

## Intramedullary Cervical Sarcoidosis as the Initial Presentation of Systemic Sarcoidosis

Rishi N. Razdan, M.D., Juan Carlos P. Lozada, M.D., and Shashi Chaddha, M.D.

We present a case of a 57-year-old African American man with systemic sarcoidosis, who initially presented with cervical spinal cord disease. Initial MRI showed an elongated intramedullary area of increased signal intensity on T2 and inversion recovery sequences within the cervical spinal cord with minimal contrast enhancement after gadolinium administration. Further radiologic evaluation led to a chest CT, which showed bilateral hilar lymphadenopathy. Thoracic lymph node biopsy revealed systemic sarcoidosis. Post treatment MRI showed improvement of the cervical spinal cord lesion, further supporting the diagnosis of systemic sarcoidosis.

### Introduction

Patients affected by intramedullary cervical spinal cord sarcoidosis typically present with neurologic symptoms associated with the location of the lesion within the cervical spinal cord. The severity of symptoms is mostly dependent on size, location, and the amount of inflammation surrounding the granuloma. If not recognized and treated early, neurosarcoidosis can be a debilitating illness leading to weakness, paresthesias, and paralysis.

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Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging

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### Case Report

A 57-year-old African American man presented to the neurologist in July, 2007 with a two month history of numbness in his finger tips, particularly of his left hand, that progressed to his torso and abdomen over a two month period. After the onset of the numbness, the patient had difficulty in ambulating, and weakness with associated falls and loss of balance. Review of systems indicated a 10-15 pound weight gain, nocturia, and tremor of his hands.

Pertinent neurological physical exam findings revealed minimal end point nystagmus on left lateral gaze and upper and lower extremity weakness with slight decrease in proprioception of both lower extremities.

MRI was obtained at this time (Fig. 1) revealing an expansile, elongated area of increased T2 signal in the cervical spinal cord on T2 and STIR sequences, extending from C4 to C7, with minimal post contrast enhancement. The findings at this time were suggestive of an intramedullary lesion, such as a demyelinating or neoplastic lesion.

At this time, due to the possibility of neoplastic

## Intramedullary Cervical Sarcoidosis as the Initial Presentation of Systemic Sarcoidosis



Figure 1A. 57-year-old man with sarcoidosis. From left to right: Short term inversion recovery, T2 weighted, and T1 weighted post gadolinium administration sagittal MRI of the cervical spine showing an elongated intramedullary area of increased signal intensity with minimal contrast enhancement after gadolinium administration.

process, CT examination of the chest (Fig. 2) was also obtained which revealed extensive right hilar lymphadenopathy with mild left hilar adenopathy and moderate mediastinal adenopathy. This does not make much sense. The patient is young and the lesion is intramedullary. Unless the patient has a known neoplasm, the possibility of metastatic disease to the is very unlikely. Consider stating that chest CT was performed as part of the diagnostic work up. Lymphoma was favored over sarcoidosis because of the asymmetry of the adenopathy.

Mediastinoscopy and biopsy of the mediastinal lymph nodes were performed by the thoracic surgeon. Pathologic analysis revealed non-caseating granulomatous inflammation consistent with sarcoidosis. The working diagnosis of Sarcoid was agreed upon and the patient was placed on steroid therapy with follow up MR recommended after one month. MRI after one months time (Fig. 3) showed significant improvement in the cervical spinal cord lesion, with decrease in size and enhancement when compared to the prior examination.



Figure 2. 57-year-old man with sarcoidosis. Coronal CT of the chest with contrast demonstrating extensive right sided hilar lymphadenopathy with mild left hilar adenopathy and moderate mediastinal adenopathy.

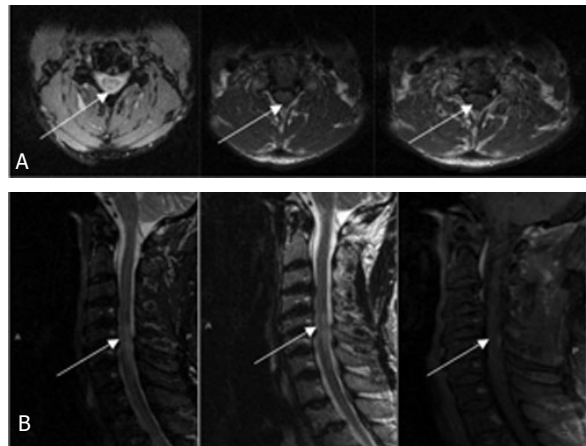


Figure 3. 57-year-old man with sarcoidosis. From left to right: Short term inversion recovery, T2 weighted, and T1 weighted post gadolinium administration sagittal MRI of the cervical spine showing a significant decrease in both signal intensity and size of the previously noted region of increased signal intensity on T2, and STIR sequences and also improvement of the amount of contrast enhancement.

## Intramedullary Cervical Sarcoidosis as the Initial Presentation of Systemic Sarcoidosis

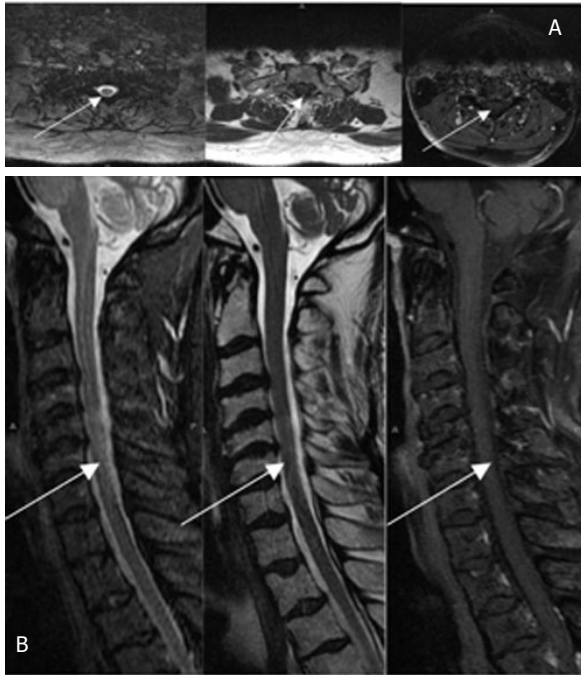


Figure 4. 57-year-old man with sarcoidosis. From left to right: short term inversion recovery, T2 weighted, and T1 weighted post gadolinium administration sagittal MRI of the cervical spine showing the previously noted elongated area of increased signal intensity again identified within the spinal cord on the STIR sequence but appearing less intense. The area extends from C5 to C7 and is without significantly increased signal intensity on the T1 and T2 sequences. There was no significant contrast enhancement on the post gadolinium T1 sequence.

The patient returned for follow up MRI four months after the initial MR (Fig. 4) , which demonstrated near complete resolution of the cervical spinal cord lesion. Although discussed by the primary clinicians, spinal cord biopsy was not necessary due to the therapeutic improvement of the patients condition and because of the risks of the procedure.

### Discussion

Sarcoidosis is an idiopathic, systemic disease characterized by granuloma formation in a variety of anatomical locations. The most common of these locations are the hilar lymph nodes. However, the disease has been

noted to involve a variety of organ systems ranging from the genitalia to the nervous system. Typically, laboratory findings include elevated angiotensin converting enzyme level (ACE), hypercalcemia, restrictive pulmonary function, elevated CSF protein, and a decreased CD4:CD8 ratio. The typical pathologic findings in sarcoidosis involve non-caseating granulomas with epithelioid cells and large, multinucleated giant cells.

The first case of sarcoidosis involving the nervous system was reported in 1948 [8]. Few documented cases of sarcoidosis of the cervical spinal cord have been reported, and even fewer are histologically documented. Sarcoidosis of the central nervous system is clinically involved in approximately 5% of cases [2]. Many articles show that cases of isolated spinal cord sarcoid (lack of systemic findings) range from 0.5-1% [6, 8]. In a report by Bhagavati and Choi, involvement of the spinal cord was presumed in approximately 0.3-0.4% of cases which predominantly affected the cervical and thoracic spinal cord [1]. Their literature review established “intramedullary involvement in 35% of cases, extramedullary involvement in 35% of cases, and both [intra and extramedullary involvement] in 23% of patients”[1]. Typically, the intramedullary lesions are longitudinally oriented within the spinal cord. The lesions demonstrate decreased signal on T2 weighted imaging. However, because of the surrounding edema caused by the granulomatous infiltration, the spinal cord is often times enlarged and exhibits high signal intensity. The differential diagnoses for longitudinally extensive myelopathies include spinal cord tumor, syringomyelia, neuromyelitis optica, and vitamin B12, vitamin E, or copper deficiency myelopathies [1].

T1 weighted imaging with Gadolinium shows enhancement of the inflammatory granuloma. Varying appearances have been described based on enhancement pattern. Junger et al describes four phases of inflammation: Phase 1 corresponds to early inflammation with contrast imaging demonstrating “linear” leptomeningeal enhancement along the surface of the spinal cord. In phase 2, an enhancing lesion may be noted due to parenchymal involvement and radiating spread of the disease thought to expand through Virchow-Robin spaces. In phase 3, there is improvement in both the inflammation and the enlargement of the spinal cord with focal or multifocal intramedullary lesions. Phase 4 is the chronic phase and atrophy of the spinal cord may be present [4].

## Intramedullary Cervical Sarcoidosis as the Initial Presentation of Systemic Sarcoidosis

Patients affected by intramedullary cervical spinal cord sarcoidosis typically present with neurologic symptoms associated with the location of the lesion within the cervical spinal cord. The severity of symptoms are mostly dependent on size, location, and the amount of inflammation surrounding the granuloma. The disease can lead to abnormal sensation or weakness in one or more limbs, or cauda equina symptoms. If not recognized and treated early, neurosarcoidosis can be a debilitating illness leading to weakness, paresthesias, and paralysis.

MRI examination with gadolinium enhancement is the superior modality to suggest the diagnosis. Although imaging is very important in establishing a diagnosis, direct tissue sampling and evaluation is mandatory in those cases where the patient has no prior history or systemic manifestation of sarcoidosis and the clinical/laboratory history is unremarkable. Tissue biopsy has proven sarcoidosis in many instances suspected to be completely different disease entities including infectious and neoplastic processes. Whereas tissue diagnosis is important, current recommendations are to engage upon less invasive modalities to establish a diagnosis if possible due to the risks of surgical manipulation of the spinal cord and the limitations associated with biopsy. Interestingly, there have been documented reports of negative proven biopsies speculated as a result of the smaller size and relative fewer number of giant cells found within spinal cord granulomas compared with systemic granulomas [1]. Furthermore, intraoperative biopsies have lead to misdiagnoses in few instances with the final correct sarcoid diagnosis made on paraffin sections. Immunohistochemistry staining and/or frozen intraoperative samples are more current modalities of pathologic evaluation and may help avoid the previously noted limitations to biopsies [7].

The combination of MRI, chest radiograph and laboratory findings including ACE levels, calcium level, and CD4:CD8 have proven the most complete and least invasive method for establishing a working diagnosis. Medical management should follow shortly thereafter including high dose steroids. Other treatment modalities include methotrexate, cyclophosphamide, cyclosporine, azathioprine, chlorambucil, chloroquine, and hydroxychloroquine [6].

Our patient who originally presented with neurologic symptoms as described above, combined with chest CT findings of hilar lymphadenopathy was initially

thought to have lymphoma. However, due to thorough follow-up by the entire medical team, a misdiagnosis was averted after further examinations were obtained leading to the final working diagnosis of systemic sarcoidosis. Interestingly, whereas most sarcoid patients present with respiratory symptoms, our patient's most prevalent symptoms manifested in the central nervous system. There is no mention in prior literature regarding the progression of disease to the central nervous system, although there is mention of isolated intramedullary lesions and their presentations. Although tissue biopsy was never performed, chest CT and laboratory findings, and the significant improvement of the clinical symptoms after medical management confirmed the diagnosis of intramedullary sarcoidosis of the cervical spinal cord.

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## Intramedullary Cervical Sarcoidosis as the Initial Presentation of Systemic Sarcoidosis

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