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CASE REPORT

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Phrenic nerve schwannoma presenting as an asymptomatic neck mass

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Abstract

Phrenic nerve schwannomas of the head and neck are exceedingly rare pathologies that can present as an asymptomatic neck mass. Surgery is the definitive treatment, and a conservative surgical approach is preferred if a benign pathology is suspected.

KEYWORDS

head and neck, neurogenic tumor, phrenic nerve, schwannoma

1 | INTRODUCTION

Schwannomas are benign, slow-growing neurogenic tumors that arise from the Schwann cells of the myelin sheath.[1] The incidence of schwannomas in the head and neck region is reported to be as high as 45%.[1] However, schwannomas of the phrenic nerve in the head and neck region are exceedingly rare.[2,3] To our knowledge, there have been 4 reports in the English literature of phrenic nerve schwannomas arising from the cervical region over the past 20 years.[1,2,4,5] Neurogenic tumors of the phrenic nerve tend to be asymptomatic and benign.[1,5] However, they have potential for malignant transformation. Management options include observation or surgery, with consideration for radiation in patients who are not surgical candidates. The literature regarding radiation for cervical schwannomas is sparse; however, it suggests radiation is an option for extracranial head and neck schwannomas if the patient is not a surgical candidate.[6] Treatment itself can cause symptoms of phrenic nerve injury such as cough, dyspnea, diaphragmatic elevation/paralysis, and/or obstructive symptoms.[2,4] Therefore, the risks and benefits of treatment options must be considered carefully. We present a case of an asymptomatic phrenic nerve schwannoma treated with surgical excision.

2 | CASE

A healthy 55-year-old female presented to Penn State Otolaryngology-Head and Neck outpatient clinic with a 10-month history of a right neck mass noticed on self-examination. The mass was initially nontender and had been gradually increasing in size. Patient denied any associated otolaryngologic or systemic symptoms, such as dysphagia, odynophagia, dysphonia, otalgia, or dyspnea.

Physical exam revealed a firm area tender to palpation, deep to the posterior sternocleidomastoid muscle in level 3. A flexible nasopharyngoscopy was unremarkable, showing no mucosal masses or vocal fold paresis. A computed tomography (CT) scan of the neck revealed a soft tissue mass in level 3 of the right neck (Figure 1A,D). Magnetic resonance imaging (MRI) of the neck revealed a T1 hypointense and T2 hyperintense, well-circumscribed mass (1.7 x 1.4 x 1.3 cm) with enhancement posterior to the carotid sheath below the level of the hyoid (Figure 1B,C,E,F). The central portion of the mass demonstrated hypointensity on T2 weighted images. The differential diagnosis at this time included but was not limited to a neurogenic tumor, lipoma, carotid body tumor, or cancer of unknown primary. An initial fine-needle aspiration (FNA) showed only fibrous tissue and skeletal muscle. A repeat FNA was again nondiagnostic with scant spindle cells that may be consistent with a neurogenic tumor. After the second FNA, the patient developed paresthesias of the right shoulder and neck, extending to the right arm. These symptoms suggested possible involvement of cervical rootlets due to their anatomic distribution.

In concert with the radiologic findings and scant spindle cells noted on FNA, these were felt to be most consistent with a benign neurogenic tumor. The origin of the tumor was indiscernible from the images alone, but possibilities included cervical rootlets, the vagus nerve, the spinal accessory nerve, or the phrenic nerve. Given the low likelihood of malignancy, treatment options of observation versus surgery were discussed. The risks and benefits of surgical resection were discussed extensively. Although the patient was generally asymptomatic, aside from the progressive growth of the tumor, she decided to proceed with surgery for definitive treatment.

Intraoperatively, the spinal accessory nerve was identified and noted to be lateral to the mass. The mass was located lateral and posterior to the internal jugular vein and was not within the carotid sheath; therefore, involvement of the vagus nerve was ruled out intraoperatively. Intraoperative stimulation of the tumor with a nerve probe

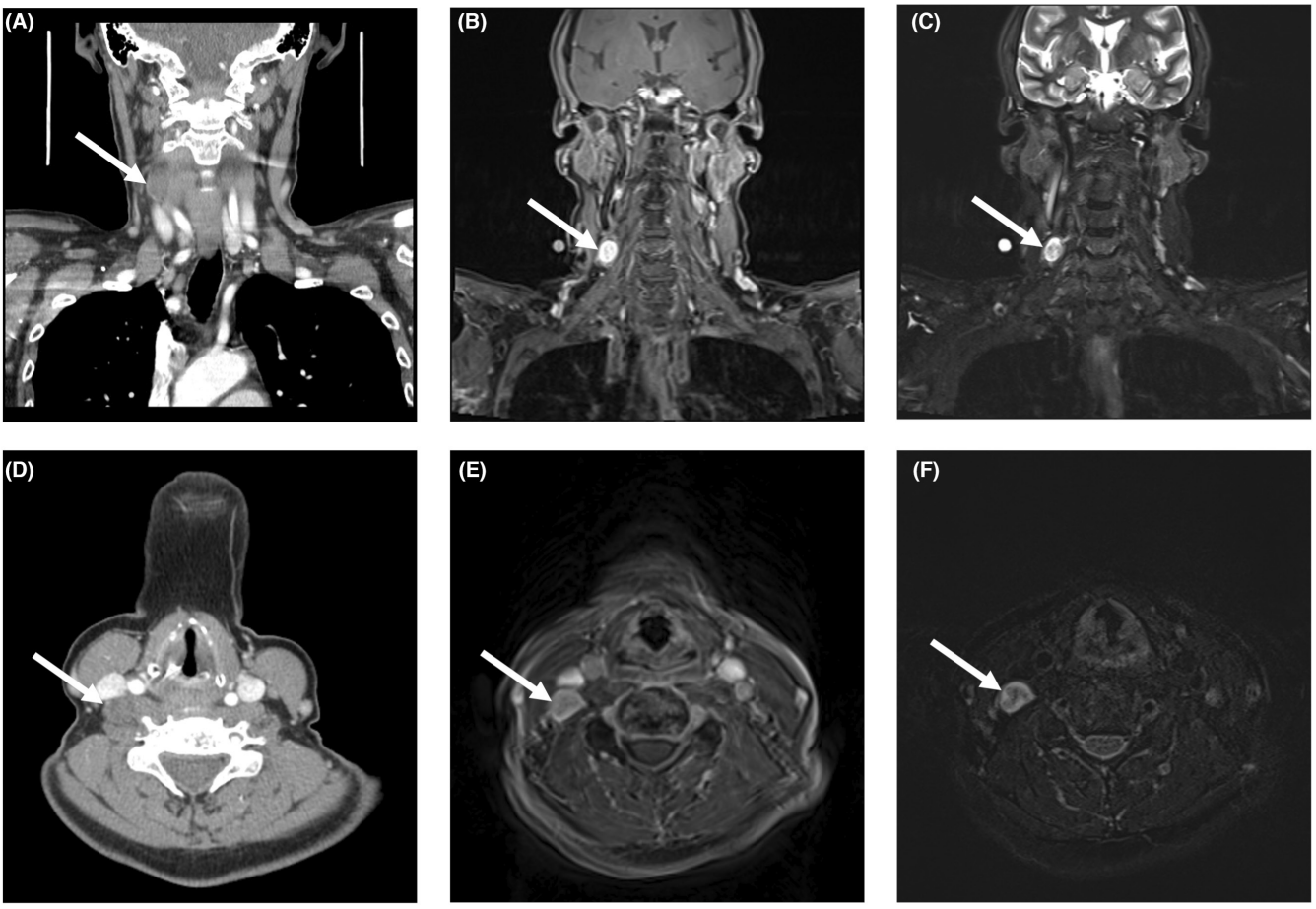

FIGURE 1 (A) Preoperative coronal CT scan showing a right-sided hypodensity at the C4-C5 level. (B) Preoperative coronal T1 Dixon MRI scan with contrast and fat saturation, showing a right-sided enhancing 17 x 14 x 13 mm mass posterior to the carotid sheath at the C4-C5 level. (C) Preoperative coronal T2 STIR (short-tau inversion recovery) MRI scan without contrast. (D) Preoperative axial CT scan. (E) Preoperative axial T1 VIBE (volumetric interpolated breath-hold examination) MRI with contrast and fat saturation. (F) Preoperative axial T2 Dixon MRI with fat saturation

at 0.5 mA resulted in activation of the diaphragm, confirming the mass was of phrenic nerve origin. Due to the origin of the mass, the decision was made for enucleation of the mass as opposed to transection in an effort to preserve the phrenic nerve function and avoid definite subsequent diaphragmatic paralysis. An incision was made in the tumor capsule, and the mass was delivered with gentle blunt dissection. Intraoperative stimulation of the phrenic nerve after enucleation showed activation of the diaphragm distal to the lesion but weakened activation proximally.

Postoperatively, a chest x-ray (CXR) was obtained to evaluate diaphragmatic function. The CXR showed signs of phrenic nerve weakness with elevation of the right hemidiaphragm (Figure 2). However, the patient did not experience dyspnea or any other respiratory symptoms. Final surgical pathology was consistent with a phrenic nerve schwannoma (Figure 3). Upon 2-month follow-up, the patient did not report any dyspnea or respiratory symptoms. Further follow-up imaging was not pursued as the patient remained asymptomatic.

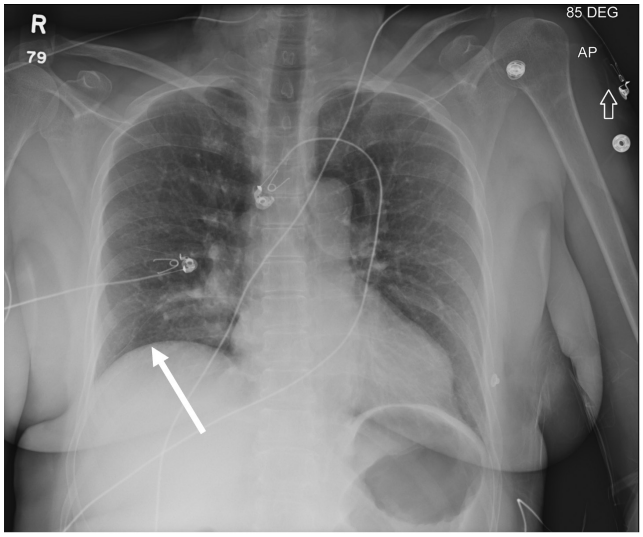

FIGURE 2 Postoperative chest x-ray showing elevation of the right hemidiaphragm. Arrow indicates elevated hemidiaphragm

3 | DISCUSSION

We describe an asymptomatic neck mass with an unusual nerve of origin. While we suspected a neurogenic tumor based on the cytopathology and imaging characteristics, the origin of the tumor was unclear. Benign neurogenic tumors are rare pathologies to begin with, and schwannomas of the phrenic nerve in the head and neck are even rarer.[1–3] In addition, the patient did not have any clinical symptoms associated with the mass with the exception of right neck and shoulder numbness following the second biopsy. The nerve of origin is important to consider because it directs the surgical approach and associated surgical risks. For example, involvement of the spinal accessory nerve can cause weakness in the shoulder and neck, while injury to the vagus nerve can cause dysphonia, blood pressure irregularities, and gastroparesis. Injury to the cervical rootlets could result in permanent skin anesthesia, or dysphagia related to strap muscle dysfunction, and injury to the phrenic nerve could result in diaphragmatic dysfunction and respiratory difficulty.

Even with a preoperative MRI, it was difficult to discern specifically where the mass was arising from. In the literature, it has been reported that phrenic nerve origin should be considered if the tumor appears to be connected to an interbody of C2-C5 vertebrae.[7] In our patient, the mass did appear to demonstrate this interbody connection at the C4-C5 level (Figure 1A–C).

In the literature, the management of phrenic nerve schwannomas in the head and neck is sparse. It has been more commonly described in the thoracic literature, where nerve preservation versus complete resection with nerve sacrifice remains controversial.[1,8–10] We believe the morbidity associated with phrenic nerve sacrifice should

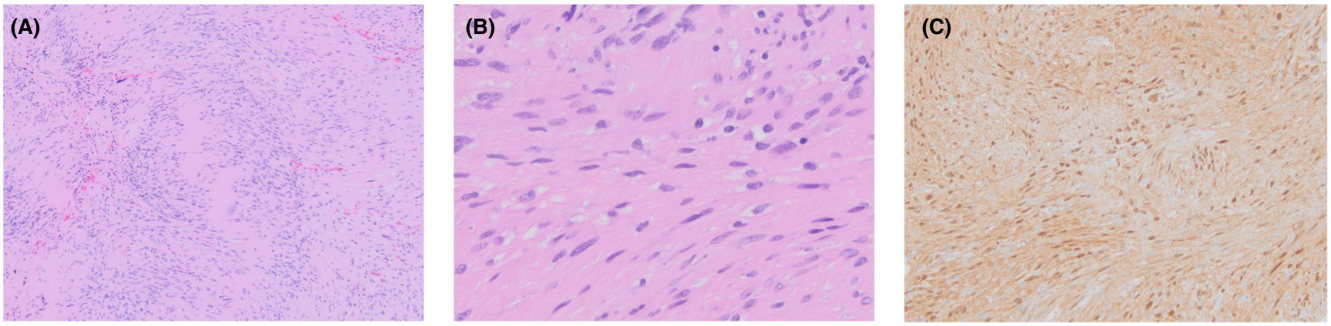

FIGURE 3 Hematoxylin and eosin (H&E) histopathology of neck mass. Hypercellular (Antoni A) and hypocellular foci (Antoni B) (A, H&E, 100x magnification). Spindled tumor cells with tapering nuclei (B, H&E, 400x magnification). Immunohistochemistry showed positivity for S100 (C, immunohistochemistry, 100x magnification)

be weighed with the malignant potential of the mass. Since neurogenic tumors tend to be benign with a low potential for recurrence, a conservative approach is usually preferred.[1] However, if malignancy is suspected or the potential for recurrence is high, such as in patients with Neurofibromatosis Type 1, then complete excision with the parent nerve should be considered.[1,10] Additionally, if preoperative assessment suggests the nerve has already been compromised, then complete resection rather than enucleation of the tumor is suggested.[2] In this case, the patient was aware of the associated risks, but desired definitive removal of the mass given its progressive growth. Given the patient's preference, the low risk for malignancy, and the origin of the mass determined intraoperatively, we opted for a conservative surgical approach with enucleation rather than nerve transection.

Despite our conservative surgical approach, our patient demonstrated asymptomatic, postoperative phrenic nerve weakness secondary to surgical manipulation of the phrenic nerve (Figure 2). Given the benign and slow-growing nature of these tumors, it is important to have an extensive discussion with patients regarding surgical intervention versus observation, as intervention may result in morbidity for the patient. In our case, despite an extensive discussion of the potential associated risks and the unknown nerve of origin preoperatively, our patient was concerned about the progressive growth and strongly preferred definitive removal of the mass. We emphasize the importance of counseling patients on these risks and to consider observation in the management of these tumors as an acceptable option.

4 | CONCLUSION

Although rare, phrenic nerve schwannomas should be considered in patients presenting with an asymptomatic neck mass. Phrenic nerve origin should be suspected when the tumor appears to have interbody connection at the C2-C5 level on imaging. Surgical excision is a definitive treatment option, but the risks and benefits of surgery must be considered carefully. Enucleation of the mass, as opposed to nerve transection, is a feasible approach to limit morbidity.

AUTHOR CONTRIBUTIONS

Kimberly Chan: Investigation; visualization; writing – original draft; writing – review and editing. **Allison Keane:** Writing – original draft; writing – review and editing. **Jason Aynardi:** Resources. **Karen Yeji Choi:** Conceptualization; supervision; writing – review and editing.

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CONFLICT OF INTEREST

The authors have no conflicts of interest to disclose.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

ETHICAL APPROVAL

The manuscript was created in compliance with the ethical standards of our institution. The patient information was not revealed.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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