Long segment spinal cord involvement as the clinical manifestation of sarcoidosis

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Practical Implications

Consider sarcoidosis in the differential diagnosis of long segment spinal cord lesions.

A 34-year-old man presented with ascending sensory changes in his legs. He reported a pins and needles sensation that started in his feet and ascended to his knees bilaterally. The patient presented to an outside hospital, where MRI scans revealed a T2 hyperintense lesion extending from C3 to C7 levels in the cervical spine. There was patchy gadolinium enhancement. MRI of the brain was unremarkable. His symptoms were thought to be secondary to transverse myelitis; he was treated with IV methylprednisolone, 1 g daily for 3 days, followed by a 5-day oral prednisone taper. The patient's symptoms resolved completely.

Two weeks later, he again noticed a change of sensation in his feet. This pins and needles feeling ascended his legs and by 2 weeks later he was unable to feel his anus and genitals. He presented to our institution for evaluation. Neurologic examination was unremarkable except for altered sensation to light touch below the waist bilaterally. MRI of the cervical and thoracic spine showed a T2 hyperintense lesion from C2 to T2 with patchy gadolinium enhancement (figure A–C). Brain MRI was unremarkable. CSF analysis showed leukocytes 2 cells/mm³ (100% lymphocytes), erythrocytes 2 cells/mm³, protein 76 mg/dL, glucose 65 mg/dL, negative bacterial, viral, and fungal cultures, and negative cytology, oligoclonal bands, and CSF neuromyelitis optica (NMO) immunoglobulin G (IgG). Serum tests were unremarkable including negative NMO IgG, HIV, rapid plasma reagin, antinuclear antibodies, angiotensin-converting enzyme, antineutrophil cytoplasmic antibodies, and serum protein electrophoresis; B₁₂ and erythrocyte sedimentation rate were normal. Admission chest roentgenogram showed hilar lymphadenopathy. CT scans of the chest, abdomen, and pelvis revealed nodules in the upper lobe of his right lung as well as bilateral hilar and mediastinal lymphadenopathy. Bronchoscopy with tissue biopsy demonstrated noncaseating granulomas consistent with sarcoidosis. He was treated with 5 days of IV methylprednisolone with lessening of his symptoms.

The patient was discharged on oral prednisone 60 mg daily and azathioprine 150 mg daily. Repeat spine MRIs were performed every 3 months for the first year; these showed a progressive decrease in signal abnormality from C5 to C7, with subjective mild focal volume loss in this area and minimal patchy gadolinium enhancement. No new lesions appeared during this time.

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Figure MRI of the cervical and thoracic spine with a long segment spinal cord lesion with patchy gadolinium enhancement as a manifestation of sarcoidosis

(A) Sagittal T2 fluid-attenuated inversion recovery image demonstrates abnormal intramedullary signal from C2 through T2 with cord expansion. (B) Patchy enhancement posteriorly from approximately C5 to C6 on a T1 postgadolinium sagittal image. (C) Triangular posterior enhancement pattern on T1 postgadolinium axial image (arrow).

The patient remained on azathioprine and a slow prednisone taper was started after 3 months. He was asymptomatic except for residual intermittent tingling below the knees.

DISCUSSION

About 5%–15% of patients with sarcoidosis have CNS involvement and 6%–8% of these patients have spinal cord lesions.¹ Patients with spinal cord sarcoidosis often have severe morbidity and frequently require aggressive immune-modulating therapy.² Sarcoidosis manifesting as a long segment spinal cord lesion is rare and may mimic multiple other disease processes including inflammatory demyelinating disease such as NMO, spinal cord infarction or compression, neoplasms including lymphoma, or autoimmune disorders such as Sjögren syndrome or lupus.^{3–5} Longitudinal myelitis puts patients at high risk for severe neurologic sequelae; therefore, prompt evaluation and treatment is essential to avoid unfavorable outcomes.

Clinical features may be used to help differentiate various causes of long segment spinal cord lesions, including time course, specific spinal cord syndromes, and associated neurologic or constitutional symptoms.⁶ MRI characteristics may also aid in diagnosis as certain patterns of signal abnormality may predict particular etiologies. Sarcoidosis of the spinal cord may appear as smooth or nodular leptomeningeal enhancement with patchy peripheral cord enhancement. Fusiform cord enlargement is typically replaced by atrophy in later stages.¹ CSF analysis is also essential to the evaluation. An elevated protein is the most common CSF finding in neurosarcoidosis, but patients may also have a pleocytosis with lymphocytic predominance.¹ ACE level is frequently obtained but is relatively nonspecific and often normal.⁷ Serologic testing should be performed, including inflammatory and autoimmune panels as well as NMO antibodies. In our patient, chest roentgenogram was key to the diagnosis. One may argue that this should be part of a standard evaluation for patients who present with long segment spinal cord lesions. Ultimately, tissue biopsy revealing noncaseating granulomas is the gold standard of a diagnosis of sarcoidosis¹; this established the diagnosis in our patient.

The etiology of long segment spinal cord lesions is frequently uncertain at presentation; therefore, empiric treatment with high-dose IV corticosteroids should be initiated while evaluation is underway.⁶ It was believed that more aggressive therapy was necessary in our case given the severity of the spinal cord lesion and recurrence of symptoms. Thus, azathioprine was also initiated. Our case highlights one possible cause of a long segment spinal cord lesion and emphasizes the importance of prompt and thorough evaluation of patients with this neuroimaging finding.

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Practical Implications

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