Functional Nerve Preservation in Extracranial Head and Neck Schwannoma Surgery

Kei Ijichi, MD, PhD; Daisuke Kawakita, MD, PhD; Shinichiro Maseki, MD, PhD; Shintaro Beppu, MD; Gaku Takano, MD; Shingo Murakami, MD, PhD

IMPORTANCE A schwannoma is an uncommon, benign neurogenic tumor of Schwann cells. Tumor enucleation is the recommended surgical method to preserve function of the original nerve, although enucleation does not guarantee completely intact nerve function after the operation.

OBJECTIVE To establish a strategy for functional preservation in extracranial head and neck schwannoma treatment by using an electromyographic (EMG) system during tumor resection.

DESIGN, SETTING, AND PARTICIPANTS A retrospective cohort study was conducted of 15 patients who underwent surgery for removal of schwannoma tumors between April 1, 2006, and March 31, 2015, at an academic tertiary referral center. Data analysis was conducted from April 3, 2006, to September 15, 2015. Neurogenic tumors were diagnosed according to preoperative findings, and during surgery tumors were exposed and given EMG-controlled electrical stimulation to analyze their origins. In motor nerve cases, the electrical activity of the muscle was measured and recorded by EMG. The tumor was then enucleated by incision along tumor fibers mapped using EMG stimulation. If a nerve bundle was visible, we incised along there and enucleated the tumor.

INTERVENTIONS A strategy using electrical stimulation to improve preservation of nerve function in extracranial head and neck schwannoma operations.

MAIN OUTCOMES AND MEASURES Frequency and duration of postoperative neurologic complications associated with functional preservation surgery with tumor enucleation was evaluated using EMG monitoring according to tumor origin.

RESULTS Of the 15 patients with extracranial schwannoma, 9 (60%) were women (mean [SD] age, 36.3 [15.3] years). All 15 patients underwent surgery using a transcervical approach. The most common nerves of origin were the vagus nerve and the sympathetic chain. In sensory or sympathetic nerve cases, the EMG response was absent. Two of 5 patients with vagus schwannoma had postoperative temporary vocal nerve palsy. These symptoms showed improvement after 1 year. There was no tumor recurrence during the follow-up period in any patient.

CONCLUSIONS AND RELEVANCE The strategy used here demonstrated a method of diagnosis and nerve preservation surgery for extracranial schwannomas. Nerve functionality was preserved in all vagus schwannoma cases. However, preservation of nerve function in sympathetic nerve schwannoma cases remains problematic and needs further investigation.
schwannoma is an uncommon, benign nerve sheath tumor arising from Schwann cells. Approximately 25% to 45% of schwannomas occur in the extracranial head and neck region and are often located in the parapharyngeal space.\textsuperscript{1,2} Schwannomas originate in the cranial (eg, V, VII, IX, XI, and XII), sympathetic, or peripheral nerves.\textsuperscript{3,4} Preoperative imaging, such as computed tomography or magnetic resonance imaging, is often used to determine tumor size, location, and origin. Surgical resection is the only curative treatment, but there is no consensus regarding the most effective surgical approach and resection method for preservation of lower cranial nerves.

The goal of surgical treatment is achieving complete resection of the tumor while preserving nerve function. In particular, if the vagus nerve is the origin of the tumor, surgical treatment should not result in postoperative vocal cord paralysis. Previous studies\textsuperscript{5} have reported vocal cord palsy to be unavoidable after resection of tumors originating from the vagus nerve, even when intracapsular enucleation was performed.

Recently, nerve monitoring with electromyographic (EMG) systems (eg, NIM [Nerve Integrity Monitor], Medtronic Xomed Inc; Lantern, The Magstim Company Ltd; and Nerveana, Neurovision Medical Products Inc) has been used routinely in thyroid and parotid gland surgery. Intraoperative nerve monitoring is reported\textsuperscript{6-8} to be useful for nerve preservation, particularly in the case of large tumors or nerve invasion.

Preservation of nerve function in extracranial schwannomas was, until recently, considered to be challenging, but preservation is important for preventing postoperative complications leading to poorer quality of life for the patient. The most challenging problem is nerve preservation with vagus schwannomas. Injury to the vagus nerve causes hoarseness and dysphagia after the operation. Hypoglossal and accessory nerve preservation is also necessary to prevent postoperative dysphagia. We established a strategy for functional preservation in extracranial head and neck schwannoma treatment by using an EMG system during tumor resection.

**Methods**

This retrospective cohort study included 15 patients who were admitted to Nagoya City University Hospital from April 1, 2006, and March 31, 2015, had an initial diagnosis of extracranial schwannoma, and underwent surgery using the NIM EMG system. Data analysis was conducted from April 3, 2006, to September 15, 2015. The study was approved by the institutional review board of Nagoya City University. All participants provided oral informed consent.

Our protocol for treatment of extracranial head and neck schwannomas is summarized in the Figure. Preoperative symptoms may provide clues to help identify the origin of the tumor when it is caused by functional failure of the nerve of origin. However, one must be aware that symptoms might also occur owing to pressure on an adjacent nerve. Coughing, hoarseness, tongue deviation, and paralysis of the soft palate can all point to the nerve of origin. However, neck discomfort is a nonspecific symptom.

**Figure. Treatment Plan for Extracranial Head and Neck Schwannomas**

Preoperative computed tomography and magnetic resonance imaging are useful to identify the tumor location. However, it is difficult to identify the nerve of origin when the tumor is located around the carotid bifurcation. Locating tumors by computed tomography, magnetic resonance imaging, and fine-needle aspiration cytology can distinguish schwannomas from parotid tumors, but the definitive diagnosis of schwannomas with fine-needle aspiration cytology is difficult. Incisional biopsy is not performed owing to the possibility of nerve damage. We inform patients before the operation that symptoms of lower cranial neurogenic deficit can arise from neural damage during tumor resection.

In the operation, use of an EMG system is essential to avoid motor nerve paralysis. A sensor can be set into the orbicularis oculi (cranial nerve VII), orbicularis oris (cranial nerve VII), thyroarytenoid (cranial nerve X), trapezius (cranial nerve XI), or lingual (cranial nerve XII) muscles. We select 2 of the 3 muscles according to the preoperative estimation of the nerve of tumor origin.

After the tumor is exposed, its anatomical location is inspected; electrical stimulation is then applied to the tumor with a stimulating electrode and muscle activity is recorded via a sensor electrode. The nerve of origin is identified as a motor
nerve when a response is recorded at the sensor electrode. If
a nerve bundle is visible on the surface of the tumor, an inci-
sion is made along the bundle and the tumor is enucleated.
When no nerve bundle is visible but there is a muscle res-
ponse, several electrical stimuli are applied to the tumor to
determine which part of the tumor surface shows no re-
sponse to electrical stimulus. After this response mapping,
an incision is made in the unresponsive tumor area to enucleate
the tumor.

If there is no response to electrical stimulation over the en-
tire tumor, the nerve of origin is expected to be a sensory or
sympathetic nerve. In this case, it is not possible to identify
the nerve fibers with an electrical stimulus. If a nerve bundle
is visible on the surface of the tumor, an incision is made along
the bundle and the tumor is enucleated using a microscope.
If no nerve bundle is visible, we incise the most swollen part
of the tumor to cut the part likely to involve fewer fibers. When
nerve strings become tangled in the tumor, piecemeal tumor
resection is used for maximum avoidance of nerve injury.

Results

Diagnosis
To diagnose the tumor origin and histologic type, preopera-
tive computed tomography, magnetic resonance imaging, fine-
needle aspiration cytology, and/or incisional biopsies were per-
formed on all patients (Table 1 and Table 2). The mean (SD)
tumor size was 42.7 (9.2) mm (range, 30-63 mm).

Treatment and Outcome
All 15 patients underwent nerve preservation surgery as de-
scribed above. All tumors were resected through a transcervi-
cal approach. The tumor was enucleated, with nerve pres-
ervation in all 15 cases. Tumors with a vagus nerve origin were
identified in 5 patients (36%) (Table 2). There was no tumor
recurrence during the follow-up period.

Vagus Schwannomas
In 4 of 5 patients (80%) with vagus schwannoma, the tumor
was located in the parapharyngeal space, and in 1 patient (20%) the
tumor was in the lower lateral neck. A nerve bundle was vis-
ible on the tumor in 3 of these 5 patients (60%) (Video, A). There
was no visible nerve bundle in the remaining 2 patients (40%),
in whom the tumor was mapped with the NIM system, and the
tumor was incised over the safe area of the tumor (the
area unresponsive to electrical stimulation) (Video, B). The tu-
mor was exfoliated and resected from the nerve sheath. Three
patients (60%) had normal vagus nerve function after the op-
eration. The remaining 2 patients (40%) had transient vocal cord
palsy, but recovered completely after 1 year. Our tumor resec-
tion protocol enabled us to preserve vagus nerve function in all
5 patients. One patient with vagus nerve schwannoma pre-
vented with preoperative cough symptoms. After the operation,
the cough symptoms showed immediate improvement.

Sensory and Sympathetic Nerve Schwannomas
For the remaining 10 patients with schwannoma whose tu-
mors were not of vagus nerve origin, the tumors were of the

Table 1. Patient Characteristics for Extracranial Head and Neck Schwannomas of Nonvagal Origin

<table>
<thead>
<tr>
<th>Age, y/sex</th>
<th>Nerve of Origin</th>
<th>Postoperative Paralysis</th>
<th>Postoperative Symptoms</th>
<th>Size, mm</th>
<th>Preoperative Cytology/Histology Findings</th>
<th>Outcome of Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>22/F</td>
<td>Sympathetic</td>
<td>−</td>
<td>−</td>
<td>80</td>
<td>Spindle cell/ND</td>
<td>NA</td>
</tr>
<tr>
<td>23/F</td>
<td>Sympathetic</td>
<td>+</td>
<td>Orthostatic hypotension, miosis</td>
<td>37</td>
<td>ND/Schwannoma</td>
<td>Resolved at 2 wk</td>
</tr>
<tr>
<td>19/F</td>
<td>Sympathetic</td>
<td>+</td>
<td>Miosis</td>
<td>45</td>
<td>Unspecified/ND</td>
<td>Resolved at 1 y</td>
</tr>
<tr>
<td>38/M</td>
<td>Sympathetic</td>
<td>−</td>
<td>−</td>
<td>45</td>
<td>Unspecified/ND</td>
<td>NA</td>
</tr>
<tr>
<td>38/F</td>
<td>Sympathetic</td>
<td>+</td>
<td>Ptosis</td>
<td>41</td>
<td>Unspecified/ND</td>
<td>Persistent at 15 mo</td>
</tr>
<tr>
<td>19/F</td>
<td>Sympathetic</td>
<td>+</td>
<td>Ptosis</td>
<td>35</td>
<td>Unspecified/ND</td>
<td>Resolved at 1 y</td>
</tr>
<tr>
<td>63/F</td>
<td>Sympathetic</td>
<td>−</td>
<td>−</td>
<td>30</td>
<td>Unspecified/ND</td>
<td>NA</td>
</tr>
<tr>
<td>33/M</td>
<td>Trigeminal</td>
<td>−</td>
<td>−</td>
<td>63</td>
<td>ND/Schwannoma</td>
<td>None</td>
</tr>
<tr>
<td>32/M</td>
<td>Cervical</td>
<td>−</td>
<td>−</td>
<td>35</td>
<td>Unspecified/ND</td>
<td>None</td>
</tr>
<tr>
<td>30/M</td>
<td>Cervical</td>
<td>−</td>
<td>−</td>
<td>54</td>
<td>Unspecified/ND</td>
<td>None</td>
</tr>
</tbody>
</table>

Abbreviations: NA, not applicable; ND, not done; −, absent; +, present.

Table 2. Patient Characteristics for Extracranial Head and Neck Schwannomas of Vagal Origin

<table>
<thead>
<tr>
<th>Age, y/sex</th>
<th>Nerve of Origin</th>
<th>Postoperative Paralysis</th>
<th>Postoperative Symptoms</th>
<th>Size, mm</th>
<th>Preoperative Cytologic/Histology Findings</th>
<th>Outcome of Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>33/F</td>
<td>Vagus</td>
<td>−</td>
<td>−</td>
<td>40</td>
<td>ND/ND</td>
<td>NA</td>
</tr>
<tr>
<td>38/F</td>
<td>Vagus</td>
<td>−</td>
<td>Hoarseness</td>
<td>44</td>
<td>Unspecified/ND</td>
<td>Resolved at 1 y</td>
</tr>
<tr>
<td>61/F</td>
<td>Vagus</td>
<td>+</td>
<td>−</td>
<td>35</td>
<td>Unspecified/ND</td>
<td>NA</td>
</tr>
<tr>
<td>24/M</td>
<td>Vagus</td>
<td>+</td>
<td>Hoarseness</td>
<td>33</td>
<td>Unspecified/ND</td>
<td>Resolved at 1 y</td>
</tr>
<tr>
<td>66/M</td>
<td>Vagus</td>
<td>−</td>
<td>−</td>
<td>53</td>
<td>Unspecified/ND</td>
<td>NA</td>
</tr>
</tbody>
</table>

Abbreviations: NA, not applicable; ND, not done; −, absent; +, present.
sympathetic nerve (7 patients [70%]), trigeminal nerve (1 [10%]) and cervical nerve (2 [20%]) origin (Table 1). All tumors originating in the sympathetic nerves were located in the parapharyngeal space. In the 2 cases (20%) of cervical nerve origin, the tumors were lateral cervical schwannomas. The trigeminal nerve tumor extended from the infratemporal fossa into the maxillary sinus and intracranial space. Because these nerves were unresponsive to the NIM system, we were not able to locate the nerve fibers by electrical stimulation. In 1 case, a nerve bundle was visible on the surface of the tumor. An incision was made along the bundle and the tumor was enucleated. The remaining 9 nonvagal schwannomas did not display visible nerve bundles; therefore, we incised the swollen part of the tumor expected to include fewer nerve fibers. Four of the 7 patients (57%) with sympathetic schwannoma experienced postoperative neurologic dysfunction consistent with Horner syndrome (orthostatic hypotension, miosis, and ptosis). The ptosis improved in 1 patient after 1 year but continued throughout the follow-up period in 1 patient. In the patients with trigeminal and cervical schwannomas, no overt neurologic symptoms were observed. The 3 patients with sensory nerve schwannoma did not show symptoms of nerve dysfunction after operation.

Discussion

We have established a method for the preservation of motor nerve function in cases of vagus nerve schwannoma. The literature suggests 2 types of surgical methods for schwannomas: complete tumor resection with nerve sacrifice and tumor enucleation preserving nerve fibers. The former method causes nerve paralysis in all cases, and the latter method causes paralysis in some cases. In a previous study, 3 70% to 80% of vagus nerve function was preserved by intracapsular piecemeal enucleation. However, to our knowledge, there are only 2 case reports on the resection of vagus nerve schwannomas with functional preservation using EMG monitoring, and those reports did not provide details on the dissection. In our cases, 2 of 5 patients with vagus nerve schwannoma had vocal cord palsy immediately after intracapsular piecemeal enucleation, but normal function returned after 1 year.

Most schwannomas originate in sensory nerve fibers. The literature reports that schwannomas arise in nerves with a sensory component and are associated with sensory ganglia. In the case of the vagus nerve, tumors are likely to involve sensory nerves and it is possible to preserve their function with our technique. All vocal cord function was preserved in our patients with vagus nerve schwannoma. Most sympathetic chain schwannomas are associated with the superior cervical ganglion, with cases reported of sympathetic ganglia becoming embedded in a meshwork of fibers. Thus, if a sympathetic nerve is the origin of the tumor, the preservation of neurologic function is difficult. Four of our 7 patients with sympathetic nerve schwannoma displayed symptoms of paralysis, and ptosis continued in 2 of these patients after surgery. It is difficult to preserve a sympathetic nerve without paralysis. However, with the sympathetic nerve, we may avoid functional nerve injury by preserving the intratumoral meshed nerve fiber with the use of an operating microscope.

Functional preservation after the operation is difficult with tumors located at the center of a nerve fiber. When nerve fibers surround a tumor, we resect the tumor piecemeal to minimize nerve injury and preserve the nerve fibers as much as possible. Moreover, because electrical devices, such as electric knives, can cause bipolar damage to neural tissue, we do not use them for intracapsular enucleation of schwannomas.

The exact recurrence rate of schwannomas after intracapsular enucleation and complete tumor resection is unknown, with few reports documenting postoperative recurrence. It has been suggested that there is little difference in the recurrence rate for total tumor resection and intracapsular enucleation. In our study, no patient experienced recurrence. However, we had a short follow-up period, with a median of 2.7 years after the operation. A longer follow-up time and evaluation of recurrence with imaging is necessary.

Preoperative diagnosis of the origin of head and neck schwannomas is difficult. The most common nerve origins of extracranial schwannomas are the vagus and cervical sympathetic nerves. When vagus nerve origin is likely, we apply electrical nerve stimulation to the top and bottom of the tumor using an EMG system; by observing the response to electrical stimulation, the origin of the tumor can be confirmed. Therefore, EMG systems are a powerful tool for identifying the nerve of origin of a tumor and its neural involvement.

As a result of our surgical strategy for extracranial schwannomas, the incidence of nerve injury can be reduced and nerve function can be preserved. Further investigation is needed to establish a method for more effective preservation of nerve function in cases of sympathetic schwannoma.

Conclusions

In cases of extracranial head and neck schwannomas, careful surgical planning is needed to avoid nerve damage and unfavorable outcomes. We need a surgical strategy that facilitates nerve preservation as well as complete tumor resection. In the present study, using an EMG system and a microscope, intracapsular resections of schwannomas were performed. We were able to preserve nerve function in all vagus nerve schwannoma cases and did not observe recurrence. However, 4 of the 7 sympathetic nerve schwannoma cases showed symptoms of paralysis after the operation. Intracapsular schwannoma resection with electrical nerve monitoring reduces the frequency of nerve injury.
Acquisition, analysis, or interpretation of data: Ijichi, Maseki, Takano, Murakami.
Drafting of the manuscript: Ijichi, Kawakita, Beppu, Takano, Murakami.
Critical revision of the manuscript for important intellectual content: Ijichi, Maseki, Takano, Murakami.
Administrative, technical, or material support: Ijichi, Kawakita, Maseki, Takano.
Study supervision: Murakami.

Conflict of Interest Disclosures: None reported.

REFERENCES