Spinal Neurosarcoidosis Mimicking an Idiopathic Inflammatory Demyelinating Syndrome

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Background: Intramedullary neurosarcoidosis may be the first and only manifestation of the disease and may mimic an idiopathic inflammatory demyelinating syndrome both clinically and on neuroimaging results.

Methods and Results: Two patients who were seen initially with a relapsing-remitting neurologic course and a cervical intramedullary lesion on magnetic resonance imaging findings are reported. Both proved to have neurosarcoidosis. A computed axial tomographic scan of the chest showed hilar adenopathy, which provided a clue to the diagnosis.

Conclusions: Symptoms due to an intramedullary cervical lesion can be the first manifestation of neurosarcoidosis. The clinical course can mimic a demyelinating illness. A high index of suspicion and a search for sarcoidosis at extraneural sites are required for an early diagnosis. Steroid treatment is associated with a favorable outcome.

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Clinical involvement of the central and/or peripheral nervous system occurs in 5% of patients with sarcoidosis.1 On autopsy, about 14% of patients with sarcoidosis have central nervous system involvement.2 Approximately 50% of patients with neurosarcoidosis initially have neurologic manifestations. In the remainder of patients, the neurologic manifestations appear within 2 years of the diagnosis of sarcoidosis.3 The most common sites of involvement in sarcoidosis are the lungs and pulmonary lymph nodes. Neurosarcoidosis is most commonly associated with granulomatous infiltrates involving the meninges, hypothalamus, pituitary gland, and cranial nerves.4 Spinal sarcoidosis is relatively uncommon and can manifest as intramedullary lesions, intradural extramedullary or extradural lesions, cauda equina syndrome, and arachnoiditis.5 We report 2 patients with a cervical cord lesion in conjunction with clinical manifestations suggestive of a myelopathy. In both patients, a diagnosis of an idiopathic inflammatory demyelinating syndrome was considered likely. Further, the results of magnetic resonance imaging (MRI) showed characteristics that were distinctively conspicuous for an intra-axial process with a concomitant leptomeningeal component.

Case 1

A 27-year-old African American woman was symptomatic with numbness involving the right arm, trunk, and leg in December 1997. This resolved spontaneously in 2 weeks. In September 1998, she developed numbness involving the medial aspect of both hands. The MRI results of the cervical spine showed a gadolinium-enhancing lesion within the spinal cord at C6-7 (Figure 1). The lesion appeared to be localized to the posterior aspect of the spinal cord. On sagittal sequences, it was difficult to differentiate whether the lesion was exclusively intra-axial or whether there was a component of leptomeningeal involvement. The MRI results of the brain were normal. Cerebrospinal fluid studies; levels of antinuclear antibodies, angiotensin-converting enzymes, and vitamin B12; syphilis and Lyme serologies; and chest radiographic and purified protein derivative skin test results were all normal. The patient was treated with intravenous methylprednisolone (1 g daily for 3 days) and improved significantly. She was diagnosed as having an idiopathic inflammatory demyelinating syndrome. In February 1999, she developed an episode of right facial numb-
ness with right facial hypesthesia, which resolved spontaneously in a few days. Her general medical and neurologic examination results were unremarkable. Repeated cervical spine MRI results showed persistent enhancement of the cervical cord lesion. Repeated brain MRI results were again unremarkable.

A computed axial tomographic scan of the chest revealed hilar adenopathy, and diffuse echogenic lesions were seen in the liver and spleen on the abdominal computed axial tomographic scan. A repeated angiotensin-converting enzyme level test showed values were elevated at 121 U/L (reference range, 7-46 U/L). Transbronchial biopsy results identified noncaseating granulomas consistent with sarcoidosis. She was prescribed oral prednisone at 60 mg/d. She developed no recurrence of neurologic symptoms; the spinal cord lesion and angiotensin-converting enzyme levels decreased, and the prednisone was tapered across 1 year.

CASE 2

A 36-year-old man had been symptomatic since July 1999 with slowly progressive numbness involving the hands and feet. Cervical spine MRI results were remarkable for an enhancing intramedullary lesion at C4-C5. A biopsy of the lesion was performed, and a review of the slides revealed evidence of mild acute inflammation and benign fibrous tissue. He was given steroids in the postoperative period, and his paresthesias showed significant improvement. In October 1999, he developed a recurrence of the paresthesias, which progressed slowly initially and then more rapidly for the 6 months prior to his evaluation. He was symptomatic with numbness below the neck and decreased ability to appreciate hot and cold sensations, on the right side more than the left. He had difficulty opening jars and some coordination difficulty with his legs. His neurologic examination results showed decreased perception of pinprick, temperature, and touch, from C4 downward, on the right side more than the left. Strength, tone, and coordination were intact. His reflexes were symmetrically brisk, and the plantar responses were flexor. His gait was normal, and Romberg test results were negative.

Cervical spine MRI results (Figure 2) revealed a 2-cm lesion in the dorsal part of the cervical cord at C4-C5. The lesion appeared to be infiltrated over the posterior cord surface and showed pronounced contrast enhancement. Also noted were anterior and posterior leptomeningeal enhancements. Brain and thoracic cord MRI results were normal. Normal values were obtained from a hemogram, and for vitamin B12, folate, liver enzymes, angiotensin-converting enzymes, immunoelectrophoresis, thyrotropin, and antinuclear antibodies. Evoked potential studies (brainstem, visual, and somatosensory) and results of spinal fluid analysis were normal.

A computed axial tomographic scan of the chest showed bilateral hilar and mediastinal adenopathy. The
bronchoalveolar lavage showed numerous macrophages and sparse inflammatory cells, predominantly histiocytes. Transbronchial biopsy results showed a compact, well-circumscribed noncaseating granuloma composed of Langerhans and foreign body–type multinucleated giant cells with abundant eosinophilic cytoplasm.

**COMMENT**

It is rare for intramedullary spinal sarcoidosis to be the initial or only manifestation of the disease. In our 2 reported cases, there were no systemic symptoms. Nevertheless, both had hilar adenopathy, which led to the diagnosis of neurosarcoidosis. The age range for intramedullary sarcoidosis is 15 to 68 years (median, 35 years). In spinal sarcoidosis, the cervical region is most commonly involved. The clinical picture is usually that of a myelopathy with paraparesis, quadriplegia, hypesthesias, and bladder and bowel dysfunction all being well-described symptoms. Intramedullary neurosarcoidosis is known to mimic a spinal cord tumor both clinically and radiologically. Spontaneous remissions and relapses and steroid-induced improvement can lead to a clinical picture that resembles an inflammatory demyelinating disease. This is well illustrated by the 2 cases presented.

Both patients had an enhancing cervical cord lesion that appeared to infiltrate over the posterior cord surface. The second case described had evidence of anterior and posterior leptomeningeal enhancement. It has been suggested that with early inflammation there is linear leptomeningeal enhancement along the surface of the spinal cord. Spread of the leptomeningeal inflammatory process to the Virchow-Robin spaces is believed to result in parenchymal involvement, which appears as diffuse cord enlargement on MRI results. With consolidation, 1 or more discrete masses form, and with resolution of the inflammatory phase, the cord returns to normal size. Ultimately, these processes can culminate in spinal cord atrophy. The MRI characteristics of spinal neurosarcoid lesions depend on the stage of the illness. Most findings can be relatively nonspecific, but the presence of an infiltrating lesion with meningeal and intramedullary enhancement should increase the suspicion for sarcoidosis or other granulomatous diseases. Enhancement patterns are variable and can include linear leptomeningeal enhancement or focal, multifocal, or diffuse intraparenchymal enhancement; often, there is a broad base on the cord surface and the enhancement does not involve the full cord thickness. A decrease in MRI enhancement can be seen as patients improve.

In neurosarcoidosis, extradural spinal disease is rare and can involve the vertebral bodies. Sarcoid spondylodiscitis and paraspinal mass have been reported. Enlarged cervical nerve roots, not evident on MRI results but evident on a myelogram, have been described. Intradural extramedullary space occupying the lesion and intraspinal epidural mass secondary to sarcoidosis can occur. Intradural extramedullary findings include pial enhancement, root nodules, and clumping of nerve roots. Enhancing cauda equina lesions mimicking subarachnoid tumor seeding can occur. Intramedullary disease can initially appear as an enhancing intramedullary lesion, cord enlargement, cord atrophy, or diffuse and focal T2-weighted signal hyperintensities. The only abnormality may be abnormal enhancement with normal cord appearance on T1- and T2-weighted images. Multiple small nodules on enhanced T1-weighted cervical cord MRI results can occur.

![Figure 2](image-url)

**Figure 2.** A, Sagittal T2-weighted cervical cord magnetic resonance image showing a hyperintense 2-cm lesion with discrete but irregular margins (arrow) within the mid and dorsal portions of the cervical cord at the C4 - C5 levels. B, Postgadolinium T1-weighted magnetic resonance image showing pronounced contrast enhancement with anterior and posterior leptomeningeal enhancement (arrow).
of spinal sarcoidosis with unique MRI findings due to hemosiderin deposition and calcification in the lesion have been described. Also described is a case of spinal sarcoidosis mimicking syringomyelia on MRI results.

In both our patients, the diagnosis was provided by transbronchial biopsy results. In fact, in our second patient, the spinal cord biopsy results did not provide the diagnosis. The granulomas in the spinal cord and brain are often poorly formed, are smaller than the systemic lesions, and have fewer giant cells. For this reason, the spinal cord lesions can mimic an intramedullary tumor on frozen section. Further attempts at wide surgical excision can lead to increased morbidity. Case reports of patients undergoing spinal cord biopsy in intramedullary spinal sarcoidosis suggest a high morbidity and mortality.

Intradural spinal sarcoidosis should be carefully considered as a possibility in a patient with known sarcoidosis who develops a radiculopathy or myelopathy. On rare occasions, intradural spinal sarcoidosis may be the initial manifestation of sarcoidosis. Manifestation mimicking a cord neoplasm is more common, but a relapsing-remitting neurologic course can occur. An infiltrating intramedullary cord lesion with leptomeningeal enhancement should bring to mind the possibility of sarcoidosis. A systematic search for evidence of extraneural sites of involvement should be mandatory in such cases. Even in the absence of systemic symptoms, targeting such sites for potential biopsy can result in the identification of the disease process with substantially less morbidity than sampling neural tissue. A high index of suspicion for the diagnosis is required because early intervention is associated with a favorable outcome. Progressive deterioration, nonresponsiveness to steroids, and an enlarging cord mass should prompt reconsideration of the diagnosis. In such cases, performing a biopsy of neural tissue may be inevitable.

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REFERENCES


