Parapharyngeal Space Schwannomas

Preoperative Imaging Determination of the Nerve of Origin

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Objectives: To determine if preoperative radiographic cross-sectional images can predict the nerve of origin of a parapharyngeal schwannoma and, specifically, whether it originates from the vagus nerve or the cervical sympathetic chain.

Design: A retrospective review.

Setting: Academic medical center.

Patients: The study population comprised 12 patients who underwent surgical resection of schwannomas of the parapharyngeal space. The nerve of origin was identified based on operative findings and postoperative physical examinations. Of the 12 patients, 11 underwent preoperative magnetic resonance imaging and 1 underwent preoperative contrast-enhanced computed tomography. A CAQ (Certificate of Added Qualification)–certified neuroradiologist reviewed the imaging studies, blinded to the surgically determined nerve of origin. For each case, it was predicted whether the tumor arose from the vagus nerve or sympathetic chain based on the location of the schwannoma with reference to the carotid sheath vessels.

Main Outcome Measure: Identification of the nerves of origin using the displacement of vessels as a marker.

Results: At the time of operation, it was determined that 5 patients (42%) had schwannomas from the cervical sympathetic chain and 7 patients (58%) had schwannomas of the cervical vagus nerve. By imaging, the nerve of origin was successfully determined in 4 of 5 cases of sympathetic chain schwannoma (80%) and in 7 of 7 cases of vagal nerve schwannoma (100%). Schwannomas of the cervical sympathetic chain were found to displace both the carotid and jugular vessels without separating them. Vagal nerve schwannomas were found to separate the carotid arteries from the internal jugular vein. A vagal nerve schwannoma may also displace the sheath vessels posteriorly, without splaying them.

Conclusions: Carotid and jugular vessel displacement, as determined by cross-sectional imaging, can predict the likely nerve of origin of a parapharyngeal space schwannoma. This determination allows for effective preoperative counseling regarding the expected sequelae of surgical resection.


Schwannomas are uncommon neurogenic tumors that are typically benign, slow growing, and asymptomatic. Up to 45% of all schwannomas originate in the head and neck region. They are reported to occur in the face, scalp, intracranial cavity, orbit, nasal and oral cavities, parapharyngeal space, middle ear, mastoid, larynx, and medial and lateral regions of the neck. In the parapharyngeal space, schwannomas most commonly arise from the vagus nerve and cervical sympathetic chain.

The accepted treatment of schwannomas is surgical resection. However, surgery is not always recommended because of the indolent nature of the tumor and the risk of postoperative neural deficits. A preoperative diagnosis of the nerve of origin would allow directed preoperative counseling as to the risks of surgery and therefore permit the patient to make an informed decision on whether to undergo surgery or adopt a course of observation.

CME course available at www.archoto.com

With a series of 9 patients in 1996, Furukawa et al reported that preoperative imaging studies revealed differences in vessel displacement patterns caused by vagal and sympathetic chain schwannomas. The vagal nerve schwannomas always resulted in a separation between the internal carotid artery (ICA) or common

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carotid artery (CCA) and the internal jugular vein (IJV). In contrast, sympathetic chain schwannomas did not produce an observable separation. With our series of 12 patients who have received surgical treatment for schwannomas of the parapharyngeal space, we tested the efficacy of this observation and investigated other imaging characteristics that may refine our ability to preoperatively diagnose the nerve of origin.

METHODS

Between 1997 and 2005, at the University of California, San Francisco, 14 patients with schwannomas of the parapharyngeal space underwent surgical excision. All resected tumors were confirmed pathologically as schwannomas. In 11 of 14 cases, the nerve of origin was determined by direct visualization during the operation. In 2 cases, the nerve of origin could not be identified during surgery, and the patients’ postoperative recovery revealed no neurologic deficits. These 2 cases were excluded from our study group. In one other case, the nerve could not be determined in surgery, but the patient developed a significant Horner syndrome postoperatively. Therefore, we determined that this schwannoma was of sympathetic chain origin, and the patient was included in the study.

Eleven patients underwent preoperative magnetic resonance imaging (MRI), and 1 patient underwent contrast-enhanced computed tomography (CT). The imaging studies were retrospectively reviewed by a CAQ (Certificate of Added Qualification)-certified neuroradiologist (C.M.G.), who was blinded to the operative findings but informed that all patients were diagnosed as having parapharyngeal space schwannomas of either the vagus nerve or the sympathetic chain. The neuroradiologist was also provided with the article by Furukawa et al3 and asked to consider the proposed criteria in making the radiographic predictions. The article by Furukawa et al3 proposes that vagal nerve schwannomas will splay the CCA and ICA away from the IJV, whereas sympathetic chain schwannomas will not result in separation of the vein and arteries (Figure 1 and Figure 2).

RESULTS

Our series consists of 12 patients with parapharyngeal space schwannomas. The schwannoma arose from the cervical sympathetic chain in 5 patients, and the vagus was the nerve of origin in 7 patients. The imaging findings, the neuroradiologist’s predicted nerve of origin, and the operatively determined nerve of origin are listed in the Table. Of the 5 cases of sympathetic chain schwannomas, 4 (80%) were correctly predicted. Of the 7 cases of vagal nerve schwannomas, the neuroradiologist correctly identified 6 (86%).

We encountered 2 cases that necessitated refinement of the criteria of Furukawa et al.3 In cases 2 and 9, the ICA and CCA were not significantly splayed apart from the IJV but were displaced posteriorly. By the criteria of Furukawa et al,3 this predicts the nerve of origin to be the sympathetic chain. The neuroradiologist’s concern was that the cervical sympathetic chain runs posterior to the carotid sheath in the parapharyngeal space and should not be expected to displace the carotid sheath vessels posteriorly. The schwannoma in case 2 was an exceptionally large tumor (4 × 4 × 5 cm), and we proposed that it could still arise from the sympathetic chain and distort the surrounding anatomy enough to displace the vessels together posteriorly and slightly laterally into the posterior cervical space (Figure 3). We therefore agreed with the criteria of Furukawa et al3 and diagnosed this schwannoma as a sympathetic chain tumor, which proved to be the correct determination. In contrast, case 9 was a more modestly sized tumor (3 × 3 × 4 cm) that would not be expected to significantly distort the surrounding tissue (Figure 4). Also, although the ICA and IJV are not splayed apart by this tumor, there is still demonstrable separation of the vessels on imaging. Thus, because of these additional considerations, it was proposed that this was a vagus nerve schwannoma with minimal vessel separation. This corresponded with operative identification of the nerve.
result, our approach correctly identified 100% (7/7) of the vagal nerve schwannomas, improving on the accuracy rate of 86% (6/7) in the article by Furukawa et al. In the 1 misdiagnosed case (case 5), the intraoperative findings definitely implicated the sympathetic chain as giving rise to the schwannoma, but the imaging studies showed splaying of the IJV and ICA, which predicted vagal nerve origin.

**COMMENT**

The parapharyngeal space surrounds the pharynx and harbors 0.5% of all head and neck neoplasms. The differential diagnosis for a mass found in the parapharyngeal space is wide and can include tumors of the deep lobe of the parotid gland, tumors of minor salivary gland origin, metastatic cervical nodes, paragangliomas, branchial cysts, lymphomas, neurofibromas, and aneurysms of the ICA. Most tumors in the parapharyngeal space are benign (70%-80%). In a series of 51 patients with parapharyngeal space tumors, schwannomas made up 18% of the series, with 6 (12%) arising from the vagus nerve and 3 (6%) from the sympathetic trunk.

Schwannomas of the parapharyngeal space are usually reported to occur in patients between the ages of 30 to 50 years. In our series, 10 of our 12 subjects were women, and all were aged 34 years or younger. Although the majority of schwannomas are benign, the incidence of malignancy is higher in the parapharyngeal space (10% vs 3%) and surgical resection is the treatment of choice for all cases.5 Schwannomas of the parapharyngeal space are usually diagnosed during the second or third trimester of pregnancy. They are frequently asymptomatic but can cause symptoms related to their location.5

### Table. Imaging Findings With Estimated Nerves of Origin vs Actual Nerves of Origin

<table>
<thead>
<tr>
<th>Patient No./Sex/Age, y</th>
<th>Imaging Type</th>
<th>Imaging Findings</th>
<th>Probable Nerve Origin as per Furukawa et al.</th>
<th>Probable Nerve Origin as per Saito et al (Present Study)</th>
<th>Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/F/34 MRI 5 × 2 × 2-cm Tumor medial to the IJV and ICA, posterior to the CCA</td>
<td>Sympathetic chain</td>
<td>Sympathetic chain</td>
<td>Vagus nerve</td>
<td></td>
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<tr>
<td>2/F/17 MRI 4 × 2 × 2-cm Tumor displaces the CCA and ICA and the IJV posteriorly</td>
<td>Sympathetic chain</td>
<td>Sympathetic chain</td>
<td>Vagus nerve</td>
<td></td>
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<tr>
<td>3/F/45 MRI 3 × 4-cm Tumor displaces the CCA and ICA and the IJV laterally</td>
<td>Sympathetic chain</td>
<td>Sympathetic chain</td>
<td>Vagus nerve</td>
<td></td>
<td></td>
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<tr>
<td>4/M/17 MRI 3 × 6-cm Tumor displaces the ICA and IJV laterally</td>
<td>Sympathetic chain</td>
<td>Sympathetic chain</td>
<td>Vagus nerve</td>
<td></td>
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<tr>
<td>5/F/59 MRI 2 × 3-cm Tumor separates the ICA and IJV</td>
<td>Sympathetic chain</td>
<td>Sympathetic chain</td>
<td>Vagus nerve</td>
<td></td>
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<tr>
<td>6/M/44 MRI 5 × 2 × 3-cm Tumor separates the CCA and IJV</td>
<td>Vagus nerve</td>
<td>Vagus nerve</td>
<td>Vagus nerve</td>
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<tr>
<td>7/F/25 MRI 3 × 4-cm Tumor separates the ICA and IJV</td>
<td>Vagus nerve</td>
<td>Vagus nerve</td>
<td>Vagus nerve</td>
<td></td>
<td></td>
</tr>
<tr>
<td>8/F/51 MRI 3 × 3-cm Tumor separates the ICA and IJV</td>
<td>Vagus nerve</td>
<td>Vagus nerve</td>
<td>Vagus nerve</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9/F/33 MRI 3 × 4-cm Tumor displaces the ICA and IJV posteriorly</td>
<td>Sympathetic chain</td>
<td>Vagus nerve</td>
<td>Vagus nerve</td>
<td></td>
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<tr>
<td>10/F/35 MRI 4 × 4 × 6-cm Tumor separates the ICA and IJV</td>
<td>Vagus nerve</td>
<td>Vagus nerve</td>
<td>Vagus nerve</td>
<td></td>
<td></td>
</tr>
<tr>
<td>11/F/17 MRI 3 × 2 × 2-cm Tumor separates the ICA and IJV, enters skull base</td>
<td>Vagus nerve</td>
<td>Vagus nerve</td>
<td>Vagus nerve</td>
<td></td>
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<tr>
<td>12/M/35 CT 3 × 2 × 2-cm Tumor separates the ICA and IJV</td>
<td>Vagus nerve</td>
<td>Vagus nerve</td>
<td>Vagus nerve</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Abbreviations: CCA, common carotid artery; CT, computed tomography; ICA, internal carotid artery; IJV, internal jugular vein; MRI, magnetic resonance imaging.
and 70 years. Most of these uncommon tumors affect the cervical vagus nerve and the cervical sympathetic chain. While the male-female ratio for cervical sympathetic chain schwannomas has been reported as either equal or at a 3:1 ratio, there does not seem to be a sex-related predisposition for vagal nerve schwannomas.

Although schwannomas are typically benign, they may affect adjacent tissues by expansion with pressure effect. The neoplasms are relatively radioresistant, so complete surgical resection remains the treatment of choice. Their slow growth, low recurrence rate, and noninvasive nature, however, often allow for an observational approach. Prior to 1931, most parapharyngeal schwannomas were resected by the transoral route, with complications such as incomplete removal, serious hemorrhage, infection, and cranial nerve damage. The transcervical surgical approach to the parapharyngeal space is the current preferred operation. If the schwannoma is completely removed, recurrence rates are extremely low.

Figure 3. T1-weighted plus gadolinium (Gd) (A) and T2-weighted (B) magnetic resonance images of a large sympathetic chain schwannoma displacing the common carotid artery (white arrow) and the internal jugular vein (IJV) (black arrow) together in a posterior and slightly lateral direction (case 2). Despite originating from a structure posterior to the carotid sheath, an especially large lesion such as this one can distort the surrounding fascial planes and push the vessels in an unpredictable posterior direction. Note that the vessels remain immediately adjacent to one another.

Figure 4. T2-weighted (A) and T1-weighted plus gadolinium (Gd) (B) magnetic resonance images of a vagus nerve schwannoma that is displacing the common carotid artery (white arrow) and internal jugular vein (black arrow) posteriorly without splaying them apart (case 9). With average-sized lesions, direct posterior displacement of vessels should suggest a vagus nerve origin given its anterior position in the carotid sheath. This diagnosis is further supported by the small separation observed between the vessels.
Because schwannomas arise outside the involved fascicle, they tend to compress the nerve fascicles to the periphery as they enlarge and can usually be dissected free of all structures except the parent fascicle. Sometimes, the involved nerve does not course through the tumor but can pass over in the tumor capsule. The literature reports rare cases of schwannoma resection with preservation of postoperative neurologic dysfunction. In most cases, however, patients should be prepared for dysfunction of the involved nerve after schwannoma resection.

It is difficult to determine the nerve giving rise to a parapharyngeal space schwannoma on the basis of the patient's symptoms. These tumors typically present as an asymptomatic neck mass, often accompanied by vague symptoms such as an awareness of mass in the throat, sore throat, or dysphagia. Patients with malignant tumors are more likely to present with a rapidly growing neck mass, pain, trismus, otalgia, or cranial nerve deficits. Preoperative Horner syndrome has been reported to be related to parapharyngeal space tumors other than sympathetic chain schwannomas and does not necessarily indicate that the sympathetic chain is the nerve of origin.

With the lack of symptoms and physical examination findings, imaging plays the central role in diagnosing and distinguishing parapharyngeal space neoplasms. Over the past 15 years, CT and, more recently, MRI have become the routine imaging studies that are used. Angiography is used selectively to assess enhancing lesions of the parapharyngeal space for evaluation of a vascular tumor and consideration for preoperative embolization. On noncontrast CT, schwannomas are hypodense as compared with muscle. Contrast administration results in some degree of enhancement, which may be homogeneously solid or heterogeneous and patchy.

It can be difficult to differentiate the rare hypervascular schwannoma from a paraganglioma. Magnetic resonance imaging allows for superior soft tissue contrast resolution and does not expose the patient to ionizing radiation. For these reasons, MRI is now considered the imaging study of choice to evaluate parapharyngeal space tumors. A recent series showed that MRI carries a 95% accuracy in delineating a parapharyngeal space mass in relation to the prestyloid vs poststyloid compartments, its relationship to the deep lobe of the parotid, and its inherent soft-tissue characteristics (vascular vs solid neoplasm). On MRI, schwannomas are well-circumscribed homogenous masses that exhibit high-signal intensity on T2-weighted images and a relatively homogeneous low-signal intensity on T1-weighted images. In contrast with paragangliomas, there are no vascular flow voids seen in schwannomas.

The parapharyngeal space is an inverted pyramid that extends from the skull base to the hyoid bone. The space is divided into an anterior and a posterior compartment by the styloid process. The poststyloid compartment contains the carotid sheath, sympathetic chain, and cranial nerves IX through XII. The carotid sheath contains the ICA, CCA, IJV, and vagus nerve. The cervical sympathetic chain runs posterior and slightly medial to the carotid sheath. Therefore, as schwannomas from this structure grow and expand, they tend to displace its contents anteriorly and laterally. Although there is collective displacement of these vessels, there is not any separation between the IJV and the ICA or CCA. The vagus nerve, in contrast, runs within the carotid sheath between the ICA and the IJV on the cranial side of the bifurcation and between the CCA and IJV on the caudal side. As a schwannoma enlarges from the vagus nerve, it tends to displace the IJV laterally and the ICA and CCA medially. Thus, imaging will reveal a separation of these vessels as the schwannoma expands.

In a series reported in 1996 of 9 patients with parapharyngeal schwannomas, Furukawa et al reported that the tumor's relationship with the carotid sheath vessels provided useful preoperative estimation of the nerve of origin. All of their patients underwent preoperative CT and MRI. The series' 5 patients with vagal nerve schwannoma all displayed separation of the IJV and the CCA or ICA. In the 4 patients with sympathetic chain schwannoma, the tumor was found posterior to the carotid sheath, and imaging failed to show any separation between the IJV and the ICA or CCA.

In our series of 12 patients, the criteria of Furukawa et al yielded good results. We correctly diagnosed 10 of 12 nerves of origin (83%) using the displacement of vessels as a marker. In addition, we observed that a schwannoma of the vagus nerve may displace the IJV and the CCA or ICA in a posterior direction without splaying them apart (as in case 9). A large schwannoma of the sympathetic chain can result in a similar picture (as in case 2) with posterior and slight lateral displacement. Hence, we propose a corollary to the paradigm of Furukawa et al. When the carotid sheath vessels are displaced posteriorly but not splayed apart by the lesion, one should also consider (1) the volume of the lesion and (2) if any distance between the vessels exists. Posterior vessel displacement would be expected from enlargement of the more anteriorly situated vagus nerve rather than a lesion from the posteriorly based cervical sympathetic chain. An especially large lesion from the sympathetic chain, however, can distort the surrounding anatomy and displace the vessels posteriorly. In this case, the vessels would be expected to stay immediately adjacent to each other in the carotid sheath (as in case 2), and slight lateral displacement may also be observed. Any observable separation of the artery and vein (as in case 9) suggests a lesion within the carotid sheath and implicates the vagus nerve.

A review of the literature yields another useful consideration. A schwannoma of the cervical sympathetic chain has been reported to splay the internal and external carotid arteries on preoperative imaging, a phenomenon that was not present in any of our cases. The CCA bifurcates into the internal and external arteries near the level of the superior cornu of the thyroid cartilage. The ICA is initially posteroslateral and then ascends posteromedially into the carotid canal of the skull base. The external carotid artery (ECA) lies anteromedial near the bifurcation and is slightly anterolateral to the ICA near the level of the skull base. Splaying of the carotid bifu-
cation (the “lyre” sign) classically suggests a carotid body tumor. The cervical sympathetic trunk, however, at the level of the carotid bifurcation, lies just posteromedial to the ICA and the carotid sheath on the prevertebral fascia. Because of limitation by the cervical vertebral column medially and the longus capitis muscle posteriorly, a cervical sympathetic chain schwannoma occasionally grows anteriorly into the space between the ICA and the ECA. Wang et al described this finding in a case report from 2004 and noted that cervical sympathetic chain schwannomas with splaying of the carotid bifurcation have been mentioned in the English language literature.

Schwannomas from the vagus nerve, on the other hand, will not splay apart the ICA and ECA. Green et al note that 9 of their 11 patients with vagal nerve schwannomas, the schwannoma occurred at the nodose ganglion, which is above the level of the carotid bifurcation. The vagus nerve fibers in the carotid sheath course lateral to the ICA above the bifurcation. Therefore, a vagus nerve schwannoma can splay the IJV posterolaterally and the ICA anteromedially but cannot separate the ICA and ECA.

As the carotid arteries were not splayed in any of our series' images, we cannot comment on the validity of this observation in the prediction of a sympathetic chain schwannoma. It may, however, prove to be another useful corollary to the criteria of vessel displacement discussed in the present report and by Furukawa et al.

The surgical resection of parapharyngeal schwannoma often results in postoperative neurologic deficits. With careful examination of vessel displacement and separation on MRI or CT, it is possible to predict with considerable precision the nerve giving rise to a parapharyngeal schwannoma. This information allows the surgeon to counsel the patient on specific anticipated postoperative neurologic deficits. In addition, patients with suspected vagal nerve schwannomas, for instance, may be referred to a laryngologist and speech or swallow therapist preoperatively to discuss rehabilitation options after surgery.

Our experience confirms the criteria for the determination of nerve origin proposed by Furukawa et al and proposes an additional consideration. Vagal nerve schwannomas will splay the carotid artery and jugular vein apart on imaging studies, while sympathetic chain schwannomas displace these vessels together. Additional criteria must be considered when a schwannoma displaces the carotid sheath posteriorly. As the vagus nerve courses through the anterior aspect of the carotid sheath, this finding typically predicts vagal origin, but only subtle splaying of the vein and artery may be observed. In the case of a very large schwannoma, posterior vessel displacement with slight lateral displacement predicts sympathetic nerve origin.

CONCLUSIONS

The surgical resection of parapharyngeal schwannoma often results in postoperative neurologic deficits. With careful examination of vessel displacement and separation on MRI or CT, it is possible to predict with considerable precision the nerve giving rise to a parapharyngeal schwannoma. This information allows the surgeon to counsel the patient on specific anticipated postoperative neurologic deficits. In addition, patients with suspected vagal nerve schwannomas, for instance, may be referred to a laryngologist and speech or swallow therapist preoperatively to discuss rehabilitation options after surgery.

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Author Contributions: Dr Saito had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Saito, El-Sayed, and Eisele. Acquisition of data: Saito, El-Sayed, and Eisele. Analysis and interpretation of data: Saito, Glastonbury, and Eisele. Drafting of the manuscript: Saito and Glastonbury. Critical revision of the manuscript for important intellectual content: Glastonbury, El-Sayed, and Eisele. Statistical analysis: Saito and Eisele. Administrative, technical, and material support: Saito and Eisele. Study supervision: Glastonbury, El-Sayed, and Eisele.

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REFERENCES