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# Surgical management of multiple schwannomas scattered on a single peripheral nerve

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## Abstract

**Background** Schwannoma is a common benign tumor. Most schwannomas are sporadic, but approximately 5% of schwannomas are multifocal. Schwannomas are sometimes present in a skip-like pattern on a single continuous peripheral nerve (Multiple schwannomas scattered on a single peripheral nerve: MSSPN). In this study, we present the clinical characteristics of MSSPN in the limbs and propose a treatment strategy based on treatment outcomes.

**Methods** The medical records of 918 patients diagnosed with schwannoma in the limbs were retrospectively reviewed. Among these cases, multiple schwannomas occurring in a single peripheral nerve and spaced more than 1 cm apart were defined as MSSPN. We investigated the clinical characteristics and surgical outcomes.

**Results** Seven patients with MSSPN in the limbs were identified, which represented 0.8% of all cases. There were six females and one male, and the mean age was 50 years. The location of MSSPN was the upper limbs in two cases and the lower limbs in five cases. After surgery, among the five cases in which all tumors were enucleated, neurological symptoms worsened in 4 cases, all of which involved deep nerve tumors. The one case among these five cases in which neurological symptoms improved involved a superficial subcutaneous tumor. Neurological symptoms improved in both of the two cases in which only the tumor causing the main complaint was enucleated; both cases involved deep nerve tumors. The average postoperative follow-up was 8 months, and no cases required reoperation or complained of recurrence.

**Conclusion** This is the first study to address the surgical strategies of multiple schwannomas occurring in a skip-like pattern on a single peripheral nerve. In surgery for MSSPN, selective enucleation of only the most symptomatic tumors may be effective in preventing the worsening of symptoms after surgery.

**Keywords** Multiple schwannomas, Segmental schwannomatosis, Multiple schwannomas scattered on a single peripheral nerve, Treatment strategy, Selective enucleation

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## Introduction

Schwannoma is a common type of benign peripheral nerve sheath tumor composed of Schwann cells. Most schwannomas are solitary, but approximately 5% of schwannomas occur multifocally (multiple schwannomas) [1]. Multiple schwannomas can occur in the context of NF2-related schwannomatosis, which is associated with acoustic schwannomas, and schwannomatosis not associated with acoustic schwannomas [2]. Plexiform schwannoma, in which multiple schwannomas grow in a plexiform pattern on a single peripheral nerve, is known as a subtype of multiple schwannomas, and there are several case reports and case series on this condition [3–5]. On the other hand, there have been very few reports focusing on the occurrence of multiple schwannomas separately on a single peripheral nerve [6, 7]. This study identifies this condition under the following name: multiple schwannomas scattered on a single peripheral nerve (MSSPN). No studies have reported on surgical methods, and the treatment strategy for this condition remains largely unknown. The purpose of this study is to clarify the clinical characteristics and treatment outcomes of cases of MSSPN in the limbs treated at our hospital and to propose a treatment strategy based on these findings. We hypothesize that selective tumor removal may result in better neurological outcomes, and present the following case series to investigate this hypothesis.

## Methods

In this study, we retrospectively reviewed the medical records of 918 patients diagnosed with schwannoma in the limbs based on the surgical case list at our hospital from 1978 to 2020. Among these cases, multiple schwannomas which occurred in a single peripheral nerve or in longitudinally continuous peripheral nerves with different anatomical names, such as the sciatic nerve and tibial nerve, and in which the tumors were more than 1 cm apart, were defined as MSSPN. Cases in which multiple schwannomas were in contact with each other and cases in which the distance between the tumors was less than 1 cm were excluded. In cases where tumors occurred in multiple non-contiguous nerves, only the nerves on which surgery was performed were focused on. Histopathological diagnosis was made by several pathologists well-acquainted with about bone and soft tissue tumors.

The following clinical characteristics were investigated: age at initial visit, sex, whether magnetic resonance imaging (MRI) was performed, number of recognized tumors occurring in the operated nerve, locations and names of nerves, symptoms at first visit, number of enucleated tumors, postoperative neuropathic symptoms, and postoperative follow-up period, and recurrence. "Tenderness" is defined as localized neuralgia on palpation, and "radiating pain" is defined as neuropathic pain that shoots along the distribution of the nerve. The surgery was performed by several senior surgeons. General anesthesia was used in all cases. The surgical procedure for schwannomas, including MSSPN, at our hospital is as follows: First, the tumor is exposed. Then, an area free of nerve fibers is identified. Finally, the capsule is incised and the tumor is removed. Neuromonitoring was not performed during surgery. Regarding the number of schwannomas enucleated, our previous policy for MSSPN was to enucleate all tumors that were identified and deemed excisable, but we observed postoperative neurological deficit in some cases. Therefore, we changed our strategy and decided to carefully examine symptoms such as tenderness, radiating pain, and Tinel sign, and to selectively remove only the tumors causing the symptoms. Postoperative complications were defined as a worsening of preoperative neurological symptoms or the onset of new neurological symptoms. The postoperative follow-up period ended when the wound healed and postoperative symptoms improved or remained unchanged.

## Results

Out of 918 schwannoma cases, 7 (0.8%) met the criteria for MSSPN (Table 1).

There were six females and one male, and the median age at first visit was 51 years (range: 28–65 years). None of the cases had a family history of schwannoma. Preoperative MRI was performed in six cases. In one case, an ultrasound diagnosis was performed because the case occurred before MRI was widely available. Multiple schwannomas, a tumor in multiple nerves, was observed in three cases. None of these cases showed acoustic schwannomas on head MRI, and no genetic testing was performed in any of the cases. The median number of tumors was three (range: 2–7). The location of MSSPN was the upper limbs in 2 cases and the lower limbs in 5 cases. In three cases, the tumors were located in skip-like locations on longitudinally continuous peripheral nerves with different anatomical names (median into digital nerve, sciatic into tibial nerve, tibial into plantar nerve). All patients had significant pain symptoms prompting surgical intervention.

Regarding the surgical method, all tumors were enucleated in five of the seven cases, and only the most symptomatic tumor was selectively enucleated in two cases. Of the five patients who had all tumors enucleated, two who had only tenderness before surgery developed new radiating pain after surgery (cases 2 and 5 (Fig. 1)). Furthermore, both of the two patients who had radiating pain before surgery required oral medication due to worsening of radiating pain after surgery, and required postoperative follow-up periods of 14 and 25 months, respectively (cases 1 and 3). In these four cases, the tumors occurred on deep nerves. The preoperative visual analogue scale

**Table 1** Seven cases of MSSPN

| | case | Age | Sex | MS | Location | Originate Nerve |
|---|---|---|---|---|---|---|
| All enucleation | | | | | | |
| | 1 | 61 | M | - | Axilla | Radial |
| | 2 | 28 | F | + | Hand, Finger | Median, Digital |
| | 3 | 65 | F | - | Thigh | Tibial |
| | 4 | 30 | F | - | Foot (medial) | Subcutaneous |
| | 5 | 51 | F | - | Foot | Tibial, Plantar |
| Selective enucleation | | | | | | |
| | 6 | 64 | F | + | Hip, Thigh | Sciatic, Tibial |
| | 7 | 50 | F | + | Leg | Peroneal |

| Preoperative symptoms | Tumors | Postoperative symptoms | VAS | Follow up (month) |
|---|---|---|---|---|
| Numbness, Radiating pain | 3 / 3 | Radiating pain (med) | 3→8 | 14 |
| Tenderness | 4 / 4 | Radiating pain | 0→4 | 2 |
| Numbness, Radiating pain | 2 / 2 | Radiating pain (med) | 3→10 | 25 |
| Tenderness | 7 / 7 | No | 3→0 | 1 |
| Tenderness | 2 / 2 | Radiating pain | 1→6 | 5 |
| Numbness, Tenderness | 2 / 3 | No | 3→0 | 2 |
| Tenderness, Radiating pain | 2 / 3 | No | 4→0 | 5 |

MS = multiple schwannomas, VAS = visual analogue scale, (med) = required medication, No = no new or residual symptoms

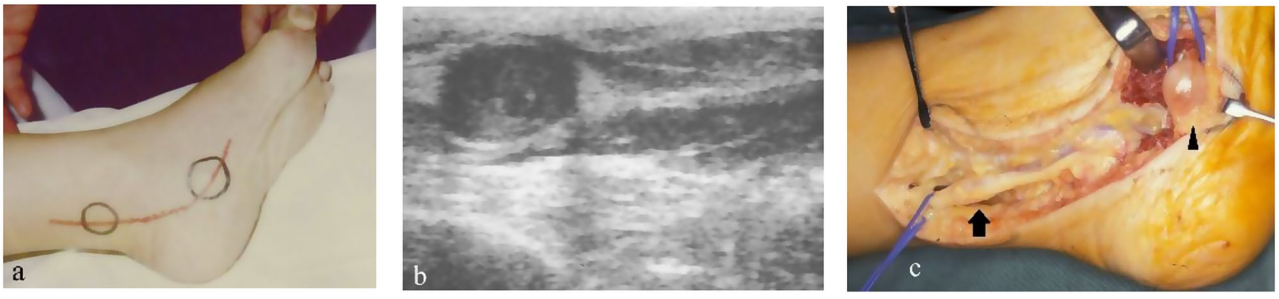


**Fig. 1** Case 5 was one of the cases in which neurological symptoms worsened postoperatively. (**a**) The patient had distal tumor tenderness preoperatively. Two tumors were palpable. (**b**) A solid tumor was observed on ultrasound. Schwannoma was suspected. (**c**) At surgery, both palpable tumors were enucleated. The tumors were located separately in longitudinal continuity (arrow = tibial nerve tumor, arrowhead = plantar nerve tumor). The patient complained of radiating pain after surgery. The radiating pain improved 5 months after surgery

(VAS) worsened in all four cases after surgery. At the end of follow-up, symptoms in case 3 were worse than before surgery, whereas the remaining three cases had improved. Among the five cases in which all tumors were enucleated, the only case in which symptoms did not worsen involved a superficial subcutaneous tumor on the foot (case 4). Three tumors were present in the two cases in which tumors were selectively enucleated (case 6 and 7). In both of these cases, the tumors occurred in deep nerves. Of the three tumors in each case, only the two with the most severe symptoms were enucleated. In both cases, the preoperative VAS was 0 after surgery, and no new neurological symptoms were observed. Worsening of neurological symptoms occurred in 80% (4/5) of patients who underwent total tumor enucleation, but in 0% (0/2) of patients who underwent selective tumor enucleation. The median postoperative follow-up period for the seven MSSPN cases was 5 months, and no cases required reoperation. None of the patients returned to our hospital after the end of follow-up complaining of mass, pain, or numbness, or had symptoms due to unenucleated schwannomas.

## Discussion

This is the first study to address the surgical strategies of MSSPN in the limbs. Four of the five cases in which all tumors were enucleated experienced worsening or new neurological symptoms after surgery. In all four of these cases, the tumors occurred in deep nerves. Enucleation of multiple tumors in MSSPN requires multiple discontinuous incisions on a single peripheral nerve. While this method ensures all tumors are removed, it markedly increases the risk of damage to functional nerves. In other words, the more tumors that are removed, the greater the risk of damage to functional nerves. We speculate that this is the reason for the poor postoperative outcomes in the cases in which all tumors were enucleated. On the other hand, in the two patients

whose tumors were removed selectively, only the causative tumors were enucleated based on symptoms such as severe tenderness and radiating pain. In these cases, the symptoms improved without any new symptoms of numbness or pain. Selective treatment may have minimized the incisions to the peripheral nerve, which may have reduced damage to the functional nerve. One implication of this study is that when tumors occur in deep major nerves such as the median nerve or sciatic nerve, it may be better to avoid removing the entire tumor.

There have been several reports on surgical indications depending on the type of schwannoma. Our findings are consistent with current recommendations for schwannomatosis, which recommend surgery only for symptomatic tumors [8]. In solitary schwannomas, surgery is generally indicated for symptomatic or growing tumors [9–12]. Similarly, our findings suggest that a more conservative surgical approach may be justified in cases of MSSPN.

In MSSPN, multiple tumors exist in one longitudinally continuous peripheral nerve, and in some cases it may be difficult to identify the tumors causing radiating pain before surgery. Current imaging methods such as MRI and ultrasound also have limited correlation with clinical symptoms. Therefore, as in this study, selective resection should only remove the tumor that is most symptomatic based on tumor size and medical history. However, these physical examinations are not reliable. It is expected that advanced neuroimaging or intraoperative nerve stimulation mapping will be utilized in future studies.

We acknowledge there are several limitations in this study. First, only 7 (0.8%) of the 918 schwannomas that underwent surgery were cases of MSSPN. The low incidence of MSSPN and the very small number of cases in this study make the analysis of surgical outcomes in this study insufficient. In addition, the period of case collection was long, more than 40 years. The surgeons were not standardized, and there may be differences in treatment outcomes depending on the surgeon. The number of tumors enucleated was based on clinical judgment and was not randomized (e.g., only tumors that were considered easier were resected, or conversely, selective resection was selected when some tumors were deemed too risky to be resected). Thus, as a retrospective study, this study lacks a uniform protocol, which may have introduced selection bias. A larger multi-center series or registry data study would be much more suitable means for investigating the hypothesis of the present study. Second, regarding the incidence, in cases treated as solitary schwannomas, not all longitudinally connected peripheral nerves were examined by MRI or ultrasound unless symptoms were present. Therefore, the incidence recorded in this study may be lower than the actual incidence. Thirdly, in this study, the postoperative follow-up period was short because treatment ended when the wound healed. In addition, objective neurological function tests such as electromyograms were not used. This may mean that long-term results such as growth of residual tumors and the appearance of symptoms may not have been recorded. In particular, careful monitoring of tumor growth and symptom appearance should be considered when tumors remain that were not selectively enucleated. Continuous surveillance of residual tumors by regular clinical or imaging examinations will be necessary in future studies. Finally, genetic evaluation was not performed in this study. Because individuals with genetic schwannomatosis may develop new tumors over time, it is suggested that genetic evaluation be performed in the management of MSSPN in the future.

Intraoperative nerve monitoring with electric stimulation is sometimes performed during surgery for schwannoma to avoid motor nerve damage. In this study, skeletal muscle action potentials were not monitored during surgery in all patients, but fortunately, no patients developed motor paralysis after surgery. Because electrical stimulation is performed only after the nerve is exposed during surgery, we believe it is not useful for preoperative selection of which tumors to enucleate. To determine which tumors should be enucleated, surgeons should thoroughly and carefully examine each patient before surgery. However, surgery for MSSPN is a concern due to the risk of neurological deficit caused by multiple incisions. Therefore, neuromonitoring may be useful when enucleating each schwannoma to prevent neurological motor fiber damage. In the future, surgeons should thoroughly and carefully examine each patient before surgery to determine which tumors should be enucleated and consider using neuromonitoring for each schwannoma as it is enucleated.

## Conclusion

This study investigated the clinical characteristics and surgical outcomes of MSSPN in the limbs. Enucleating all tumors is advantageous for removing the tumors. However, multiple discontinuous incisions on a single peripheral nerve increase the risk of damaging functional nerves unrelated to the tumors. Therefore, enucleating all tumors may lead to an increase in postoperative new neurological deficit. When treating MSSPN, our findings suggest that postoperative neurological deficit may be reduced by carefully examining patients preoperatively and selecting the tumors to be excised, rather than blindly excising all tumors.

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**Author contributions**
TK and SM designed the study; MS, YF, KH and TT collected data; KA supervised the experiments; TK and SM wrote the manuscript.

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**Data availability**
The data used during the current study are available from the corresponding author on reasonable request.

## Declarations

**Ethics approval and consent to participate**
This study followed the Declaration of Helsinki. This study was approved by the Institutional Review Board at the Cancer Institute Hospital of the Japanese Foundation for Cancer Research (IRB No.2020-GA-1168). Informed consent for participate in this study was obtained by all participants.

**Consent for publication**
Informed consent for publication was obtained by all participants in this study.

**Competing interests**
The authors declare no competing interests.

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**References**

1. Antinheimo J, Sankila R, Carpen O, Pukkala E, Sainio M, Jaaskelainen J. Population-based analysis of sporadic and type 2 neurofibromatosis-associated meningiomas and schwannomas. Neurology. 2000;54:71–6. https://doi.org/10.1212/WNL.54.71.
2. Scott RP, Ludwine M, Eric L, et al. Updated diagnostic criteria and nomenclature for neurofibromatosis type 2 and schwannomatosis: an international consensus recommendation. GenetMed. 2022;24:1967–77. https://doi.org/10.1016/j.gim.2022.05.007.
3. Agaram NP, Prakash S, Antonescu CR. Deep-seated plexiform Schwannoma: a pathologic study of 16 cases and comparative analysis with the superficial variety. Am J Surg Pathol. 2005;29:1042–8.
4. Araghi F, Tabary M, Kamyab K, Forouzanfar MM, Robati RM. A rare case of plexiform Schwannoma on the foot. Clin Case Rep. 2021;9:04234. https://doi.org/10.1002/ccr3.4234.
5. Ejiyooye TF, Dirisanala S, Makky AH, Mahjabeen SS, Sajjad T, Khan A. A rare case of plexiform Schwannoma of the little finger and its management: A case report. Cureus. 2022;14:26391. https://doi.org/10.7759/cureus.26391.
6. Min HJ, Kim KC, Jun SH, Lee YG. Schwannomatosis on a single foot: a case report. Foot Ankle Spec. 2015;8:226–9. https://doi.org/10.1177/1938640014565047.
7. Wang ZX, Chen SL, Yi CJ, Li C, Rong YB, Tian GL. Segmental schwannomatosis in upper-extremity: 5 cases report and literature review. Beijing Da Xue Xue Bao Yi Xue Ban. 2013;45:698–703.
8. Gonzalvo A, Fowler A, Cook RJ, et al. Schwannomatosis, sporadic schwannomatosis, and Familial schwannomatosis: a surgical series with long-term follow-up. J Neurosurg. 2011;114:756–62. https://doi.org/10.3171/2010.8.JNS091900.
9. Siqueira MG, Socolovsky M, Martins RS, et al. Surgical treatment of typical peripheral schwannomas: the risk of new postoperative deficits. Acta Neurochir (Wien). 2013;155:1745–9. https://doi.org/10.1007/s00701-013-1818-6.
10. Kim SM, Seo SW, Lee JY, Sung KS. Surgical outcome of schwannomas arising from major peripheral nerves in the lower limb. Int Orthop. 2012;36:1721–5. https://doi.org/10.1007/s00264-012-1560-3.
11. Ujigo S, Shimose S, Kubo T, Fujimori J, Ochi M. Therapeutic effect and risk factors for complications of excision in 76 patients with Schwannoma. J Orthop Sci. 2014;19:150–5. https://doi.org/10.1007/s00776-013-0477-z.
12. Fujibuchi T, Miyawaki J, Kidani T, Miura H. Risk factors for neurological complications after operative treatment for schwannomas. J Clin Neurosci. 2017;46:136–40. https://doi.org/10.1016/j.jocn.2017.09.002.

## Publisher's note