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Original article

Myelopathy in systemic lupus erythematosus: clinical, laboratory, radiological and progression findings in a cohort of 1,193 patients

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ABSTRACT

Objective: To describe clinical, laboratory, radiological and progression characteristics of myelopathy in systemic lupus erythematosus (SLE).

Patients and methods: A retrospective analysis was performed on a cohort of 1193 patients with SLE (ACR criteria) in order to identify patients with myelopathy (neuropsychiatric ACR). Disease activity was assessed by the SLE activity index (SLEDAI) on the date of the event and functional capacity was assessed by the Expanded Disability Status Scale (EDSS) at the last visit.

Results: We identified 14 (1.2%) patients with myelopathy. All were women with a mean age of 30 ± 11.5 years. Myelopathy occurred at the diagnosis of SLE in four (28%) patients; and nine (64%) patients had another type of neuropsychiatric manifestation associated. Neurological recurrence was observed in one (7%) patient. Disease activity was observed in 2 (14%) patients. Cerebrospinal fluid presented pleocytosis on 7 (53%) patients; antiphospholipid antibodies were positive in 5 (45%). Magnetic resonance imaging (MRI) showed T2 hyperintensity with a predominance of longitudinal involvement in 6 (86%) patients. Most were treated with intravenous corticosteroids and cyclophosphamide. No patient had full recovery and four (36%) had high EDSS scores. Three (21%) patients died from sepsis early in the course of their myelopathy, during or after immunosuppressive therapy.

Conclusions: Myelopathy occurred in 14 (1.2%) of the patients in our cohort and this may be the first manifestation of the disease occurring independently of systemic disease activity. Although rare, myelopathy shows great morbidity and mortality, can be recurrent and MRI is critical for diagnosis.

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Mielopatia no lúpus eritematoso sistêmico: achados clínicos, laboratoriais, radiológicos e evolutivos em uma coorte de 1.193 pacientes

Palavras-chave:
Lúpus eritematoso sistêmico
Mielopatia
Mielite transversa
Ressonância magnética

RESUMO

Objetivo: Descrever características clínicas, laboratoriais, radiológicas e evolutivas de mielopatia no lúpus eritematoso sistêmico (LES).

Pacientes e métodos: Foi realizada análise retrospectiva de uma coorte de 1193 pacientes com LES (critérios ACR), para identificar os pacientes com mielopatia (ACR neuropsiquiátrico). A atividade de doença foi analisada pelo Índice de Atividade do LES (SLEDAI) na data do evento e a capacidade funcional pela Escala Expandida do Estado de Incapacidade (EDSS) na última consulta.

Resultados: Foram identificados 14 (1,2%) pacientes com mielopatia. Todas eram mulheres com média de idade de 30 anos (DP ± 11,5 anos). A mielopatia ocorreu no diagnóstico do LES em quatro (28%) e em nove (64%) havia outro tipo de manifestação neuropsiquiátrica associada. Recorrência do quadro neurológico foi observado em uma (7%) paciente. Atividade de doença foi observada em 2 (14%) pacientes. O líquido cefalorraquidiano apresentava pleocitose em 7 (53%) pacientes anticorpos antifosfolípides eram positivos em 5 (45%). A ressonância magnética (RM) demonstrou hipersinal em T2 com predomínio do comprometimento longitudinal em 6 (86%) pacientes. A maioria foi tratada com corticosteroides e ciclofosfamida endovenosos. Nenhuma paciente teve completa recuperação e quatro (36%) tinham escores altos da EDSS. Óbito foi observado em 3 (21%) durante episódio de mielopatia, por septicemia durante ou após terapia imunossupressora.

Conclusões: A mielopatia ocorreu em 14 (1,2%) dos pacientes da nossa coorte e pode ser a primeira manifestação da doença ocorrendo independentemente de atividade sistêmica da doença. Embora rara, é de grande morbimortalidade, pode ser recorrente e a RM é fundamental para o diagnóstico.

Introduction

Neuropsychiatric manifestations in systemic lupus erythematosus (SLE) have an important impact on the prognosis of the disease by their frequency and severity.[1] Myelopathy is a manifestation of the central nervous system (CNS) that occurs only rarely in SLE, affecting about 1–2% of patients.[1–3]

In 1999, the American College of Rheumatology (ACR) established the criteria of neuropsychiatric manifestations in SLE, including myelopathy. This possibility should be considered if the patient shows a rapid progression (hours or days) of one or more of the following signs/symptoms: bilateral muscle weakness in lower limbs, with or without involvement upper limbs; a sensory disorder with similar level of motor impairment, with or without bowel or bladder involvement. Expansive injury causing spinal cord compression and *cauda equina* injury should be ruled out.[4]

In 2002, the Transverse Myelitis Consortium Working Group proposed diagnostic criteria for idiopathic transverse myelitis, defined with the clinical manifestations described above, associated with inflammation within the spinal cord demonstrated by cerebrospinal (CSF) pleocytosis or increased IgG index or gadolinium enhancement in magnetic resonance imaging (MRI).[5]

Myelopathy may present as a transverse myelopathy with sectional involvement of one level of the spinal cord, or as a longitudinal myelopathy in which more than three segments are affected, continuously or not.[6]

The term myelitis is still used by many authors; however, myelopathy is more suitable to characterize spinal cord changes associated with inflammatory diseases such as SLE, this being the nomenclature recommended by ACR.[4]

The cause of myelopathy in SLE is not well understood and the participation of both thrombosis and vasculitis have been implicated in this process.[3] Some authors suggest that there is a relationship between antiphospholipid antibodies and myelopathy, which would augment the possibility of thrombosis; but other studies do not confirm this association.[7–10]

Although rare, thanks to its importance this condition was recently included in the new classification criteria of the disease.[11] The literature only presents case reports, with the publication of case series only by a few authors.[8,10,12–19]

The aim of this study is to describe cases of myelopathy in SLE, from a cohort of a single university hospital, describing their clinical picture, laboratory results, imaging findings on MRI of the spinal cord, treatment and outcome.

Patients and methods

Medical records of a cohort of 1193 patients with SLE,[20] followed at the Rheumatology outpatient clinic of the Hospital das Clínicas, Universidade Estadual de Campinas (UNICAMP) were analyzed retrospectively.

Patients with myelopathy were identified by the presence of acute clinical manifestations suggestive of spinal cord