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Case Report

Thoracic intramedullary neurosarcoidosis with thoracic disc herniation: Diagnostic importance of intramedullary contrast enhancement

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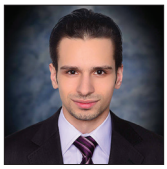

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ABSTRACT

Background: Thoracic intramedullary neurosarcoidosis is an uncommon but serious manifestation of spinal cord disease. Its concomitant occurrence with thoracic disc herniation can mislead the physician into attributing neurologic and radiographic findings in the spinal cord to disc pathology rather than inflammatory disorder. Here, we present such a rare case of concomitant thoracic disc and spinal neurosarcoidosis.

Case Description: A 37-year-old male presented with progressive right lower extremity weakness and numbness. Magnetic resonance imaging (MRI) of the thoracic spinal cord revealed a T6-T7 paracentral disc eccentric to the right with T2 signal change extending from T2 to T10 level. This prompted acquiring a contrasted MRI that also depicted intramedullary enhancement around the T6-T7 disc bulge. Computed tomography scan of the chest showed mediastinal lymphadenopathy concerning for sarcoidosis. Lymph node biopsy confirmed the diagnosis of sarcoidosis, and high-dose steroid treatment was initiated. The patient had significant symptomatic improvement with steroids with full neurological recovery and improvement of his symptoms.

Conclusion: While stenosis from thoracic disc disease could potentially suggest a mechanical etiology for the patient's symptoms, attention must be paid to the imaging findings as well as the degree and extent of cord signal change and intramedullary contrast enhancement. Appropriate and timely diagnosis is essential to avoid unnecessary invasive procedures.

Keywords: Cord edema, Intramedullary, Neurosarcoidosis, T2 signal change, Thoracic disc; mimickers

INTRODUCTION

Sarcoidosis is a well-described inflammatory granulomatous disease that affects multiple organ systems. Involvement of the central nervous system is present in approximately 25% of cases; however, it remains subclinical in the majority of patients.[8] Spinal involvement occurs in <1% of sarcoidosis cases, of which the cervical cord is most commonly involved, while the thoracic cord is affected in approximately 20–30% of cases.[7,9,12]

Spinal neurosarcoidosis is particularly difficult to diagnosis due to its rarity and nonspecific clinical presentation, which can be indistinguishable from other etiologies. Radiographic findings

can frequently be nondiagnostic. However, the classic pattern that suggests a diagnosis of spinal neurosarcoidosis is T2 hyperintensity and contrast enhancement within the central spinal cord.[1,4] Similarly, direct mechanical compression of the spinal cord from a disc herniation may also lead to cord signal change and edema, making a discrete distinction between the two etiologies difficult. However, intramedullary cord contrast enhancement is not typically seen with cord compression secondary to disc herniation. Herein, we report a rare case of concomitant spinal neurosarcoidosis and a compressive thoracic disc disease leading to cord signal change and intramedullary enhancement. We review the imaging findings of spinal neurosarcoidosis and important key factors to differentiate between the two processes for timely diagnosis and treatment to avoid unnecessary invasive procedures.

CASE REPORT

Clinical presentation

A 37-year-old otherwise healthy African-American male presented with 3 months of progressive right lower extremity weakness and numbness. The patient noticed that he had been walking with a limp and was unable to urinate or ejaculate despite preserved erection. On neurological examination, he had decreased strength in the right hip flexion (4/5), knee extension (4/5), foot dorsiflexion (3/5), extensor hallucis longus (2/5), and plantar flexion (2/5). He also reported decreased sensation in the right lower extremity in nondermatomal distribution.

Imaging

Computed tomography (CT) imaging of the spine revealed a calcified T6-T7 paracentral disc eccentric to the right [Figure 1a and b]. Magnetic resonance imaging (MRI) of his spine depicted again the T6-T7 paracentral disc with T2 signal change extending from T2 to T10 level [Figure 1c and d]. Given that, the extent of T2 cord signal change was out of proportion to the disc disease, contrasted MRI was acquired afterward demonstrating intramedullary enhancement around the T6-T7 disc bulge [Figure 1e].

Histopathologic findings

The patient was started on high-dose dexamethasone. The patient noticed significant improvement in his strength over the subsequent 3 days. Given the extent of T2 signal change and contrast enhancement in the spinal cord and response to steroids, the suspicion for another primary etiology was investigated. CT chest revealed diffuse mediastinal lymphadenopathy. Angiotensin-converting enzyme level was within normal limits. He underwent an ultrasound-guided endobronchial biopsy of the enlarged lymph nodes. Histopathological examination demonstrated noncaseating granulomas consistent with a diagnosis of sarcoidosis [Figure 1f].

Treatment and follow-up

The patient received 3 days of high-dose methylprednisolone therapy followed by prednisone (40 mg twice a day) at the time of discharge along with methotrexate and infliximab infusion. His right lower extremity returned to nearly full strength, with only mild weakness in the right extensor halluces longus and plantar flexion (4+/5) at his last follow-up visit.

DISCUSSION

Neurosarcoidosis can occur anywhere along the neuroaxis but most commonly affects the meninges of the skull base. The occurrence of spinal neurosarcoidosis is uncommon, with the cervical spine being the most common location followed by the thoracic spine.[10,11] Similarly, intramedullary manifestation of the disease is also uncommon with the disease usually spanning >3–4 intramedullary levels.[10,11] As seen in this case, the presence of T2 signal change out of proportion to the disc size/compression as well as the rapid and significant improvement in the neurological examination following steroid administration prompted investigation of another etiology before pursuing surgical intervention.

Spinal neurosarcoidosis is associated with a spectrum of clinical manifestations that could be nonspecific. Common presentations include cranial nerve palsy (31–55%), chronic aseptic meningitis (16–37%), spinal cord disease/myelitis, cerebral parenchymal disease (21%), neuroendocrine involvement (6–9%), hydrocephalus (9–10%), cerebral infarction (6%), and peripheral nervous system symptoms such as polyneuropathy (17%).[13] Due to the variability in symptoms, other confirmatory diagnostic tools must be used in conjunction with the clinical examination to establish the diagnosis and tailor management.

Diagnostic workup of neurosarcoidosis involves imaging, serum and cerebrospinal fluid (CSF) analysis, and biopsy of enlarged lymph nodes. Imaging findings in spinal neurosarcoidosis can also be found in other conditions. Contrast enhancement on MR imaging provides evidence of breakdown of the blood–brain barrier, which has been reported as a highly sensitive marker of neuroinflammation.[6] The presence of extensive T2 signal change within the spinal cord can be often seen. However, intramedullary contrast enhancement spanning >3–4 levels can point towards the diagnosis. In our patient, although a compressive thoracic disc herniation can lead to T2 signal change in the cord, the extensive and diffuse nature of the T2 signal change was

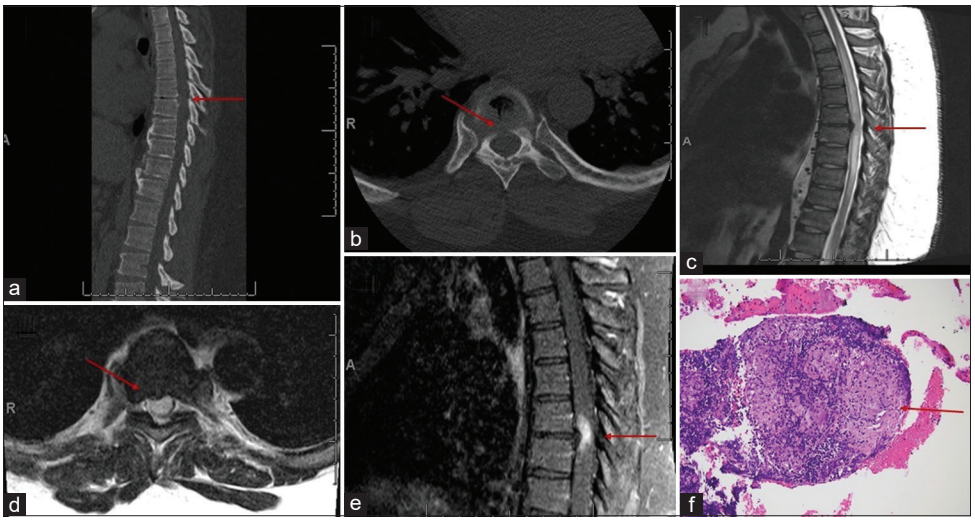

Figure 1: Sagittal (a) and axial (b) computed tomography scan of the thoracic spine showing a calcified T6-T7 paracentral disc eccentric to the right. Sagittal (c) and axial (d) T2-weighted magnetic resonance imaging (MRI) demonstrating the T6-T7 paracentral disc with T2 signal change extending from the T2 to T10 levels. Sagittal T1-weighted MRI with gadolinium (e) demonstrating enhancement around the T6-T7 disc bulge. Histopathological examination demonstrated noncaseating granulomas consistent with the diagnosis of sarcoidosis (f).

reflective of a more inflammatory etiology, which prompted us to acquire a contrasted MRI that revealed intramedullary contrast enhancement.

A thorough diagnostic workup is critical to providing the appropriate treatment therapy in this patient. Furthermore, establishment of a diagnosis of spinal neurosarcoidosis prevented this patient from undergoing an unnecessary surgery for his T6-7 paracentral disc. This is important as neurological surgical procedures are not without risk; the overall morbidity following surgery to correct a thoracic disc herniation is reported as high as 29% (medical 21%, surgical site 11%, CSF related 8%, and neurologic complications 5%).[2] Thus, accurate diagnosis is of a paramount importance before entertaining any surgical intervention.

Timely and appropriate treatment of neurosarcoidosis is essential to achieve reversal of symptoms and to prevent progression of disease. Specific treatment of spinal neurosarcoidosis is not well described in the literature and is adapted from general management of multiorgan sarcoidosis. The goal is to treat the underlying inflammatory process and to reverse the symptomatic effects of the neuroinflammation. Initial therapy often starts with oral steroids, or an intravenous equivalent may be used for severe disease.[13] Failure of steroid treatment is common, with >80% of patients progressing to second- or third-line therapies. Second-line treatment includes nonsteroidal disease-modifying agents such as methotrexate or mycophenolate.[13] However, these medications have extensive side effect profiles and are frequently not tolerated. Finally, third-line treatment agents include tumor necrosis factor-alpha inhibitors such as infliximab. Recent studies have shown its effectiveness despite treatment failure with steroids.[2,5,13] In this case, our patient responded appropriately to methylprednisolone high pulse therapy followed by prednisone, methotrexate, and infliximab infusion.

CONCLUSION

We present a rare case of concomitant compressive thoracic disc and spinal neurosarcoidosis in a patient presenting with leg weakness. While a thoracic disc on spinal imaging may potentially suggest a mechanical etiology for the patient's symptoms, careful attention must be paid to radiographic characteristics, specifically the extent of spinal cord signal change and intramedullary enhancement. This report highlights the importance of a complete diagnostic workup to identify the underlying etiology and provide the appropriate therapeutic intervention. Due to its variable presentation, spinal neurosarcoidosis should remain a differential diagnosis in patients presenting with progressive weakness.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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