

When Lupus Hits the Spine: A Case of Extensive Transverse Myelitis

J. Diogo^{1,2,3}, N. Nagle⁴, K. Murray^{1,2,3,5}, J. Devlin^{1,2,3}.

1. School of Medicine, University of Limerick, Castletroy, Co. Limerick, Ireland.
2. Rheumatology Department, Health Service Executive MidWest, Co. Limerick, Ireland.
3. Atlantic Research Collaboration, University of Limerick, Castletroy, Co. Limerick, Ireland
4. Radiology Department, Health Service Executive MidWest, Co. Limerick, Ireland.
5. Health Research Institute, University of Limerick, Castletroy, Co. Limerick, Ireland.

Abstract

Presentation

The patient presented with acute urinary retention, fever (39°C), and rapidly progressive bilateral lower limb weakness progressing to flaccid paraplegia (0/5 MRC) with a T6 sensory level.

Diagnosis

MRI spine demonstrated longitudinally extensive T2-hyperintense intramedullary lesions from T6 to the conus. CSF analysis showed lymphocytic pleocytosis and elevated protein; autoimmune serology confirmed active systemic lupus erythematosus (SLE). SLE-associated longitudinally extensive transverse myelitis (LETM) was diagnosed following exclusion of other differential diagnoses.

Treatment

High-dose intravenous methylprednisolone, intravenous immunoglobulin (IVIg), and rituximab were administered. Corticosteroids precipitated psychosis, managed with antipsychotics. Maintenance therapy with mycophenolate mofetil and hydroxychloroquine was continued. Despite aggressive immunotherapy, there was no neurological recovery, and the patient remained paraplegic with neurogenic bowel and bladder dysfunction.

Discussion

This case underscores LETM as a rare, aggressive manifestation of SLE that can occur even in patients of European ancestry. Despite prompt multimodal immunotherapy, neurological recovery was absent, reflecting the poor prognosis of extensive cord involvement. Early recognition, exclusion of mimics, and coordinated multidisciplinary care are vital to optimise outcomes and quality of life.

Introduction

Transverse myelitis (TM) is a rare neuroinflammatory disorder of the spinal cord characterised by neurological motor, sensory, and autonomic dysfunction below the level of the lesion^{1,2}. Longitudinally extensive transverse myelitis (LETM), defined by inflammation spanning three or more vertebral segments, is often associated with autoimmune diseases such as systemic lupus erythematosus (SLE)². SLE affects predominantly women of childbearing age, with neuropsychiatric manifestations in up to 60% of cases^{3–6}. LETM is a rare but severe form involving immune complex deposition, complement activation, vasculitis, and disruption of the blood-spinal cord barrier^{1,4–6}. TM occurs in 1–8 people per million annually, with LETM comprising 30–40% of SLE-related CNS involvement^{3,4}. It is a neurological emergency requiring rapid recognition, aggressive immunosuppression, and exclusion of mimics including NMOSD, multiple sclerosis, and infection^{5–7,10}.

Case Report

A 26-year-old woman of European ancestry with a known diagnosis of systemic lupus erythematosus (SLE) since 2010 had been living abroad, where she experienced two years of intermittent symptoms including alopecia, polyarthralgia, mood disturbance, hallucinations, and recurrent fevers. She later developed lower limb weakness and impaired mobility, prompting her family to bring her back to Ireland, and on the same day she arrived, she presented to University Hospital Limerick with acute urinary retention.

On admission, she was not taking immunosuppressive therapy. Within 24 hours, she developed rapidly progressive bilateral lower limb weakness, progressing from Medical Research Council (MRC) grade 4/5 to flaccid paraplegia (0/5), accompanied by T6 sensory level and fever (peak 39°C).

Spinal MRI revealed a longitudinally extensive T2-hyperintense intramedullary lesion from T6 to T11, later extending from T2 to the conus (Figure 1). Axial imaging at T10 confirmed intramedullary hyperintensity with associated cord edema.

CSF analysis revealed marked lymphocytic pleocytosis (WCC 1365/μL, 92% polymorphs), elevated protein (0.58–1.28 g/L), and matched oligoclonal bands, while infectious PCR panels were negative. Autoimmune serology revealed positive ANA, markedly elevated anti-dsDNA titres, and low complement (C3/C4) (Table 1).

Neuromyelitis optica spectrum disorder (NMOSD) was considered due to the longitudinal extent of the spinal cord lesion; however, aquaporin-4 and MOG antibodies were negative,

and brain MRI was normal. Comprehensive cerebrospinal fluid testing, including bacterial cultures and multiplex PCR, excluded infectious causes.

Differentials included systemic lupus erythematosus (SLE)-associated longitudinally extensive transverse myelitis (LETM) was identified as the most likely cause, supported by the patient's longstanding SLE, active serology (positive ANA, high anti-dsDNA, low complement), and exclusion of alternative diagnoses^{4,6}. Other considerations included spinal ischaemia and infectious myelitis; however, absence of vascular risk factors and negative cerebrospinal fluid cultures supported an inflammatory autoimmune aetiology^{5,7,10}.

High-dose intravenous methylprednisolone, intravenous immunoglobulin (IVIg), and rituximab were administered, but neurological recovery did not occur. Corticosteroids precipitated psychosis, managed with antipsychotics, and immunosuppression was transitioned to mycophenolate mofetil and hydroxychloroquine.

At four-year follow-up, a persistent T6 sensory level is documented. She remains paraplegic with neurogenic bladder and bowel but clinically stable, off corticosteroids, with normalised inflammatory markers and good quality of life.

This case illustrates LETM as a rare, devastating manifestation of SLE requiring early recognition, exclusion of mimics, and multidisciplinary management.



Figure 1: Sagittal MRI of the thoracic spine demonstrating longitudinally extensive intramedullary T2 hyperintensity extending from approximately T6 to T11 (red arrows), consistent with spinal cord inflammation. Post-contrast images show patchy intramedullary enhancement, while STIR sequences demonstrate associated cord oedema across the same segments.

Test	Result	Reference Range	Interpretation
Appearance	Slightly yellow	Clear	Abnormal
WBC (Leucocytes)	1365 / μ L	<5 / μ L	Marked pleocytosis
Differential	92% polymorphs, 8% mononuclears	—	Neutrophilic pleocytosis
Protein	0.58–1.28 g/L	0.15–0.45 g/L	Elevated
Glucose	3.4 mmol/L	2.2–3.9 mmol/L	Normal
Oligoclonal Bands	Present (CSF + serum)	Absent	Abnormal
Culture / PCR	Negative	—	Normal
ANA (IIF)	Positive	Negative	Abnormal
Anti-dsDNA	738 $\rightarrow$ 667 $\rightarrow$ 618 IU/mL	0–100 IU/mL	Strongly positive
Anti-RNP/Sm	Positive	Negative	Abnormal
Lupus anticoagulant	Positive	Negative	Abnormal
Complement (C3/C4)	C3 0.7 g/L (low); C4 <0.02 g/L (very low)	C3: 0.9–1.8; C4: 0.10– 0.40 g/L	Low
ESR	>120 mm/hr	<20 mm/hr	Markedly elevated
CRP	47 mg/L (initial); <1 mg/L (follow-up)	<5 mg/L	Elevated then normalized
ANA (IIF)	Positive	Negative	Abnormal

Table 1. CSF and Blood Analysis

Discussion

Longitudinally extensive transverse myelitis (LETM) represents a rare but severe neuropsychiatric manifestation of systemic lupus erythematosus (SLE), often associated with high morbidity if not promptly recognised and treated⁶. Our patient's rapid progression to paraplegia, combined with radiological involvement that initially extended from T6 to T11 and subsequently progressed from T2 to the conus medullaris, illustrates the aggressive end of the SLE myelitis spectrum. The marked CSF pleocytosis, elevated protein, and active serology (ANA, anti-dsDNA, hypocomplementemia) strongly supported an inflammatory autoimmune process⁷.

Although severe neuropsychiatric manifestations of systemic lupus erythematosus are reported more frequently in non-European populations, this case highlights that longitudinally extensive transverse myelitis can also occur in patients of European ancestry, underscoring the heterogeneity of lupus manifestations.

Furthermore, similar outcomes have been reported, with poor recovery despite intensive immunosuppression^{8,9}. In this case, the combination of longstanding SLE, active serology, and systematic exclusion of mimics confirmed SLE-associated longitudinally extensive transverse myelitis^{4,6}. Despite treatment with high-dose corticosteroids, IVIg, and rituximab, no neurological recovery occurred. Persistent MRI abnormalities were observed despite clinical stability, a recognised feature of lupus myelitis⁷. This case underscores the need for early recognition, aggressive immunotherapy, and multidisciplinary management to optimise outcomes.

Declarations of Conflicts of Interest:

None declared.

Corresponding Author:

Jany Diogo,
School of Medicine,
University of Limerick,
Castletroy,
Co. Limerick,
Ireland.

E-Mail: janydiogo12@gmail.com

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