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MENINGOMYELORADICULITIS WITH EXTENSIVE BRAINSTEM INVOLVEMENT SECONDARY TO SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT

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BACKGROUND

As many as 40–50% of patients with systemic lupus erythematosus (SLE) have neurological involvement during the course of their disease. Neuropsychiatric lupus may present with several neurological and psychiatric syndromes, including headache, seizures, stroke, myelitis, movement disorders, psychosis, cognitive disorders, and depression. Diagnosis is often difficult and requires a high degree of clinical suspicion.

CASE REPORT

A 25-year-old male patient presented to the emergency department with a 7-day history of paraparesis, urinary retention, intestinal constipation, dysautonomia and headache. He reported a history of additive, symmetrical polyarthritis in the past 8 months and daily fever and night sweats in the previous month. Shortly after the hospitalization, he developed acute respiratory failure and required endotracheal intubation. On physical examination, he presented paraplegia with lower limbs areflexia, muscle atrophy, hypotonia, involvement of III, VII, VIII and XII cranial nerves, and T4 sensory level. Neuroaxis T2-weighted magnetic resonance imaging (MRI) demonstrated hyperintense lesions in brainstem and left nucleocapsular/thalamus region and longitudinally extensive transverse myelitis, associated with leptomeningeal, cranial nerves and cauda equina roots enhancement. Cerebrospinal fluid (CSF) analysis showed neutrophilic pleocytosis (240 cells/mm3; neutrophils 75%, lymphocytes 22%, macrophages 3%), elevated protein (192 mg/dL) and low glucose (13 mg/dL) levels; but gram stain, cultures and polymerase chain reaction (PCR) assays for herpesvirus and *Mycobacterium tuberculosis* were negative. Laboratory tests revealed chronic disease anemia and normal urinalysis. Chest X-ray was normal. Laboratory tests for autoimmune diseases showed positive ANA, anti-Ro, anti-RNP/Sm, high-titer anti-dsDNA antibodies and low complement levels. Antiphospholipid and anti-aquaporin-4 antibodies were negative. Salivary gland biopsy was not suggestive of Sjögren's syndrome (focal score 0). Diagnostic hypothesis was lupus meningomyeloradiculitis with brainstem involvement, and the patient received high dose of glucocorticoid (methylprednisolone 1 g/day for 5 days, followed by 1 mg/kg/day), IVIG, cyclophosphamide, plasmapheresis and rituximab therapies. The patient showed a gradual clinical and radiological improvement, but he still has lower limbs weakness and urinary retention, and was referred to a rehabilitation center.

CONCLUSION

Longitudinally extensive transverse myelitis is a severe and rare manifestation of SLE. The concomitant meningeal, radicular and extensive brainstem involvement described herein reminds us of the wide spectrum of neuropsychiatric lupus presentations. High degree of clinical suspicion, with prompt diagnosis and treatment, is crucial in reducing sequelae and preserving functional capacity and quality of life.