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BRIEF REPORT

Longitudinally extensive transverse myelitis with neuro-lymphatic protein expressionin systemic lupus erithematosus patient. An autopsy case report

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KEYWORDS
Myelitis;
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Abstract Acute transverse myelitis (inflammation across one or more segments of spinal cord) is a rare complication of systemic lupus erythematosus (SLE) although its frequency is greater than in the general population. Even less common is longitudinal extensive transverse myelitis (LETM), (inflammation affects three or more vertebral segments). The pathogenesis of LETM is unclear and the management uncertain.

We present a case of a 34-year-old woman with SLE and LETM of the whole spine, with rapid progression despite intensive treatment. Autopsy revealed a spine with liquefactive necrosis; some vessels showed fibrinoid necrosis and there were thrombi and an infiltration of lymphocytes and neutrophils in both the grey and white matter. Histological examination of brain revealed necrosis and oedema in the cortex and around the lateral ventricles. The immunohistochemistry showed CD3-positive T-lymphocytes in the wall of the spinal blood vessels, and a prominent D2-40 immunostaining, mainly localized at perivascular inflammatory regions.

PALABRAS CLAVE
Mielitis;
Vasculitis lúpica;
Sistema nervioso central;
Autopsia

Mielitis transversa longitudinal extensa con expresión proteica neurolinfática en un paciente con lupus eritematoso sistémico. Reporte de una autopsia

Resumen La mielitis aguda transversa (inflamación en uno o más segmentos de la médula espinal) es una complicación muy rara, con mayor prevalencia en los pacientes con lupus eritematoso sistémico que en la población general. Mucho menos frecuente es que esta inflamación afecte a 3 o más segmentos espinales. Su patogénesis no está bien definida y el tratamiento es incierto. Presentamos un caso de autopsia de una mujer de 34 años de edad con lupus

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eritematoso sistémico y mielitis transversa longitudinal extensa con un evolución clínica rápidamente desfavorable. Se encontró una médula espinal con necrosis licuefactiva, necrosis fibrinoide de la pared de los vasos, trombos en los mismos y una infiltrado inflamatorio de linfocitos y neutrófilos en la sustancia gris y blanca. En el cerebro había necrosis y edema de la corteza y alrededor de los ventrículos laterales. El estudio inmunohistoquímico mostró linfocitos T CD3 positivos en la pared de los vasos, con una fuerte expresión de D2-40 perivascular.

Introduction

Central nervous system involvement in systemic lupus erythematosus (SLE) is reported to be 24–51%. Acute transverse myelitis (inflammation across one or more segments of spinal cord) is a rare complication, found in only 1–2% patients with SLE; however, this is a 1000 times greater frequency than idiopathic myelitis in the general population (1–2). Even less common is longitudinal extensive transverse myelitis (LETM), a devastating inflammatory syndrome of the spinal cord, affecting three or more vertebral segments, causing weakness, numbness and sphincteric deficits. The pathogenesis of LETM is unclear and the management uncertain.

Here we present a case of SLE and LETM in the entire spine; the disease progressed rapidly despite intensive treatment and the patient died 33 days after admission.

Case presentation

A 34-year-old woman presented with a 1-week history of fever and acute urinary retention. She had been diagnosed with SLE 10 years previously at the age of 20 when diagnostic criteria for SLE, such as arthralgia, rash and multiple biochemical markers, were positive. She was initially treated with immunosuppressants but had suffered complications since 2011, including chronic pancreatitis, haemolytic anaemia, diabetes mellitus and iatrogenic Cushing, for which she was treated with mycophenolate.

On admission she had cushingoid appearance, her vital signs were normal, she was alert and her body temperature was 39 °C. Two hours later she had flaccid leg paralysis and areflexia, with paraesthesia below the level of T1. Neurological examinations revealed sensory deficits, predominantly of deep sensation, and sphincter dysfunction with urinary retention. Her neurological functions continued to deteriorate and a spinal MRI was performed, after which she had a cardiac arrest and needed advanced resuscitation.

All haematological and biochemical parameters were normal, including a complete blood count, liver enzymes levels, total bilirubin level, coagulation profile, renal function and urinalysis. Levels of anti-Sm, anti ANCA, anti RNP, anti Jo 1, anti Scl.70, anti-SSA and anti-SSB antibodies were negative, and serum levels of complement C3 (19 mg/dl; normal 75–140) and C4 (3 mg/dl; normal 10–34) were low.

Spinal MRI showed extensive intramedullary abnormal intensity from the cervical cord to conus medullaris, mainly involving all the central grey matter, but also the lateral autonomic white matter (Fig. 1). The heterogeneous abnormal hyperintensive signal extended to the brainstem. These findings were consistent with a diagnosis of longitudinally extensive transverse myelitis. Brain MRI with contrast showed no obvious abnormality in the brain parenchyma or intracranial arteries.

A lumbar puncture showed pleocytosis (leucocyte count of 432 cells/mm^3; normal 0–5), with 90% neutrophils and 10% lymphocytes, a mildly low glucose level of 37 mg/dl (normal 40–70) and an elevated total protein level of 408.70 mg/dl (normal 15–40). The patient's cerebrospinal fluid sample was negative for bacterium and syphilis antibodies. The results of viral serology of the cerebrospinal fluid for enterovirus, varicela zoster, HIV, herpes simplex

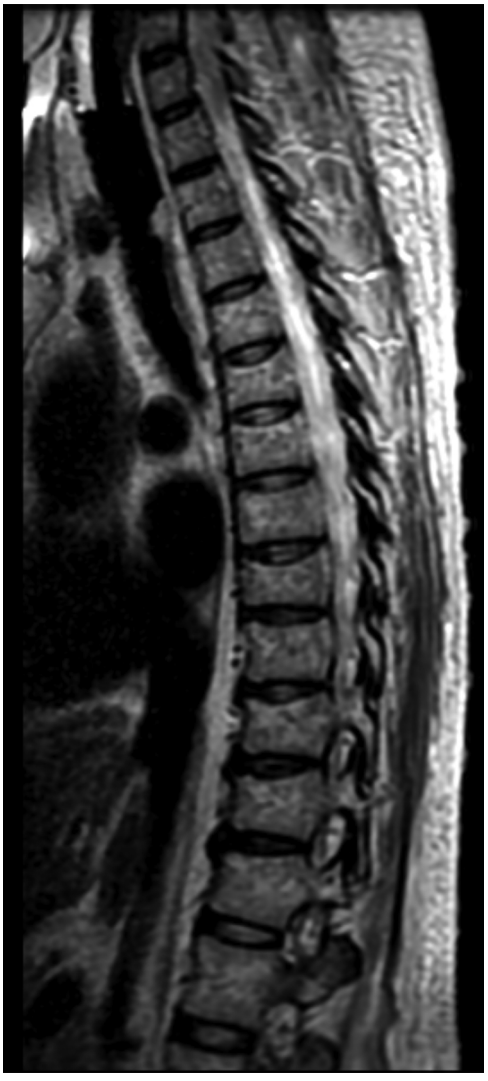

Figure 1 MRI shows T2W1 bright in the spinal cord.