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Case report

IgA nephropathy in systemic lupus erythematosus patients: case report and literature review

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ABSTRACT

Systemic erythematosus lupus (SLE) is a multisystemic autoimmune disease which has nephritis as one of the most striking manifestations. Although it can coexist with other autoimmune diseases, and determine the predisposition to various infectious complications, SLE is rarely described in association with non-lupus nephropathies etiologies. We report the rare association of SLE and primary IgA nephropathy (IgAN), the most frequent primary glomerulopathy in the world population. The patient was diagnosed with SLE due to the occurrence of malar rash, alopecia, pleural effusion, proteinuria, ANA 1: 1280, nuclear fine speckled pattern, and anticardiolipin IgM and 280 U/mL. Renal biopsy revealed mesangial hypercellularity with isolated IgA deposits, consistent with primary IgAN. It was treated with antimalarial drug, prednisone and inhibitor of angiotensin converting enzyme, showing good progress. Since they are relatively common diseases, the coexistence of SLE and IgAN may in fact be an uncommon finding for unknown reasons or an underdiagnosed condition. This report focus on the importance of the distinction between the activity of renal disease in SLE and non-SLE nephropathy, especially IgAN, a definition that has important implications on renal prognosis and therapeutic regimens to be adopted in both the short and long terms.

Nefropatia por IgA em paciente portadora de lúpus eritematoso sistêmico: relato de caso e revisão de literatura

Palavras chave:
Lúpus eritematoso sistêmico
Nefropatia por IgA
Glomerulonefrite

RESUMO

O lúpus eritematoso sistêmico (LES) é uma doença autoimune multissistêmica que tem como uma das manifestações mais marcantes a nefrite. Apesar de poder coexistir com outras doenças autoimunes e determinar a predisposição a diversas complicações infecciosas, o LES raramente é descrito em associação a nefropatias de etiologia não lúpica. Relatamos

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o caso da rara associação entre LES e nefropatia por IgA (NIgA) primária, a glomerulopatia primária mais frequente na população mundial. A paciente foi diagnosticada com LES pela ocorrência de eritema malar, alopecia, derrame pleural, proteinúria, pancitopenia, FAN 1:1.280 padrão nuclear pontilhado fino e anticardiolipina IgM 280 U/mL. A biópsia renal revelou hipercelularidade mesangial com depósitos isolados de IgA, compatível com NIgA primária. Foi tratada com antimalárico, prednisona e inibidor da enzima conversora de angiotensina e apresentou boa evolução. Por consistirem em doenças relativamente frequentes, a coexistência de LES e NIgA pode ser de fato um achado incomum por motivos desconhecidos ou uma condição subdiagnosticada. Este relato atenta para a importância da distinção entre a atividade de doença renal do LES e nefropatias não lúpicas, em especial a NIgA, definição que tem implicações importantes sobre o prognóstico renal e regimes terapêuticos a serem adotados em curto e longo prazo.

Introduction

Systemic lupus erythematosus (SLE) is a chronic disease of the connective tissue characterized by a number of immunological disorders which result in the onset of inflammatory lesion in various organ tissues. Lupus nephritis (LN) is the most common visceral manifestation of SLE, being diagnosed in approximately 37–45% of the patients at some time during the course of the disease in the Brazilian population.[1,2] The description of nephritis of other etiologies in patients diagnosed with SLE, however, is an uncommon finding.[3] IgA nephropathy (IgAN), although being the most common cause of glomerulopathy in general population,[4] is rarely associated with SLE.[5–10] We reported a case of rare coexistence of IgAN in a patient with SLE.

Case report

Forty-year-old female patient, complaining of generalized edema six months ago, associated with polyarthralgias, intermittent fever, alopecia, weight loss of 7 kg, and ulcerated lesion on the right leg. On examination the patient was pale, with bilateral periorbital edema, facial flushing, reduced vesicular murmur on the right, lower limb edema (2+/4+) and pyoderma gangrenosum in the middle third of the right leg, associated with stiffness and swelling of the right calf. Additional assessment indicated hemoglobin of 8.9 g/dL, leukopenia, mild thrombocytopenia, ANA 1:1.280 of nuclear fine speckled pattern, CH50 of 88 U/mL, C3 of 46 mg/dL, C4 of 9 mg/dL, negative anti-dsDNA, anticardiolipin IgM of 211 U/mL, proteinuria of 1045 mg/24 h, endogenous creatinine clearance of 162 mL/min, dysmorphic hematuria, and granular urinary casts. Chest computed tomography showed bilateral pleural effusion, mild pericardial effusion, and ascites. Lower limb Doppler ultrasonography ruled out thrombotic event. Renal biopsy was performed, showing 16 intact glomeruli with mesangial granular deposition of IgA, negative for other immune deposits, and mesangial hypercellularity (Fig. 1). The patient was diagnosed with SLE associated with IgAN, and she was initially treated with prednisone 60 mg/day, hydroxychloroquine 400 mg/day, enalapril 10 mg/day, and supplementation of calcium and vitamin D, and antibiotics for skin lesion. After initiating treatment, the patient showed improvement of the joint, cutaneous, hematologic, and renal status, being ready for hospital discharge.

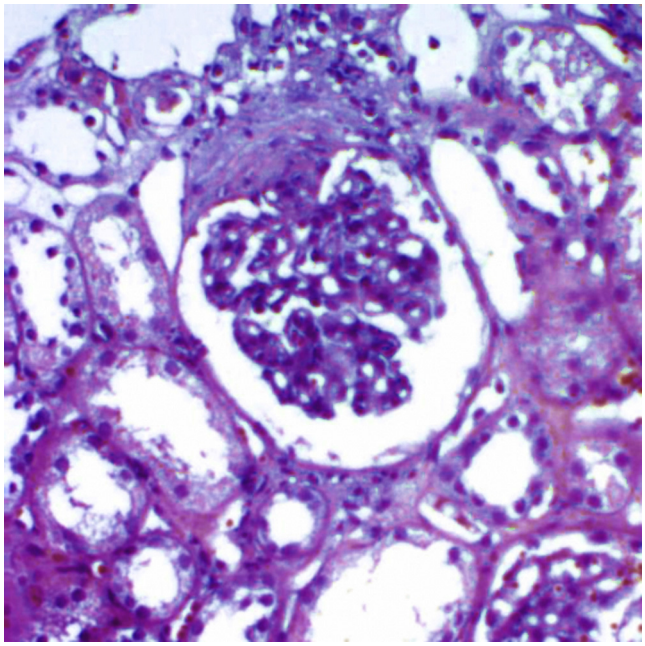

Fig. 1 – Optical microscopy of specimen obtained by renal biopsy revealing mesnagial expansion and hypercelullarity (HE, 200×).

Discussion

SLE is a disease marked by heterogeneity of clinical phenotypes and unpredictable course.[1,2] These properties make the characterization of the disease, the acknowledgment of its complications, and the detection of overlying conditions a constant challenge in the routine of rheumatology services. The classic clinical presentation of LN is persistent proteinuria and/or cellular casts, or active urinary sediment, i.e., five or more red blood cells or leukocytes per high power field.