Bilateral Cervical Masses: Wolf in Sheep's Clothing

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A 45-year old woman presented with bilateral painless slowly-progressing cervical lumps for the past 8 years. She never had chronic cough, loss of weight or trauma. Her appetite and bowel habits were satisfactory throughout this period. Her pitch and caliber of voice was normal, without any dysphagia, loss of taste or ear ache. Her family history was insignificant. General physical and systemic examinations were normal. Local examination revealed 3 x 2 cm and 3.5 x 3 cm globular firm rubbery non-tender cervical masses located anterior to the sternocleidomastoid muscles just below the angles of mandibles on the right and left sides, respectively. (Figure 1) They were well-mobile horizontally but had restricted mobility in

vertical direction. Non-compressible, they were pulsatile on deep palpation and they did not move with deglutition. No other lumps were felt in any triangles of the neck. Her ear, nose, throat and indirect laryngoscopic examinations did not reveal any abnormality. Her bimanual oral examination was normal.

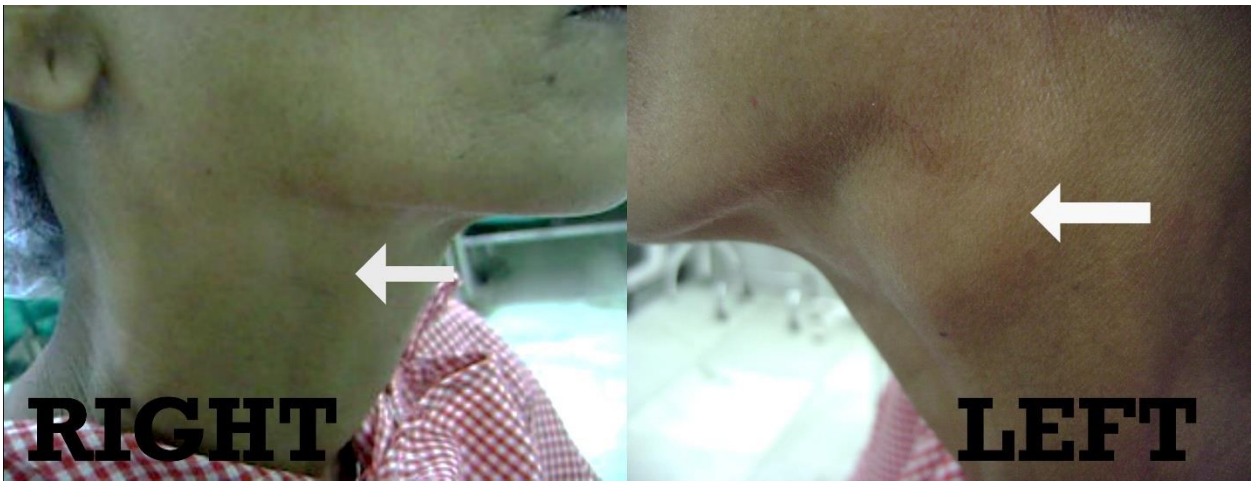

Fig. 1 Clinical picture of carotid body tumor: wolf in sheep's clothing. Note, the typical site *(white arrow)* could simply prompt a clinician to make not only erroneous but hazardous diagnosis of cervical lymphadenopathy before advising a fatal biopsy.

Local ultrasonography revealed solitary well-defined hypoechoic lesions on either side of neck located at the carotid bifurcations. On addition of color Doppler, they exhibited intense tumor-blush. A contrast-enhanced computed tomography (CECT) scan with digital subtraction angiography (DSA) demonstrated two distinct well-demarcated masses—3.2 x 2.3 x 1.5 cm and 3.7 x 3.2 x 2 cm at the carotid bifurcations on right and left sides, respectively. They were homogenously enhancing, loosely abutting against ECA for about 80^{o} on either side without infiltration. Maintaining the calibers, tumors splayed respective ECAs and ICAs distinctly. (Figure 2)

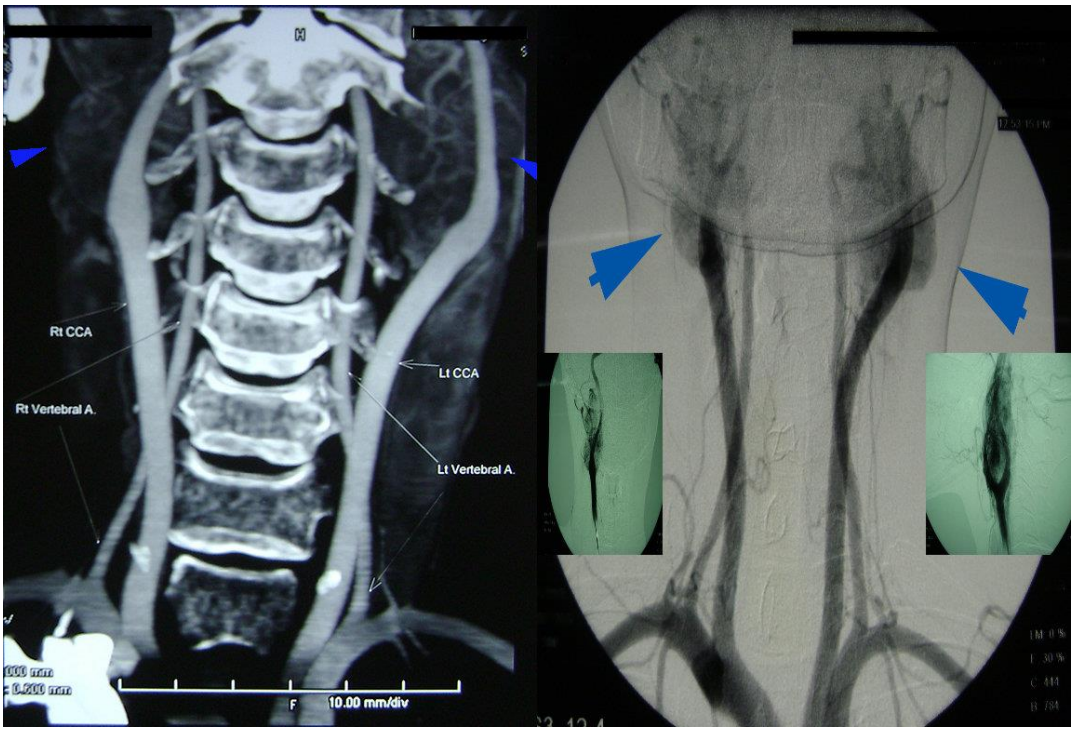

Fig. 2 CECT (3D reconstruction) with DSA of carotid body tumor. Note, bilateral intensely hypervascular masses (*blue arrow-heads and arrows)* at but not encircling the carotid bifurcation, splaying ECA and ICA—the pathognomonic lyre sign. Also note the absence of tumor thrombus or vascular dilation.

CECT, contrast-enhanced computed tomogram; 3D, 3 dimensional; DSA, digital subtraction angiogram; ECA, external carotid artery; ICA, internal carotid artery

Question

1. What is the probable diagnosis?

a. Tuberculous lymphadenitis

b. Carotid body tumors

c. Thyroid nodules

d. Enlarged submandibular glands

Answer

b. The clinical presentation and the radiological findings are typical of carotid body tumor. However, the patient refused staged excision and opted for surveillance. Over one year follow-up, she is doing well and the lumps have remained unchanged in size and character.

Discussion

Carotid body, a relatively small but extremely vital organelle situated at the carotid artery bifurcation, was first described by Von Haller in 1743.[1,2] A part of paraganglion system, it contains a conglomeration of highly vascular and innervated chemoreceptor tissue specialized in ameliorating hypoxia, hypercapnea and acidosis by modulating the autonomic nervous pathway.[2-4] These carotid bodies produce catacholamines and are said to be essential during fetal development.[2]

Carotid body tumor (CBT), better nominated as carotid body paraganglioma, was first reported by Von Luschka in 1862.[2] It could be sporadic (commonest), familial (due to mutations in succinate dehydrogenase gene) or hyperplastic (secondary to chronic hypoxia like at high altitudes).[2-4] As such, bilateral disease is less common—seen more frequently in familial (30%) than in sporadic (5%) variety.[2-4] And, only 5-10% eventually undergoes malignant transformation.[3]

The most important “rate limiting” step in its management is keeping a high index of suspicion generated after a sound physical examination and rational judgment; or else, the clinician could easily fall to its deceptive clinical appearance and tend to offer commoner differentials like lymphadenopathy or thyromegaly (others being salivary gland tumors or carotid aneurysms) leading to disastrous consequences like advising needle aspiration or even a biopsy.[1,2,4] Here,

though its typical clinical presentation (painless cervical lump) could be inherently confusing, the Fontaine sign—free mobility in horizontal rather than cranio-caudal plane, could clinch the clinical diagnosis in favor of CBT.[1,2]

Duplex ultrasound, a handy initial investigation, reveals a hypervascular tumor at carotid bifurcation.[2,4] However, Computerized Tomography and Magnetic Resonance angiography are modern non-invasive vascular mapping tool that produce reliable high-quality images depicting homogenously-enhancing lesion splaying ICA and ECA (the classic lyre sign), extent of infiltration and degree of encircling carotid artery along with its precise size and cranio-caudal extent.[1,2,4,5] Based on these criteria, Shamblin et al grouped CBTs into type I (small size, < 180^{o} carotid encircling), type II (medium size, 180^{o}-270^{o} carotid encircling) and type III (large size, > 270^{o} carotid encircling).[1,2] While early lesions (type I and II) merit immediate curative surgical excision by sequential cranio-caudal or retro-carotid dissection techniques, it is considered prudent to administer preoperative chemo-embolization and radiation to advance tumors (type III) before subjecting them to extirpative surgery involving complicated vascular reconstruction.[1,2]This reduces peroperative bleeding and major cranial nerve (X-XII) deficits as high as 40%.[3] However, for bilateral disease, a staged approach is recommended in which larger tumor is excised before smaller at two separate sittings to avoid labile hypertension.[3]

Thus, CBTs demand high degree of suspicion and optimum imaging for pin-pointing the diagnosis. This ultimately incurs multi-disciplinary involvement much critical to achieve better results.[1]

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