



Contents lists available at ScienceDirect

## International Journal of Surgery Case Reports

journal homepage: [www.casereports.com](http://www.casereports.com)

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

Austin T. Baker<sup>a,\*</sup>, Tyler J. Homewood<sup>a</sup>, Terry R. Baker<sup>b,c</sup><sup>a</sup> Texas College of Osteopathic Medicine, University of North Texas Health Science Center, 3500 Camp Bowie Blvd., Fort Worth, TX, 76107, USA<sup>b</sup> Mountain View Hospital 2325 Coronado St, Idaho Falls, ID 83404, USA<sup>c</sup> Idaho Falls ENT, 3200 Channing Way Ste A105, Idaho Falls, ID 83404, USA

## ARTICLE INFO

## Article history:

Received 12 February 2018

Received in revised form 16 May 2018

Accepted 4 June 2018

Available online 9 June 2018

## Keywords:

Schwannoma

Cervical sympathetic chain

Vagus nerve

Horner's syndrome

Persistent Idiopathic Facial Pain

Case report

## 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.

© 2018 The Author(s). Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## 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

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

\* Corresponding author.

E-mail addresses: [ab0706@my.unthsc.edu](mailto:ab0706@my.unthsc.edu) (A.T. Baker),

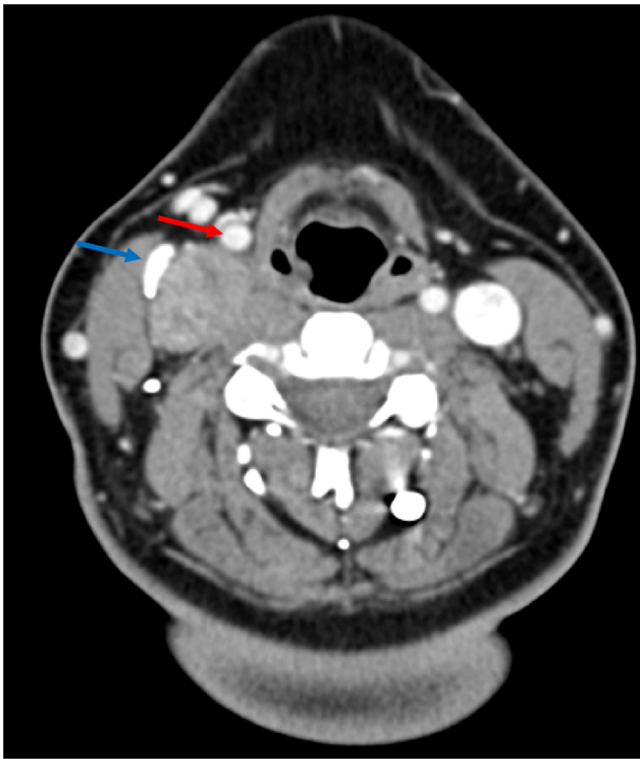
[Tyler.Homewood@my.unthsc.edu](mailto:Tyler.Homewood@my.unthsc.edu) (T.J. Homewood), [tjbakermd@cableone.net](mailto:tjbakermd@cableone.net) (T.R. Baker).

<https://doi.org/10.1016/j.ijscr.2018.06.001>

2210-2612/© 2018 The Author(s). Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

(CSCS) have been known to masquerade as other neoplasms on computed tomography (CT) and magnetic resonance imaging (MRI). Postoperative complications and sequelae 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.



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

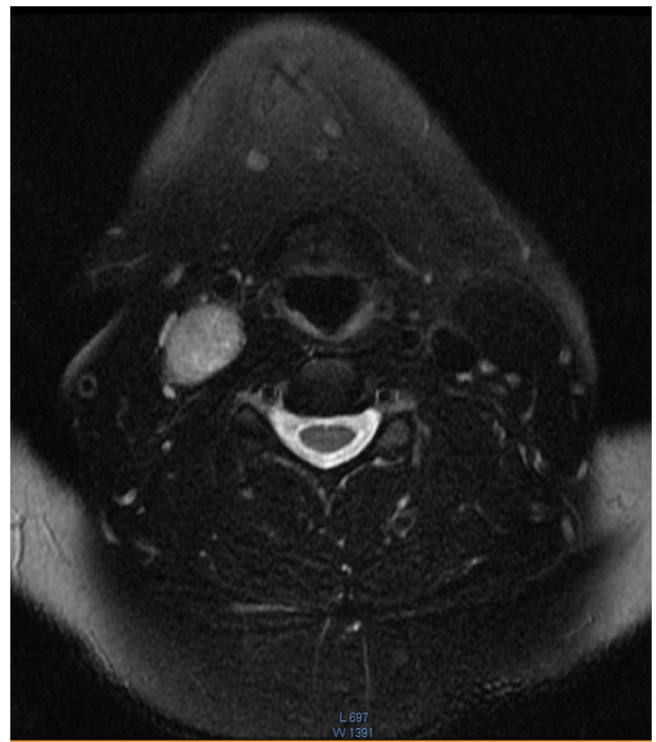
This work has been reported in line with SCARE criteria [3].

## 2. PRESENTATION OF CASE

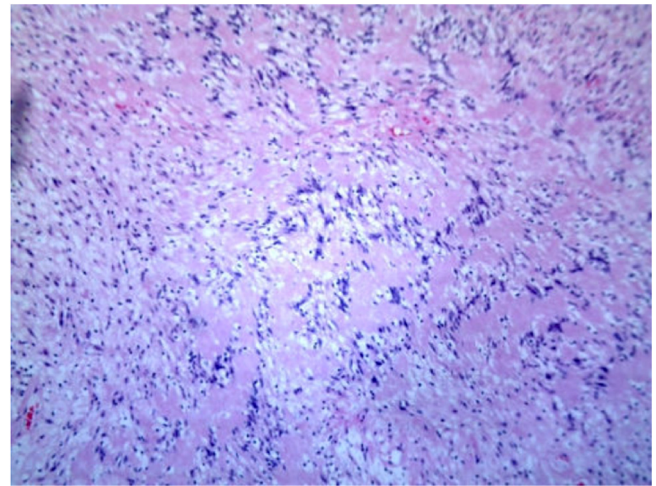
A 36-year-old Caucasian female was referred to otolaryngology with an eight-month history of a painful, slow-growing, right neck mass and a two-month history of significant, patient-reported esophageal dysphagia. The patient denied weight loss, hoarseness, loss of sensation, and upper limb weakness. Physical examination was inconsequential with the exception of a  $4 \times 3$  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  $35 \times 24 \times 28$  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. AT1/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



**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.

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

throughout the right half of the face enduring for several hours each day. Ophthalmology and neurology were consulted for management of the Horner's syndrome and facial pain respectively. Two-months postoperatively, the ptosis had resolved with surgical intervention, but the visual disturbances and facial pain persisted. Two years following surgery, the patient demonstrated no further improvement and was deemed unable to work due to pain and difficulty concentrating. The patient demonstrated understanding and has maintained great rapport with her doctors despite her frustration with the postoperative sequelae. She currently receives multi-disciplinary management and varying doses of topiramate, amitriptyline, and gabapentin, cognitive behavioral therapy and other lifestyle modifications with mild relief.

### 3. DISCUSSION

CSCS is an unusual diagnosis. Although the schwannoma in this case atypically developed in the mid-cervical portion of the carotid space, cervical schwannomas are commonly reported in the parapharyngeal space [4]. The vagus nerve is the most common nerve of origin [1]. According to one study, VNS make up 6% of parapharyngeal space tumors while CSCS make up just 3% [5]. Cervical schwannomas are typically seen in patients between the ages of 30 and 60, usually presenting as an asymptomatic, slow-growing mass [6]. Occasionally, symptoms consistent with local invasion can increase clinical suspicion.

Imaging with CT and MRI is the preferred diagnostic modality for evaluating cervical schwannomas. Paragangliomas can be differentiated by their vascular flow voids on MRI [7]. Schwannomas tend to be avascular [8]. According to a study done by Furukawa et al. and later confirmed by Saito et al., splaying of the carotid artery and IJV is a sign of VNS [[6], [9]]. They also noted that the absence of a gap between the carotid artery and IJV combined with anterior displacement of both vessels on imaging frequently occurs in CSCS [[6], [9]]. This phenomenon has been termed Furukawa's rule. Although CSCS have been known to masquerade as different tumors, it has rarely been documented to cause the characteristic vessel splaying seen in VNS as exemplified in this case [8].

In a recent literature review of 106 patients, Graffeo et al. noted variations in Furukawa's rule. They found that the splaying of the carotid artery and the IJV on imaging carried only a 75% probability of VNS while the absence of such splaying with anterior displacement of the carotid artery and IJV provided an 87% probability of CSCS [10]. This leaves significant room for misdiagnosis. An uncertain nerve of origin makes informing the patient of specific surgical outcomes difficult. However, Graffeo et al. also noted two new criteria in their study that create a multivariate strengthening of Furukawa's rule. A medially displaced carotid artery was found to be more determinate of VNS and a laterally displaced carotid artery was found to be more commonly associated with CSCS [10]. In addition, they found that peripheral enhancement of the mass on T2-weighted MRI was significantly associated with VNS while homogenous enhancement was isolated to CSCS [10]. When these criteria were used together with Furukawa's rule, predictive probability drastically increased to 97% in VNS and 99% in CSCS [10]. It is important to have a healthy knowledge of the imperfections of Furukawa's rule in an isolated setting as well as its improved diagnostic accuracy when combined with the additional criteria provided by Graffeo et al. Interestingly, the only criteria specific to CSCS seen in this case was the homogenous enhancement on MRI. Nevertheless, a more certain diagnosis significantly aids in informing the patient of specific postoperative sequelae.

Although observation can be considered due to their slow-growing and non-infiltrative nature, surgical enucleation by transverse cervical approach is the recommended therapy for cervi-

cal schwannomas. However, these tumors frequently require nerve excision causing dysfunction related to the involved nerve [11]. Nerve resection is required in 87% of CSCS surgeries as compared to 52% of VNS cases [10,11]. Despite this, most complications and sequelae are well tolerated. FBS and a transient, asymptomatic Horner's syndrome are the most common complications and sequelae in CSCS [8,11,12]. Conversely, the persistent, symptomatic Horner's syndrome and the facial pain observed in this case are rarely reported and deserve further inspection.

Mild postoperative Horner's syndrome has been described to occur in 91.1% of CSCS cases, most commonly as a consequence of sympathetic nerve resection [11]. The classic triad of miosis, anhidrosis, and ptosis generally goes unnoticed by patients. Postoperative Horner's syndrome rarely requires treatment and is ordinarily asymptomatic, but symptomatic patients commonly show gradual improvement [8,12]. Our case exemplifies rarely reported and severe visual symptoms that persisted despite treatment.

CSCS resection provides additional nerve-related risks. To our knowledge, the facial pain observed in this case has not been demonstrated in the literature. The most comparable recorded complication seen in CSCS resection is FBS, occurring in 21.1% of cases [11]. FBS presents with pain radiating throughout the jaw elicited on the initial bite of each meal that resolves with ensuing mastication [13]. The pathophysiology of FBS is likely due to sympathetic nerve damage [14,15]. In this case, the dissociation of pain from meals and mastication distinguishes it from FBS. Although the pain appears to be aligned with the distribution of the trigeminal nerve, the duration of this patient's pain and the lack of contact with the trigeminal nerve during surgery makes trigeminal neuralgia an unlikely diagnosis. However, the appearance of the pain shortly after the procedure makes some form of surgical trauma likely. A specific diagnosis was elusive despite multi-disciplinary management. The symptoms did not meet criteria for other forms of facial pain. The symptoms most closely align with persistent idiopathic facial pain (PIFP). PIFP is described as persistent facial pain recurring daily for more than 2 hours per day for greater than 3 months, in the absence of clinical neurological deficit [16,17]. PIFP is often associated with post-traumatic or post-surgical onset [16]. Although the underlying mechanism is unknown, we hypothesize that in this case, the pain may be due to surgical trauma to an unusual anatomical distribution of the trigeminal nerve or an atypical presentation of pain from sympathetic nerve damage.

### 4. CONCLUSION

This case presents a rare scenario of a CSCS masquerading as a VNS that serves to improve clinician awareness of the potential for misdiagnosis, its negative effects on preoperative counseling, and the recent strides in screening methodology. The outcomes associated with this case were unusual, and although the correlation with an initial misdiagnosis may be coincidental, this serves as an area for further research. This case highlights recently enhanced diagnostic criteria to improve preoperative accuracy, unusual postoperative sequelae, and opportunities to improve quality doctor-patient counseling when developing a treatment plan.

#### Conflicts of interest

None.

#### Funding

None.



## 7. Ethical Approval

N.A. This study is exempt from ethical approval at UNT Health Science Center.

## 8. CONSENT

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is attached and available for review by the Editor-in-Chief of this journal on request.

## 9. Author contribution

Austin Baker: Data collection, data analysis and interpretation, writing and revision of paper.

Tyler Jay Homewood: Writing and final revision of paper.

Terry Baker: Concept of the paper, data analysis and interpretation, final edit and revision of the paper, check for accuracy.

## 10. Registration of Research Studies

Registry: researchregistry.com.

UIN: researchregistry3492.

## 11. Guarantor

Austin Baker.

## References

- [1] M.F. Sheridan, D.W. Yim, Cervical sympathetic schwannoma: a case report and review of the English literature, *Otolaryngology Head Neck Surg.* 117 (December (6)) (1997), S206–10. Review. PMID: 9419150.
- [2] H.S. Gilmer-Hill, D.G. Kline, Neurogenic tumors of the cervical vagus nerve: report of four cases and review of the literature, *Neurosurgery* 46 (2000) 1498–1503.
- [3] R.A. Agha, A.J. Fowler, A. Saetta, I. Barai, S. Rajmohan, D.P. Orgill, for the SCARE Group, The SCARE Statement: Consensus-based surgical case report guidelines, *International Journal of Surgery* 34 (2016) 180–186.
- [4] F. Riffat, R.C. Dwivedi, C. Palme, B. Fish, P. Jani, A systematic review of 1143 parapharyngeal space tumors reported over 20 years, *Oral Oncol.* 50 (May (5)) (2014) 421–430, <http://dx.doi.org/10.1016/j.oraloncology.2014.02.007>, Epub 2014 Feb 28. Review. PMID: 24589290.
- [5] F.R. Miller, J.R. Wanamaker, P. Lavertu, B.G. Wood, Magnetic resonance imaging and the management of parapharyngeal space tumors, *Head Neck.* 18 (January–February (1)) (1996) 67–77, PMID: 8774924.
- [6] D.M. Saito, C.M. Glastonbury, I.H. El-Sayed, D.W. Eisele, Parapharyngeal space schwannomas: preoperative imaging determination of the nerve of origin, *Arch Otolaryngol Head Neck Surg.* 133 (July (7)) (2007) 662–667, PMID: 17638778.
- [7] A.B. Rao, K.K. Koeller, C.F. Adair, From the archives of the AFIP. Paragangliomas of the head and neck: radiologic-pathologic correlation, *Armed Forces Institute of Pathology. Radiographics.* 19 (November–December (6)) (1999) 1605–1632, Review. PMID: 10555678.
- [8] Anil G, Tan TY Imaging characteristics of schwannoma of the cervical sympathetic chain: a review of 12 cases. *AJNR Am J Neuroradiol.* 2010 31 September (8) 1408–1412 doi: 10.3174/ajnr.A2212 Epub 2010 Jul 8. PMID: 20616174.
- [9] M. Furukawa, M.K. Furukawa, K. Katoh, M. Tsukuda, Differentiation between schwannoma of the vagus nerve and schwannoma of the cervical sympathetic chain by imaging diagnosis, *Laryngoscope* 106 (December (12 Pt 1)) (1996) 1548–1552, PMID: 8948621.
- [10] C.S. Graffeo, K.M. Van Abel, J.M. Morris, M.L. Carlson, J.J. Van Gompel, E.J. Moore, D.L. Price, J.L. Kasperbauer, J.R. Janus, K.D. Olsen, M.J. Link, Preoperative diagnosis of vagal and sympathetic cervical schwannomas based on radiographic findings, *J Neurosurg.* 126 (March (3)) (2017) 690–697, <http://dx.doi.org/10.3171/2016.1.JNS151763>, Epub 2016 Apr 22. PMID: 27104848.
- [11] M. Navaie, L.H. Sharghi, S. Cho-Reyes, M.A. Keefe, B.A. Howie, G. Setzen, Diagnostic approach, treatment, and outcomes of cervical sympathetic chain schwannomas: a global narrative review, *Otolaryngol Head Neck Surg.* 151 (December (6)) (2014) 899–908, <http://dx.doi.org/10.1177/0194599814549550>, Epub 2014 Sep 11. Review. PMID: 25214550.
- [12] M.K. Wax, S.G. Shiley, J.L. Robinson, J.L. Weissman, Cervical sympathetic chain schwannoma, *Laryngoscope.* 114 (December (12)) (2004) 2210–2213, PMID: 15564847.
- [13] O. Laccourreye, A. Werner, D. Garcia, D. Malinvaud, P. Tran Ba Huy, P. Bonfils, First bite syndrome, *Eur Ann Otorhinolaryngol Head Neck Dis.* 130 (November (5)) (2013) 269–273, <http://dx.doi.org/10.1016/j.anorl.2012.11.003>, Epub 2013 Jul 23. Review. PMID: 23890788.
- [14] Y. Kawashima, T. Sumi, T. Sugimoto, S. Kishimoto, First-bite syndrome: a review of 29 patients with parapharyngeal space tumor, *Auris Nasus Larynx.* 35 (March (1)) (2008) 109–113, Epub 2007 Sep 11. PMID: 17851003.
- [15] K. Ijichi, D. Kawakita, S. Maseki, S. Beppu, G. Takano, S. Murakami, Functional Nerve Preservation in Extracranial Head and Neck Schwannoma Surgery, *JAMA Otolaryngol Head Neck Surg.* 142 (May (5)) (2016) 479–483, doi: 10.1093/otolaryng/142.5.479.
- [16] J.M. Zakrzewska, Differential diagnosis of facial pain and guidelines for management, *Br J Anaesth.* 111 (July (1)) (2013) 95–104, <http://dx.doi.org/10.1093/bja/aet125>, Review. PubMed [citation] PMID: 23794651.
- [17] H. Forssell, S. Jääskeläinen, T. List, P. Svensson, L. Baad-Hansen, An update on pathophysiological mechanisms related to idiopathic oro-facial pain conditions with implications for management, *J Oral Rehabil.* 42 (April (4)) (2015) 300–322, <http://dx.doi.org/10.1111/joor.12256>, Epub 2014 Dec 8. Review. PubMed [citation] PMID: 25483941.

## Open Access

This article is published Open Access at [sciendo.com](http://sciendo.com). It is distributed under the [IJSCR Supplemental terms and conditions](#), which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.