Intramedullary spindle cell hemangioma: case report

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The authors describe the case of a 48-year-old man found to have the first reported intramedullary spinal cord spindle cell hemangioma. Previous research indicates that spindle cell hemangiomas are rarely found in the spine. Only 3 previous cases exist, all in the intradural, extramedullary space. In the present case, gross-total resection of the tumor was possible with no loss of function from baseline. This report presents the successful resection of the first reported intramedullary spindle cell hemangioma and reports 4-month follow-up, demonstrating the biological behavior of this rare tumor.

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Spinle cell hemangioendothelioma, alternately referred to as spindle cell hemangioma, is a vascular tumor mainly affecting young adults. Initially described by Weiss and Enzinger in 1986,12 these tumors typically appear in the distal extremities.10 In this report we describe the case of a 48-year-old man who presented with signs of myelopathy and was found to have an intramedullary spinal cord spindle cell hemangioma, the first such tumor reported. This case report brings attention to a rare diagnosis for an intramedullary mass and discusses the biological behavior of the spinal spindle cell hemangioma.

Case Report
History and Presentation
A 48-year-old man presented with gait ataxia, bilateral lower extremity numbness, and subacute urinary retention. He did not have a history of sarcoma or any prior malignancy. A neurological examination revealed diminished strength in the left iliopsoas muscle (4/5 on the Medical Research Council scale).5 A sensory level at approximately the level of T-10 was apparent. The patient exhibited clinical findings consistent with American Spinal Injury Association (ASIA) Grade D.1 In addition, deep tendon reflexes were subtly enhanced symmetrically in the legs, and a Babinski sign was present on the left.

MRI of the thoracic spine with intravenous Gd demonstrated an enhancing intramedullary lesion that measured approximately 0.7 × 0.6 × 1.7 cm eccentric to the left side of the cord at the T6–8 level (Fig. 1). Moreover, extensive T2 cord signal abnormalities extended superiorly and inferiorly from the enhancing lesion. At the time of diagnosis, the initial concern was that the patient had an intramedullary astrocytoma or hemangioblastoma.

Operation
Following a T6–8 laminectomy and midline dural opening, a red-purple lesion was apparent on the spinal cord surface and was concerning for a possible vascular lesion (Fig. 2). A cleavage plane was identified between the tumor and the spinal cord permitting a gross-total resection. Intraoperative bleeding was readily controlled. Small motor-evoked potentials (MEPs) were present throughout in the right abductor hallucis muscle, but there were no consistent MEPs in any of the other monitored leg muscles. Posterior tibial nerve somatosensory evoked potentials were also not adequate for intraoperative monitoring. Therefore, an epidural electrode was placed caudal to the tumor for recording of D-waves during tumor resection. These were less well-formed following right hemisphere stimulation, but aside from one change related to repositioning of the recording electrode, they remained stable during tumor resection (Fig. 3).

Postoperatively, the patient experienced mild lower ex-
tremity weakness, but was ambulatory (ASIA Grade D). However, the patient recovered to his neurological baseline within several days postoperatively, during a 7-day taper off steroids. An immediate postoperative MR image was obtained and revealed no evidence of residual tumor (Fig. 4).

Histological Analysis

Upon microscopic inspection, the lesion had endothelial-lined channels of variable size. Histological analysis was positive for both CD31 and CD34. Moreover, many cells expressed vimentin (VIM). Furthermore, smooth muscle actin (SMA)-stained cells adjacent to the endothelial cells were consistent with pericytes and smooth muscle cells of the vascular wall. The final pathological analysis was consistent with spindle cell hemangioma.

Postoperative Course

At the 2-month follow-up evaluation, the patient continued to improve clinically with resolution of his urinary retention and marked improvement in gait. The patient at this time was found to have only very mild weakness in left foot dorsiflexion. At the 4-month follow-up, the patient had full strength in all extremities and had complete resolution of his numbness. The patient exhibited improvement from ASIA Grade D to ASIA Grade E. Postoperative MRI at the 4-month interval showed no recurrence of the tumor (Fig. 5). Unfortunately, the patient was lost to follow-up after the 4-month office visit.

Discussion

Spindle cell hemangiomas are vascular epithelioid
Intramedullary spindle cell hemangioma

These tumors are rarely seen in the spine, with only 3 cases previously reported, all in the intradural, extramedullary space. These tumors are genetically associated with syndromes such as Ollier disease, Klippel-Trenaunay syndrome, and Maffucci syndrome. The patient presented did not have a genetic predisposition toward developing such a lesion. Histologically, spindle cell hemangiomas demonstrate many dilated vascular channels with interspersed spindle cells. Immunohistochemical analysis is significant for positive staining for VIM, SMA, and CD34.

Although the natural history of this tumor is not fully understood, spindle cell hemangiomas are regarded as indolent in nature. In the present case, there is no evidence of recurrence. Ding et al. described these tumors as arising from abnormal vasculature, while others implicate variations in blood flow creating alternating areas of expansion and collapse. Although the lesion is of vascular etiology and preoperative embolization has been described for spindle cell hemangiomas in the vertebral column, our experience in this case was that intraoperative bleeding was readily controlled and preoperative embolization would have been unnecessary.

Gross-total resection remains the preferred treatment modality. Postoperatively, there is a 60% recurrence rate according to Fukunaga et al. Ideally, patients with these tumors should be followed closely because of this high recurrence rate and the unknown tumor biology they may exhibit. Unfortunately in the present case, the patient was lost to follow-up after 4 months. Radiosurgery has been used by some authors, especially for postoperative tumor recurrence. However, this would not likely be appropriate for an intramedullary lesion. Mahdavi et al. described the use of alpha interferon therapy in instances of recurrence.

Spindle cell hemangiomas are vascular epithelioid tumors that occur very rarely in the spine. In this paper we report the first instance of an intramedullary spindle cell hemangioma in the thoracic spinal cord. Intraoperative...
appearance was similar to hemangioblastoma, and gross-total resection was possible. Follow-up showed excellent recovery of neurological function with no progression of the tumor at the 4-month mark.

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References

Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
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FIG. 5. Four-month postoperative sagittal (upper) and axial (lower) MR images showing no evidence of recurrent tumor.