK: Revealed at 3.5x2.9x2.3cm sized, well circumscribed lesion in the right side of the neck at the bifurcation of the right common carotid artery, which is iso to hypo intense on T1WI & hyperintense on T2WI with intense post contrast enhancement and characteristic "salt and pepper appearance".

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CASE REPORT

A 21 year old male patient presented to OPD with swelling on the right side of the neck, which was gradually progressing without any other



Figure 2: Left side mass causing splaying of internal and external



Figure 6: MR ANGIO - splaying of internal and external carotid arteries

concomitant lesion measuring 2×1.3×1.1cm lesion note in left side of the neck at the bifurcation of the left common carotid artery, which is iso to hypo intense on T1WI & hyperintense on T2WI with intense post contrast enhancement.

MR Angiography revealed splaying of internal and external carotid arteries.

Histopathology: FNAC from mass on right side of neck suggested diagnosis as carotid body tumour .

DISCUSSION

Extra-adrenal paragangliomas are neoplasms of the paraganglia located within the paravertebral sympathetic and parasympathetic chains. Paragangliomas may arise anywhere along these tracts and common sites of occurrence include the abdomen, retroperitoneum, chest and mediastinum and various head and neck locations such as jugulotympanic membrane, orbit, nasopharynx, larynx, vagal body and carotid body. The most common site in head and neck is the carotid body. The average age at diagnosis is usually the fifth decade of life, and the sex incidence is known to be nearly equal. In approximately 10% of cases they are bilateral. They may occur sporadically or autosomal dominant familial inheritance and associated with MEN II a and MEN II b & phakomatoses (tuberous sclerosis, NF 1, VHL).^[1]

Carotid body tumours (CBTs) are slow-growing hypervascular tumors arising from paraganglionic cells of the carotid body which acts as a vascular chemoreceptor organ and is usually located in the posteromedial wall of the CCA at the carotid bifurcation^[2]

Carotid body paragangliomas often manifest as slow-growing, nontender masses in the neck, located anterior to the sternocleidomastoid muscle at the level of the hyoid bone. They are movable in the horizontal plane, but their mobility is limited in the vertical plane (Fontaine sign). Occasionally, a carotid pulse, a bruit or thrill may be associated with the mass. Symptoms such as fluctuating hypertension, blushing and palpitations suggest the tumour may be producing catecholamines. Bilateral involvement is uncommon but occurs more frequently in familial cases.^[3]

These lesions splay apart the internal (ICA) and external carotid arteries (ECA), and as it enlarges, it will encase, but not narrow the ICA and ECA. Upon contrast administration the lesions avidly enhance reflecting their vascular nature. Flow voids from the numerous vessels are typically seen on



Figure 3: T1WI – isointense mass in carotid triangle on right side of

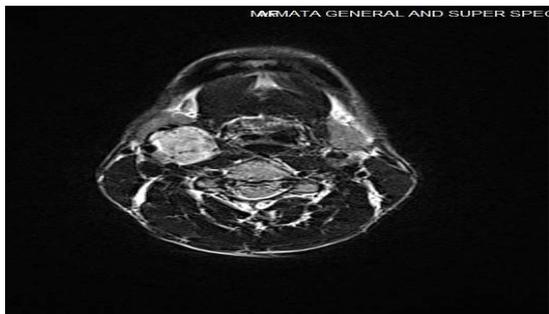


Figure 4: T2 WI – Hyperintense mass in carotid triangle on both sides of neck with “salt & pepper appearance”



Figure 5: T1 FS PC – intense enhancement of masses in carotid triangle on both sides of neck

MR imaging, and this finding is part of the classic “salt and pepper” imaging appearance of these lesions seen on T2 weighted images. The lesions tend to be isointense relative to muscle on T1 weighted imaging and hyperintense on T2. Avid enhancement is seen on post contrast imaging. The pathognomonic angiographic finding is widening of the carotid bifurcation by a well defined tumour blush (lyre sign).

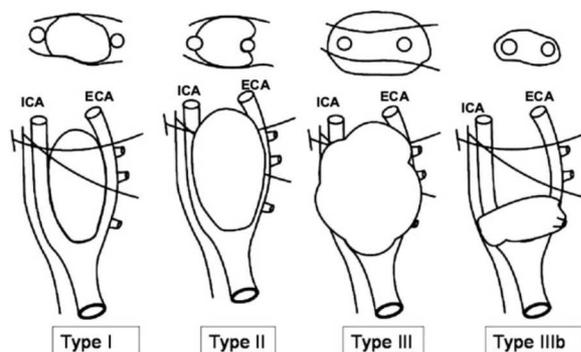


Figure 7: Shows a schematic diagram of the Shamblin grouping of CBTs into I, II, and III, as well as IIIb (as proposed in the modification to Shamblin’s classification by Luna-Ortiz et al⁴). The surgical grouping is chiefly based on the relationship of the tumor to the carotid vessels, ICA and ECA. The class IIIb tumors include tumors of any size that are intimately adherent to the carotid vessels. The oblique lines shown represent the X and XII nerves, which are intimately related to the tumors and have to be carefully dissected along with the vessels

Cather angiography and CT angiography reveal a hypervascular mass with enlarged feeding arteries (typically the ascending pharyngeal or ascending cervical artery), intense tumour blush and early draining veins. Octreotide scanning shows a focal area of early intense radiotracer uptake will be seen in the region of the paraganglioma, and is sensitive for detecting tumors greater than 1.5 cm. It is also useful for detecting the presence of multicentric or metastatic paragangliomas, and for distinguishing scar from residual tumor after surgery.^[4]

Shamblin et al. ² classified these tumors into 3 groups based on the operative notes and gross specimen examination.^[5]

The preferred method of treatment is surgery, which can be challenging because the tumor is a highly vascular mass that is often densely adherent to the carotid bifurcation. Meticulous preoperative planning and careful patient selection are essential for a successful surgical outcome. A landmark in the assessment of resectability of these tumors is the surgical classification by Shamblin et al.^[6]

Differential Diagnosis :

Vagal schwannoma: tends to displace both vessels together rather than splaying them

Vagal neurofibroma: tends to displace both vessels together rather than splaying them

Lymph node mass : may look similar if hypervascular

Glomusvagaletumour: same pathology but located more rostrally.

CONCLUSION

The carotid body tumour is the most common paraganglioma of the head and neck which are slow-growing hypervascular tumors arising from paraganglionic cells of the carotid body which acts as a vascular chemoreceptor organ and is usually located in the posteromedial wall of the CCA at the carotid bifurcation. Bilateral involvement is uncommon but occurs more frequently in familial cases.

Our patient presented with slow growing neck mass & MRI showed masses with low signal intensity in T1-weighted sequences and a hyperintense signal in T2-weighted sequences with characteristic “salt and pepper” appearance. Contrast-enhanced MRA revealed the pathognomonic angiographic finding of widening of the carotid bifurcation by a well defined tumour blush (lyre sign). FNAC suggested diagnosis of carotid body tumours.

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