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Case Report

Surgical management of intramedullary cervical spinal sarcoidosis complicated by transient unilateral weakness: A case report

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ABSTRACT

Background: Sarcoidosis, a multisystem inflammatory non-caseating granulomatous disease, can present with neurologic lesions in up to 10% of patients.

Case Description: A 57-year-old male presented with three months of worsening upper extremity radicular pain associated with dysmetria, hyperreflexia, bilateral Hoffman's, and positive Babinski signs. The contrast magnetic resonance imaging (MRI) showed a diffuse T2 signal hyperintensity and T1-enhancing 2.5 cm lesion extending sagittally between C4 and C6. The cerebrospinal fluid analysis showed a high protein level and lymphocytic pleocytosis. A cardiac positron emission tomography scan was consistent with the diagnosis of cardiac sarcoidosis. With the diagnosis of multisystemic/probable neurosarcoidosis, the patient was unsuccessfully treated with intravenous methylprednisolone, followed by infliximab. Due to severe cord compression/myelopathy, a C3–C6 laminectomy and C3–C7 posterior spinal fusion were performed. Postoperatively, the patient developed a transient right-sided hemiparesis. Over nine postoperative months, the patient had four relapses of transient repeated episodes of paresis, although follow-up cervical MRI scans revealed adequate cord decompression with a stable intramedullary hyperintense lesion.

Conclusion: Patients with neurosarcoidosis respond unpredictably to surgical decompression and require prolonged medical care, which is often unsuccessful.

Keywords: Cervical intramedullary lesion, Laminectomy, Neurosarcoidosis

INTRODUCTION

Sarcoidosis, a chronic multisystem inflammatory non-caseating granulomatous disease of unknown etiology, can present with neurologic lesions in up to 10% of patients, including intramedullary or extramedullary spinal cord lesions.[12] As patients with spinal sarcoidosis may exhibit inconsistent responses to steroids and other medications, surgery is often warranted. Here, we described a patient with a C4–C6 intramedullary cervical sarcoid lesion and the patient's variable responses to surgical decompression and subsequent medical management.

CASE PRESENTATION

Clinical findings

A 47-year-old African American male with a past medical history of glucose-6-phosphate dehydrogenase deficiency presented with three months of worsening bilateral upper extremity radicular pain/numbness and tingling radiating to both shoulders. On examination, he had bilateral hyperreflexia, bilateral Hoffmann's and Babinski's signs, all reflecting the presence of significant cervical myelopathy.

Radiological diagnosis of sarcoidosis

The cervical magnetic resonance imaging (MRI) demonstrated congenital spinal stenosis and a 2.5 cm intramedullary C4–C6 spinal cord lesion that enhanced with contrast on the T2 image and was diffusely hyperintense on the T2-weighted studies [Figures 1-4]. The abdominal-chest computed tomography and endobronchial ultrasound-guided fine-needle aspiration (i.e., of mediastinal hilar lymphadenopathy) revealed non-caseating granulomas consistent with sarcoidosis. The cardiac positron emission tomography scan also confirmed cardiac sarcoidosis characterized by a patchy abnormal increased signal u in the left ventricle. Further, the cerebrospinal fluid (CSF) protein level was high (>390) [Table 1].

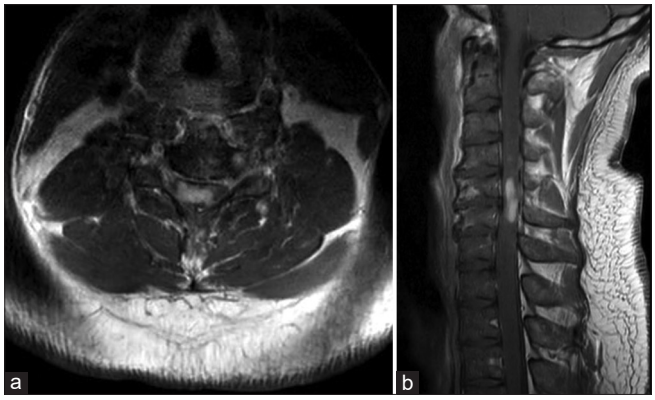

Figure 1: (a) Preop axial T1-weighted magnetic resonance imaging (MRI) of the cervical spine at C5. (b) Preop sagittal T1 weighted MRI of the cervical spine with an enhanced intramedullary spinal cord lesion from C4 to C6.

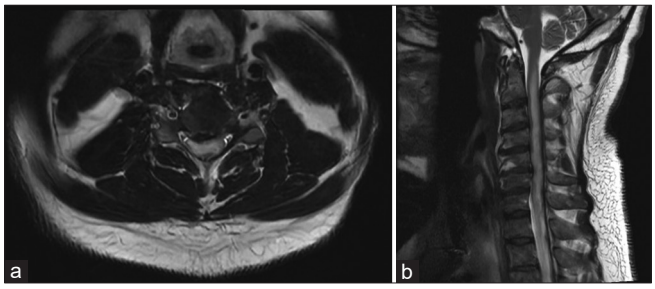

Figure 2: (a) Preoperative axial T2-weighted magnetic resonance imaging (MRI) of the cervical spine at C5. (b) Preoperative sagittal T2-weighted MRI of the cervical spine revealed spinal cord hyperintensity from the corticomedullary junction to the T1 level.

Initial failed medical management

With the initial diagnosis of multisystemic sarcoidosis/probable neurosarcoidosis, the patient was started on daily IV methylprednisolone (dose of 1 g). After developing a left punctate pontine stroke (i.e., resulting in left internuclear

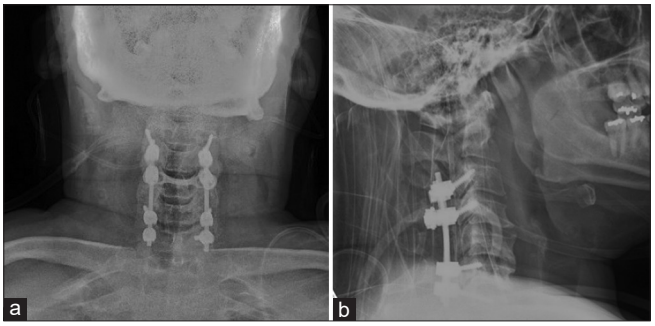

Figure 3: (a) Postoperative C3–C7 fusionanterior–posterior X-ray of the cervical spine. (b) Postoperative C3–C7 fusion lateral X-ray of the cervical spine.

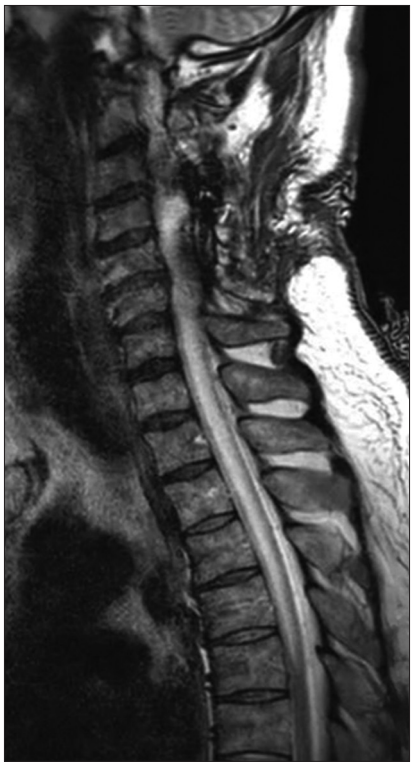

Figure 4: Postoperative mid-sagittal T2-weighted magnetic resonance imaging of the cervical spine at C5.

ophthalmoplegia), vertical nystagmus, and mild left ptosis, he was started on clopidogrel and aspirin. However, as the subsequent contrast brain MRI revealed pachymeningitis, leptomeningitis, and a T2-hyperintense lesion in the pons, the patient was subsequently started on infliximab (5 mg/kg) and referred to neurosurgery with the diagnosis of severe cervical myelopathy secondary to a cervical C4–C6 intramedullary neurosarcoid lesion.

Cervical surgery

To decompress but not biopsy the C4–C6 intramedullary mass, the patient underwent a C3–C6 laminectomy and C3–C7 posterior spinal fusion. Although one day postoperatively, he developed a new right-sided motor hemiparesis that lasted for two weeks; the repeat MRI scan did not show any new cord lesions or increased intrinsic/extrinsic cord compression. Nine months later, the patient experienced four transient relapses of hemiparesis/quadriparesis (i.e., none of which warranted or were treated surgically due to no new MR findings) for which he received varying doses of rituximab, methotrexate, and corticosteroids [Table 2].

Table 1: Patient-specific clinical findings suggestive of neurosarcoidosis.

Clinical findings	Results
Complete blood count	
Hematocrit	46.5%
WBC	9.7 k/uL
Platelets	248 k/uL
Comprehensive metabolic panel	
Calcium	10.3 mg/dL
Glucose	209 mg/dL
Miscellaneous serum	
ACE	15.9 u/L
Tuberculosis	Negative
Cerebrospinal fluid analysis	
Cell count	32/uL
Lymphocytes	28/uL
RBC	65/mm^3
Protein	392 mg/dL
Glucose	79 mg/dL
Flow cytometry	Normal cell populations
Gram stain	1+PMN cell (s)
Culture	No growth
Meningitis/encephalitis panel	Negative
Lymph node biopsy	
Pathology	Non-necrotizing granuloma present
EBUS findings	
Pathology	Benign endobronchial mucosa with submucosal edema and hyalinization. No granulomas seen
Bronchoalveolar lavage	
Mycobacterial culture	Negative
Fungal culture	Negative
Anaerobe culture	Negative
Respiratory culture	Negative

WBC: White blood cell, ACE: Angiotensin-converting enzyme, RBC: Red blood cell, PMN: Polymorphonuclear, EBUS: Endobronchial ultrasound

DISCUSSION

Diagnosis of neurosarcoidosis and systemic sarcoidosis

The diagnosis of neurosarcoidosis is confirmed by laboratory evidence of central nervous system inflammation (i.e., elevated levels of CSF protein and/or cells) and MRI-documented neural lesions (i.e., T2 enhancement of

Table 2: Summary of clinical events experienced by the patient.

Post-operative period	Findings
Hospitalization period	
Clinical course	Transient right-sided hemiparesis that subsided progressively over 2 weeks. Started on Infliximab and underwent intensive physical therapy.
Imaging	No acute intracranial process or large vessel occlusion was detected on repeat brain MRI. No additional lesions on the cervical spine MRI.
2 Weeks post-operation	
Clinical course	Worsening left-sided hand weakness. Started on corticosteroids and given an additional dose of Infliximab.
Imaging	Stable postoperative changes. Stable multifocal cervical spinal cord enhancement.
1 Month post-operation	
Clinical course	New left-sided hemiparesis that improved following corticosteroids
Imaging	Stable postoperative changes. Stable multifocal cervical spinal cord enhancement.
2 Months post-operation	
Clinical course	Tetraparesis, hyperreflexia, and torsional nystagmus with an anti-infliximab titer and undetectable infliximab activity. Discharged on rituximab, methotrexate, and corticosteroids.
Imaging	No updated imaging was obtained.
7 Months post-operation	
Clinical course	Upper extremity paresis following corticosteroid taper. Discharged with near resolution of symptoms on discharge.
Imaging	Adequate mechanical decompression with stable hyperintensity of the cervical spinal cord. Worsening periventricular enhancement.

MRI: Magnetic resonance imaging

Table 3: Summary of the literature on cases of intramedullary cervical neurosarcoidosis treated with decompression and no biopsy.

Study (Ref No.)	Age, gender	Presentation	Surgical procedure	Postoperative clinical status
Our study	47/M	C3–C7 radiculopathy and myelopathy: Dysmetria, hyperreflexia, and positive Babinski and Hoffmann	C3–C6 laminectomy+C3–C7 posterior fusion	Transient hemiplegia with improvement followed by multiple episodes of relapse.
Nurboja *et al.*[10]	43/F	C4–C6 radiculopathy+myelopathy: Bilateral hand paresthesia, hyperreflexia and spastic gait	C3–C7 laminectomy with mass screws	No improvement in clinical symptoms
Oe *et al.*[11]	56/F 58/F 59/M	Cervical myelopathy: Bilateral hand paresthesia and weakness, bilateral leg weakness, gait difficulties, hyperreflexia –56F with additional urination difficulties	Laminoplasty Case 1: C3–7 Case 2: C2–T1 Case 3: C3–5	• Case 1: No clinical deterioration in symptoms but a marked reduction in MRI findings Case 2: Mild improvement of hand function, but worsening gait and lower limb symptoms; unable to stand at 3 years • Case 3: Mild deterioration in hand function with unchanged gait; no reduction in signal intensity
Mathieson *et al.*[7]	56/F	Cervical myelopathy: Bilateral lower extremity paresthesia, paraparesis, urinary incontinence, hyperreflexia, bilateral ankle clonus	C5–C7 laminectomy	Persistent paresthesia at 6 months, but strength returned to normal
Kwon *et al.*[5]	56/F	Bilateral hand paresthesia and hypoesthesia, gait disturbance 3 months postoperatively	Symptoms appeared 3 months after C3–C7 Laminoplasty -Isolated Biopsy performed on readmission	Despite steroid therapy at 3 months post-biopsy, weakness worsened, and urinary incontinence appeared. 6 months after biopsy, mild improvement of motor symptoms and gait
Jefferson[3]	66/F (case 7)	Focal epileptic seizures of L side, Horner's syndrome, progressive hemiparesis and dysesthesia, hyperreflexia, spasticity	Laminectomy No biopsy was done during the procedure, but done for the autopsy	Resolution of pain but persistent hemiparesis. Died 1 week postoperatively following a seizure

F: Female, M: Male, C: Cervical, T: Thoracic, MRI: Magnetic resonance imaging.

meningeal or parenchymal tissues). In addition, patients may have evidence of systemic sarcoidosis and demonstrate cardiopulmonary, dermatologic, or ophthalmologic symptoms/signs and/or elevated inflammatory laboratory markers reflective of sarcoidosis requiring further medical management.[12,15]

No consensus regarding pharmaceutical management of neurosarcoidosis

There is presently no consensus regarding the pharmaceutical management of neurosarcoidosis. The first line of treatment is typically the administration of corticosteroids (i.e., varying dosage, duration, and method). However, if this fails, alternative immunomodulatory therapies are offered (i.e., tumor necrosis factor alpha inhibitors, infliximab). For instance, in Moravian and Segal's series, seven patients with glucocorticoid-refractory neurosarcoidosis responded favorably to infliximab.[9]

Surgery

A decline in neurological function has been reported following extensive decompressive surgeries, underscoring the unpredictable course of neurosarcoidosis.[4,7] Patients with cervical neurosarcoidosis variably respond to surgical biopsy, decompression alone, laminoplasty alone, or laminectomy with fusion in addition to preoperative/postoperative corticosteroids and immunotherapy.[1,2,4,6,7,10,11,13,14] In one study, 4 of 12 patients with cervical neurosarcoidosis improved for two months following cervical decompression alone (i.e., without biopsy); notably, preoperative non-myelopathic patients had better outcomes.[13] Another series also successfully managed intramedullary neurosarcoidosis/cervical stenosis with decompression only.[10] In a review study, 50% of patients undergoing decompressions without biopsy worsened postoperatively versus a lower 28% in worsening after laminectomy with biopsy.[7] Furthermore, cervical laminoplasty without biopsy performed for neurosarcoidosis

also resulted in new postoperative neurologic deficits that failed to respond to corticosteroids.[11] Our patient, following a decompressive cervical laminectomy without biopsy, developed fluctuating postoperative neurological deterioration that was not controlled/mitigated by the administration of corticosteroids and/or immunotherapy. Posterior instrumented fusion was performed to preserve the cervical alignment and avoid post-laminectomy kyphosis.[8] Table 3 summarizes the literature of intramedullary cervical neurosarcoidosis treated with decompression without biopsy.

CONCLUSION

Patients with neurosarcoidosis may respond unpredictably, with marked fluctuations/exacerbations and remissions, to either medical (i.e., primarily to corticosteroids and secondarily to immunotherapies) and/or surgical management (i.e., decompression with/without fusion).

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflict of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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