

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Bilateral sporadic carotid body tumors—A rare case report

Jipsa Anand, MBBS, DNB radiology resident*, Janender P. Singh, MBBS, MD

Max Super Speciality Hospital, Mohali, Punjab, India

ARTICLE INFO

Article history:

Received 9 May 2018

Revised 15 June 2018

Accepted 8 July 2018

Available online 9 August 2018

Keywords:

Carotid body tumor

Hypoechoic

Homogeneous enhancement

ABSTRACT

Carotid body tumor presents as palpable mass in anterolateral aspect of the neck. However, bilateral sporadic carotid body tumor is a rare anomaly, reported to occur in 5% of population. We are going to report the case of a 60-year-old patient who presented to our department with painless, palpable mass bilaterally in anterolateral aspects of the neck. He did not have any family history of neck masses. This paper summarizes the topic of carotid body tumor and discusses the radiological and clinical implications of this condition.

© 2018 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license.

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

Introduction

Carotid body tumors occur frequently in adults averaging 45–50 years of age and are uncommon in children. Familial forms account for 10% with bilateral tumors seen in 32% of cases. In sporadic cases, bilateral tumors are detectable in only 5% of patients. While the majority of these tumors are benign, 2%–13% pursue a malignant course with metastases to regional lymph nodes, lungs, and bones [3–4]. Medical history and physical examination are essential for the diagnosis of carotid body tumor. As direct biopsy is not suitable for the diagnosis of carotid body tumor due to the vascular nature of carotid body tumor, diagnostic imaging modalities are important in the diagnosis and differential diagnosis of this condition. Based on vascularity and location, ultrasound scan, CT (computed tomography), and MRI (magnetic resonance imaging) are able to diagnose carotid body tumor and differentiate it from many other masses in the neck [10–12].

Case report

A 60-year-old patient presented to our department with painless, palpable mass bilaterally in anterolateral aspects of the neck. His other complaints included dizziness, headache, flushing, palpitations, and tachycardia. Physical examination revealed a mass which was pulsatile, located below the angles of the mandible and was mobile laterally but fixed vertically. The mass was nontender, firm, and incompressible.

Ultrasonography demonstrated a solid, well-defined, hypoechoic mass located within the carotid bifurcation which was causing splaying of the bifurcation and separation of the internal and external carotid arteries (ECA) bilaterally. On color flow imaging, hypervascularity of the tumors were seen as irregular color signals with flow direction being predominantly cephalad. Both tumors had low resistance character with a high diastolic component. Power Doppler imag-

* Corresponding author.

E-mail addresses: drjipsa.anand@gmail.com (J. Anand), Janender.Singh@maxhealthcare.com (J.P. Singh).

<https://doi.org/10.1016/j.radcr.2018.07.001>

1930-0433/© 2018 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license. (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

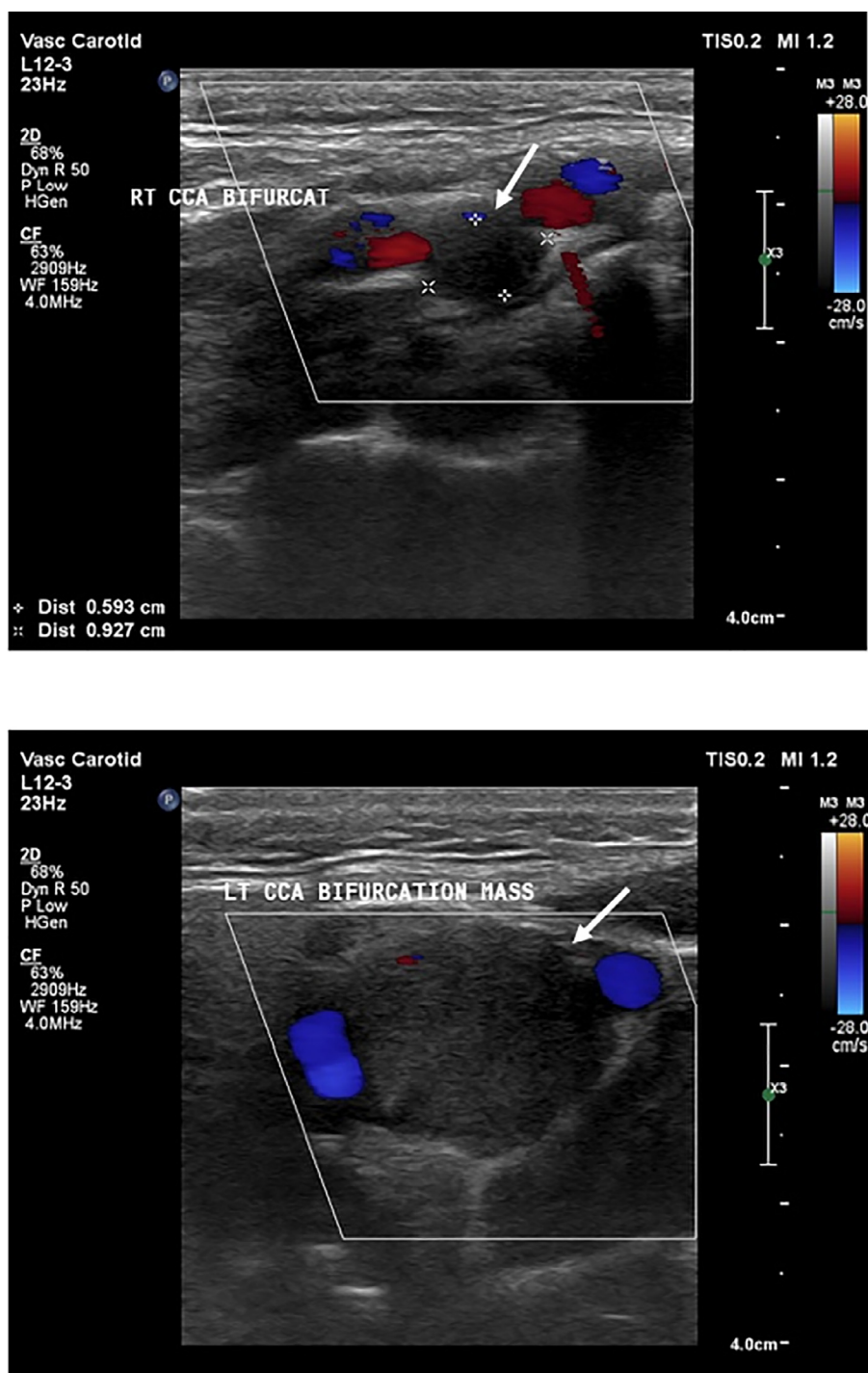


Fig. 1 – (A and B) Ultrasound shows solid, well-defined, hypoechoic mass located within the carotid bifurcation on right and left side, respectively.

ing shows abundant flow, characterized as intense blush, throughout the tumors. On the basis of location, ultrasonographic appearance and Doppler finding other solid non-hyper vascular neck masses such as lymph nodes, metastases, salivary gland tumors, or cervical cysts were ruled out.

CT demonstrated a well-defined solid mass that showed homogeneous enhancement on intravenous contrast administration in the regions of carotid bifurcation on both side causing splaying of internal carotid artery (ICA) and ECA approximately measuring $2.7 \times 1.9 \times 1.8$ cm on left side and $1.4 \times 0.9 \times 0.8$ cm on right side. The presence of uniform contrast enhancement and large feed-



Fig. 2 – (A and B) Sagittal section of CECT shows well-defined solid mass with homogeneous enhancement in carotid bifurcation region on both side causing splaying of ICA and ECA on right and left side, respectively.

ing vessels helped it to differentiate from lymph node masses and schwannomas. It was differentiated from the glomus vagale tumors, which is the paraganglioma of vagus nerve by its relative location in the neck, with glomus vagale tumors arising slightly more cephalad and presenting as an intensely vascular, enhancing mass on contrast enhanced computed tomography (CECT), displacing the ICA and ECA anteriorly and the internal jugular vein posteriorly. Differentiation between vagal schwannomas and carotid body tumor was done, as vagal schwannomas displaces the internal jugular vein laterally and the carotid artery medially. Also, schwannomas from the cervical sympathetic chain displace both the carotid and internal jugular vein without



Fig. 3 – Coronal section of CECT shows well-defined solid mass with homogeneous enhancement in carotid bifurcation regions bilaterally.

separating them, whereas carotid body tumor causes splaying of ICA and ECA. So, after ruling out other differentials, diagnosis of carotid body tumors was made (Figs. 1–5).

Discussion

The carotid body was first described by Von Haller in 1743, who described it is a chemoreceptor located in the adventitia of the carotid bifurcation [2]. The carotid bodies (glomera carotica) are reddish brown, ellipsoidal structures 5-7 mm in length and 2.5-4 mm in width, embedded in the outer fibrous layer of the common carotid arteries. When the carotid body detects decreasing levels of oxygen (hypoxia), increasing levels of carbon dioxide (hypercapnia), and decreasing pH (acidosis), it increases respiratory rate, tidal volume, heart rate, and blood pressure together with vasoconstriction and the production of circulating catecholamines. Embryologically, carotid body is derived from the mesodermal elements of the second arch artery and elements originating from the neural ectoderm.

Carotid body tumor is hypertrophy of the carotid body tissue, which is the most common type of the paraganglioma. It is also known as a chemodectoma, endothelioma, glomus caroticum, perithelioma, chromaffinoma, and nonchromaffin paraganglioma [1]. These tumors may be functionally active (ie, catecholamine secreting) or nonactive. Carotid body tumors may be familial (10%-50%) or nonfamilial. The incidence of bilateral carotid body tumors is 32% in the familial group and 5% in the nonfamilial group. Most carotid body tumors are slow growing and benign. Approximately 5%-10% of carotid body tumors may progress to malignancy, with local vasculo-

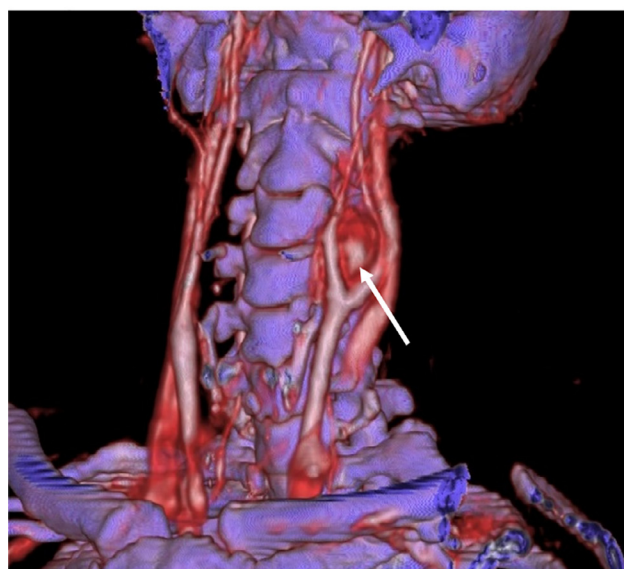


Fig. 4 – (A and B): On 3D reconstruction, highly vascular masses are seen bilaterally in carotid bifurcation regions with splaying of external and internal carotid arteries. 3D, 3-dimensional.

lar and/or lymph node invasion, and rarely distant metastases. The initial presentation of carotid body tumor is usually a painless, palpable, slow-growing, pulsatile rounded neck masses anterior to the sternocleidomastoid at the angle of the mandible at the level of hyoid bone [7–9]. Involvement of the cranial nerves that travel through the carotid sheath (the glossopharyngeal, vagus, accessory, and hypoglossal nerves) can lead to associated signs and symptoms including dyspha-

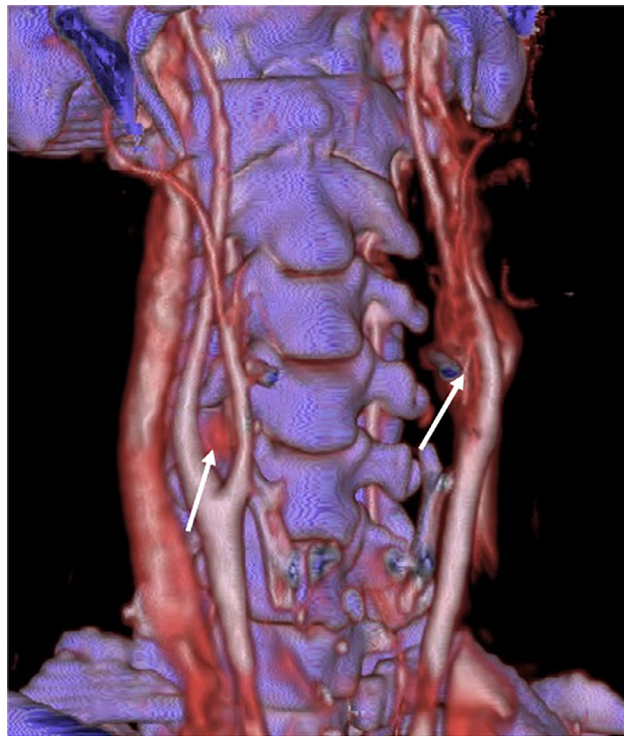


Fig. 5 – On 3D reconstruction, highly vascular masses are seen bilaterally in carotid bifurcation regions causing splaying of external and internal carotid arteries. 3D, 3-dimensional.

gia, hoarseness, dysarthria, swallowing difficulties, and shoulder drop. Other symptoms may include dizziness, headache, flushing, palpitations, tachycardia, arrhythmias, diaphoresis, and photophobia.

Physical examination reveals a mass which may be pulsatile, located below the angle of the mandible, typically laterally mobile but vertically fixed. The mass is usually nontender, rubbery, firm, and noncompressible. A bruit may be audible. Neurologic abnormalities caused by vagal or hypoglossal nerve involvement and Horner's syndrome are unusual but may appear in some patients [3].

The characteristic splaying of the internal and ECA is known as the lyre sign. On contrast-enhanced CT, the carotid body tumor appears as an avidly enhancing hypervascular mass at the carotid bifurcation with splaying of the ECA anteriorly and ICA posteriorly. MRI may demonstrate serpentine or punctate vascular flow voids (“pepper”) on T2 MR sequences, corresponding with vascular structures. The ascending pharyngeal artery is the typical arterial feeder [6]. Arteriovenous shunting may be seen on angiography. Surgical excision is the treatment of choice with preoperative embolization of larger lesions. Surgical outcome is related to the Shamblin classification of carotid body tumor (CBTs), which predicts tumor resectability and vascular morbidity. MRI findings can be used to predict the Shamblin surgical classification by noting the degree of contact of the tumor relative to the ICA with regard to $<180^\circ$, between 180° and 270° , and $>270^\circ$;

additionally, vascular splaying $>90^\circ$ indicates a less easily resectable tumor [5–6].

Conclusion

When presented with an enhancing carotid space mass, imaging is the primary diagnostic work-up, since biopsy of a highly vascular tumor is not recommended. Duplex ultrasound is a noninvasive, inexpensive, and readily available diagnostic tool, capable of diagnosing carotid body tumor based on its vascularity and location, and can be used as a first-line imaging modality for the diagnosis and assessment of carotid body tumors. Contrast-enhanced CT readily identifies an enhancing neck mass; the pattern of vascular displacement or splaying may suggest a particular diagnosis. Carotid body tumor is diagnosed by a high index of suspicion and confirmed by investigations.

Competing Interests

I had full access to all study data, take fully responsibility for the accuracy of the data analysis, and have authority over manuscript preparation and decisions to submit the manuscript for publication.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2018.07.001](https://doi.org/10.1016/j.radcr.2018.07.001).

REFERENCES

- [1] Pellitteri PK, Rinaldo A, Myssiorek D, Gary Jackson C, Bradley PJ, Devaney KO, et al. Paragangliomas of the head and neck. *Oral Oncol* 2004;40:563–75.
- [2] Irons GB, Weiland LH, Brown WL. Paragangliomas of the neck: clinical and pathological analysis of 116 cases. *Surg Clin North Am* 1977;57:575–83.
- [3] Lees CD, Levine HL, Beven EG, Tucker HM. Tumors of the carotid body: experience with 41 operative cases. *Am J Surg* 1981;142:362–5.
- [4] Romanski R. Chemodectoma (non-chromaffinic paraganglioma) of the carotid body with distant metastases with illustrative case. *Am J Pathol* 1954;30:1–13.
- [5] Silver AJ, Mawad ME, Hilal SK, Ascherl GF, Chynn KY, Baredes S. Computed tomography of carotid space and related spaces. Part II: neurogenic tumors. *Radiology* 1984;150:729–35.
- [6] Dhiman DS, Sharma YP, Sarin NK. US and CT in carotid body tumor. *Indian J Radiol Imaging* 2000;10:39–40.
- [7] Power AH, Hallett JW. Carotid body tumors. In: Stanley JC, Veith FJ, Wakefield TW, editors. *Current therapy in vascular and endovascular surgery*. 5th ed. Philadelphia: Elsevier; 2014. p. 141–5.
- [8] La Muraglia GM, Patel VI. Carotid artery. In: Cronenwett JL, Johnston KW, editors. *Rutherford's vascular surgery*. 8th ed. Philadelphia: Elsevier; 2014. p. 1598–614.
- [9] Day TA, Bewley AF, Joe JK, et al. Neoplasms of the neck. In: Flint PW, Haughey BH, Lund V, et al., editors. *Cummings otolaryngology*. 6th ed. Philadelphia: Elsevier; 2015. p. 1787–804.
- [10] Arya S, Rao V, Juvekar S, Dcruz AK. Carotid body tumors: objective criteria to predict the Shamblin group on MR imaging. *Am J Neuroradiol* 2008;29(7):1349–54.
- [11] Wieneke JA, Smith A. Paraganglioma: carotid body tumor. *Head Neck Pathol* 2009;3(4):303–6.
- [12] Ibeh Chinwe, Potigailo Valeria. Enhancing carotid space mass. *J Am Osteopath Coll Radiol* 2015;4(3):24–6.