



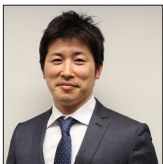
Case Report

# Laminectomy triggers symptomatic growth of spinal schwannoma in a patient with schwannomatosis

Takahiro Oyama<sup>1</sup>, Yusuke Nishimura<sup>1</sup>, Yoshitaka Nagashima<sup>1</sup>, Tomoya Nishii<sup>1</sup>, Masahito Hara<sup>2</sup>, Masakazu Takayasu<sup>3</sup>, Ayako Sakakibara<sup>4</sup>, Ryuta Saito<sup>1</sup>

<sup>1</sup>Department of Neurosurgery, Nagoya University, Nagoya, <sup>2</sup>Department of Neurosurgery, Aichi Medical University, Nagakute, <sup>3</sup>Department of Neurosurgery, Inazawa Municipal Hospital, Inazawa, <sup>4</sup>Department of Pathology and Laboratory Medicine, Nagoya University, Nagoya, Japan.

E-mail: Takahiro Oyama - oyama.takahiro.0504@gmail.com; \*Yusuke Nishimura - yusuken0411@med.nagoya-u.ac.jp; Yoshitaka Nagashima - nagashima4251@gmail.com; Tomoya Nishii - tomoya.nishii@gmail.com; Masahito Hara - masahara219@hotmail.co.jp; Masakazu Takayasu - mtakayas@aichi-med-u.ac.jp; Ayako Sakakibara - ayasak@med.nagoya-u.ac.jp; Ryuta Saito - ryuta@med.nagoya-u.ac.jp



\*Corresponding author:

Yusuke Nishimura,  
Department of Neurosurgery,  
Nagoya University, Nagoya,  
Japan.

yusuken0411@med.nagoya-u.ac.jp

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## ABSTRACT

**Background:** Schwannomatosis (SWN) is genetically similar to neurofibromatosis type 2 (NF2) and represents a NF2 gene mutation. Previous studies have shown that these mutations in both neurons and Schwann cells can lead to the development of schwannomas after nerve crush injuries. Here, we reviewed the potential pathoanatomical mechanisms for the development of a trauma-induced spinal schwannomas in a 55-year-old male with SWN.

**Case Description:** A 49-year-old male had originally undergone a L3–L5 lumbar laminectomy for stenosis; the schwannomas seen on the preoperative magnetic resonance imaging (MRI) were not resected. Now at age 55, he newly presented with low back pain and numbness in the left L5 dermatome, and he was diagnosed with an L4 vertebral level cauda equina tumor on MRI. Following gross-total resection, the histopathological assessment revealed a Ki-67 labeling index 5–10% in hotspots (i.e., slightly higher than the normal range of schwannomas) and a 20% mosaic loss of SMARCB1. Based on these criteria, he was diagnosed as having SWN.

**Conclusion:** In this patient with SWN, compression/physical trauma to nerves of the cauda equina during the L3–L5 laminectomy 6 years ago likely caused the progression of schwannoma.

**Keywords:** Laminectomy, Nerve injury, Neurofibromatosis, Schwannomatosis, Spinal schwannoma

## INTRODUCTION

Schwannomatosis (SWN) is a rare subtype of neurofibromatosis (NF) and is characterized by multiple schwannomas.<sup>[5,14]</sup> SWN is genetically similar to NF type 2 (NF2), and patients present with NF2 gene mutations.<sup>[13]</sup> NF2 gene mutations may involve both neurons and Schwann cells and potentially trigger the development of schwannomas after nerve crush injuries.<sup>[4,10]</sup> Spinal nerve schwannomas account for a quarter of all primary spinal cord tumors.<sup>[9]</sup> Here, a 55-year-old male with SWN had a prior L3–L5 laminectomy for lumbar canal stenosis 6 years ago and now presented with a new or further enlarged L4 vertebral level schwannoma.

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## CASE DESCRIPTION

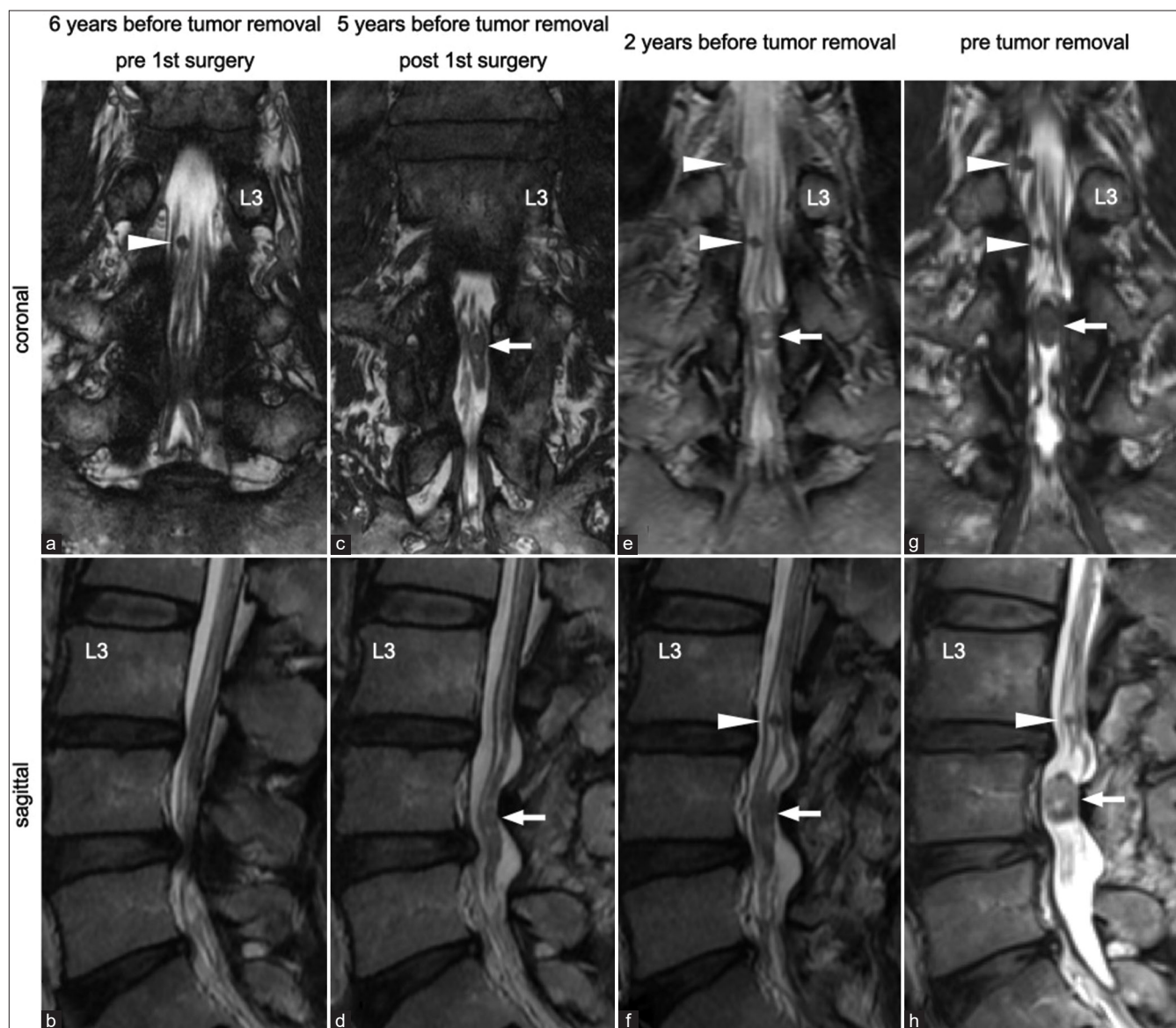
### History

Six years ago, a 49-year-old male had undergone a L3–L5 laminectomy for lumbar canal stenosis; notably, the small intradural schwannomas identified on the preoperative magnetic resonance imaging (MRI) were not removed but helped establish the underlying diagnosis of SWN. Now at age 55, he newly presented with low back pain and numbness in the left L5 dermatome, and he was diagnosed with a L4 vertebral level cauda equina tumor (i.e., either

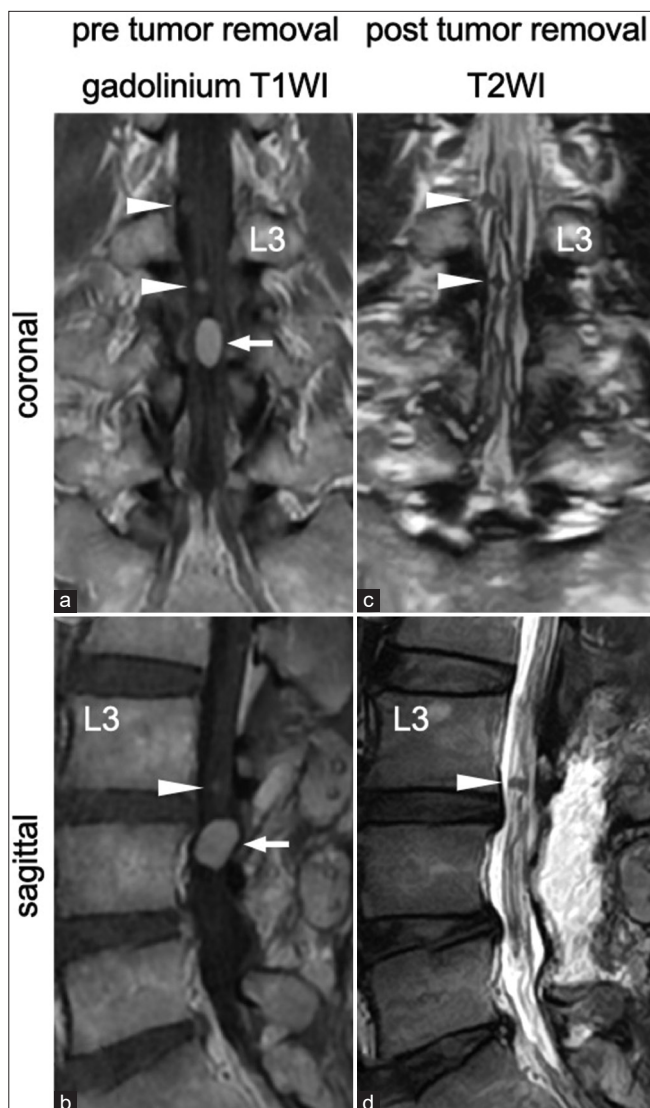
newly developed or progressed) on MRI [Figures 1a-h arrow]. The lesion demonstrated the following classical MRI findings, such as a low-intensity mass on T1-weighted images, isointensity mass on T2-weighted images, and homogeneously enhancing mass on gadolinium T1-weighted images [Figures 2a and b arrow].

### Surgical procedure

A revision L3 and L4 laminectomy were performed. With ultrasound guidance, the tumor was visualized within the cauda equina at the L4 vertebral level and was found to be



**Figure 1:** (a and b) He had originally undergone posterior laminectomy for lumbar canal stenosis at L3–L5 levels at another hospital 6 years before his first visit to us. There seemed no tumors at the L4 vertebral level. Several small tumors (arrowhead) had already existed in the cauda equina before laminectomy. (c and d) He had another tumor pointed out at the L4 vertebral level (arrow) 1 year after surgery. (e-h) Only the newly detected tumor (arrow) had gradually grown in size and had become symptomatic despite other small tumors (arrowhead) remaining the same in size.

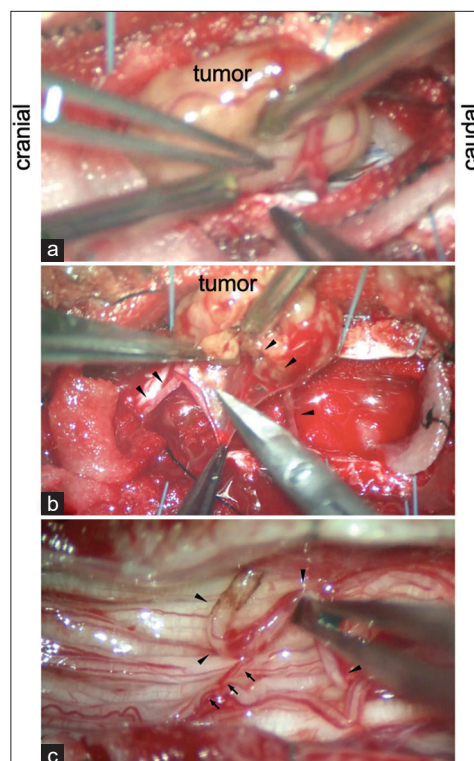


**Figure 2:** (a and b) The newly detected tumor (arrow) and several small tumors (arrowhead) were homogeneously enhanced on gadolinium-enhanced T1-weighted images. (c and d) The newly detected tumor was resected completely in the second surgery. The several small tumors (arrowhead) remained the same in size.

markedly adherent to the surrounding nerve roots. The tumor was totally resected *en bloc* with careful circumferential tumor dissection using intraoperative neurophysiological monitoring [Figures 2c, d and 3a-c].

#### Postoperative course

The hematoxylin and eosin stained sections revealed coexistent hypercellular and hypocellular areas identified as Antoni A and B, respectively, consistent with the diagnosis of a schwannoma [Figures 4a and b]. Ki-67 labeling index was 5–10% in hotspots, which was slightly higher than the normal range of schwannomas [Figure 4c]. About 20% mosaic loss of

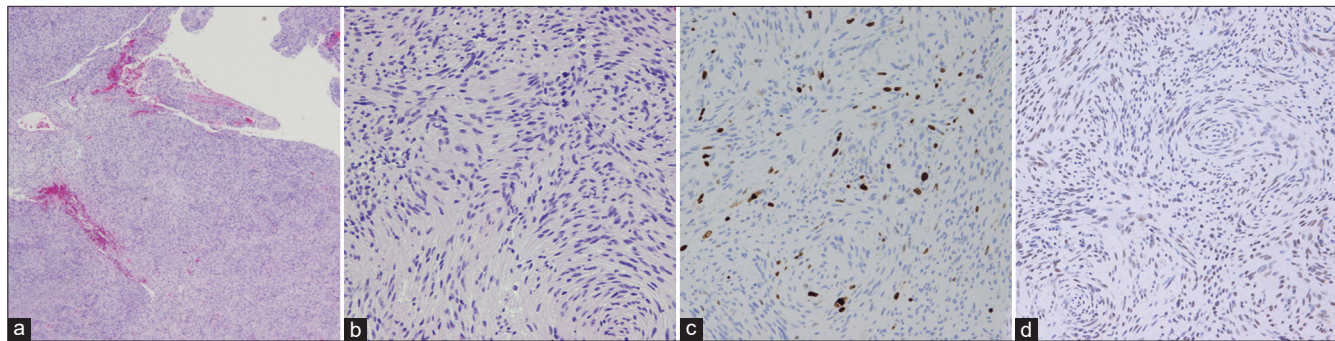


**Figure 3:** (a) The tumor adhered to the irrelevant surrounding nerve roots. (b) Careful circumferential and subcapsular dissection allowed us to isolate the tumor from the irrelevant surrounding nerve roots. The nerve of origin was identified (arrowhead). (c) The tumor was resected *en bloc* by cutting the nerve of origin (arrowhead) following negative confirmation on direct nerve stimulation.

SMARCB1 was observed [Figure 4d]. The patient's condition was diagnosed as SWN despite his declining genetic testing. He significantly recovered immediately postoperatively, and the lumbar MRI 1 week later confirmed complete tumor removal.

#### DISCUSSION

The diagnosis of SWN, as in this patient, was classically based on the identification of two or more nonintradural schwannomas, one with pathological confirmation without bilateral vestibular schwannoma on high-quality MRI obtained after the age of 30 years and no symptoms of 8<sup>th</sup> nerve deficits.<sup>[6]</sup> SWN is a rare subtype of NF that is more similar to NF2 than to NF1.<sup>[3]</sup> NF2 normally produces merlin, located at 22q12.2, which regulates multiple proliferative signaling pathways.<sup>[13]</sup> Tumorigenesis in SWN is attributed to mutations of the tumor suppressor genes, SMARCB1 or LZTR1, as well as the NF2 gene.<sup>[1,11,12]</sup> In immunohistochemical staining, mosaic loss of SMARCB1 is observed with SWN and NF2 as seen in the present case, although there is no loss with isolated schwannoma.<sup>[2]</sup>



**Figure 4:** (a and b) Histopathological analysis of the hematoxylin and eosin stained sections revealed coexistent hypercellular areas and hypocellular areas identified as Antoni A and B, respectively, which are indicative of schwannoma. (c) Ki-67 labeling index was 5–10% in hotspots, which is slightly higher than the normal range of schwannoma. (d) About 20% mosaic loss of SMARCB1 was observed. (a) Objective lens  $\times 5$  and (b-d) objective lens  $\times 20$ .

### SWN has genetic overlap with NF2

It is difficult to determine the true pathomechanisms of the spinal schwannoma in this patient. However, it is possible that it was due to the underlying diagnosis of SWN and the likely genetic overlap with NF2. Here, the newly detected schwannoma 6 years after the original L3–L5 lumbar laminectomy, although potentially a small lesion on the original preoperative MRI, potentially reflected an elevated Ki-67 labeling index.<sup>[8]</sup>

### Nerve injuries may promote development of schwannomas

Various types of nerve injuries possibly caused by laminectomy might promote the development of schwannomas.<sup>[4]</sup> Schulz *et al.* showed that an NF2 gene mutation in both neurons and Schwann cells could lead to the development of schwannomas after a single nerve crush injury.<sup>[10]</sup> Schwann cells may easily fail to redifferentiate into myelinating cells with the upregulation of proliferative signaling pathways, leading to schwannoma formation under the influence of NF2 gene mutation. Helbing *et al.* concluded clinical evidence suggests that schwannomas may preferentially appear or grow in locations that are prone to nerve injury by compression or physical trauma.<sup>[4]</sup> The concept of schwannomas arising from failure in the recovery process of nerve injury may provide clinical evidence regarding the development of spinal schwannoma in the present case. The vestibular nerves are most often affected by schwannomas intracranially, because their anatomical course is located within a confined environment that is amenable to physical stress and injury.<sup>[4,7]</sup>

### CONCLUSION

Compression or physical trauma to nerves occurring during the L3–L5 laminectomy 6 years ago likely caused the

development of an L4 vertebral level schwannoma in this patient with SWN.

### Declaration of patient consent

The authors certify that all appropriate patient consents have been obtained.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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