

CLINICAL VIGNETTE

A Unique Presentation of a Spinal Cord Tumor

Adam Cavallero, M.D., and Luke Macyszyn, M.D.

Case Presentation

A 36-year-old Caucasian male presented to his primary care physician's office complaining of progressively worsening paresthesias and a sensation of "coldness" in his torso and bilateral feet. He also voiced intermittent numbness from the waist down, excessive diaphoresis of the scalp/anterior thorax/bilateral arms, as well as an unsteady gait. These symptoms were occurring in the setting of chronic upper back pain. The patient did not experience any fevers, chills, weight loss, or other symptoms. He also denied any associated saddle anesthesia, bowel, or urinary incontinence. Over the preceding year, he was managed by an orthopedic surgeon and given the diagnosis of "postural kyphosis." This was treated with conservatively with pain medications and NSAIDS.

Vital signs were unremarkable. Physical exam was notable for grossly intact cranial nerves 2-12. Motor strength was normal in the bilateral upper extremities and slightly (4+/5) diminished throughout the bilateral lower extremities. Sensation to pinprick and light touch was diminished in the bilateral lower extremities. The hypoesthesia was especially pronounced from the level of the gastrocnemius to the foot. Deep tendon reflexes were normal and there was no ankle clonus. The patient's fine motor control and coordination was intact; however, his gait was slow, wide based, and unsteady.

A neurological condition was suspected and confirmed with an MRI of the thoracic spine showing a heterogeneous enhancing intramedullary T3-T5 spinal cord lesion with dimensions of 4.3 cm cranio-caudal and 1.1 cm in diameter. There was associated extensive edema involving the cervical cord through the T8 level. These radiological findings were most consistent with a neoplasm, and the differential diagnoses included hemangioblastoma, astrocytoma, or ependymoma. A subsequent MRI of his entire neural axis did not demonstrate any other lesions or metastasis. The patient was promptly evaluated by neurosurgery, and surgery was recommended for definite diagnoses and resection. He underwent a T3-T5 laminectomy with gross total resection of the intramedullary tumor. Pathology was consistent with an ependymoma, WHO grade II. Repeat surveillance MRIs were performed at 3 and 5 months postoperatively demonstrating resolution of the cord edema with no new or residual enhancement surrounding the resection cavity.

The patient's lower extremity sensation and proprioception was affected postoperatively, which initially limited his ability to ambulate. However with intense physical and occupational therapy, the patient progressed from using a wheelchair to a walker, and now only requires a cane to ambulate safely. He has no residual upper back pain, but he does experience mild neuropathic pain affecting his lower extremities. These symptoms are managed well with daily gabapentin and baclofen.

Discussion

Spinal cord tumors are rare oncologic diseases that represent less than 2% of central system tumors. The annual incidence of spinal ependymomas in adults is approximately 600 cases per year according to the Collaborative Ependymoma Research Network.¹ The large majority of ependymomas occur in the brain, but this disease makes up a quarter of all spinal cord malignancies, second only to meningiomas. Over half of these tumors are of the classic subtype, which are classified by the WHO as grade two.

Thus, ependymomas are not frequently at the top of the differential diagnosis when patients present with chronic back pain. In our case, the patient was an otherwise healthy male with no neurological symptoms on first presentation. Hence, it was appropriate to evaluate the patient without obtaining an MRI and institute a course of conservative therapy. It is only after the patient developed rather unique and vague symptoms of "coldness" and excessive diaphoresis in his torso that a neurological process was suspected. Eventually, the patient evolved more classic symptoms of a spinal cord tumor, including back pain, intermittent numbness, and an unsteady gait.

The gold standard diagnostic method for a patient with a suspected spinal cord lesion is MRI, with and without contrast. Spinal ependymomas tend to be hypointense on T1 sequences and hyperintense in T2. These tumors typically have avid but heterogeneous enhancement and frequently have associated cysts at the poles of the lesion. If the lesion is centered around the conus, this raises the possibility of a myxopapillary ependymoma and complete cranio-spinal imaging should be obtained to evaluate for dissemination.

The treatment of choice for spinal ependymomas is gross total surgical resection.² The feasibility of resection is dictated by

the size of the lesion, location, and the plane surrounding the tumor. For small- to moderate-sized lesions that have an identifiable capsule, gross total resection is much more likely than for large or infiltrating tumors. Any residual disease is treated with adjuvant radiotherapy, while the role for chemotherapy is quite limited at this time.

Finally, a patient's neurological outcome following surgery is dictated by the location of the tumor and pre-operative deficits. In other words, patients that present with minimal symptoms are at lower risk for post-operative neurological deficits. Nonetheless, due to the surgical approach, a midline myelotomy where the posterior columns are split, patients frequently develop sensory and proprioceptive deficits after surgery due to dorsal column dysfunction.³ These deficits can range from mild to debilitating and patients frequently require extensive post-operative rehabilitation to maximize their functional outcome.

Figures

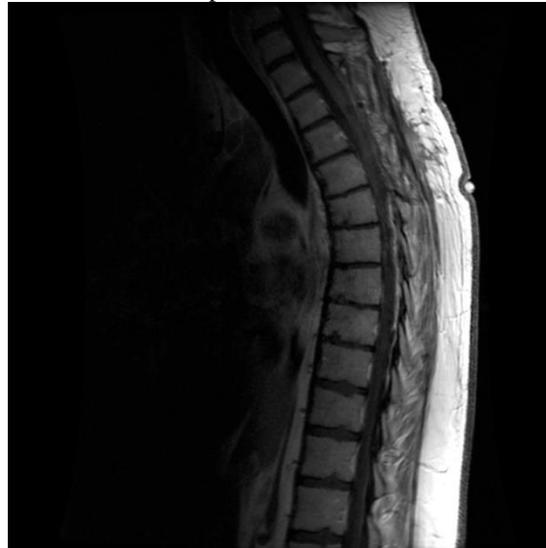
Figure 1. Pre-operative sagittal T2 MRI demonstrating T3-5 intramedullary lesion with cord expansion and significant edema extending cranially and caudally.



Figure 2. Pre-operative sagittal T1 MRI with gadolinium demonstrating heterogeneous enhancement of the thoracic lesion.



Figure 3. Post-operative sagittal T1 MRI with gadolinium obtained 9 months after surgery. There is no residual enhancement and spinal cord edema has resolved.



References

1. **Ostrom QT, Gittleman H, Farah P, Ondracek A, Chen Y, Wolinsky Y, Stroup NE, Kruchko C, Barnholtz-Sloan JS.** CBTRUS statistical report: Primary brain and central nervous system tumors diagnosed in the United States in 2006-2010. *Neuro Oncol.* 2013 Nov;15 Suppl 2:ii1-56. doi: 10.1093/neuonc/not151. Erratum in: *Neuro Oncol.* 2014 May;16(5):760. PubMed PMID: 24137015; PubMed Central PMCID:PMC3798196.
2. **Klekamp J.** Spinal ependymomas. Part 1: Intramedullary ependymomas. *Neurosurg Focus.* 2015 Aug;39(2):E6. doi: 10.3171/2015.5.FOCUS15161. PubMed PMID: 26235023.
3. **Nagasawa DT, Smith ZA, Cremer N, Fong C, Lu DC, Yang I.** Complications associated with the treatment for spinal ependymomas. *Neurosurg Focus.* 2011 Oct;31(4):E13. doi: 10.3171/2011.7.FOCUS11158. Review. PubMed PMID: 21961857.

Submitted May 18, 2016