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Case Report

Longitudinally Extensive Transverse Myelitis in a Systemic Lupus Erythematosus Patient

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Abstract

Longitudinally extensive transverse myelitis (LETM) is a rare and devastating type of Transverse myelitis (TM) that involves at least three contiguous vertebrae on T2-weighted magnetic resonance imaging (MRI). It occurs in different infectious, traumatic and autoimmune diseases such as Systemic lupus erythematosus (SLE). Herein, we report a case of lupus flare with a severe outcome as LETM with literature review. A 32-year-old woman with eleven-year-history of well controlled SLE, was hospitalized in our department with chronic fever evolving for two months and appearing 48 hours after uncomplicated vaginal delivery. The patient experienced tetra paresis with paresthesia and urinary retention. MRI of the spine showed intramedullary T2–weighted hyper-intense signals extended longitudinally to more than 9 vertebral segments and enlargement of the spinal cord, suggestive of LETM. Cerebral MRI in axial flair image showed nonspecific bilateral fronto-parietal white matter hyperintensities. The patient received methylprednisolone 1 g/day for 5 days, relayed by oral prednisolone 1 mg/kg/d and cyclophosphamide (1g/month) during 6 months with functional physiotherapy. The evolution was marked by a complete regression of paresis and paresthesia in limbs and urinary sphincter disorder.

Keywords: Longitudinally Extensive Transverse Myelitis; Transverse Myelitis; Systemic Lupus Erythematosus

Introduction

Systemic lupus erythematosus (SLE) is an autoimmune inflammatory disease with multisystemic involvement. Its clinical presentation varies widely from mild muco-cutaneous manifestations to multiorgan and severe central nervous system involvement.

More than half of SLE patients will present neuropsychiatric SLE (NPSLE) symptoms during the course of their disease [1,2]. The presentations of NPSLE may range from mild manifestations to acute life threatening involvements. Myelitis is a very rare manifestation of SLE and occurs in 1–2% of patients [3,4]. Transverse myelitis (TM) is an inflammatory lesion of the spinal cord, causing significant morbidity and disability. It occurs in different infectious, traumatic and autoimmune diseases such as SLE. Longitudinally extensive transverse myelitis (LETM) is a rare and devastating type of TM that involves at least three contiguous vertebrae on T2-weighted magnetic resonance imaging (MRI) [4]. Herein, we report a case of lupus flare with a severe outcome as LETM with literature review.

Case Presentation

We report the case of a 32-year-old woman with eleven-year-history of well controlled SLE. When the diagnosis was originally made, at the age of 20-year-old, she had cutaneous lesions, polyarthritis, pleurisy, positive immunologic tests, low complement levels (C3 and C4) and hemolytic auto-immune anemia. She was treated with steroids and hydroxychloroquine.

In May 2022, she was hospitalized in our department with chronic fever evolving for two months and appearing 48 hours after uncomplicated vaginal delivery. On examination, the patient had a body temperature at 39.5 °C, with stable vital signs.

The patient experienced tetra paresis with paresthesia and urinary retention. MRI of the spine showed intramedullary T2–weighted hyper-intense signals extended longitudinally to more than 9 vertebral segments and enlargement of the spinal cord, sug-

gestive of LETM (Figures 1, 2). Cerebral MRI in axial flair image showed nonspecific bilateral fronto-parietal white matter hyperintensities (Figure 3).

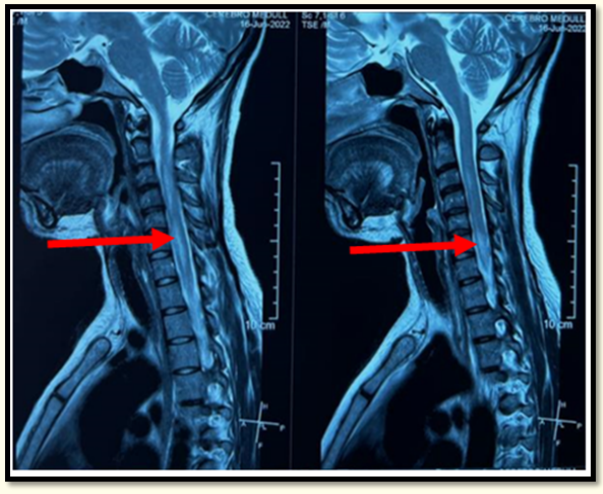

Figure 1: Spinal cord MRI in sagittal T2 weighted image, showing enlargement with increased signal intensity extended longitudinally to more than 9 vertebral segments.

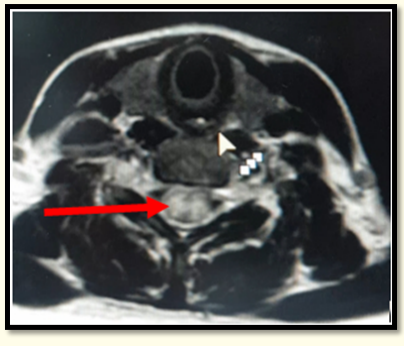

Figure 2: MRI of the spinal cord, in axial T2 weighted MRI, showing increased signals extended transversally to more than 50% of the large diameter of the spinal cord.

The lumbar puncture revealed increased leukocyte count of 150 (normal range: 0–5/mm^3) with 56% of lymphocytes and 44 % of neutrophils, hypoglycorrhachia (*Cerebrospinal fluid*/serum glucose ratio =4), a total protein level of 0.91 (normal 0.18–0.58 g/L) and chloride was within normal ranges (Table 1).

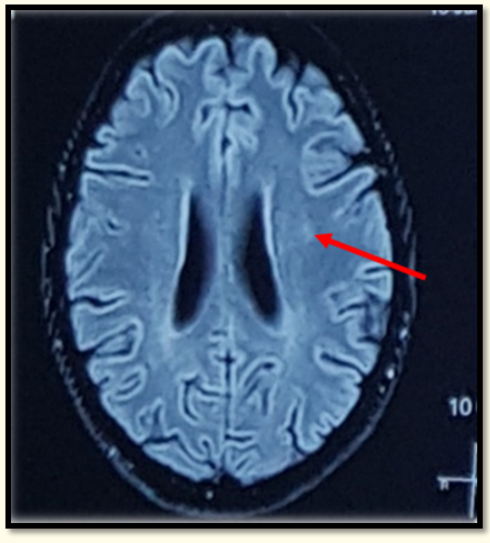

Figure 3: Cerebral MRI showing in axial flair nonspecific bilateral frontoparietal white matter hyperintensities.

Table 1: Laboratory data.

Laboratory tests		Count
Lumbar puncture	Leukocyte	150 (normal range: 0–5/mm^3) with 56% of lymphocytes and 44 % of neutrophils
	Glycorrhachia	Ratio 4
	Protein	0.91 (normal 0.18–0.58 g/L)
Complete blood count	White blood cells	2900/mm^3
	Hemoglobin	9g/dl
	Platelets	219. 10^3/mm^3
ESR	102 mm/hr	
Serum albumin	28 g/L	
Twenty for hours proteinuria	0,2 g/24hours	
Immunological tests	ANA	Positive 1:1280
	Anti-dsDNA	Positive
	APLA	Negative
	C3	0.33 g/L (normal range: 0.9-1.9 g/L)
	C4	C4=0.09 (normal range : 0.1 -0.4
	Anti-Aquaporin-4 and anti-MOG antibodies	Negative

The patient's cerebrospinal fluid sample was negative for tuberculosis, listeria monocytogenes, *Neisseria meningitidis*, *Streptococcus agalactiae*, *Streptococcus pneumoniae* and *Escherichia coli* K1.

Laboratory tests revealed a regenerative and normocytic anemia (9 g/dL, reticulocytes=134000/mm^3), with positive Direct *Coombs Test* for *IgG antibodies*, leukopenia with a total leucocyte count of 2900/mm^3, lymphopenia = 900/mm^3, platelets count of 219. 10^3/mm^3; erythrocyte sedimentation rate (ESR =102 mm/hr), and serum albumin was low at 28 g/L (normal range: 35–50) Twenty-four hours proteinuria was 0.2g/24 hours.

The serology of Hepatitis B virus (HBV), hepatitis C virus (HCV), Human immunodeficiency virus (HIV), Herpes simplex virus (HSV), cytomegalovirus (CMV), Epstein-Bar virus (EBV), syphilis, and brucellosis were found to be negative.

Immunological tests revealed reactive antinuclear antibodies (ANA) = 1: 2800, positive anti-dsDNA, and negative antiphospholipid antibodies (APLA) with hypocomplimentemia C3= 0.33 g/L (normal range: 0.9-1.9 g/L) and C4=0.09 (normal range : 0.1 -0.4). Immunological research for anti-Aquaporin-4 and anti-MOG antibodies was negative.

The diagnosis of an acute LETM secondary to SLE was retained. The Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) =22 indicating a severe flare.

The patient received methylprednisolone 1 g/day for 5 days, relayed by oral prednisolone 1 mg/kg/d and cyclophosphamide (1g/month) during 6 months with functional physiotherapy.

The evolution was marked by a complete regression of paresis and paresthesia in limbs and urinary sphincter disorder with a total removal of radiological damage (figure 4).

The outcome of the patient was deemed favorable during the follow-ups over a period of 2-years in our outpatient consultation.

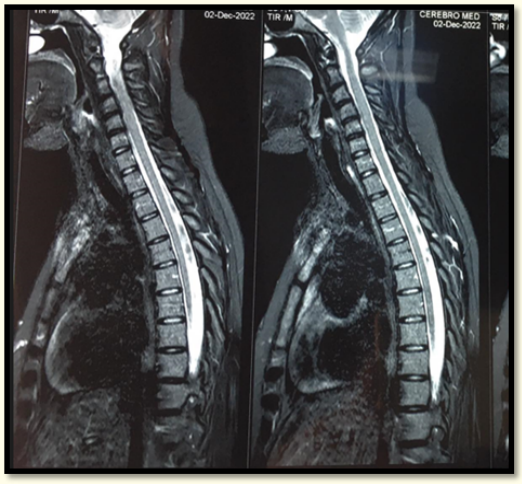

Figure 4: Spinal cord MRI in sagittal T2 weighted image, showing a total removal of neurological damage.

Discussion

SLE is a worldwide distributed autoimmune disease, affecting multiple systems. Its clinical presentation is varied, unpredictable, and sometimes challenging, evolving symptoms that mimic other diseases.

Fever is one of the most common manifestations in SLE. It is also the main cause of admission as seen in our case. Stahl, et al. reported that among 83 febrile episodes in SLE patients, 60% resulted from SLE activity, 23% from infection and 17% from other causes [5]. In their study, Rovin, et al found that fever may be completely suppressed under 20- 40 mg of prednisone, within 24 hours, in 22 SLE patients [6]. Moreover, some clinical and laboratory findings may be helpful in attributing fever to lupus flare, such as lower serum complement C3 and higher SLEDAI score [6]. These features were present in our case.

Nevertheless, NPSLE is one of the major causes of morbidity and mortality in patients with SLE and LETM could be one of NPSLE manifestations. It consists on a substantial spinal cord lesion that extends to three or more vertebral segments on spinal MRI. Its clinical presentations may encompasses sensory, motor and autonomic disorders. It may range from mild extremity numbness, dysesthesia, total sensory loss, paraplegia, bowel and anal sphincter dysfunctions.

Multiple etiologies were incriminated in this disease [4,7]. It most frequently occurs in association with neuromyelitis optica (NMO). LETM may be caused also by spinal cord infarction, infectious myelopathy, compressive myelopathy or paraneoplastic myelitis. In addition, LETM could appear in the course of inflammatory or autoimmune conditions like SLE as shown in our case, Sjögren syndrome, sarcoidosis, or Behçet's disease. Rarely, This disease may be an isolated or idiopathic condition [4,7].

NMO is a rare inflammatory neurologic disease, characterized by severe optic neuritis and LETM. It is associated with NMO immunoglobulin G. We distinguish antibodies bind to the water channel aquaporin 4 (AQP4), which are highly specific, occurring in 70% to 90% of patients in former NMO series [8]. Their presence helps to distinguish NMO from other autoimmune neurologic disorders. Our patient did not present any clinical specific sign of NMO and anti-AQP4, anti-MOG antibodies were negative.

As SLE-related LETM is a rare entity, data in the literature about its clinical course, outcome, and treatment is still missing. Its prevalence is higher in young women to 77% of cases and the mean age is 30 years [9].

Nevertheless, three hypotheses may explain the different courses of myelitis in SLE. It may be a vascular dysfunction, ranging from perivasculitis to thrombosis, a subdural hematoma without vasculopathy or a peripheral white-matter degeneration at multiple spine levels [4,7]. The antiphospholipid syndrome may be associated with LETM in SLE. Antiphospholipid antibodies were negative in our case.

European League Against Rheumatism (EULAR) published in 2010, recommendations for the management of NPSLE based upon a systematic review of over 1000 published studies and expert opinion and updated them recently in 2019.

Specific therapy depends on the pathogenesis (inflammatory or thrombotic disease) [10].

Therefore, the combination of intravenous methylprednisolone and intravenous cyclophosphamide can be effective as shown in our case, with clinical and biological improvement. Plasma exchange therapy, i.v. immunoglobulin and rituximab were used in severe and refractory cases and showed good results [10].

The course of LETM depends, not only, on the severity of initial neurological manifestation, but also, on the extent of the spinal cord lesion on MRI and on the speediness of adequate treatment. Sphincter disorders, gray matter involvement and delayed therapy predict worst outcome. Our patient had a favorable outcome due to prompt and adequate therapy.

Conclusion

LETM is a rare manifestation among NPSLE patients. Its assessment requires excluding other etiologies especially NMO and infectious disease. Given the severity of the disease, it is of paramount importance to know the clinical features of LETM to be swiftly recognized in order to laboratory and imaging diagnosis be promptly made, and the adapted treatment should be immediately started to prevent even life-threatening outcomes.

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