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Case Report

Isolated spinal neurosarcoidosis: An enigmatic intramedullary spinal cord pathology-case report and review of the literature

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Abstract

Isolated spinal cord neurosarcoidosis (NS) in the absence of systemic disease or intracranial involvement is exceptionally rare. Adjunctive laboratory tests though useful may not be reliable and the absence of any pathognomonic radiological features makes the diagnosis difficult. As spinal cord NS may be a presenting feature of systemic sarcoidosis which may be occult on routine workup, ^{18}F-fluorodeoxyglucose-positron emission tomography (FDG-PET) may be of value in unraveling this systemic involvement avoiding biopsying the spinal cord. A case of truly isolated NS is described with review of literature on this enigmatic pathology. Long segment intramedullary signal changes with focal parenchymal along with dural/meningeal enhancement in the absence of significant cervical stenosis in a young patient of northern European or African-American decent is very suggestive of NS and although may be presumably treated with steroids; there should be a low threshold for spinal cord biopsy especially in the absence of response to steroids to confirm isolated spinal cord NS in a patient with clinical neurological deterioration.

Key words: Biopsy, diagnosis, management, neurosarcoidosis, spinal

INTRODUCTION

Sarcoidosis is a systemic, multisystem granulomatous disease of unknown cause most commonly involving the lungs, lymph nodes, skin, and eyes.[1,2] While subclinical central nervous system (CNS) involvement may be more common as reported based on postmortem studies,[3] clinically obvious neurosarcoidosis (NS) is seen in only up to 15% of cases with 6-8% of these patients having spinal cord involvement.[1,2,4-6] According to Zagilek's criteria, NS can be diagnosed as definite, probable and possible based on clinical symptomatology, magnetic resonance imaging (MRI) features, laboratory findings, presence or absence of systemic sarcoidosis, exclusion of alternative diagnosis, and positive nervous system histology.[7] Most patients with NS have concomitant systemic involvement often obviating the need of surgical intervention relating to the CNS for diagnosis or therapeutic purposes and can be treated based on possible and probable criteria per Zagilek's.[4-10] This however is not always true and sarcoidosis albeit rarely; can manifest as true isolated spinal cord involvement in less than 0.5% cases.[9,11-27] The rarity of this presentation with nondiagnostic imaging features, clinical presentation, and absence of systemic involvement often makes it challenging to diagnose this condition leading to delay in diagnosis and often inappropriate management with concomitant morbidity.[23] Blood and cerebrospinal fluid (CSF) markers are mostly adjunct and have very low sensitivity and specificity for NS confounding the diagnosis.[1,5,28] While development of neurological syndromes in a patient with biopsy proven active systemic sarcoidosis, helps the diagnosis of NS, obtaining a tissue

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diagnosis at present represents the only way to diagnose isolated spinal NS which is invasive and can result in significant morbidity secondary to the critical location of the pathology within the intramedullary spinal cord.[23,29] Though sarcoidosis is a medically treatable disease, neurosurgeons may encounter it first hand as it can masquerade cervical spondylotic myelopathy[30] or may be consulted for possible spinal cord biopsy in neurological patients with features suspicious of NS/intramedullary spinal cord tumor for confirmation of diagnosis.[23] In fact, most of the cases with isolated NS in the study by Nozaki *et al.*,[9] were diagnosed as some other pathology with lymphoma being the most common before biopsy of the CNS and confirmation of the diagnosis of NS. Truly isolated spinal cord NS can only be definite or possible per Zagilek's criteria[7] as there is absence of systemic involvement. Probable NS is not truly isolated as there is evidence of systemic involvement and is more common than isolated NS, where CNS involvement is a part of systemic sarcoidosis and is fairly well-described.[31,32] Though these cases initially presented with features of spinal cord involvement, workup revealed presence of extraneural involvement,[31-33] as against very few cases described in the literature wherein true isolated spinal cord NS was described and mandated biopsy.[2,9,11,12,14-18,20-27,31,33-38] We encountered a case of isolated cervical intramedullary NS which presented as cervical myelopathy and could be diagnosed only after performance of spinal cord biopsy meeting the definite Zagilek's criteria. The clinical picture with nuances of diagnosis of this condition from the present case is discussed with comprehensive review of the literature and characteristic MRI features of spinal NS which might be useful for diagnosis/workup.

CASE REPORT

A 44-year-old African American man presented at an outside hospital with history of neck pain and left arm pain in a nondermatomal distribution associated with left hand numbness/tingling and left-sided weakness for a couple of months. Examination revealed presence of subtle left sided hemiparesis (4+/5 on Medical Research Council (MRC) grade) and considering a diagnosis of cervical spondylosis, a plain MRI of cervical spine was ordered which revealed presence of moderate cervical stenosis associated with T2 signal changes with no significant focal compression and he was treated conservatively. He however had progression of symptoms over the next few weeks due to which he was referred to our institution. Examination revealed presence of worsening weakness involving the left side (3/5) with brisk reflexes in bilateral upper and lower extremities. Sensory exam showed no gross dermatomal sensory deficits with presence of Hoffman's reflex. These neurologic findings prompted us to repeat an MRI with contrast enhancement due to presence of signal changes on the MRI without significant compression and disconcordance between clinical examination and imaging findings. MRI revealed presence of an approximately 1.5 cm patchy enhancement involving the left half of the cervical spinal cord at C4 level with presence of T2 signal changes spanning three to four segments in the cervical spine along with presence of moderate to severe spinal canal stenosis [Figures 1 and 2]. In light of the clinical symptoms and neuroimaging abnormalities, the diagnosis of spondylotic compressive myelopathy was questioned and a differential diagnosis of a neoplasm (astrocytoma/ependymoma), inflammatory, ischemic, and demyelinating lesions was entertained. Initial laboratory workup including lumbar puncture revealed that the patient had a mildly elevated protein level (43.5 mg/dl), a normal immunoglobulin (Ig)G/albumin CSF ratio (0.2), negative oligoclonal bands, and no malignant cells on CSF cytology studies. His CSF also demonstrated 110 red blood cells and 15 white blood cells per ml with 90% lymphocytes. Quantiferon gold tuberculosis (TB) test, aerobic, anaerobic, fungal, and TB cultures were all negative making infection an unlikely diagnosis. Erythrocyte sedimentation rate and C-reactive protein were slightly elevated with values being 19 (N: 0-17 mm/h) and 17 (N: 0.0-8 mg/dl). He was started on pulse dose of steroids with a likely diagnosis of myelitis and his clinical examination improved with improvement in motor strength to 4+/5 and he could ambulate better. However, while the steroids were being tapered, he had recurrence of symptoms with worsening weakness (3/5). The steroids were restarted, but because it was not possible to make a definitive diagnosis based on laboratory studies, in the

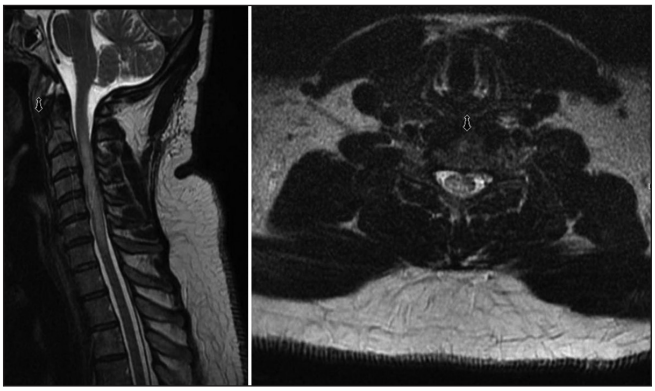

Figure 1: Sagittal (left) and axial (right) T2-weighted magnetic resonance imaging (MRI) demonstrating intramedullary hyperintensity without any enlargement of the spinal cord extending from the C3 to C6 level with superimposed cervical stenosis secondary to degenerative changes

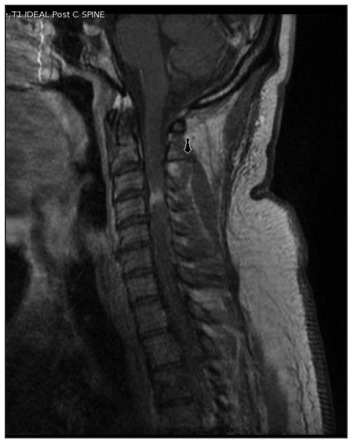

Figure 2: Sagittal post-constrast MRI showing patchy intramedullary enhancement at the C4 level with anterior dural/leptomeningeal enhancement

face of neurological worsening on tapering steroids, despite the risks of an intramedullary biopsy, this was thought to be the best option for diagnosis. A C3 through C6 laminoplasty and excisional biopsy of the C4 intramedullary lesion was performed to address the cervical stenosis and biopsy the intramedullary lesion [Figure 3]. Intraoperatively, the cord appeared normal except that it was quite swollen laterally on the left-hand side at C4 level and a dorsal root entry zone myelotomy was performed and abnormal tissue was immediately encountered which was quite distinct from the normal spinal cord tissue. This was sent for frozen section, which revealed gliosis versus neoplasm. Given the fact that this could be neoplastic tissue, further dissection was then carried out and a dissectible plane was found separate from normal spinal cord and a gross total resection could be achieved. The patient initially had worsening of his strength on left side and became essentially hemiplegic, but started improving gradually. Final pathology demonstrated non-necrotizing granulomatous disease consistent with sarcoidosis [Figure 4]. After the diagnosis was confirmed, an MRI of the brain [Figure 5] and CT of the chest abdomen and pelvis were done to rule out any other site of involvement which demonstrated slightly, but nonsignificant mediastinal and retroperitoneal lymph nodes. Serum and CSF angiotensin converting enzyme (ACE) levels performed were within normal limits. He was subsequently treated with pulse dose of intravenous (IV) methylprednisolone, methotrexate, and induction infliximab (chimeric monoclonal antibody against tumor necrosis factor (TNF)-α) therapy and was discharged home with significant improvement in his motor strength. He remained in clinical follow-up with rheumatologist and neurologist and was treated on an outpatient basis with prednisolone 60 mg/day for 6 months along with methotrexate and infliximab. At his last follow-up at 9 months, he was on tapering dose of prednisone with continued weekly oral methotrexate and eight weekly maintenance infliximab therapy. He maintained neurological improvement in his motor strength except residual left ankle dorsiflexion weakness required an ankle foot orthosis and could ambulate independently with occasional use of walker for stability.

DISCUSSION

Sarcoidosis is a multisystem granulomatous disease usually diagnosed between 20 and 40 years of age; the pathological hallmark of it being the presence of non-caseating epitheloid granuloma.[1] In the US, the adjusted annual incidence is approximately three times higher among individuals of African descent than among individuals of European descent with peak age being between ages 20 and 40 years.[1,5,16] NS was first reported in 1904 and is often a part of systemic sarcoidosis with superimposed neurological manifestations.[4-6,28,39] Isolated NS is uncommon; with that restricted to the spinal cord being exceptionally rare.[9,11-27] When involved, the cervical, thoracic, and lumbar spinal cord is involved in that sequence. Spinal sarcoidosis can present with varied symptoms, including weakness; paresthesias; myelopathy; demyelinating syndrome; facial nerve paralysis; bowel, bladder, or sexual disturbances; and back pain or even radicular pain which are no different from other spinal cord pathologies which are much more common than spinal sarcoidosis.[7,21,35-37]

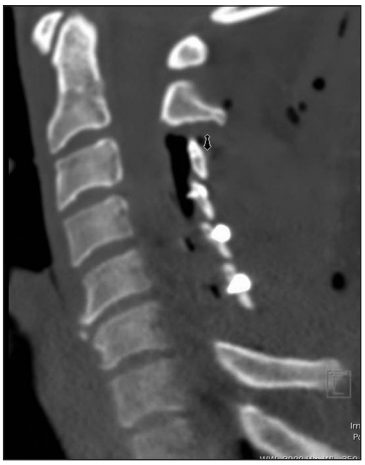

Figure 3: Postoperative sagittal CT spine showing expanded spinal canal and postoperative changes after spinal cord biopsy

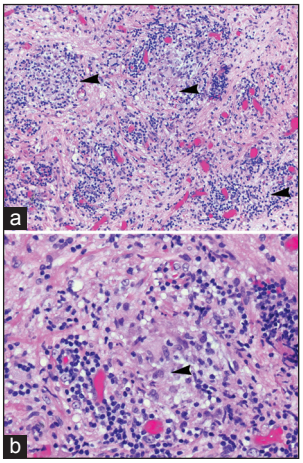

Figure 4: Low power microphotograph of the spinal cord (a) showing non-necrotizing granulomas (arrowheads). High magnification of a granuloma (b) shows central epithelioid cells (arrowheads) surrounded by a lymphocytic infiltrate and lack of caseous necrosis (hematoxylin and eosin, (a) ×10, (b) ×20)

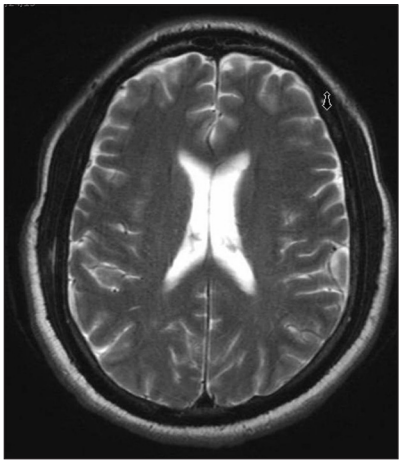

Figure 5: T2-weighted axial MRI of the brain showing no obvious abnormality

Saleh *et al.*,[33] reported eight cases of spinal cord NS of which there was only one case of true isolated spinal cord NS with no other organ involvement which required spinal cord biopsy for diagnosis highlighting the rarity of the disease and role of obtaining tissue diagnosis to unravel this rare disease.[33] Similarly, all the cases in the series by Varron *et al.*, and Sakushima *et al.*, were probable NS as systemic involvement was present in all of them.[31,32] Though spinal sarcoidosis most commonly demonstrates smooth or nodular leptomeningeal enhancement with patchy peripheral cord enhancement because of infiltration of the perivascular spaces; spinal sarcoidosis has a myriad of imaging manifestations from diffuse intramedullary T2 hyperintensity to mass-like intramedullary enhancement.[8,15,18,19,21,23] The imaging findings of spinal sarcoidosis can mimic other disease processes such as neoplasm, demyelinating processes (such as multiple sclerosis (MS)), post-infectious myelitis, spondylotic compressive myelopathy, lymphoma, cord ischemia, and paraneoplastic processes.[8,15,18,19,21,23,30] The lack of characteristic clinical or imaging findings makes the radiological/clinical diagnosis of isolated spinal NS challenging often necessitating obtaining tissue for confirmation of diagnosis.[11-19,23] Often, a noncontrast scan is performed in patients who have myelopathy and the findings are no different from that visualized in cervical spondylotic myelopathy which is way more common than sarcoidosis.[30] This is further compounded by the fact that often these patients have associated cervical stenosis which leads to consideration of CSM and are often subjected to decompressive surgery with no improvement.[23] Disconcordance between clinical symptoms and neurological findings and rapid deterioration in neurologic symptoms should lead to performance of contrast MRI scan to unravel other pathologies like malignancies or spinal cord sarcoidosis. There is no diagnostic feature of NS on imaging, but review of reported cases in the literature and the present case demonstrates that suspicion of sarcoidosis should be high in patients with contagious multisegmental intramedullary T2 signal hyperintensity with combination of leptomeningeal and/or focal patchy intramedullary enhancement restricted to one segment/fewer segments than the T2 signal change without significant cord expansion in patients in the absence of significant stenosis.[11-19] The distinction of NS from MS can be very difficult and although none of the MRI appearance is specific for NS, leptomeningeal and dural enhancement and if present, persistent parenchymal enhancement is very suggestive of NS as compared to MS.[10] Performance of systemic study such as CT of the chest abdomen and pelvis should be considered to reveal any systemic malignancies or other manifestations of sarcoidosis which are often coexistent. Patients with systemic sarcoidosis who have symptomatic nervous system involvement typically come to medical attention as a result of their neurological deficits, rather than because of symptoms referable to other tissues which might be subclinical but may provide other organ systems like lymph nodes to biopsy and confirm the diagnosis.[31,33] Varron *et al.*,[31] suggested performing a ^{18}F-fluorodeoxyglucose-positron emission tomography (FDG PET) to identify occult sites of disease for diagnostic biopsy to avoid biopsying the neural tissue and its associated morbidity as the spinal cord is frequently the first manifestation of systemic sarcoidosis and 3/7 patients in their study had evidence of systemic involvement with sarcoidosis with no obvious manifestation providing extraneural tissue for diagnosis. Whole-body gallium scanning remains a useful indicator of systemic disease, which although relatively nonspecific measure, may be of diagnostic value. Though Ga-67 citrate is taken up by sites of active sarcoidosis and other inflammatory and malignant diseases, like tuberculoma and lymphoma, the pattern of uptake in sarcoidosis is well-recognized.[40,41] Recently, FDG-PET has been increasingly used to unravel systemic sarcoidosis in patients with NS and can detect extraneural involvement for biopsy and diagnosis and is superior to gallium-67 scanning, which has been used in the past for similar purposes, in terms of image resolution (particularly in the CNS) and faster uptake of the radiopharmaceutical agent.[1,5,31] Findings on lumbar puncture may demonstrate raised protein level, pleiocytosis (i.e., increased presence of both lymphocytes and neutrophil granulocytes), and oligoclonal bands with elevated serum and CSF ACE levels. Elevated CSF oligoclonal bands and serum and CSF ACE levels are all supportive of the diagnosis, but due to the varied and nonspecificity of the test results, the sensitivity is very low and the absence of these does not rules out the diagnosis and are neither confirmatory even when positive.[42-45] Further, various series have demonstrated the that leukocytes and ACE levels are not elevated in CSF of spinal cord sarcoidosis patients limiting the diagnostic role of these tests in patients with true spinal cord NS with no other evidence of sarcoidosis.[31,32,42-46]

The diagnosis of isolated NS is challenging. There may be increasing role of performing Ga-67 scan and/or FDG-PET to detect presence of occult systemic sarcoidosis. However, they might be normal in cases of truly isolated spinal NS where performance of spinal cord biopsy has to be done to confirm the diagnosis. Though invasive, the importance of it cannot be overemphasized because there is no pathognomonic feature of NS on imaging and the laboratory finding though useful; are only adjunct.[42-45] Also, though the initial treatment of sarcoidosis is steroid which can be administered on a presumptive diagnosis based on certain imaging features as described above in the appropriate clinical context,[24] not all patients my respond to that or may relapse which might need administration of immunomodulatory and/or immunosuppressive agents as described in the present case and in other cases in the literature, which makes obtaining a tissue diagnosis mandatory. Studies have reported that spinal cord sarcoidosis is one of the steroid-refractory lesions among NS and often requires additional immunosuppressant therapy which suggests that absence of response to steroid does not rules out NS highlighting the importance of biopsy.[38] Most typically, clinical and/or radiological relapse occurs during steroid weaning, despite maintenance on adjunctive immunosuppressants.[5] The present case clearly highlights these important clinical observations. Most neurosurgeons that see these cases rarely are reluctant to biopsy

the spinal cord due to failure to recognize this presentation of NS considering the rarity of this diagnosis and understanding the risks of biopsying the spinal cord. Nevertheless, in the appropriate clinical context, this should be considered given its importance in clinical management. If performed, intraoperative pathological evaluation can be useful as the diagnosis of sarcoidosis can avoid aggressive resection with increased chances of operative morbidity if it can be confirmed during surgery. However, CNS lesions may be smaller and have fewer giant cells within poorly formed granulomas, which may increase the likelihood of a negative/nonspecific intraoperative biopsy result.[21] Also, frozen sections can be mimicking tumor as happened in our case. Since the proposed mechanism by which sarcoidosis becomes intramedullary is via extension through perivascular spaces, the leptomeningeal biopsies may be of value as the arachnoid is often infiltrated with lymphocytes and the typical granulomas increasing the diagnostic yield of frozen section.[37]

The management of NS is based on immunosuppressant therapy, with a goal of suppression rather than cure. Corticosteroids, such as prednisone, are the mainstay of immunosuppressant therapy, with stronger immunosuppressant agents, such as methotrexate, and chlorambucil being reserved for refractory cases of in cases with relapse on steroids.[2,5,8,10] There have been increasing role of immunomodulatory agents like infliximab (monoclonal antibodies to TNF-α); the exact duration and effectiveness is not proven due to the rarity of the disease with only few cases being described in the literature.[2,5,10] The value of surgical intervention is mainly diagnostic and direct appropriate medical treatment and its value cannot be overemphasized due to difficulty diagnosing isolated spinal NS secondary to its rarity and also the lack of characteristic laboratory or radiological findings.

To conclude, intramedullary sarcoidosis is very uncommon, and it is exceedingly rare without systemic sarcoidosis. Its variable imaging appearance and inconsistent clinical manifestations can pose a diagnostic dilemma. There has been some uniformity in radiological presentation of spinal cord NS and although rare, isolated spinal cord NS should be high in the differential diagnosis when evaluating young patients with myelopathy, especially in people of northern European or African American descent. Contrast MRI may help suspecting the presence of NS and in these patients, open biopsy may be indicated if there are no other sites of sarcoidosis on the workup to direct appropriate treatment.

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