

4. Conclusion

Patients with moyamoya disease who present with aneurysmal intracerebral hemorrhage should be treated to prevent rebleeding complications. Endovascular embolization using Onyx can be an effective treatment for aneurysms associated with moyamoya disease that would otherwise be difficult to treat surgically.

Conflicts of Interest/Disclosures

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

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Malignant peripheral nerve sheath tumor arising in the setting of cervical nerve root schwannomas



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ABSTRACT

We present a 23-year-old woman who was diagnosed with a malignant peripheral nerve sheath tumor (MPNST), 17 months following the resection of a schwannoma. MPNST is rare and is usually associated with neurofibromatosis. The typical treatment of resection and radiation is difficult to achieve in the spine.

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1. Introduction

Malignant peripheral nerve sheath tumors (MPNST) are commonly associated with neurofibromatosis 1 (NF-1), seen in up to 5% of patients [1,2]. MPNST is extremely rare in the general population with an incidence of 0.001% [1,2]. Almost exclusively, neurofibroma acts as the source of malignant transformation in NF-1. Transformation of schwannomas into MPNST is exceedingly rare [1,3]. We present a transformation of a schwannoma to a low grade MPNST.

2. Case report

A 23-year-old woman, with no significant past medical history, presented with 4–6 months of intermittent neck pain and

numbness. She denied that she had weakness or bowel or bladder dysfunction. On examination, she had 4/5 strength on right and left dorsiflexion and plantarflexion, as well as decreased sensation below T1–T2 on the left and T5–T6 on the right. She had 4+ patellar reflexes bilaterally with positive clonus. The remainder of her neurologic examination was unremarkable. An MRI showed a spinal cord lesion at the level of C7–T1, suspicious for schwannoma (Fig. 1). She underwent a C7–T1 laminectomy and gross total resection, with care taken to preserve the right C8 nerve root. The histology at this time established the diagnosis of schwannoma (Fig. 2).

Due to financial constraints, the patient missed her planned follow-up and returned approximately 17 months later to the emergency department with acute upper back pain radiating to her right upper extremity and involving the posterior neck accompanied by decreased sensation of the right upper extremity. On examination, she had 4/5 strength in her bilateral upper extremities, 3+ patellar reflexes, and gait instability. An MRI of the cervical spine showed recurrence at the C7–T1 level with significant cord

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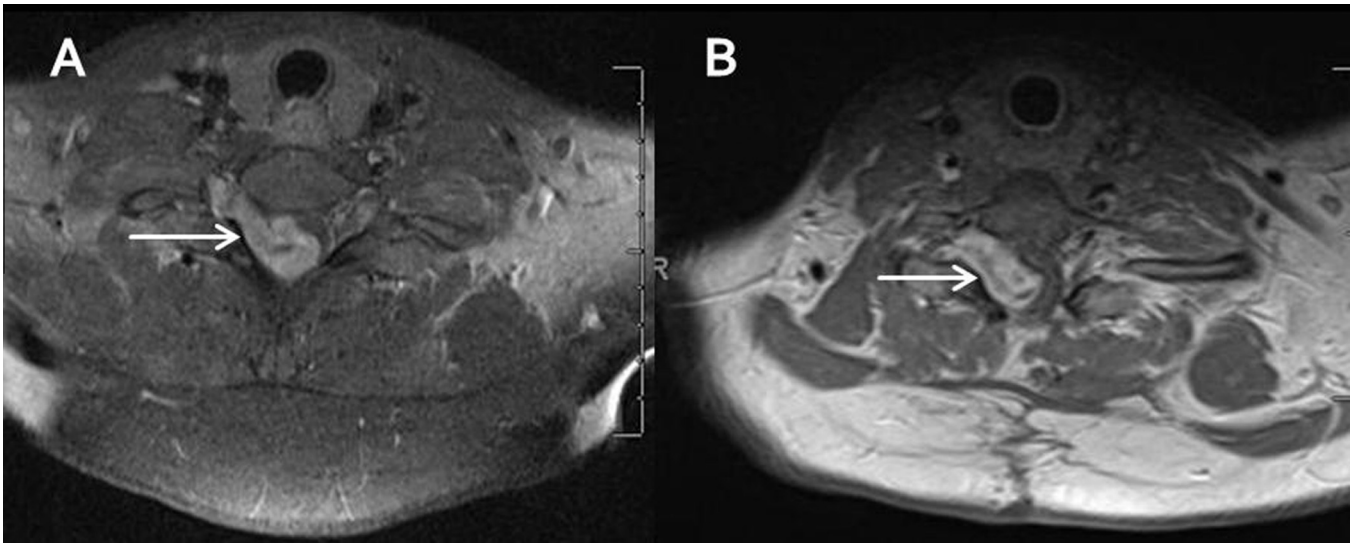


Fig. 1. (A) Axial T1-weighted postcontrast MRI (July 2012) showing an extramedullary intradural mass at C7–T1 (arrow) extending through the neural foramina. (B) Axial T1-weighted postcontrast MRI (February 2014) showing the recurrent heterogeneously enhancing extramedullary intradural lesion expanding through the C7–T1 neural foramen.

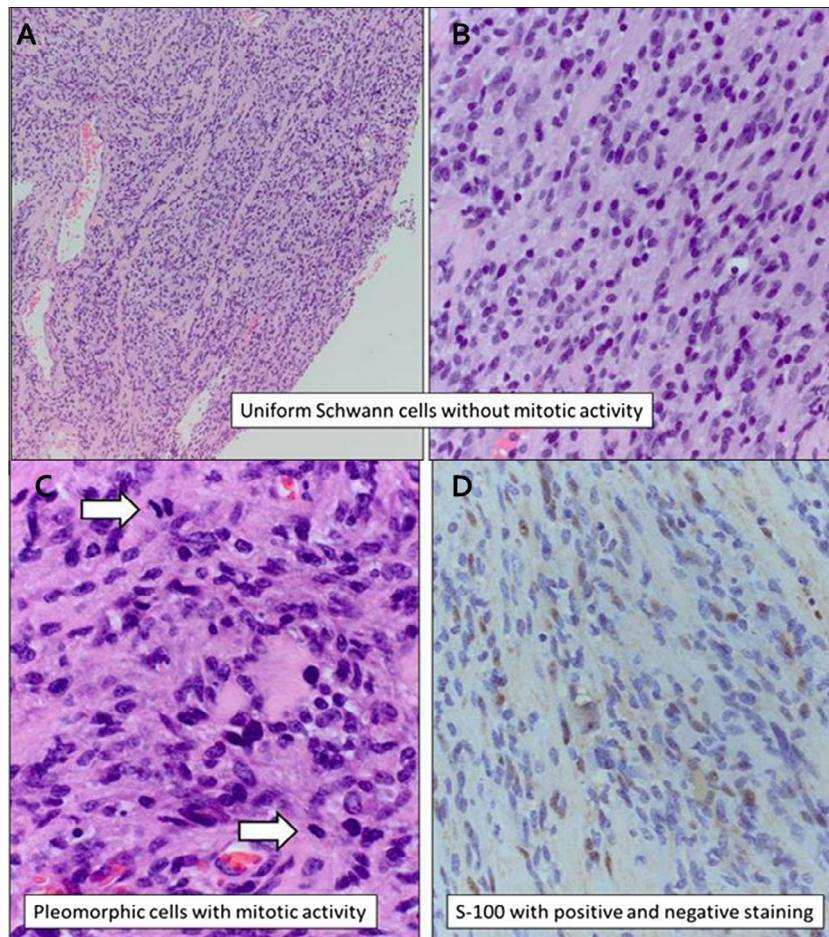


Fig. 2. First resection: (A) Schwann cell proliferation seen at low power magnification (hematoxylin and eosin [H&E]; 10 × magnification); (B) high power magnification (H&E; 20 × magnification). Second resection: (C) Low grade malignant peripheral nerve sheath tumor showing mitotic activity and pleomorphic cells (arrow; H&E; 40 × magnification); (D) S-100 positivity is variably lost (20 × magnification). This figure is available in colour at www.sciencedirect.com.

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