Extramedullary Spinal Neurosarcoidosis: Report of Two Cases

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Dear Sir,

Sarcoidosis is a chronic systemic disorder of unknown etiology characterized in affected organs by an accumulation of epithelioid granulomas without caseation or staining for infectious agents and derangement of the normal tissue architecture [1]. These granulomas often incorporate multinucleated giant cells and lymphocytes. In the past decade, there has been significant progress in our understanding of the immunopathogenesis of the disease, but the etiology of this enigmatic condition still eludes us.

Clinical neurological involvement occurs in approximately 5% of patients. However, autopsy results suggest that subclinical involvement may be present in up to 25% of patients. Neurosarcoidosis is a great mimicker and an uncommon presentation of sarcoidosis; it is therefore a diagnostic challenge, especially when there is no prior history of systemic sarcoidosis [2–8].

A typical imaging feature is thickening and enhancement of the basilar leptomeninges of the brain. Other imaging findings, such as enhancing or nonenhancing parenchymal lesions, dural and bone lesions may occur in the head and spine.

Spinal sarcoidosis is a rare condition, whose natural history and therapeutic outcome are still not fully known, and extramedullary extradural mass formation is even rarer. We describe 2 additional cases of spinal extramedullary sarcoïd lesions and discuss the diagnostic approach and therapeutic management of this condition.

Case Reports

Patient 1. An 80-year-old woman was admitted to our department for a sudden onset of a sensorimotor deficit in her right upper and lower limbs; she also complained of neck pain radiating into her right shoulder. The patient presented a sarcoidosis of the lung with hilar and mediastinal lymphadenopathy, and anterior uveitis, diagnosed in 1994. Sarcoid pathology of noncaseating granuloma with giant cells was detected in the lung biopsy specimen. She was treated with oral steroids for 1 year; for the last 10 years there had been no signs of disease.

Neurological examination revealed right-sided hemiplegia and severe hypesthesia over the left side. The plantar response was extensor on the right side. Computed tomography of the brain and cervical spine was normal; magnetic resonance imaging (MRI) of the cervical spinal cord revealed a mass that was extramedullary in the right lateral aspect of the spinal canal at the level C2–C3 and compressing the spinal cord (fig. 1 a–c). Surprisingly the plasma level of angiotensin-converting enzyme (ACE) was low (3 IU/l). An intravenous corticosteroid therapy (250 mg/day methylprednisolone for 5 days) was started, followed by a 40-mg daily dose of oral prednisone (approx. 1 mg/kg per day), then gradually tapered to 5 mg/day over 2 months. A dramatic improvement was seen on follow-up MRI (fig. 2 a, b); neuroradiological findings corresponded to this clinical improvement.

Patient 2. A 52-year-old woman presented with a 2-month history of progressive back pain, numbness and weakness in her lower extremities. Difficulty in micturition was noted. Neurological examination revealed right-side dominant spastic paraparesis and a left-side dominant decrease in sensation to pain and touch below the level of T4. Vibration and position senses were also disturbed, especially on her right side. Deep tendon reflexes were hyperactive on the lower limbs, especially on the right side. Plantar responses were extensor on both sides.

No evidence of systemic illness was noted concomitant with the patient’s onset of neurological symptomatology.

MRI revealed an extramedullary and extradural lesion extending at the level T2–T6 and encasing the dorsal cord from the anterior, posterior and right lateral aspects, mimicking an en plaque meningio-
appropriate testing did not reveal systemic involvement of sarcoidosis. ACE was 22 IU/l (normal limit 20 IU/l). The cerebrospinal fluid ACE level was elevated (4.8 μmol/l/min).

A high-dose methylprednisolone therapy (1 g/day for 5 days) was given, followed rapidly by almost complete neurological recovery; successively a daily dose of 60 mg prednisone (approx. 1 mg/kg per day) was orally administered for 7 days, then reduced to 20 mg daily.

The plasma ACE level decreased to a normal level of 11 IU/l. Follow-up MRI examination after 1 year did not show any abnormalities.

Discussion

The cases that we described are 2 examples of the protean and difficult-to-recognize presentation that neurosarcoidosis may adopt. Neurosarcoicosis has a variable expression, which tends to mimic other neurological diseases. Neurological manifestations occur usually as a part of the spectrum of the systemic disease; the diagnosis is based on the documentation of systemic sarcoidosis in the absence of other neurological diseases.
Our report confirmed the role of MRI in diagnosing intraspinal sarcoidosis, but the findings are nonspecific and must be considered with the clinical course of the patient in arriving at the correct diagnosis. Corticosteroids are commonly used to treat systemic sarcoidosis; however, the effectiveness of corticosteroid therapy for spinal cord sarcoidosis has not yet been established. It has been previously reported that, in patients in whom initial corticosteroid therapy was delayed, MRI findings were dramatically improved after therapy, while the clinical course was not favorable [16, 17]. In agreement with these findings, the use of intravenous cortisone has resulted in an excellent outcome for both patients who started the therapy in the relatively early phase. Additional immunosuppressive drugs were also not necessary.

It can be hypothesized that, after the therapy, inflammation abates and granulomas disappear, but irreversible changes, such as gliofibrosis of the lesion and destruction of the neural pathways, may lead to prolonged myelopathic symptoms. Therefore, corticosteroid therapy should be initiated as early as possible before the spinal cord parenchymal damage becomes irreversible, to enhance the chance of a complete recovery of the spinal cord.

Unlike intramedullary sarcoid lesions, an extramedullary lesion can be totally removed. However, our report indicates that, in the unusual case of spinal extramedullary sarcoidosis, corticosteroid therapy should be considered as the first therapeutic option and has to be recommended prior to aggressive surgical management involving the removal of the extradural mass. To conclude, it is important to consider and establish the diagnosis of neurosarcoidosis as early as possible, because early recognition of this condition may lead to subsequent immediate and appropriate treatment with steroids.
Fig. 3a–d. T1-weighted sagittal, T2-weighted sagittal, gadolinium-enhanced DTPA T1-weighted sagittal and T1-weighted axial magnetic resonance images from patient 2 show an extramedullary and extradural lesion extending at the level T2–T6 and encasing the dorsal cord from the anterior, posterior and right lateral aspects, mimicking an en plaque meningioma.
References