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INTRODUCTION

Paragangliomas have a neuroectoderm origin and arise from the extra-adrenal paraganglia of the autonomic nervous system.\(^1,2\) Paragangliomas of the head and neck are rare, and most frequently benign, slow growing, and nonsecretory.\(^1,3\) These tumors are often associated with neurovascular structures given the role of paraganglia as chemoreceptors.\(^4\) In the head and neck, the most frequent locations these tumors arise include the carotid body, jugular bulb, vagus nerve, tympanic branch of the glossopharyngeal nerve, and sympathetic chain.\(^4\) Additional reported locations in the literature include the sinonasal cavity, skull base, orbit, and larynx.\(^1,2,5\)–\(^8\) To our knowledge, there is only one report of a single case of a paraganglioma arising from the recurrent laryngeal nerve in the literature.\(^9\) Here we present the second known case.

CASE REPORT

The patient is a 50-year-old male with a 5-month history of waxing and waning hoarseness. He was initially treated with steroids with improvement in voice, although after several months his hoarseness recurred. The patient presented to an outside otolaryngologist and was found to have right vocal fold paralysis. Neck and chest computed tomography with contrast was ordered to evaluate the course of the recurrent laryngeal nerve, which demonstrated a 1.8 × 1.3 × 1.9-cm avidly enhancing, well-circumscribed soft tissue nodule posterior to the inferior pole of the right thyroid gland in the right tracheoesophageal groove (Fig. 1). No other cervical or thoracic pathology was identified. An ultrasound-guided fine-needle aspiration was again nondiagnostic (Fig. 2).

The patient was taken to the operating room for resection of the right tracheoesophageal groove mass. The right thyroid lobe was mobilized and a distinct rubbery mass completely separate from the thyroid lobe was palpated within the underlying tissue near the tracheoesophageal groove. The right recurrent laryngeal nerve was identified inferior to the mass. The mass, which did not extend to the paraglottic space, was invested and integrated with the right recurrent laryngeal nerve. The superior aspect of the recurrent laryngeal nerve was identified as it entered near the

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cricothyroid joint. The mass was completely excised, thereby sacrificing the RLN inferolateral to the insertion.

Pathologic examination demonstrated a well-circumscribed 2.5-cm mass composed of nests (zellballen) of polygonal cells with amphophilic granular cytoplasm, round to oval nuclei with single inconspicuous nucleoli, and a rich vascular plexus in the surrounding stroma (Fig. 3). Immunohistochemical stains showed that the tumor cells were positive for synaptophysin, insulinoma-associated protein 1, and GATA3; S100 highlighted sustentacular cells. Additionally, tumor cells demonstrated a loss of succinate dehydrogenase iron-sulfur subunit B (SDHB) immunohistochemical expression with preserved staining in background sustentacular cells and vasculature, consistent with a succinate dehydrogenase (SDH)-deficient paraganglioma.

A week after surgery the patient was seen by a laryngologist and underwent transcervical injection medialization laryngoplasty with Restalyne with notable improvement in voice. Five months later the patient then underwent definitive operative laryngoplasty medialization with a Gore-Tex implant. He has been doing well since with noted improvement in voice.

DISCUSSION

Paragangliomas of the head and neck are rare, with an overall incidence of 1:30,000 to 1:100,000.4 Given the neural crest origin, paragangliomas may arise along migratory routes of neural crest tissue.1 However, these tumors typically arise along neurovascular structures. In the head and neck, these tumors most frequently arise in the 5th to 6th decade of life, and the most frequent location of origin is the carotid body (most common), jugular bulb, and vagus nerve.3 Interestingly, only one other case of recurrent laryngeal nerve paraganglioma has been reported in the literature.9 In that case, an 87-year-old female with a history of left hemithyroidectomy 25 years prior for an adenoma presented with cough and a 6.5-cm nodule of the left thyroid consistent with recurrent thyroid adenoma. In that report, during resection, the recurrent laryngeal nerve could not be fully delineated due to its direct involvement with the tumor.9 The specimen contained a larger mass consistent on pathologic analysis and immunostaining with paraganglioma and abutted thyroid tissue, whereas a smaller nodule located within the thyroid tissue itself was found to be a Hürthle cell adenoma.9 Here we describe the second known case of recurrent laryngeal nerve paraganglioma, which in contrast did not have a history of head and neck surgery or prior pathology, and had a presenting symptom of ipsilateral vocal fold paralysis. Screening imaging may be important in patients who present with vocal fold paralysis of unknown etiology and should be considered prior to performing interventions.10,11

Interestingly, this specimen demonstrated a loss in SDHB expression consistent with SDH deficiency. Up to 40% of paragangliomas of the head and neck are associated with a hereditary predisposition for paraganglioma development most frequently through germline mutations in the succinate dehydrogenase complex.4 In one study, SDH germ-line mutations were discovered in 36 of 79 (45.6%) paragangliomas of the head and neck.12 Of these, 29 of 36 (80.5%) SDH mutations were in SDHD.12 Mutations in any of the SDH complex members results in loss of SDHB expression, thus SDHB expression is often used as a screening tool for SDH genes.3 Another factor known to predispose to carotid body paraganglioma
development is living at high altitude, which does not appear to be a factor in the case of recurrent laryngeal nerve paraganglioma presented here.

Vocal fold paralysis often causes dysphonia but can also have additional severe impact on quality of life and has been correlated with loss of income and reduced socialization. Thus, it is important to appropriately address treatment options. Surgical options can be frequently categorized into medialization versus reinnervation options. Medialization techniques were used in this patient and included a temporary Restalyn injection shortly after surgery followed later by operative medialization with an implant. Medialization could also alternatively be performed with an arytenoid adduction. In the setting of known operative sacrifice of the recurrent laryngeal nerve, one could also consider permanent methods of medialization soon after surgery. Medialization techniques have the advantage of rapid improvement in patient symptoms. Reinnervation techniques are another option and utilize a donor nerve, often the ansa cervicalis, to strive for a reinnervation and improvement in muscle bulk. Reinnervation often takes a prolonged period of time prior to an observed benefit and is often combined with a temporary injection medialization. Thus, multiple medialization and reinnervation methods are available to aid in relieving symptoms due to recurrent laryngeal nerve function deficits.

Management of paragangliomas of the head and neck is adapted to the specific presenting symptoms, imaging findings, tumor size/location, age, and patient risk profile. Although surgical excision of paragangliomas of the carotid body, jugular bulb, and vagus nerve may offer a high cure rate, there can be considerable morbidity and cranial nerve deficits as a result. Other treatment options for paragangliomas of the head and neck include fractioned radiation or stereotactic radiosurgery, which can aid in control of tumor growth, without reducing the size of the paraganglioma. However, in this case of paraganglioma of the recurrent laryngeal nerve, with preexisting nerve deficit, need for pathologic diagnosis, low morbidity of surgery, the benefits of radiosurgery appeared low. In many cases, observation may be considered with repeat imaging to determine tumor growth rate. Thus, treatment options must be individualized for each patient.

CONCLUSION

Paragangliomas of the head and neck are rare, often benign, slow growing, and nonsecreting, with a hereditary predisposition for those with SDH germline mutations.
Here we present the second known case of paraganglioma of the recurrent laryngeal nerve. This case is unique given the patient presentation due to ipsilateral vocal fold paralysis, which has not previously been reported, lack of previous surgery, and demonstration of loss of SDHB expression.

BIBLIOGRAPHY