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Cervical Paraganglioma Presenting as Thyroid Neoplasia

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Keywords

inferior laryngeal, cervical, carotid body, mediastinal, paraganglioma, thyroid, neoplasia, computed tomography, contrast media, iodine, gadolinium, magnetic resonance imaging, positive emission tomography, radioiodine, radiotherapy

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We report an unusual location of cervical paraganglioma, presenting as thyroid neoplasia, and discuss its possible origin from ectopic, laryngeal, or mediastinal paraganglia. Misdiagnosis of neck lesions may expose the surgeon and patient to inappropriate procedures.¹ Paragangliomas are rare tumors derived from the neural crest of the autonomous nervous system,² with an incidence of 0.2 to 1 per 100,000.² Head and neck paraganglioma are commonly associated with carotid body, jugulotympanic, and vagus nerve origins.³ They are usually non-secretory and benign. Preoperative diagnosis is often difficult through clinical findings and radiology.³ Surgery and radiotherapy represent the main treatment modalities.³

Institutional Review Board approval was obtained from the Western Health Low Risk Ethics Panel.

Case Report

A 42-year-old woman was referred with 8 weeks of increasing neck mass and hoarse voice. The referring physician had organized all imaging. Ultrasonography revealed a large 50 × 38 × 84 mm solid hypervascular nodule of the right thyroid with retrosternal extension. Nuclear thyroid scan showed a cold solitary nodule. Noncontrast computed tomography (CT) demonstrated homogenous soft-tissue attenuation, adjacent to the posterior right thyroid lobe, down to the level of aortic arch (**Figure 1a**). Fine-needle aspirate biopsy showed cells with mildly pleomorphic nuclei that favored an epithelial neoplasm, probably follicular in origin. Nasendoscopy revealed normal bilateral vocal cord movement.

At surgery for right hemithyroidectomy, the mass was found to be continuous with the carotid sheath, not involving the thyroid capsule (**Figure 2**), and converted to an open incisional biopsy. Immunohistochemical staining was

positive for CD56, synaptophysin, and focal chromogranin and negative for TTF-1 and calcitonin. A histological diagnosis of paraganglioma was made. Urgent contrast CT (**Figure 1b**) and magnetic resonance imaging were requested (**Figure 1c**). Both showed a hypervascular nodule that arose separately from the thyroid. Positron emission tomography showed moderate heterogeneous fluorodeoxyglucose uptake (**Figure 1d**).

The patient underwent surgical excision and right hemithyroidectomy via a hemisternotomy approach. The thyroid appeared normal with no invasion. Right recurrent laryngeal and phrenic nerves were identified and preserved. None of the 7 mediastinal lymph nodes were involved, and no tumor was seen in a tracheal biopsy. The paraganglioma was excised partially encapsulated, 85 mm in diameter, with foci of extension to resection margins. After discussion at an oncology multidisciplinary meeting, the patient received external beam radiotherapy of 45 Gy in 25 fractions (1.8 Gy/fraction) at 3 months, with ENT surgery review for residual hoarse voice.

Discussion

Nodular neck lesions that mimic thyroid pathology are rare but represent a challenge for the surgeon to obtain the correct diagnosis.¹ Regarding surface anatomy, the paraganglioma was palpable above the right sternoclavicular articulation, lateral to the thyroid. Differential diagnoses include an inferior derivative of carotid body paraganglia, ectopic or novel tissue not yet described.² Paragangliomas that arise within the thyroid and surrounds, between thyroid cartilage and first tracheal ring, may be derived from inferior laryngeal paraganglia.² Called subglottic or laryngeal paraganglioma, they are extremely rare.² While the patient's female gender, middle-age group, and right-sided laterality² was associated with laryngeal origin, the paraganglioma appeared more inferior on radiology (**Figure 1**). It arose at

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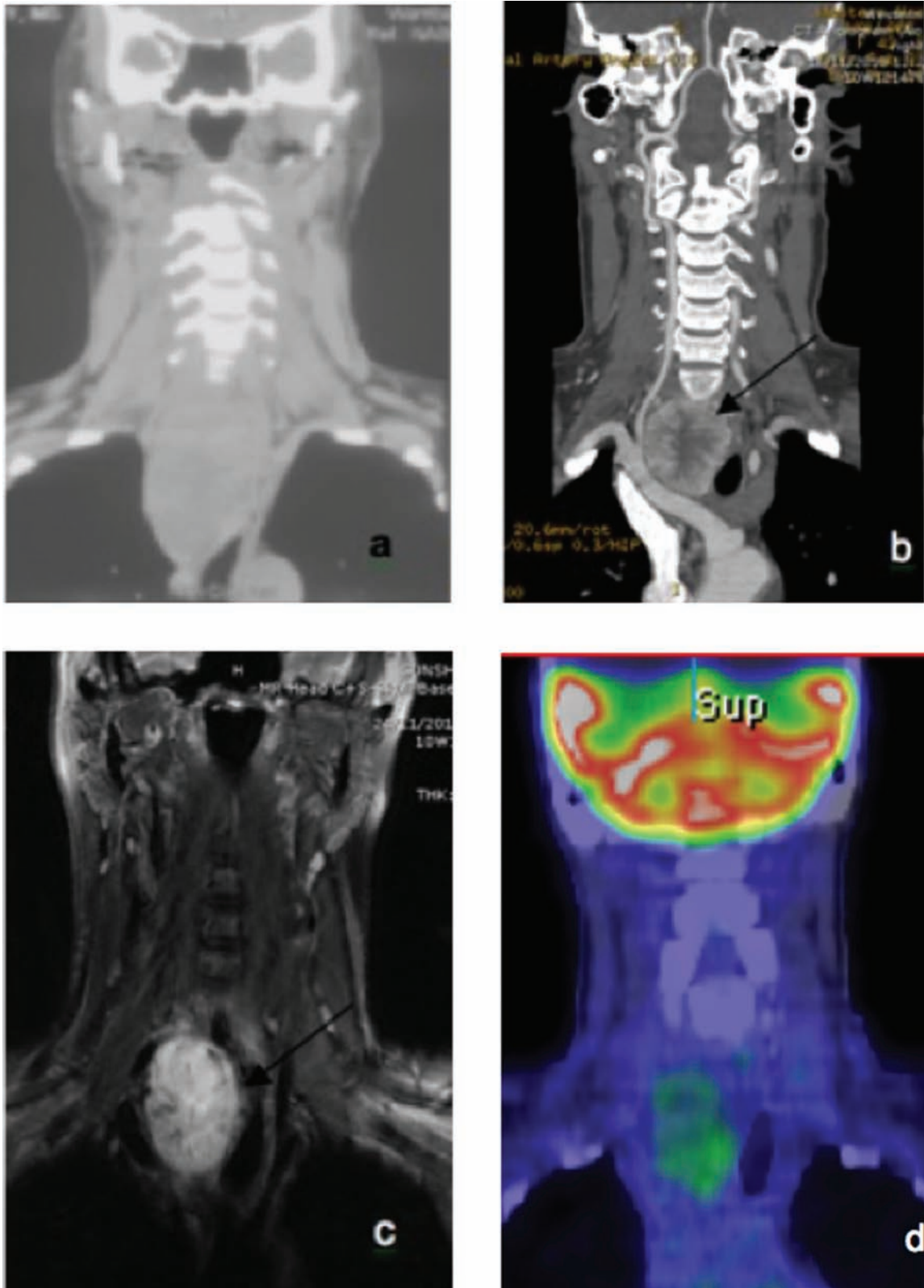


Figure 1. Neck computed tomography (CT) without contrast (a) showing homogenous attenuation, compared with CT with iodine contrast (b) and magnetic resonance imaging with gadolinium contrast (c), both of which demonstrate a well-defined, highly contrast-enhanced tumor (arrow) medial to the right common carotid artery and (d) positron emission tomography scan showing heterogeneous fluorodeoxyglucose uptake.



Figure 2. Photograph taken at incisional biopsy, demonstrating right carotid sheath lateral (1) continuous with paraganglioma (2) and separate from right thyroid lobe superior medial (3).

the level of bifurcation of the brachiocephalic artery, medial to the right common carotid, at the root of the neck.

This raises the further differential diagnosis of mediastinal origin, such as from para-aortic or paravertebral sympathetic chain ganglia.⁴ Paragangliomas are usually located in the bifurcation of great vessels,⁴ as in this case, and biopsy can be hazardous because of such proximity and vascularity.⁴

Noncontrast CT was unable to completely characterize the pathology. Iodine contrast is preferentially avoided to prevent the Wolff-Chaikoff dilution effect and delay of radioiodine therapy for thyroid neoplasia. Iodine contrast, however, is a relative and not absolute contraindication to radioiodine. Contrast media can therefore be used to delineate difficult anatomy.

In conclusion, surgeons should be aware of unusual locations of cervical paraganglioma. Complete resection is the standard of care and is associated with excellent survival.⁴ Radiotherapy, as a treatment option, is often reserved for

residual disease following surgery, unresectable tumors, and the medically inoperable⁵ and is delivered using modern techniques at an appropriate dose (40-50 Gy) and fractionation (1.8-2 Gy/fraction).⁵ The highly vascular nature and strategic anatomical locations of cervical paraganglioma make complete resection demanding and lifelong surveillance for local recurrence and metastatic spread mandatory.⁴

Author Contributions

Violet Kieu, corresponding author, data analysis, manuscript writing; **Anita Yuen**, data analysis and surgical consultation; **Peter Tassone**, surgical consultation and manuscript revision; **Chris G. Hobbs**, surgeon supervisor and final manuscript.

Disclosures

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