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Case Report

Longitudinal Myelopathy in a Child with Systemic Lupus Erythematosus (SLE) – A rare case report and review of the literature

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Abstract:

Systemic lupus erythematosus (SLE) is a chronic, multisystem autoimmune disease with a highly variable clinical course. Myelopathy is one of the neuropsychiatric lupus syndromes. Although infrequent, it is manifested severely, leading to motor and sensory deficits, and sphincter dysfunction. The exact pathogenesis is still unknown, but may be related to arterial thrombosis and vasculitis. Diagnosis is based on clinical findings, laboratory tests and the use of Magnetic resonance imaging. Here we discuss the clinical presentation, the magnetic resonance imaging findings, and other relevant laboratory studies of this rare but serious complication of systemic lupus erythematosus.

Keywords: *SLE, children, Longitudinal myelopathy, MRI, NPSLE.*

Introduction

Systemic lupus erythematosus (SLE) is a chronic autoinflammatory disease that affects multiple systems. It is characterized by hyperactive autoreactive B and T cells, and immune complex induced inflammation.[1] Approximately 20% of all cases of SLE occur in the pediatric age group.[2]

American College of Rheumatology (ACR) defines 19 neuropsychiatric syndromes (NPSLE) associated with SLE.[3] The syndromes can affect up to 80% of patients with SLE with headache, cognitive dysfunction, seizure being the most common.[4] Myelopathy is one of the least common NPSLE which have been reported in only 1-2% of patients.[5] It is particularly rare in children with only a few cases reported worldwide.[6-10]

Transverse myelopathy is a process involving the gray and white matter in one or more adjacent spinal cord segments. Although rare, it has a dramatic and severe manifestation leading to rapidly involving motor and sensory deficits and sphincter dysfunction.[3] The exact pathogenesis is still unknown. But it may be related to vasculitis and arterial thrombosis.[11] Diagnosis is based on clinical findings, laboratory tests, and the use of gadolinium-enhanced magnetic resonance imaging (MRI). The standard therapy is the combination of intravenous glucocorticoids and cyclophosphamide.[12] The SLE-related myelopathy may often become challenging to clinicians due to its broad differential diagnosis and rapid initiation of immunosuppressive therapy.

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Our reported patient was a girl of 12 years with lupus myelopathy. Herein we discussed the clinical presentation, MRI findings, and other relevant laboratory test results of the reported case. The literature concerning SLE-related myelopathy was also reviewed.

Table 1 shows Diagnostic Criteria for Myelitis in Systemic Lupus Erythematosus.

Diagnostic criteria for myelopathy in SLE proposed in 1999 by the American College of Rheumatology (ACR)[3]
Sudden onset (days or hours) of the following signs/symptoms
Bilateral weakness of lower limbs that may or may not include upper limbs (paraplegia or tetraplegia). It can be asymptomatic
Change in sensory sensitivity corresponding to a motor impairment, with or without intestinal-bladder dysfunction
Exclusion criteria
Compressive spinal cord lesion (e.g., disc prolapse)
Cauda equina syndrome

Table 2 shows nineteen neuropsychiatric syndromes (NPSLE) associated with SLE divided in two groups[3]

Central NPSLE	**Peripheral NPSLE**
Aseptic meningitis	Guillain Barre syndrome
Cerebrovascular disease	Autonomic neuropathy
Demyelinating syndrome	Mononeuropathy
Headache	Myasthenia gravis
Movement disorder	Cranial neuropathy
Myelopathy	Plexopathy
Seizure disorders	Polyneuropathy
Acute confusional state	
Anxiety disorder	
Cognitive dysfunction	
Mood disorder	
Psychosis	

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Case Presentation

A 12 year old girl presented with fever for 12 days which was low grade and continued in nature; not associated with chills and rigor. Two days later she developed weakness of all limbs and urinary retention. She became unable to walk within 24 hours of developing weakness. On further query, she gave a history of generalized body ache, joint pain, recurrent fever, and a discoid itchy rash all over the body for the last 12 months. She faced frequent oral ulcers and rash on her face which was photosensitive. The child gave no history of trauma to the spine, back pain, seizure, visual disturbances, abnormal behavior and movement, headache, or the same type of attack previously. She is the first issue of her nonconsanguineous parents and her antenatal, natal, and postnatal periods were uneventful. Her development was age-appropriate and now she is a student of class five. On examination, the girl was anxious, mildly pale, her vitals were within normal limit except temperature which was 100^0 F. We noticed oral ulcer, malar rash, and multiple discoid, scaly rashes all over her body. Her higher psychic function and cranial nerves were intact. She had quadriparesis evident by hypotonia, decreased muscle power (MRC 3/5 in upper limb, 0/5 in both lower limbs), upper limbs hyporeflexia, and lower limbs areflexia without any definite sensory level. None of the joints were swollen or tender.

Laboratory results showed leukopenia of 3×10^9/L, microcytic hypochromic anemia, and an elevated ESR of 90 mm/h. Total platelet count, serum electrolytes, liver, and renal function tests were within normal range. CSF findings revealed a normal opening pressure, mild leukocytosis (16 cells/mm^3), and mildly elevated protein (0.9 g/l). MRI of the cervico-dorsal spine revealed diffuse enlargement of cervical cord in T1 sequence (Figure 1) and increased signal intensity images in the T2 sequence with longitudinal involvement of the spinal cord from C3 to T1 levels (Figure 2). Axial T2-weighted images of the spinal cord at C4 level showing intramedullary multiple increased signal lesions. We also performed additional laboratory testing, which revealed low C3, C4 levels, positive antinuclear (ANA), and anti double stranded DNA (anti-dsDNA). The patient was treated with intravenous methylprednisolone (30 mg/kg/day as a single daily dose for five days). After this, she was maintained with 1 mg/kg daily oral prednisone. She was gradually improving her spinal shock and developed spasticity without any improvement in her bladder function. Thereafter, further treatment strategies were settled with the consultation of the department of pediatric rheumatology.

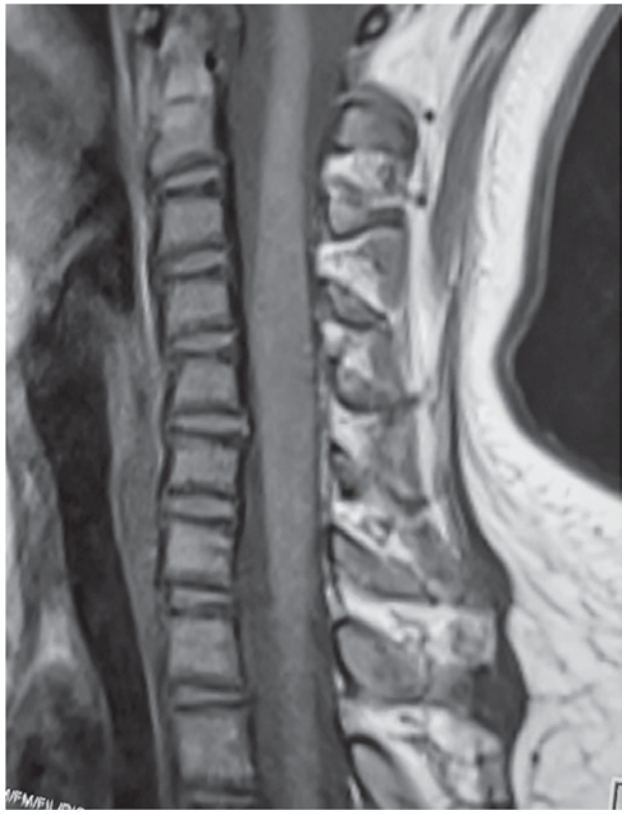

Figure 1:

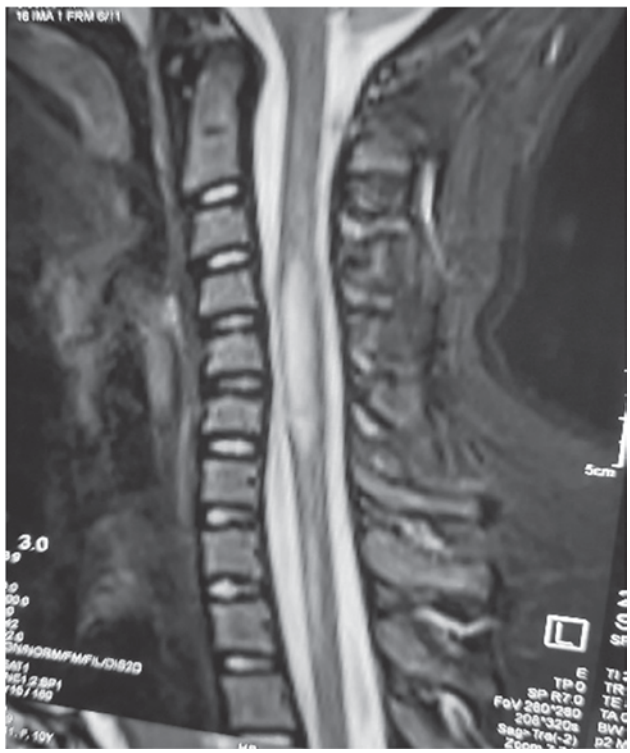

Figure 2:

Discussion

SLE-related myelopathy was first described in 1959 as a rare complication of systemic lupus erythematosus and is particularly rare in children with only 13 cases reported in patients younger than 18 years of age, mostly occurring in adolescents. [6-10] Published reports stated that lupus myelopathy had a stereotyped presentation in children. There were a variable time of onset of the systemic disease, acute flaccid paralysis, inflammatory cerebrospinal fluid profile with pleocytosis, positive antiphospholipid antibodies, extensive abnormality of spinal cord signal on MRI, and a variable prognosis for recovery.[7-10] The differential diagnosis of transverse myelopathy in patients with SLE includes idiopathic transverse myelitis, viral infection, medullar compression due to vertebral fractures, epidural or subdural lipomatosis, epidural or paraspinal abscess complicating disc space infection, and atlantoaxial subluxation.[13]

We considered that our patient's disease was systemic lupus erythematosus complicated with myelopathy. Although a prolonged febrile disease with a rash and limb pain could be caused by several viral infections; typical photosensitive malar rash, joint pain, and a discoid scaly itchy rash all over the body raised the suspicion of SLE. The positive ANA, anti-dsDNA, and depressed C3 and C4 serum levels favoured the diagnosis of SLE.

MRI is the preferred tool in the evaluation of spinal cord pathology including SLE-related myelopathy. Two subtypes of myelopathy are seen in patients of SLE.[12] Firstly, the more prevalent one is transverse myelopathy involving one level of the spinal cord. Secondly, longitudinal myelopathy in which more than four levels of the spinal cord are affected, either continuously or separately.[14] The most frequent presentation of patients with transverse myelopathy consists of paraesthesia (usually at levels T5-T8), which can be accompanied by paraparesis and/ or sphincter dysfunction. A small subset of patients may have normal sensory functions.[15] The reported MRI experience in patients with SLE-related myelopathy is heterogeneous and includes normality, cervical cord enlargement, and increased signal intensity in the T2-weighted images at the thoracic segments.[15-17] Deodhar et al[18] reported a case with continuous involvement of the spinal cord from C3 to T2 levels and from T7 to the conus medullaris and they described this as the first case report of a 'longitudinal myelitis'. The longitudinal involvement was also found in a case series of 6 patients.[19] Our patient had longitudinal myelitis with urinary retention. But sensory function was intact. Although the pathogenesis of myelitis is poorly understood, literature indicate that longitudinal myelitis might be related to either a vascular occlusive phenomenon of the spinal cord or to a direct interaction between anti phospholipid and spinal cord phospholipids.[19,20]

CSF examination and cultures should be performed to exclude infectious diseases. In 50-80% of NPSLE patients may have mild, non-specific abnormalities (e.g. pleocytosis, elevated protein levels, and decreased glucose levels).[12].Our case showed pleocytosis and mildly elevated proteins in CSF analysis. However, these findings may also be observed in other diseases.

Whenever SLE-associated myelopathy is suspected, treatment with immunosuppressive drugs should be initiated. The literature reviewed that initial treatment should consist of intravenous methylprednisolone for three consecutive days followed by intravenous cyclophosphamide (750 mg/m2). This regimen should be followed by monthly infusions of cyclophosphamide for 6-12 months, combined with oral prednisolone 1 mg/ kg for three months, which should be tapered afterward according to disease activity.[15,21] Our case was treated by intravenous methylprednisolone followed by oral prednisolone. She showed clinical improvement after the start of immunosuppressive therapy but still had a grade IV paresis in the lower extremities at discharge. A monthly infusion of cyclophosphamide was planned further. The overall outcome of patients with SLE-associated myelopathy seems to be favourable. Complete recovery occurs in up to 50% of the cases, whereas 21-31% of the patients show no clinical improvement or deterioration after immunosuppressive therapy.[15]

In conclusion, we have presented a case of SLE-related myelopathy which can be classified as longitudinal myelitis. The clinical features and laboratory findings are similar to those presented by acute transverse myelitis patients. Diagnosis is often challanging due to the broad differential diagnosis and lack of disease-specific findings. Aggressive immunosuppressive therapy should be initiated early in the course of the disease for better outcome. SLE-related myelopathy should always be considered in the differential diagnosis of transverse myelitis.

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