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Case Report

Paraganglioma of the Recurrent Laryngeal Nerve

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ABSTRACT

Background/Objective: Paragangliomas are rare neuroendocrine tumors that primarily arise in the adrenal gland. Head and neck paragangliomas comprise approximately 3% of all extra-adrenal paragangliomas, with a majority of those being found in the carotid body. Recurrent laryngeal nerve paragangliomas are exceedingly rare, with only 2 reported cases found in literature review. Here, we will present the third.

Case Report: The patient is a 46-year-old woman with a history of a right thyroid nodule that had been previously biopsied benign with "paucity of diagnostic material." Neck ultrasonography revealed a 7.4 cm nodule that demonstrated interval growth over a 2-year period, so it was recommended to proceed with right thyroid lobectomy and isthmusectomy. During resection, the recurrent laryngeal nerve appeared to "disappear" into the nodule, and it was resected along with the nodule to ensure proper margins. The nerve was reconstructed with an ansa cervicalis interposition graft, and the nodule was sent to pathology. Pathology revealed that the nodule was a 4.8 cm paraganglioma of the recurrent laryngeal nerve.

Discussion: Paragangliomas of the head and neck are exceedingly rare. In patients who present with symptoms of dysphagia or dysphonia, further workup, including laryngoscopy and magnetic resonance imaging, could potentially identify and allow for appropriate planning for surgical resection.

Conclusion: In rare cases, consideration of paraganglioma as part of the differential for thyroid nodules may assist with planning of surgery but will unlikely alter treatment.

Introduction

Paragangliomas are rare tumors that arise from neuroendocrine tissue. Approximately 90% arise from the adrenal gland and are traditionally referred to as pheochromocytomas, with the remaining 10% originating from the abdomen (85%), thorax (12%), and head and neck (3%) regions.[1] Head and neck paragangliomas generally present in mid–adult life, and a majority are benign, asymptomatic, and nonfunctional, with the most common location being the carotid body, followed by jugular foramen and vagal nerve; there are also documented cases in the nasopharynx, nasal cavities, paranasal sinuses, larynx, thyroid, and orbit.[1,2] Literature search on PubMed revealed 2 other reported cases of paraganglioma arising from the recurrent laryngeal nerve.[3,4] Here, we will present the third case.

Case Report

The patient is a 46-year-old woman with a history of hypothyroidism and a right thyroid nodule. Fine needle aspiration (FNA) was interpreted as "favoring benign follicular nodule. Paucity of diagnostic material precluded definitive diagnosis." Interval enlargement was noted over a 2-year period (from 5.6 cm to 6.3 cm), and the patient was referred for endocrine surgical evaluation. Neck sonography performed in our office revealed a 7.4 cm hypoechoic mass within the right thyroid lobe (Fig. 1 through 3). The patient denied dysphonia or dysphagia with liquids; therefore, no direct or indirect laryngoscopy was performed prior to recommending to proceed with right thyroid lobectomy and isthmusectomy to rule out malignancy. Intraoperatively, the thyroid was

Abbreviations: FNA, fine needle aspiration; MRI, magnetic resonance imaging; SDHB, succinate dehydrogenase complex B.

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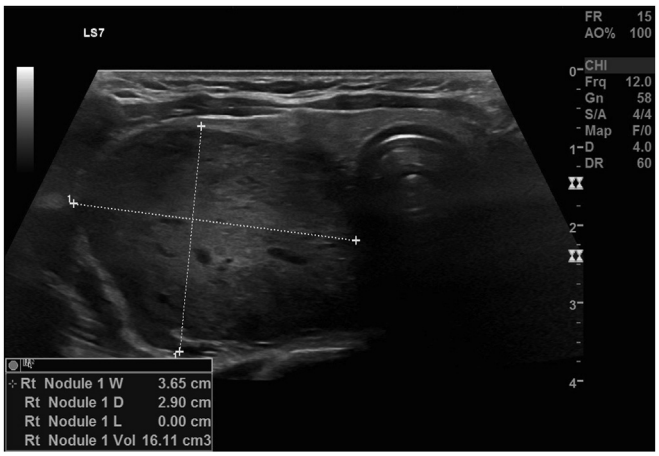

Fig. 1. Axial view of preoperative paraganglioma on ultrasound.

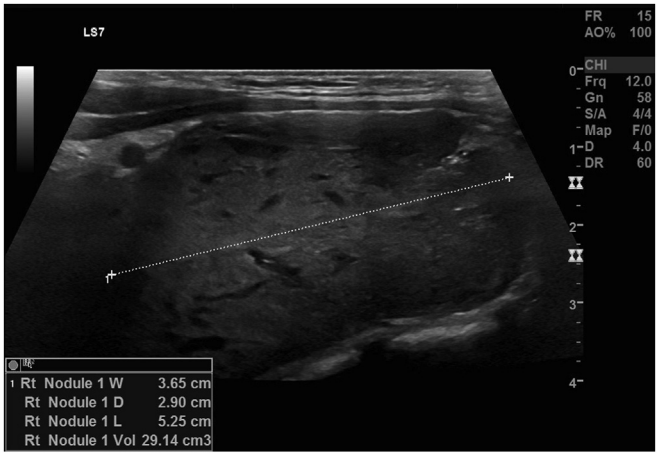

Fig. 2. Sagittal view of preoperative paraganglioma on ultrasound.

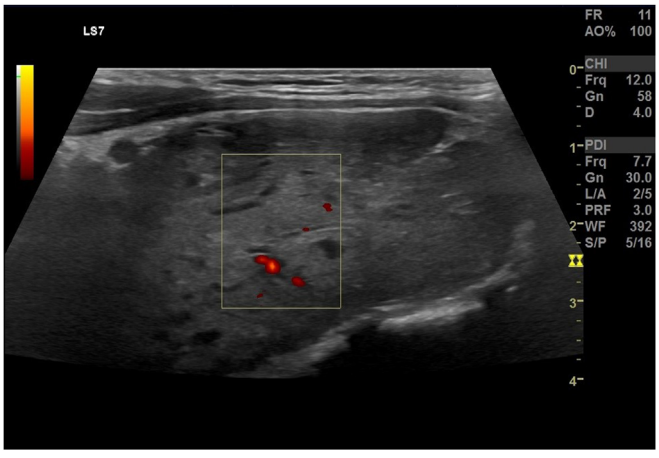

Fig. 3. Color Doppler of preoperative paraganglioma on ultrasound.

mobilized, the superior and inferior arteries were ligated along with the superior, middle, and inferior veins, and the distal recurrent laryngeal nerve was identified; however, upon attempts to trace the nerve retrograde, it appeared to "disappear" into the presumed thyroid nodule. The proximal recurrent laryngeal nerve was then identified and also appeared to disappear into the "thyroid nodule." In order to resect the potentially malignant nodule, the recurrent laryngeal nerve was divided proximally and distally and reconstructed with an ansa cervicalis interposition graft.

Routine pathology revealed that the "thyroid nodule" was in actuality a 4.8 cm paraganglioma of the recurrent laryngeal nerve. The mass was S-100 and synaptophysin and chromogranin–positive (Fig. 4) and was found to have normal expression of succinate dehydrogenase complex B (SDHB).[5] Despite this, the patient was sent for genetic testing at an outside institution, which identified no clinically significant variants for hereditary paragangliomas.

Postoperatively, the patient experienced hoarseness and, prior to discharge home, underwent a swallow study that did not identify aspiration with thin liquids. At the time of this publication, she is undergoing speech therapy and ear, nose, and throat evaluation.

Highlights

- Paragangliomas can present at the recurrent laryngeal nerve, with or without hoarseness of voice
- Paragangliomas present as well-circumscribed, vascular, hypoechoic lesions
- Fine needle aspiration (FNA) of paragangliomas can show paucity of diagnostic cells
- Consider cross-sectional imaging in large well-circumscribed lesions with paucity of cells on FNA

Clinical Relevance

Paragangliomas of the recurrent laryngeal nerve are extremely rare but can present without symptoms and should be considered in cases of large, well-circumscribed, vascular, hypoechoic nodules with paucity of cells on fine needle aspiration.

Discussion

Paragangliomas of the head and neck are rare, with an incidence of 1:30 000 to 1:100 000 and only 2 documented cases involving the recurrent laryngeal nerve. One of those cases was a patient with a previous history of left thyroid lobectomy that was believed to be a recurrent adenoma, whereas the other presented with hoarseness and vocal cord paralysis.[3,4] Our patient was asymptomatic, and ultrasound showed a hypoechoic mass with sharp, well-defined borders and grade 2 vascularity that had significantly increased in size, warranting surgical excision for diagnosis. This imaging was reviewed by multiple physicians who specialize in endocrine

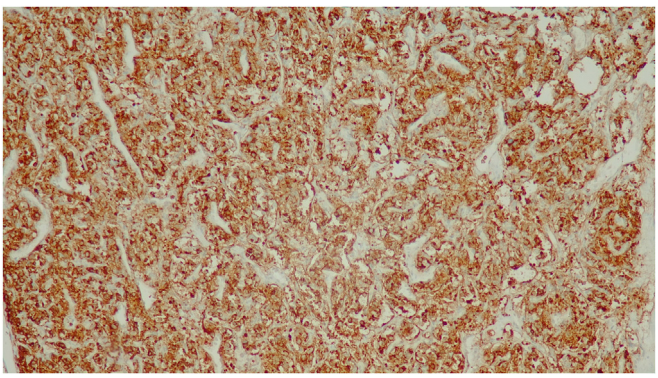

Fig. 4. Chromogranin stain of specimen.

oncology, and paraganglioma was never considered in the differential diagnosis. Retrospective review of the workup, imaging, and biopsy shows that there were scant follicular cells favoring a benign tumor, with a paucity of cells preventing definitive diagnosis on FNA. Although it has been described that diagnosis of paraganglioma can be difficult via FNA biopsy, this lack of cells and size of the mass could have warranted further workup, such as magnetic resonance imaging (MRI), which potentially could have allowed diagnosis of paraganglioma prior to planning surgery.[1,6] A preoperative diagnosis would have allowed further imaging, laryngoscopy to assess for vocal cord function, and earlier initiation of the appropriate genetic testing.[1,2,5] Traditionally, asymptomatic paragangliomas of the head and neck did not require biochemical testing unless there were symptoms to suggest they were secretory.[1] However, more recent recommendations suggest that every patient should undergo biochemical testing with plasma metanephrines or 24-hour urine metanephrines, as it can signify a more aggressive tumor.[7,8] Due to the location and ability for excision, surgical excision would still have been the recommended treatment; however, radiation therapy is another acceptable treatment option for paragangliomas that are tough to resect or when resection would leave the patient with significant morbidity. Observation can also be considered in patients in whom tumors are stable over time.[1,2] With this being said, the rarity of paragangliomas of the recurrent laryngeal nerve and the expense of MRI does not warrant this workup without high clinical suspicion.

Once in the operating room, upon discovery that the lesion is involved with the recurrent laryngeal nerve, resection with reinnervation using the ansa cervicalis is a good option for treatment, as it has been shown to improve phonation and reduce risk of aspiration equivalently to thyroplasty. Ansa cervicalis reinnervation also helps maintain muscle bulk.[9,10]

Following pathological discovery that the patient has a paraganglioma, it is important to test for SDHB to rule out inherited paraganglioma, which has increased chance at malignancy because SDHB, in particular, has been shown to be more commonly associated with head and neck paragangliomas.[5]

Conclusion

Paragangliomas of the head and neck are very rare, particularly those of the recurrent laryngeal nerve. Here, we present the third case of recurrent laryngeal nerve paraganglioma found in the literature. However, our patient presented with the unique feature of having no hoarseness or history of prior thyroid surgery. Because of the lack of confounding variables, cross-sectional imaging was not done; although, in hindsight, for patients with large well-circumscribed tumors in which a paucity of cells prevents definitive diagnosis, cross-sectional imaging, such as MRI or computed tomography scan, could be considered to provide additional detail prior to surgery.

Disclosure

The authors have no multiplicity of interest to disclose.

Author contributions

S.V.G., R.M.H., D.B., C.E., and G.A. diagnosed and operated on the lesion. T.D. wrote the initial case report. G.A. and D.B. revised it. All authors contributed to the final case report.

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