

Malignant peripheral nerve sheath tumors arising in *SMARCB1*-associated schwannomatosis: illustrative cases

Taha Khalilullah, BS,¹ Xinlan Yang, MS,¹ Jonathan Yao,² Abdul Karim Ghaith, MD, PhD,¹ Jawad M. Khalifeh, MD, MS,¹ Tej D. Azad, MD,¹ A. Karim Ahmed, MD,¹ Yuanxuan Xia, MD,¹ Nicholas Theodore, MD, MS,¹ Carlos G. Romo, MD,³ Jean-Paul Wolinsky, MD,^{1,4} Allan J. Belzberg, MD,¹ and Daniel Lubelski, MD¹

¹Department of Neurosurgery, Johns Hopkins University School of Medicine, Baltimore; ²Johns Hopkins University, Baltimore; ³Division of Neuro-Oncology, Department of Neurology, Johns Hopkins University School of Medicine, Baltimore, Maryland; and ⁴Department of Neurological Surgery, Feinberg School of Medicine, Northwestern University, Chicago, Illinois

BACKGROUND Schwannomatosis is a rare tumor predisposition syndrome characterized by benign peripheral nerve sheath tumors in the absence of bilateral vestibular schwannomas. In rare cases, malignant peripheral nerve sheath tumors (MPNSTs) may arise with schwannomatosis, particularly with *SMARCB1* mutations. These malignancies pose significant diagnostic and therapeutic challenges and a poor prognosis.

OBSERVATIONS The authors present 2 cases of high-grade MPNST with *SMARCB1*-associated schwannomatosis. Both presented with rapidly enlarging and concerning imaging features, including heterogeneous enhancement, central necrosis, and low apparent diffusion coefficient values. Histopathological analysis confirmed high-grade MPNST with elevated mitotic activity and loss of H3K27me3. Both patients underwent en bloc resection with negative margins. One patient remains disease free at 5 years; the other patient developed metastatic recurrence and died. A systematic review identified 20 cases in the literature.

LESSONS These cases emphasize clinical vigilance for malignant transformation in schwannomatosis, particularly with rapidly enlarging lesions or new neurological symptoms. MRI and FDG-PET can aid in identifying concerning features. Despite margin-negative resection, recurrence and metastasis may occur, reflecting the tumor's aggressive biology. Genetic counseling may be considered in *SMARCB1*-associated cases. While malignant transformation is much more common in NF1 lesions, rare cases do arise in schwannomatosis, requiring close surveillance and multidisciplinary management.

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KEYWORDS MPNST; *SMARCB1*; schwannomatosis; illustrative case

Schwannomatosis is a rare tumor predisposition syndrome within the neurofibromatosis spectrum, primarily characterized by the development of multiple benign schwannomas affecting the cranial, spinal, and peripheral nerves, often associated with germline mutations in *SMARCB1* or *LZTR1*.^{1–3} This pathology is clinically and genetically distinct from neurofibromatosis type 1 (NF1), which results from mutations in the *NF1* gene and is characterized by cutaneous neurofibromas, café-au-lait macules, and optic pathway gliomas, and from neurofibromatosis type 2 (NF2), caused by mutations in the *NF2* gene and defined by bilateral vestibular schwannomas and other intracranial tumors such as meningiomas and ependymomas.⁴ Although the tumors are typically benign, malignant peripheral nerve sheath tumors (MPNSTs) can rarely arise in this context, posing a substantial diagnostic and therapeutic challenge due to their aggressive clinical course and limited treatment responsiveness.^{5–7} The diagnosis of MPNST in

patients with schwannomatosis is complicated by the overlapping clinical and radiological features shared with benign lesions, necessitating high clinical suspicion when patients present with rapid tumor growth, escalating pain, and new neurological deficits.⁸ Advanced imaging modalities such as FDG-PET and diffusion-weighted MRI, showing features like central necrosis, heterogeneous enhancement, and low apparent diffusion coefficient (ADC) values, are critical for early identification of malignant transformation.^{9,10} Histologically, these tumors often exhibit high-grade epithelioid morphology, elevated mitotic activity, necrosis, and immunohistochemical features including loss of H3K27me3 expression and variable S100 and SOX10 positivity.^{11,12}

The standard of care of MPNST remains complete resection with negative margins, as this offers the best chance for local control and cure; however, recurrence and metastasis are still relatively common, even in cases managed with aggressive multimodal therapy, including

ABBREVIATIONS ADC = apparent diffusion coefficient; MPNST = malignant peripheral nerve sheath tumor; NF1 = neurofibromatosis type 1; NF2 = neurofibromatosis type 2.

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radiation therapy and chemotherapy.⁵ These outcomes may highlight the need for improved diagnostic tools and more effective multimodal systemic therapies to better manage high-risk MPNSTs. In this report, we present 2 illustrative cases of high-grade MPNST arising in the setting of *SMARCB1*-associated schwannomatosis. These cases were selected to represent distinct clinical outcomes: one with durable disease control and another with progressive, treatment-refractory disease, highlighting the challenges and critical considerations in the diagnosis, treatment, and surveillance of this rare pathology.

Illustrative Cases

Case 1

A 65-year-old woman with a confirmed history of familial schwannomatosis presented with a progressively enlarging, painful mass in the right posterior thigh. The patient had a prior history of multiple schwannomas involving the brain, spine, and peripheral nerves, particularly a lesion of the trigeminal nerve treated years earlier with stereotactic radiotherapy without bilateral vestibular schwannomas involved and confirmed schwannomas in the finger and thoracic spine. The clinical picture was consistent with *SMARCB1*-associated schwannomatosis. Germline molecular genetic testing identified a mutation in *INI1/SMARCB1*, altering the splicing of exon 8.

She first noticed the thigh mass several months prior to presentation and initially presumed it to be another benign lesion. However, the mass began to enlarge more rapidly and was associated with worsening neuropathic pain radiating to the gluteal region and medial shin. There were no constitutional symptoms. Examination was nonfocal with full strength and sensation in the lower extremities.

MRI demonstrated a large, centrally necrotic, peripherally and heterogeneously enhancing mass in the gracilis muscle measuring approximately $6.2 \times 6.2 \times 10.2$ cm, possibly concerning for MPNST and with surrounding stable nodular lesions along the sciatic nerve consistent with the patient's known background of schwannomatosis (Fig. 1). A core needle biopsy confirmed a grade 3 MPNST based

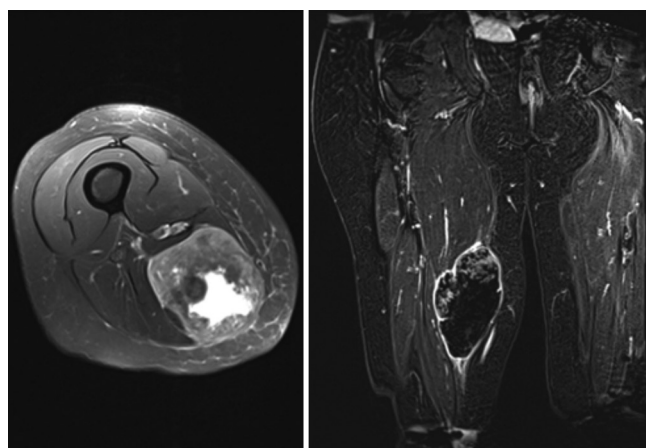


FIG. 1. Axial T2-weighted MR image (**left**) and coronal postcontrast T1-weighted VIBE image with subtraction (**right**) of the left lower extremity, demonstrating a $6.2 \times 6.2 \times 10.2$ -cm heterogeneously enhancing, centrally necrotic mass within the gracilis muscle, concerning for MPNST. Multiple smaller nodular lesions are seen along the sciatic nerve, consistent with the patient's underlying schwannomatosis.

off the 2021 WHO classification. Histopathological analysis showed sheets of high-grade epithelioid cells with abundant mitoses and necrosis. Immunohistochemical evaluation was negative for S100, AE1/AE3, and myogenin. Loss of H3K27me3 expression with retained INI1 was observed, consistent with MPNST arising in the setting of *SMARCB1*-related schwannomatosis.

At an external institution, the patient initiated neoadjuvant chemotherapy (2 cycles of doxorubicin + Ifex + Mesnex, followed by 2 cycles of ifosfamide + etoposide), but treatment was complicated by early-onset neurotoxicity, ultimately limiting further systemic therapy. She then completed neoadjuvant intensity-modulated radiation therapy (50 Gy over 25 fractions) to the right thigh local site.

At our institution, the patient underwent en bloc resection of the tumor with negative margins. Intraoperatively, the tumor was dissected off the adductor compartment with preservation of major vasculature. Tumor-involved branches of the saphenous nerve were resected. Her postoperative course was uncomplicated, and she reported complete resolution of preoperative pain, with preserved strength and expected numbness in the medial knee distribution consistent with the saphenous nerve.

She remained free of local recurrence over a 5-year surveillance period. However, 2 years after her initial surgery, a 1.7-cm pulmonary lesion was detected and resected via segmentectomy. Pathological review determined this to be a metastatic schwannoma-associated sarcoma, and she underwent adjuvant radiation therapy (60 Gy over 8 fractions) to the right lung. The patient is medically stable and without significant neurological deficits at the 5-year follow-up.

Case 2

A 26-year-old man with a long-standing diagnosis of *SMARCB1*-associated schwannomatosis presented with new left-sided back and leg pain. His history included more than 25 tumor resections and functional quadriplegia diagnosed in adolescence. He had been maintained on bevacizumab for symptom and disease control. Surveillance MRI and FDG-PET revealed substantial growth of a left-sided L2–3 paraspinal/psoas mass measuring 7.6×4.3 cm with concerning features: central necrosis, heterogeneous enhancement, erosion of the L2 vertebral body, invasion into the left L2–3 neural foramen, arterial-phase enhancement, and a low ADC value (0.4), decreased from 1.4 in imaging 6 months prior (Fig. 2). Biopsy of the left psoas

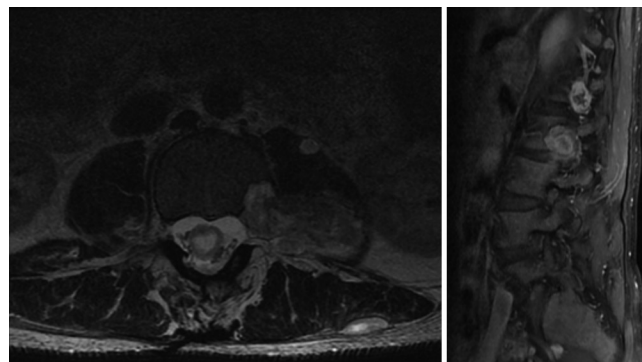


FIG. 2. Axial T2-weighted (**left**) and sagittal postcontrast fat-suppressed T1-weighted (**right**) MR images of the lumbar spine, demonstrating a heterogeneously enhancing left-sided paraspinal mass centered at L2–3 with central necrosis, invasion of the left L2–3 neural foramen, and osseous erosion of the L2 vertebral body, concerning for MPNST.

muscle confirmed high-grade malignant transformation in the setting of schwannomatosis with epithelioid morphology, S100 and AE1/AE3 positivity, and a Ki-67 index of approximately 30%, consistent with MPNST on immunohistochemical analysis. Whether H3K27me3 was lost or retained was not ascertained by pathology. The molecular genetic analysis identified a mutation in the *SMARCB1* gene with deletion in exons 8–9 following germline testing.

At our institution, the patient underwent a two-stage en bloc resection. Stage I involved a posterior approach with T11–L3 laminectomy, left-sided facetectomies, rhizotomies of L1–3 nerve roots, posterior fusion and instrumentation, and partial L2 vertebrectomy. Stage II, via a retroperitoneal approach, included coronal and axial osteotomies through L1–3; resection of L1–3 vertebral bodies; en bloc resection of involved nerve roots, the iliopsoas muscle, and the lumbar plexus; and anterior column reconstruction with structural allograft and instrumentation. Negative margins were achieved. There was no prior local radiation therapy.

Surgical biopsy of the tumor of the L2 vertebral body revealed a 7.0-cm high-grade MPNST with epithelioid features, necrosis, and bony erosion. Postoperative imaging showed no evidence of recurrence. There was a presence of preexisting residual schwannomas, including multiple stable intradural masses resulting in mass effect on the distal spinal cord, with no evidence of malignant transformation or interval progression. The patient resumed bevacizumab following wound healing complications. Bilateral dorsiflexion and left eversion were 1/5 in strength, and dysesthesia was noted along the left flank and thigh. No other complications or neurological deficits were noted.

Nine months postoperatively, follow-up imaging demonstrated interval growth of a new enhancing paraspinal mass at L2 and an enlarging left suprarenal lesion. Biopsies of both lesions confirmed recurrent MPNST. The patient received image-guided intensity-modulated radiation therapy to the L2 paraspinal and suprarenal lesions, alongside temozolomide. Toward the end of radiation administration, CT imaging revealed a malignant pleural effusion and multiple bilateral pleura-based masses, the largest measuring 7.8 × 5.0 cm. Thoracentesis confirmed metastatic sarcoma. Despite further therapy, the disease progressed, and the patient died several months later.

Informed Consent

The necessary informed consent was obtained in this study.

Discussion

Schwannomatosis is a primarily benign tumor predisposition of neurofibromatosis characterized by the development of multiple intracranial, spinal, and peripheral schwannomas in the absence of vestibular schwannomas, representative of NF2.¹³ MPNSTs in the context of schwannomatosis are exceedingly rare but aggressive neoplasms with limited reported associations with schwannomatosis, posing significant diagnostic, therapeutic, and prognostic challenges (Table 1).¹⁴ In this case series of 2 patients, we provide insights into recognizing malignant transformation, optimizing surgical strategies, managing long-term outcomes, and understanding genetic implications associated with schwannomatosis, particularly involving *SMARCB1* mutations.

Observations

The rarity of MPNST associated with schwannomatosis necessitates heightened clinical vigilance, especially given the early clinical

TABLE 1. Summary of published cases of MPNSTs in the setting of *SMARCB1*-related schwannomatosis

Authors & Year	No. of Patients	Mean Age at Diagnosis (yrs)	Sex		Tumor Location (n)	Survival Status	Adjuvant Radiotherapy or Chemotherapy	<i>SMARCB1</i> Gene Mutation
			Male	Female				
Ding et al., 2018 ²¹	1	30	0	1	Cervical, thoracic, thoracolumbar, & lumbar spine	Dead*	—	Yes
Evans et al., 2012 ⁶	3	29.7	2	1	—	—	—	Yes
Carler et al., 2012 ¹⁶	1	31	0	1	Lt common peroneal nerve, rt ulnar nerve	Alive	Radiotherapy	Yes
Eeloo et al., 2019 ¹⁴	1	51	0	1	Retroperitoneum abutting the lt kidney	Alive	None	Yes
Nakayama et al., 2013 ²⁷	1	18	0	1	Rt Meckel's cave, intracranial portion of lt oculomotor nerve	Alive	None	Yes
Schaefer et al., 2019 ²⁸	13	42.8	7	6	Abdominal wall, axilla, chest wall (3), back, thigh, arm (2), knee (2), pleura, urinary bladder, vertebral column	—	—	Yes

* Died 1 month after disease onset.

manifestations and aggressive disease course observed in our cases. Both patients demonstrated rapid lesion growth and neurological deterioration, which are critical clinical indicators for malignancy. This observation aligns with existing literature emphasizing rapid tumor enlargement, increased pain, and neurological deficits as hallmark signs prompting aggressive diagnostic evaluations.^{6,14} Additionally, the rarity of the condition may be due in part to the lack of specificity in identification and standardized diagnostic criteria for MPNSTs with schwannomatosis.^{13,15,16}

Radiological features indicative of malignant transformation were observed, including heterogeneous enhancement, central necrosis, high metabolic activity on FDG-PET, and notably low ADC values on diffusion-weighted imaging, reflecting high cellularity and aggressive biology.^{17,18} Additionally, the tumor presents in a dumbbell shape with hypo- to moderate intensity on T1-weighted MRI and hyperintensity on T2-weighted and STIR MRI.¹⁸ Such imaging features are crucial for healthcare practitioners to promptly identify high-risk lesions warranting biopsy and expedited surgical and multimodal therapeutic intervention. Furthermore, the criteria for schwannomatosis outlined by Plotkin et al. following the 2011 International Schwannomatosis Workshop included the presence of two or more pathologically confirmed schwannomas, absence of vestibular schwannomas on high-quality MRI, and no constitutional NF2 mutation, with genetic testing for *SMARCB1* and *LZTR1* mutations supporting diagnosis.¹⁷

Histologically, case 2 showed epithelioid features, high Ki-67 proliferation indices, and extensive necrosis, consistent with high-grade MPNST. Immunohistochemical evaluation demonstrated characteristic findings including positivity for S100 and AE1/AE3 in case 2, which may reinforce the aggressive and malignant nature of these tumors, aligning with established molecular and clinical diagnostic criteria for epithelioid MPNSTs.^{15,18–20}

From a surgical perspective, our cases underscore the importance of aggressive, en bloc resections with negative surgical margins, which currently represents the cornerstone of therapy for symptomatic patients and for compressive spinal cord schwannomas. However, in the context of MPNSTs, even successful negative-margin resection may be insufficient, as evidenced by recurrence and metastasis in both cases. Such findings highlight the biological aggressiveness of MPNSTs and the limitations of current treatment paradigms. In the second case, disease progression necessitated multimodal therapies, including targeted radiation therapy and chemotherapy, which achieved transient disease control but ultimately inadequate efficacy in preventing disease progression. While adjuvant radiotherapy and chemotherapy are traditionally reserved for lesions with malignant behavior or those not amenable to complete resection due to anatomical constraints, their limited effectiveness in our case reflects the need for improved systemic therapies.¹⁸ Similar outcomes have been reflected in studies by Ding et al. and Hasegawa et al. that documented rapid disease progression and mortality within 1 month of diagnosis.^{21,22} These findings demonstrate the urgency for rapid diagnosis and the development of more effective treatment options.

Genetic implications associated with schwannomatosis, particularly *SMARCB1* and *LZTR1* mutations, underline the heterogeneous, aggressive, and malignant potential observed in our series.²³ *SMARCB1*-associated schwannomatosis has been increasingly recognized for its potential for malignant transformation, necessitating comprehensive genetic counseling and regular surveillance for affected patients and their families.^{14,17,24} Studies such as the one by Paganini et al. demonstrated a notable relation between *SMARCB1* mutation and oncogenic pathogenesis in schwannomas and other

sarcomas.^{6,25–28} Such findings further emphasize the critical role of genetic predisposition in malignant transformation and support the integration of genomic testing into routine evaluation for patients with MPNST with schwannomatosis.

Lessons

Clinicians must maintain a high index of suspicion for malignant transformation in patients with schwannomatosis, particularly when tumors enlarge rapidly, neurological symptoms worsen, or imaging demonstrates concerning features. Comprehensive evaluation with MRI and FDG-PET is essential for early recognition of aggressive characteristics, facilitating timely biopsy and surgical intervention. When feasible, surgical management should prioritize en bloc resection with negative margins to maximize local control, although the aggressive biology of MPNST often necessitates vigilant postoperative surveillance and multimodal adjuvant therapy. Germline testing and genetic counseling are advisable, especially in patients with multiple schwannomas or a family history suggestive of a hereditary predisposition, given the potential association with *SMARCB1* mutations and the rare but important risk of malignant transformation. Because of the poor prognosis and aggressive course of MPNST in schwannomatosis, further multicenter research is urgently required to define effective systemic therapies and explore personalized approaches. Ultimately, this series highlights the importance of heightened clinical vigilance, multidisciplinary coordination, and continued research to improve outcomes for patients with MPNST arising in the context of schwannomatosis.

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Author Contributions

Conception and design: Lubelski, Khalilullah, Ghaith, Khalifeh, Belzberg. Acquisition of data: Khalilullah, Yao, Ghaith, Wolinsky. Analysis and interpretation of data: Lubelski, Khalilullah, Yang, Ghaith, Khalifeh, Azad, Romo, Wolinsky, Belzberg. Drafting the article: Khalilullah, Yang, Ghaith, Khalifeh, Azad, Romo. Critically revising the article: Lubelski, Khalilullah, Yang, Ghaith, Khalifeh, Azad, Ahmed, Xia, Theodore, Romo, Wolinsky, Belzberg. Reviewed submitted version of manuscript: Lubelski, Khalilullah, Yang, Ghaith, Ahmed, Xia, Theodore, Wolinsky. Approved the final version of the manuscript on behalf of all authors: Lubelski. Statistical analysis: Khalilullah, Ghaith. Administrative/technical/material support: Khalilullah, Ghaith, Theodore. Study supervision: Khalilullah, Ghaith, Theodore.

Correspondence

Daniel Lubelski: Johns Hopkins University School of Medicine, Baltimore, MD. dlubelski@jhmi.edu.