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Case Report

Thoracic Intramedullary Sarcoidosis Masquerading as Thoracic Spondylotic Myelopathy: A Case Report and Review of the Literature

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Abstract

We report a rare case of biopsy proven, thoracic intramedullary neurosarcoidosis in a 52- year-old male, who was thought to have progressive paraparesis as a result of degenerative spondylosis of the thoracic spine. Despite decompressive surgery, he would, over the course of 21 months, become irreversibly paraplegic and unresponsive to steroid treatment. To our knowledge, this is the only reported case of thoracic intramedullary neurosarcoidosis masked by severe thoracic stenosis and spondylotic changes.

Keywords: Thoracic Intramedullary Sarcoidosis; Thoracic Spondylotic Myelopathy

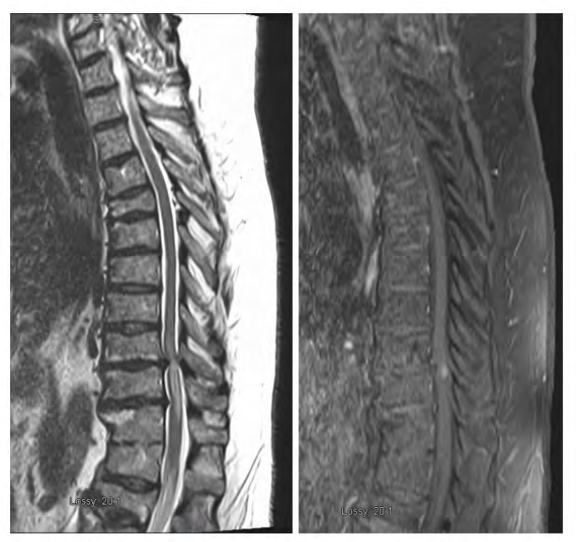
Introduction

Sarcoidosis is a systemic granulomatous disease that can manifest within the spinal cord, potentially leading to irreversible neurological decline and death. Intramedullary neurosarcoidosis is an infrequent diagnosis that shares a radiographic differential diagnosis with central nervous system (CNS) malignancy, demyelinating disease, spinal cord ischemia, infectious or post-infectious myelitis, spondylotic myelopathy, vitamin B12 or copper deficiency, and paraneoplastic syndrome [1-4]. The diagnosis can therefore be elusive, and as such, should be considered early in the workup of expansile intramedullary lesions. The mainstay of therapy is high-dose intravenous corticosteroids and immunosuppressants, which can lead to a cessation or amelioration of symptoms [2,5]. Though clinical improvement is often variable, the potential for a favorable response creates an impetus for prompt recognition and treatment.

Case Report

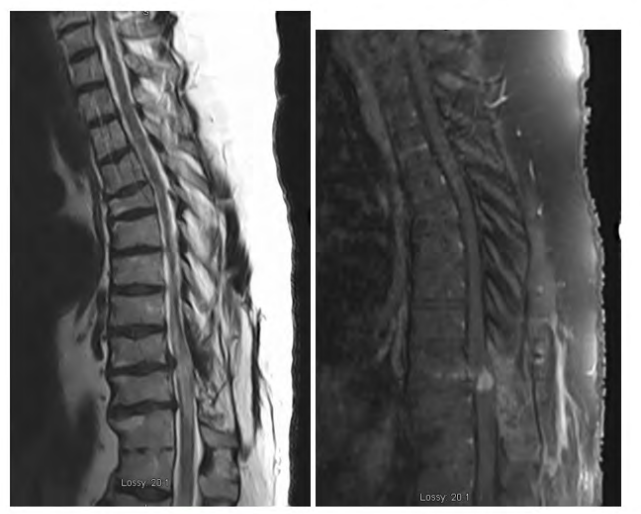
A 52-year-old male was referred with an intramedullary spinal cord lesion. His symptoms began nearly 2 years prior with sharp back pain, radicular symptoms, and difficulty with ambulation. Within 2 weeks of symptom onset, he developed bilateral lower extremity weakness with rapid progression. Magnetic resonance imaging (MRI) showed disc-osteophyte complexes at T9-10 and T10-11, causing severe canal stenosis and cord compression. A 5 cm intramedullary hyper-intensity centered at the T9-10 level was seen on T2-weighted MRI (Figure 1a). A small 0.7 cm intradural enhancing focus was seen along the posterior margin of the cord at T9-10 on T1-weighted MRI after gadolinium injection (Figure 1b).

Decompressive laminectomies were performed emergently at an outside institution from T8 to T10. This was undertaken 21 months prior to presentation at our hospital. Notable residual neurological deficits were observed in both lower extremities



(Figure 1a) (Figure 1b)

Figure 1: Pyrolysis products from microwave pyrolysis of agro-residue.



(Figure 2a) (Figure 2b)

Figure 2: Sagittal T2-weighted MRI (Figure 2a) and T1 -weighted MRI with gadolinium enhancement (Figure 2b) following thoracic laminectomy, depicting progressive expansile thoracic intramedullary lesion with increased enhancement.

postoperatively despite adequate decompression. Serial MRI scans showed markedly increased T2-signal abnormality with focal enhancement within the thoracic cord (Figure 2a and 2b). A 5-day course of high-dose intravenous Solu-Medrol was started for a presumptive diagnosis of transverse myelitis, which had a favorable clinical and radiographic response. A period of waxing and waning neurological symptoms ensued characterized by motor weakness, spasticity, and urinary retention. Subsequent radiographic and serological workups for multiple sclerosis (MS) and neuromyelitis optica (NMO) were unrevealing (Figure 3). Computerized tomography (CT) scans of the chest demonstrated paratracheal lymphadenopathy. Right paratracheal lymph node biopsy showed numerous noncaseating epithelioid granulomata with negative staining for acid-fast bacilli and fungi, morphologically consistent with a diagnosis of sarcoidosis (Figure 4). Angiotensin converting enzyme level was within normal limits at 17 U/L.

Lower extremity strength eventually deteriorated despite ongoing treatment with low- dose oral cortico-steroids and plasma exchange therapy. The patient presented to the senior author 19 months after the onset of symptoms with spastic diplegia and incontinence.

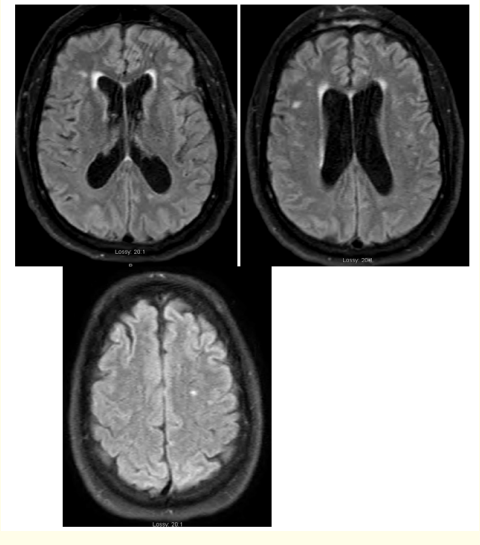


Figure 3: Axial flair (fluid-attenuated inversion recovery) sequence MRI depicting scattered periventricular and subcortically based areas of T2 signal abnormality that likely represent areas of gliosis that are not disease specific.

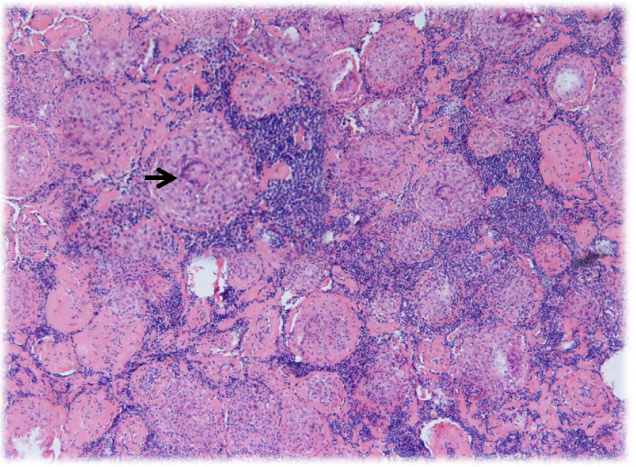


Figure 4: H&E stained microphotograph (40X) of a paratracheal lymph node biopsy shows multiple well formed non-caseating epithelioid granulomas in a inflammatory background, a characteristic feature of sarcoidosis. Note the presence of a well developed multi-nuclear giant cell histiocyte (arrow).

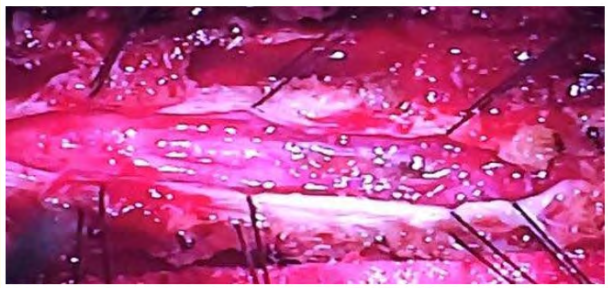


Figure 5: Thoracic mid-line myelotomy with necrotic intramedullary mass.

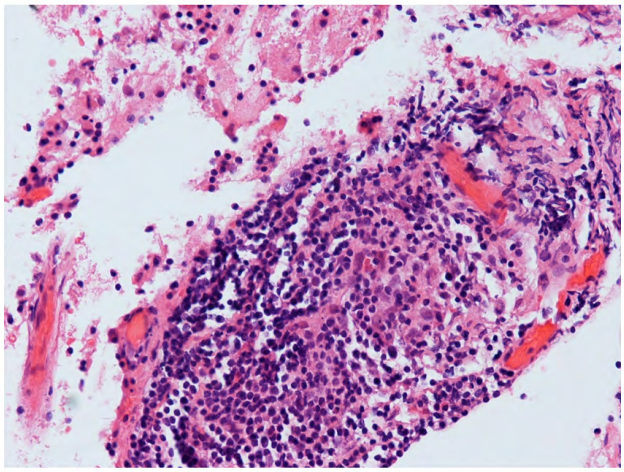


Figure 6: H&E stained microphotograph (200X) of a thoracic spinal cord biopsy shows a non- caseating epithelioid granuloma with adjacent lymphocytes.

The patient underwent a midline myelotomy and biopsy of the thoracic intramedullary lesion (Figure 5). A necrotic, expansile, infiltrative intramedullary lesion was encountered. Pathology showed focal and ill-defined granulomata with no evidence of neoplastic tissue (Figure 6). No clinical improvement was noted at three month follow- up.

Discussion and Conclusion

Sarcoidosis was first described in 1877 by British surgeon, Jonathan Hutchinson, as being an affliction of the skin. It is now widely accepted as a multi-organ system affliction which stems from a type-1 immune response [6]. This leads to the formation of noncaseating granulomas, the hallmark pathological finding [1,6]. Although up to 90% of cases involve the lungs, granulomata can be widely dispersed in the skin, lymphatics, liver, spleen, eyes, parotid glands, phalangeal bones and the nervous system, lending to tremendous heterogeneity in clinical presentation and delayed or missed diagnosis [1,6-8]. Diagnostic methods have also varied historically, and as a result, have lead to discordance in epidemiological data [6]. Biopsy of affected tissue is the gold standard for accurate diagnosis [1]. In the US, risk among whites is 0.85% with an incidence of 35.5 cases among 100,000 whites. Among blacks, risk is 2.4% with an incidence of 10.9 cases among 100,000 blacks [6].

Central and peripheral nervous system involvement occurs in 5 to 17% of patients with sarcoidosis [1,9,10]. CNS involvement alone can occur in up to 15%, with leptomeningeal disease and cranial neuropathies being most prevalent [9-11]. Spinal cord involvement occurs in 6 to 8% of patients, and among this group, a cervical lesi-

on can be found in 56%, a thoracic lesion in 37% and a lumbosacral lesion in 7% [1,9]. Isolated spinal cord sarcoidosis, occurring in the absence of systemic disease, is exceedingly rare and occurs in less than 1% [1,12].

Biopsy proven cervical intramedullary neurosarcoidosis was described in a case report of a 59-year-old female with a large disk-osteophyte complex at C4-5 and adjacent spinal cord enhancement on contrast MRI. Symptoms improved transiently after anterior cervical discectomy and fusion, however, recurrence of weakness lead to a posterior decompression and biopsy, which unveiled the diagnosis [13]. There was another report of a patient who presented with cervical spondylosis and adjacent cord signal abnormality on T2-weighted MRI and 'electric-shock' like pain accompanied by weakness in both arms. Diagnosis was reached with transbronchial biopsy and the patient improved with corticosteroids [14]. Progressive paraparesis owing to thoracic intramedullary sarcoid has been described, however none in context of severe thoracic canal stenosis related to spondylotic disease [1,15].

Duhon., *et al*. recently described the case of a 48-year-old male whose symptoms were predominated by back pain with classic Lhermitte's phenomena and proprioceptive loss in the legs. MRI showed an expansile intramedullary lesion in the distal thoracic cord and adjacent disc-osteophyte complexes that effaced the ventral cord. Despite thoracic decompression and fusion, symptoms worsened. The patient improved after treatment with steroids and immunosuppressants for histologically proven neurosarcoidosis [1].

The case presented in our report is distinguished by concomitant thoracic disc herniation and stenosis causing complete obliteration of the arachnoid space as well as cord compression. This necessitated urgent surgical decompression in light of a rapid clinical deterioration. It is, however, important to note the limitations of performing a thoracic laminectomy alone for treatment of thoracic disc herniation. This treatment has been associated with an exceedingly high rate of morbidity by way of rapid neurological deterioration. Transpedicular or costovertebral approaches, with removal of the herniated disc, are instead favored and enable dorsal and ventral decompression of the spinal cord [16-18].

Our patient also experienced a waxing-waning clinical course despite continued oral steroids. Kumar., *et al*. described 2 patients, each with a cervical intramedullary lesion and relapsing-remitting symptoms that resembled an idiopathic demyelinating syndrome.

Both patients were diagnosed with transbronchial biopsy and one had a favorable response to prolonged oral steroids [3]. Of note, ACE levels appeared to be normal in the patients described by Duhon., *et al*. and Kumar., *et al*. however both reported elevated ACE levels on repeat testing [1,3]. CSF analysis is recognized as being unreliable for diagnosis of neurosarcoidosis [3,11,19].

Thoracic intramedullary neurosarcoidosis can cause a relentlessly progressive paraparesis that is not easily detectable by non-invasive methods. Prompt histopathological diagnosis must be pursued. The use of plasma-exchange therapy has not been well established [20-22]. Unfortunately, 25% of patients with spinal neurosarcoidosis will not improve with steroid treatment, and up to 40% of non-responders will also not improve with immunosuppressants [23]. Nevertheless, practitioners must remain mindful of the evasive nature of this treatable condition. The patient herein presented with acute painful paraparesis and initially had a waxing a waning clinical course, which is not characteristic of thoracic stenosis attributed to spondylotic changes. Early attention to the underlying, concomitant neurosarcoidosis could have potentially influenced outcome.

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