

## Case Report

### Cervical Sympathetic Chain Schwannomas

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**Abstract** : Surgery for cervical sympathetic chain schwannomas may result in post-operative Horner's syndrome. Thus, appropriate informed consent and adequate surgical experience are required to prevent any decrease in patients' quality of life (QOL) following such surgery. Here we report three cases of cervical sympathetic chain schwannomas, focusing on diagnosis and postoperative neuroparalysis. A schwannoma is suspected on pre-operative imaging when the common carotid artery (internal and external carotid artery) and internal jugular vein are located in front of or alongside the tumor, and cases in which the artery and vein are not separated are considered to be of sympathetic nerve origin. Since the origin nerve comes from the back anatomically, great care is needed during surgery. None of the patients in this study had symptoms of nerve paralysis postoperatively.

**Key words** : schwannoma, cervical sympathetic chain, intercapsular resection

## Introduction

Schwannomas are comparatively rare neck tumors, but still a diagnosis that must always be kept in mind in clinical practice. Surgery designed to preserve nerve function is the standard choice for treating schwannoma and many enucleations of schwannomas have now been reported (REF)<sup>1-18)</sup>; however, due to the rarity of these tumors, reports of the same surgical technique being carried out by the same surgeon are few. In addition, since any such surgery may reduce a patient's quality of life (QOL), appropriate informed consent and adequate surgical experience are needed before conducting Schwannoma resections. Indeed, surgical treatments involving the cervical sympathetic chain may cause ipsilateral Horner's syndrome postoperatively. Herein, we report three cases of cervical sympathetic chain schwannomas.

## Patients and methods

This study reviewed three patients with neck schwannomas that arose from the cervical sympathetic chain who underwent intercapsular resection by the same surgeon. These cases were

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Table 1. Cases of cervical sympathetic chain schwannoma

	Age (years)/sex	Tumor length (mm)	Symptoms before operation	Symptoms after operation	Location of tumor relative to vessels	Separation of the artery and the vein*
1	26 / F	40	None	None	Front	None
2	65 / F	44	None	None	Front	None
3	52 / F	48	None	None	Outside	None

\*common artery and internal jugular vein

selected from 32 neck schwannomas treated in our center from April 2005 to March 2014. Age, sex, pre-operative neurological symptoms, size of the tumor, tumor and vessel findings on MRI, and postoperative neurological symptoms were examined.

## Results

Of 32 cases of neck schwannoma treated at our center during the study period, three arose from the cervical sympathetic chain. Table 1 lists the details of these cases.

### *Age / sex*

The patient age ranged from 26 to 65 years; the average age was 47.7 years. All patients were women.

### *Neurological symptoms before the operation*

All cases were asymptomatic prior to surgery, and no cases had neurological symptoms when the tumor was touched or had pressure applied to it.

### *Size of tumors*

The maximum diameter of the enucleation specimens ranged from 40 to 48 mm, with an average of 44 mm.

### *Sites of the tumors and vessels on MRI*

In two of the three cases, the common carotid artery (internal and external carotid artery) and the internal jugular vein were located anterior to the tumor, while in the remaining case they were alongside the tumor. No cases showed separation of the artery and vein.

### *Postoperative neurological symptoms*

None of the cases in this series had symptoms of nerve paralysis after surgery.

The imaging and surgical findings for each case are described below.

### *Cases*

Case 1 : A 26-year-old woman

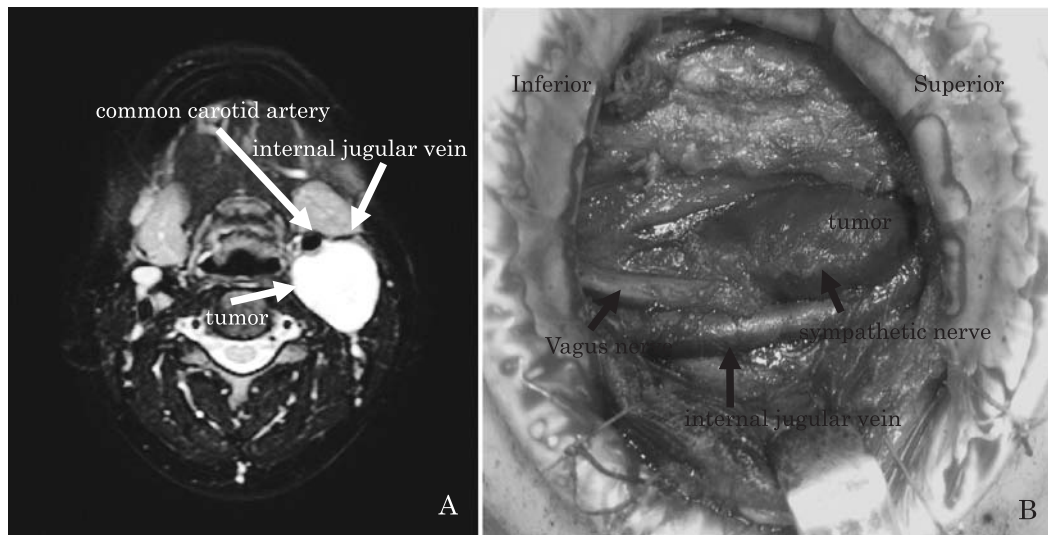


Fig. 1.

- A: MRI findings. The tumor is 40 mm in size with a clear border and located behind the left submandibular gland. The tumor showed high intensity on T2-weighted imaging, and had pushed the common carotid artery and the internal jugular vein anteriorward.
- B: Findings at surgery. A sympathetic nerve fiber is visible emerging from the back of the tumor.

MRI findings (Fig. 1A): The tumor was 40 mm in size with a clear border and was behind the left submandibular gland. The tumor showed high intensity on T2-weighted imaging and it pushed the common carotid artery and the internal jugular vein anteriorly.

Findings at surgery (Fig. 1B): A sympathetic nerve fiber was visible at the back of the tumor.

Case 2: A 65-year-old woman

MRI findings MRI (Fig. 2A): The tumor was 44 mm in size with a clear border and was behind the left submandibular gland. The tumor showed high intensity on T2-weighted imaging and it also had pushed the common carotid artery and the internal jugular vein anteriorly.

Findings at surgery (Fig. 2B): The sympathetic nerve fiber was proven to be the origin from the back of the tumor, and it was identified running ahead of the tumor.

Case 3: A 52-year-old woman

MRI findings (Fig. 3A): The tumor was 48 mm in size with a clear border and was behind the left submandibular gland. The tumor showed non-uniform high intensity on T2-weighted imaging and it had pushed the common carotid artery and the internal jugular vein to the side.

Findings at surgery (Fig. 3B): The sympathetic nerve stem was identified as leading to the tumor from the backside, and the nerve fiber was observed running to the backside of the tumor.

## Discussion

A schwannoma is a tumor that originates from a Schwann cell. The most frequent stem nerves involved in neck schwannomas are the vagus, brachial plexus, cervical, and sympathetic

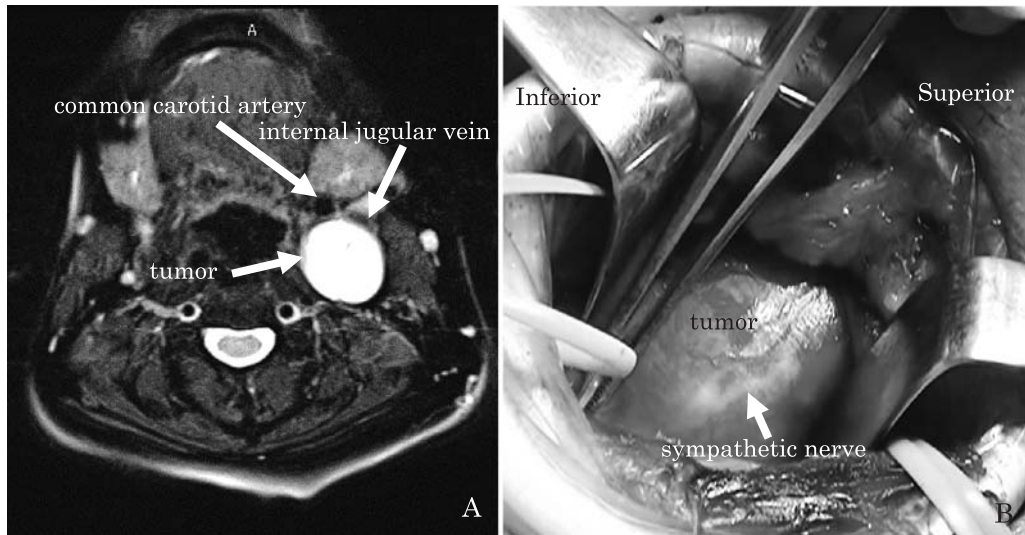


Fig. 2.

- A: MRI findings. The tumor is 44 mm in size with a clear border and located behind the left submandibular gland. The tumor showed high intensity on T2-weighted imaging and had pushed the common carotid artery and the internal jugular vein anteriorward.
- B: Findings at surgery. It was proven that the sympathetic nerve fiber was the nerve stem coming from the back of a tumor, and it could be identified running in front of the tumor.

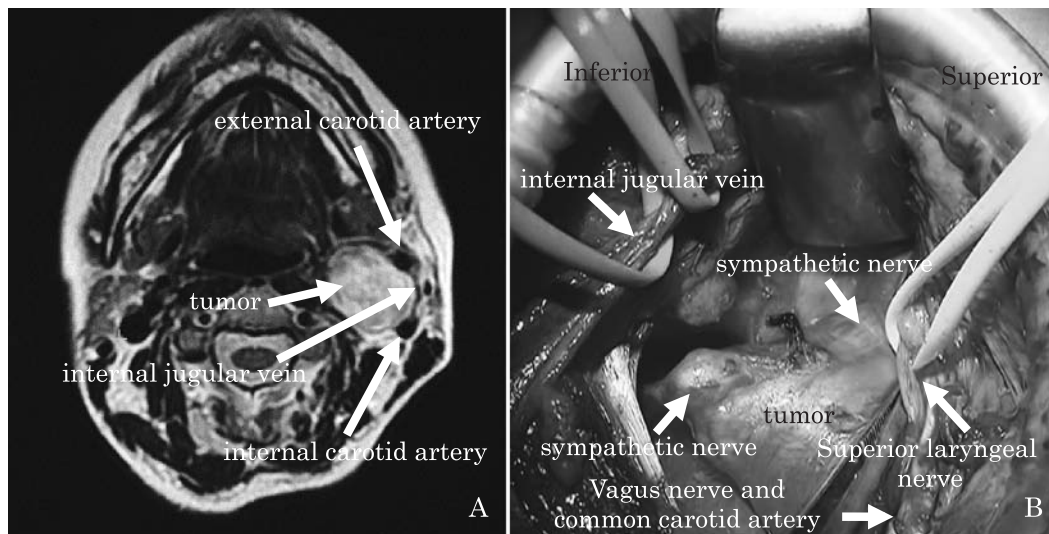


Fig. 3.

- A: MRI findings. The tumor is 48 mm in size with a clear border and located behind the left submandibular gland. The tumor showed non-uniform high intensity on T2-weighted imaging, and had pushed the common carotid artery and the internal jugular vein anteriorward.
- B: Findings at surgery. The sympathetic nerve stem leading to the tumor has been identified to originate from the backside. The nerve fiber has been identified as running to the dorsum of the tumor.

nerves<sup>1-3</sup>). In 32 neck schwannomas treated in our department, the origin was most commonly a vagus nerve, followed by the brachial plexus, a cervical nerve, and then a sympathetic nerve. This incidence of tumor location is consistent with previous reports<sup>2-5</sup>). Such tumors are often indolent preoperatively, and all were asymptomatic before operation.

In the preoperative diagnosis, a cervical sympathetic chain schwannoma appears similar to a vagus nerve case on imaging. Surgery then reveals whether the vagus nerve is involved, and the final identification of the stem nerve is sometimes not definitive until Horner's syndrome occurs after the operation<sup>6-8</sup>). Cases have been reported in which the common carotid artery and the internal jugular vein were located anterior to sympathetic nerve tumors, and separation from the artery and vein was not seen on MRI<sup>9</sup>). However, if no such presentation is observed on imaging, it is often difficult to recognize the stem nerve in neck schwannoma cases<sup>7</sup>). A sympathetic chain schwannoma can look like a vagus schwannoma on ultrasound examination, although a sympathetic chain schwannoma is suspected if the vagus nerve is separate from the tumor<sup>10</sup>). In two of the present cases, MRI showed the tumor located posterior to the common carotid artery and the internal jugular vein, while in the third case it was located to the alongside of these vessels.

Separation of the artery and the vein was not visible in any of the cases presented herein. Anatomically, sympathetic nerves are located in the back and middle of the common carotid artery and the internal jugular vein; thus, these vessels are located either anterior to or alongside of sympathetic chain schwannomas. In addition, when attempting to differentiate a cervical chain schwannoma from that of a vagus nerve, an important finding that points to a sympathetic origin is separation of the artery and the vein.

With respect to diagnosis during surgery, the main part of the cervical sympathetic chain exists on both sides of the vertebrae with many branches. We consider it difficult to find a narrow nerve stem connected from the back of a tumor in such cases, because the common carotid artery (internal and external carotid artery), the internal jugular vein, and the vagus nerve are located anterior to the tumor and the nerve. Therefore, when the nerve stem cannot be identified during the operation, a sympathetic origin is often only confirmed when postoperative neurological deficits appear. We reported previously that for such patients, we exfoliate carefully so that the surrounding tissues are not cut at the time of tumor enucleation and are removed only after identifying the nerve stem and the course of the nerve fiber with respect to the tumor<sup>11</sup>). In all present cases, we confirmed at surgery that the tumors were not contiguous with the vagus nerve and that the nerve was connected from the back of the tumor; intercapsular resection was then performed. Therefore, it was possible to remove tumors without causing postoperative neurological symptoms.

The surgical methods for neck schwannomas include total enucleation, intercapsular resection, and undercapsular resection<sup>10, 12, 13</sup>). Prior to 2000, the most common technique performed was total enucleation, with this modality selected for 55 of 85 cases by Haraguchi and Okuno<sup>5</sup>), and for 40 of 55 cases by Kida *et al*<sup>4</sup>). In addition, Valentino *et al*<sup>14</sup>) reported the appearance of permanent neuroparalysis in 64% of 146 cases of neck schwannoma. Because neuropreservation

was difficult, Valentino et al suggested total enucleation and nerve transplantation. However, in the mid-2000s, Hashimoto<sup>12, 13)</sup> reported intercapsular resection as a postoperative nerve function-preservation technique that achieved good results, and after 2000, reports of no neuroparalysis after surgery have been more frequent<sup>2, 3, 11-13, 15-18)</sup>.

We attempt to identify the stem nerve before surgery and understand the course of the nerve anatomically. With intercapsular resection, the tumor is resectable without postoperative paralysis, the appearance of which can mask recurrence of the tumor during follow-up. In cases of neck schwannomas, careful follow-up is therefore necessary that also considers each patient's postoperative QOL.

## Conclusion

Three cervical sympathetic chain schwannomas treated by intercapsular resection by the same surgeon were reviewed. No cases showed symptoms of nerve paralysis postoperatively. A schwannoma is suspected based on the preoperative imaging findings of the common carotid artery (internal and external carotid artery) and the internal jugular vein located anterior to or alongside of the tumor, and when the artery and vein are not separated. It is necessary in such cases to consider a sympathetic nerve origin, and since the nerve origin comes from the back anatomically, extreme care is needed to avoid postoperative nerve deficits.

## Conflict of interest disclosure

The authors declare no conflict of interest.

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