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Cervical Sympathetic Chain Schwannoma Masquerading as a Vagus Nerve Schwannoma Complicated by Postoperative Horner's Syndrome and Facial Pain: A Case Report

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ABSTRACT

INTRODUCTION: Cervical Sympathetic Chain Schwannomas (CSCS) of the carotid sheath are rare neoplasms that can be misdiagnosed on imaging. The following case documents a rare incident of a misdiagnosed CSCS with unusual outcomes of permanent Horner's syndrome and facial pain.
PRESENTATION OF CASE: A 36-year-old female presented with a slow-growing neck mass. CT and MRI led to a preoperative diagnosis of vagus nerve schwannoma (VNS). However, surgical treatment revealed the mass to be involved with the cervical sympathetic chain rather than the vagus nerve. The diagnosis was corrected to CSCS and the nerve was resected with the mass. The patient presented postoperatively with Horner's syndrome and severe facial pain. These symptoms persisted despite two years of medical management.
DISCUSSION: Studies indicate that imaging trends used for distinction between VNS and CSCS show inconsistencies in making preoperative diagnoses. Recent literature reveals helpful criteria for improving diagnostic standards that assist with preoperative patient counseling. In addition, postoperative outcomes, such as temporary, asymptomatic Horner's syndrome are common in CSCS. The following case report exemplifies the difficulties in diagnosis and addresses the unique complications of facial pain and permanent Horner's syndrome.
CONCLUSION: This case report examines postoperative outcomes and improves clinician awareness of the potential for misdiagnosis of a rare neoplasm and the recently improved diagnostic measures, providing for higher quality preoperative counseling. Future research is recommended to confirm and improve diagnostic guidelines and accuracy. Additional studies may focus on evaluating the effects of incorrect preoperative diagnosis on postoperative complication rates.

1. INTRODUCTION

Schwannomas are benign neoplasms of the nerve sheath derived from Schwann cells. Cervical schwannomas most frequently arise from the vagus nerve but an uncommon subgroup arises from the cervical sympathetic chain [1]. Despite predictive radiologic patterns, Cervical Sympathetic Chain Schwannomas (CSCS) have been known to masquerade as other neoplasms on computed tomography (CT) and magnetic resonance imaging (MRI). Postoperative complications and sequalae are common and specific to the nerve of origin. Postoperative risks include hoarseness and dysphagia in vagus nerve schwannomas (VNS) or Horner's syndrome and first bite syndrome (FBS) in CSCS [2]. Therefore, preoperative counseling is an important step in patient care.

This case details an interesting presentation of a CSCS misdiagnosed preoperatively as a VNS which presented with unique postoperative symptoms of permanent facial pain and persistent Horner's syndrome. It is important to be aware of the diagnostic insufficiencies associated with these tumors. Misdiagnosed nerves of origin hinder preoperative patient counseling. Our aim is to draw attention to recent developments in diagnosis that improve preoperative accuracy and patient counseling by examining the diagnostic difficulties and postoperative outcomes seen in this case.

Abbreviations: CSCS, Cervical sympathetic chain schwannoma; CT, Computed tomography; FBS, First Bite Syndrome; IJV, Internal jugular vein; MRI, Magnetic resonance imaging; VNS, Vagus verve schwannoma; PIFP, Persistent Idiopathic Facial Pain.

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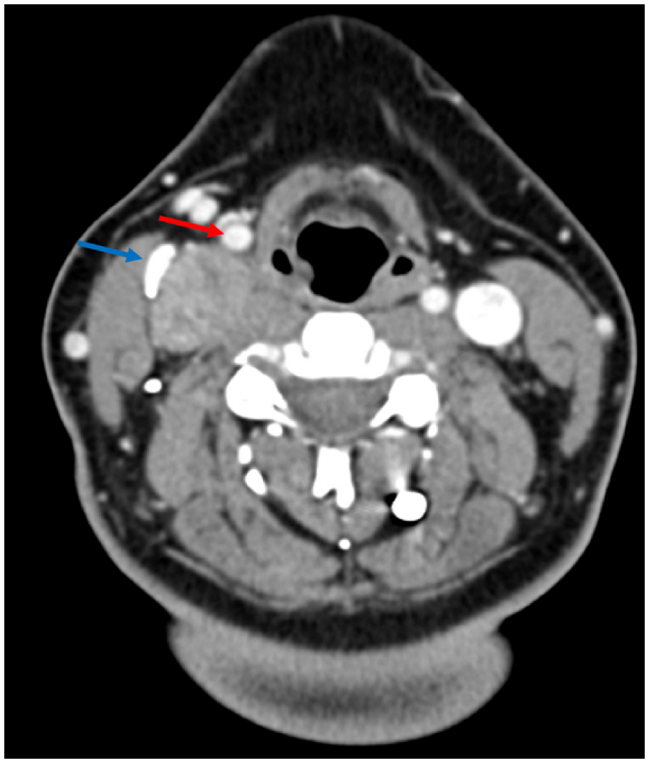

Fig. 1. An axial CT of the neck revealing an oval heterogeneous mass in the right carotid space measuring 35 × 24 x 28 mm. The internal jugular vein (blue arrow) is displaced laterally and the common carotid artery (red arrow) is displaced anteriorly leading to the spaying seen. This is often a characteristic sign of VNS.

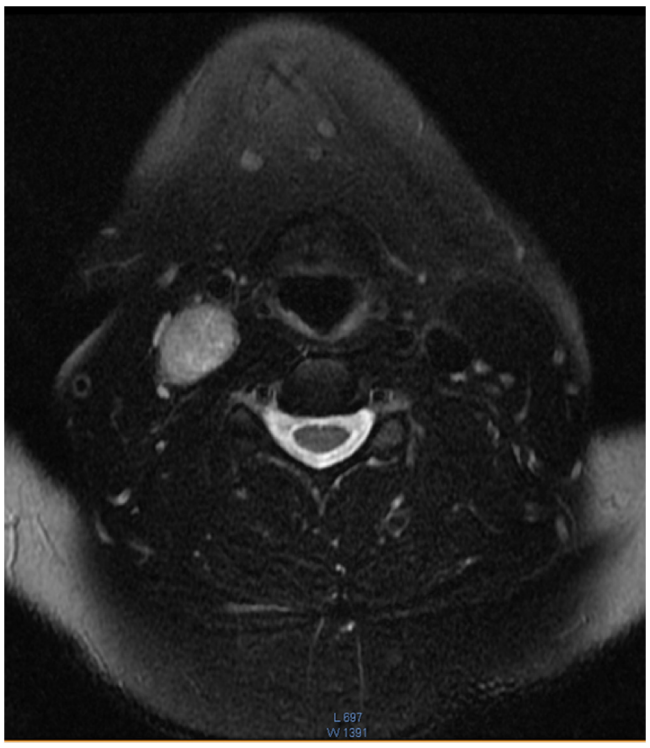

Fig. 2. An axial T2-weighted, fat saturated MRI with post gadolinium contrast further demonstrating a right sided mass in the carotid space with homogenous enhancement and a lack of vascular flow voids.

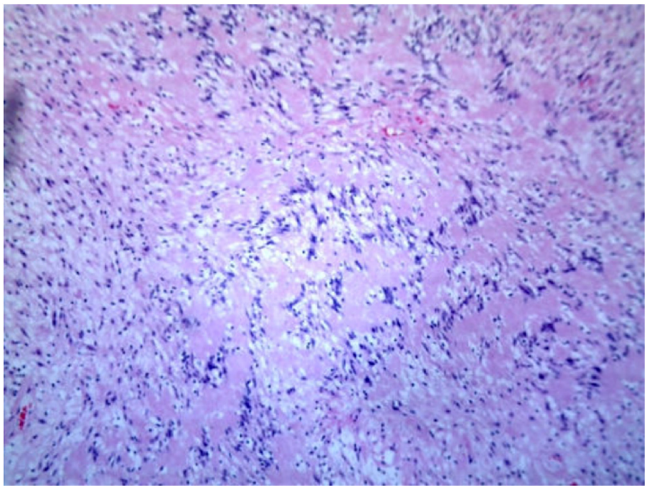

Fig. 3. Postoperative pathologic findings demonstrating spindle cells with both Antoni A and Antoni B areas with Verocay bodies consistent with a diagnosis of schwannoma.

This work has been reported in line with SCARE criteria [3].

2. PRESENTATION OF CASE

A 36-year-old Caucasian female was referred to otolaryngology with an eight-month history of a painful, slow-growing, right neck mass and a two-month history of significant, patient-reported esophageal dysphagia. The patient denied weight loss, hoarseness, loss of sensation, and upper limb weakness. Physical examination was inconsequential with the exception of a 4 x 3 cm painful, firm, mobile mass in the anterior triangle of the right neck, inferior to the hyoid bone and superior to the thyroid gland. No cranial nerve deficits were noted. The patient's history and medication list were noncontributory.

CT of the neck revealed an oval heterogeneous mass in the right carotid space measuring 35 × 24 x 28 mm, extending from vertebral levels C3 to C5 (Fig. 1). The mass displaced the common carotid artery anteriorly and the internal jugular vein (IJV) anterolateral causing splaying of the two vessels. A T1/T2 weighted MRI with contrast demonstrated a homogenous enhancement of the mass and no signs of vascular flow voids (Fig. 2). These radiological findings aligned most closely with a diagnosis of VNS, although CSCS could not be excluded. Malignancy was deemed unlikely. The treatment plans of surgical excision versus repeating imaging after six months were discussed at length with the patient. The patient advocated strongly for surgery due to her symptoms. With consideration to the growth of the mass and the progressive symptoms described, surgical resection was agreed upon. After demonstrating a clear understanding of the potential outcomes, the patient elected to proceed with surgical therapy.

Surgical dissection was performed utilizing a transverse cervical approach. The vagus nerve was visualized running over the top of the mass, uninvolved in the tumor formation. This proved the preoperative diagnosis of VNS incorrect. Instead, the cervical sympathetic chain was identified superiorly and inferiorly to the mass, making CSCS the most likely diagnosis. Because resection is the recommended treatment for both CSCS and VNS, the intraoperative management plan remained unchanged. Enucleation was attempted, but nerve excision was required due to the contiguous nature of the mass with the nerve. Pathology reports showed schwannoma characteristics, including spindle cells with Antoni A and Antoni B areas with Verocay bodies (Fig. 3).

One-week postoperatively, a right-sided Horner's syndrome was observed with ptosis and visual disturbances described as "difficulty concentrating." In addition, right-sided facial pain without sensory loss manifested as a constant sharp headache radiating