**Intradural Extramedullary Sarcoidosis**  
**Case report and review of literature**

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Spinal sarcoidosis represents a rare subgroup of neurosarcoidosis. Most spinal sarcoid lesions are intramedullary, and only a few cases of Intradural Extramedullary (IDEM) sarcoidosis have been reported till date.

A thirty years-old female patient with intradural extramedullary sarcoid lesion in the cervico-dorsal spinal canal (C7-D1) without any systemic involvement of sarcoidosis is described. The patient presented with rapidly progressive compressive myelopathy with bladder involvement. She was treated with surgical removal of the lesion coupled with the administration of steroid and showed marked improvement.

Keywords: spinal cord, sarcoidosis, intradural extramedullary, cervico-dorsal, laminectomy

**Introduction:**

Sarcoidosis is a rare manifestation of systemic disease, characterized by a non-caseating, granulomatous infiltration.

The presenting symptoms of Sarcoidosis of the nervous system are either of intracranial origin, primarily related to leptomeningeal involvement or, of peripheral neuropathic origin. However, the disease may affect any part of the nervous system. The overall incidence of neurological involvement ranges from 4% to 17%.¹,² Douglas and Maloney, in a survey of 500 patients from the Edinburgh Sarcoidosis registry, found only 6 cases with neurological involvement in 1973.³

Spinal cord sarcoidosis is a rare condition, that is difficult to diagnose and intradural extramedullary mass formation is even rarer.⁴

Involvement of the spinal cord including meninges in sarcoidosis is about 1% clinically. It can present as three entities: Multiple intramedullary (IM) lesions with focal arachnoiditis; large IDEM tumors with marked mass effects with focal neurologic deficits; or, as an extradural (ED) mass from sarcoid infiltration of the spinal cord and dura.⁵

In 1978, Baruah et al. found only 4 cases of histologically proven sarcoidosis of the cervical canal that had presented as mass lesion.⁶ Schaller B. et al. in 2006, found 8 cases of IDEM sarcoid lesions in the literature before publishing their case report.⁷

We are reporting another case of IDEM sarcoid lesion that, presented with features of compressive myelopathy, at the level of C7-D1 mimicking a meningioma with no other known evidence of the disease elsewhere.

**Case note:**

A 30 years-old lady presented with one month history of progressive weakness and stiffness of her both lower limbs along with occasional neck pain without any radiation. Within 2 weeks of the initial symptoms she became completely bed ridden with difficulty on bladder evacuation. There was history of irregular flexor spasms of her both lower limbs especially at night. She also felt occasional girdle pain sensation around her upper chest region for the last 2 weeks.

Her past and family histories were not significant.

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**MRI of the spine revealed an IDEM mass iso-intense in T1, mixed intensity in T2 and readily enhanced with contrast with a dural tail sign, almost encircling the cord at C7/D1 region suggesting a meningioma.**

Her higher mental function and cranial nerves were normal on examination including the fundi. She was completely bed ridden on admission. Both lower limb power were 1/5, hypertonic without any wasting or, fasciculation. All lower limb jerks were exaggerated, plantars were extensor in response. Abdominal reflexes were absent in all quadrants and pin prick & temperature sensations were impaired from D4 dermatome downwards (including peri-anal region) bilaterally; left side was more affected than the right. Both upper limbs were normal on examination except both radial jerks that were depressed. Bladder had been catheterized due to acute retention of urine. She was also constipated.

Routine examinations didn’t show any abnormality including chest X-ray.

She underwent C6 – D1 laminectomy in prone position. Dura was tense and not pulsating. After opening the dura, the arachnoid was found to be adherent to its inner surface and appeared thickened. A grayish gritty mass was encountered, which was almost encasing the cord except the right lateral portion. The mass differed in consistency from that of a meningioma as suspected preoperatively from MRI imaging. A gross total removal of the mass could be achieved under operating microscope and few bits of the tissue, adherent to the anterior dura were cauterized and had been left to avoid injury to the neural structures. She had been on intravenous Dexamethasone (4mg, 6hr.ly) in a tapering dose from the first postoperative day, and showed

**Figures:** MRI of the patient (T1, T2 and T1 with Gd-DTPA) Pre-operative

Histological preparation of the lesion - showing chronic granulomatous lesion with chronic inflammatory cells and Langhan’s giant cells consistent with Sarcoidosis (Hematoxylin & eosin staining)
rapid steady progressive improvement of the neurological deficits including sphincters.

Histopathology of the resected tissue showed a non-caseating granulomatosis consistent with sarcoidosis. Postoperative screening didn’t reveal any systemic involvement of sarcoidosis.

She was discharged on the 10th postoperative day with minimal residual weakness & stiffness of her both lower limbs without any sphincter disturbance. On lieu of the histology report she had been advised to continue on oral steroids.

She became absolutely normal at her first follow up (4 weeks later) and steroid was tapered off. After 6 months, patient again developed difficulty on walking. Serum Angiotensin converting enzyme (ACE) was raised this time and other tests were negative. Patient rapidly improved with oral steroid preparation and didn’t need any hospital admission. At her last follow up (one year) she had no complaints except increase in her body weight.

Discussion:

Neurological lesions in sarcoidosis were first reported by Winkler in 1905.8

In 1941 Longcope documented the spinal sarcoidosis at autopsy9. Since 1941, about 22 cases of sarcoidosis (proven by either biopsy or, autopsy) affecting the spinal cord including cauda equina at various sites had been described till 1978.10,11,12,13

IDEM mass formation in sarcoidosis is a rare event and also difficult to diagnose. Most of the cases are intramedullary.4,7

Typical presentation is progressive, painless limb weakness (according to the involvement of the cord segment). Thoracic spine is the most common site of affection. Serum and CSF levels of angiotensin converting enzyme (ACE) can be followed to assess the progression of the disease.11,14

In our case, the area of affection was in the cervico-dorsal region and the serum ACE level was not significantly raised initially.

Dense adhesions, arachnoiditis and multiplicity of lesion make complete surgical removal often impossible, even with the use of operating microscope.11,15 Sometimes IDEM lesions can be removed totally4,7. We could achieve a near total resection. Decompression of the granuloma coupled with the administration of corticosteroids, is known to be an effective treatment in this disease.6,10,11,14 However, several other factors influence the overall prognosis in a given case: degree of dissemination, multiple sites of cord involvement and secondary vascular changes producing further cord damage.

In MRI, the lesion is usually iso-intense in T1, hypo or hyper-intense in T2 and also enhanced by Gd-DTPA. Preoperative impression thus sometimes makes diagnosis a bit confusing. Meningioma4, schwannoma15, even glioma12,16 were considered the likely diagnosis at or before the operation.

Authors suspected the mass to be a meningioma in the present case but, the mass differed in consistency from that of a meningioma peroperatively.

Spinal cord compression in sarcoidosis may result not only from mass lesions but also from arachnoiditis and meningeal thickening, as demonstrated by biopsy or autopsy findings.17,18,19 The cord changes may vary, ranging from localized necrosis to atrophy or, even replacement of wide segments by homogenous, firm, grey, gelatinous tissue.12 In our patient the cervico-dorsal cord showed no change other than mild pallor, but the arachnoid was thick and was adherent to the inner surface of the dura in few places.

Literature review suggests that, in sarcoidosis of the nervous system, the prognosis is better when there is no widespread systemic involvement20,21, although an excellent recovery was also achieved by some authors even in presence of systemic involvement.7

Postoperative screening didn’t reveal any systemic involvement in our patient, though she developed recurrence of symptoms six months after the operation. This time, the serum ACE was raised. Probably, at the first time the enzyme level was suppressed, as the patient was getting steroids.
Spinal cord sarcoidosis is a rare condition that is difficult to diagnose. It is important to remain open to the possibility of spinal cord sarcoidosis when a mass is detected in the spinal canal. It can progress rapidly and relentlessly, and carries a serious prognosis if left untreated. IDEM location is a much more rare entity and resection is much easier than intramedullary lesions. Positive histological examination is required to establish the diagnosis as other diagnostic tests are unspecific. The natural history of spinal sarcoidosis is remission and relapse; and corticosteroids are the cornerstone of continuing medical treatment. Evidence of systemic sarcoidosis has to be excluded meticulously to institute appropriate treatment for this rare disease.

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