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"Full-House" Nephropathy in a ANA Negative Patient

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Introduction

Antinuclear antibody (ANA) is considered a constant in the diagnosis of systemic lupus erythematosus (SLE), mainly due to its high sensitivity. However, a few recent studies have suggested that ANA may be negative in patients with established SLE, partly because of the variation in immunofluorescence assays that may influence its outcome. The frequency of test negativity ranged from 4.9% to 22.3% in some studies [1-4].

Case report

A 31-year-old female was admitted on Public Hospital in Recife in January 2018 with fever, bilateral ventilatory-dependent chest pain, arthritis on knees and erythema malar. During hospitalization, ANA, anti-DNA, anti Ro, CRP, Rheumatoid factor, C3, C4, anticardiolipin, proteinuria of 24 hrs, echocardiogram, cultures, serology for HIV, syphilis, hepatitis, thyroid hormone were normal. Later, the patient developed lymph nodes enlargement in the cervical and inguinal regions, with acute renal failure (creat 5.7), anemia (Hb 6.4) with direct coombs positive, plaquetopenia (43,000), increased bilirubin, schizocytes in peripheral blood,hematuria ++/4+, proteinuria +/4+ and dyspnea, received dialytic therapy and pulse therapy with methylprednisolone 1 g/day for 5 days. Thoracic tomography demonstrated hilar lymph node enlargement and areas of ground glass in pulmonary bases. **Myelogram:** absence of parasites and foreign cells. Plasmapheresis was performed and patient had clinical and laboratory improvement, with a return to baseline renal function, with no more need for hemodialysis and prednisone 1 mg/kg/day was done. Renal biopsy was suggestive for SLE with Full-house immunofluorescence with optical microscopy showing focal and segmental mesangio proliferation (Figure 1), and the patient received cyclophosphamide 1 g (NIH regimen).

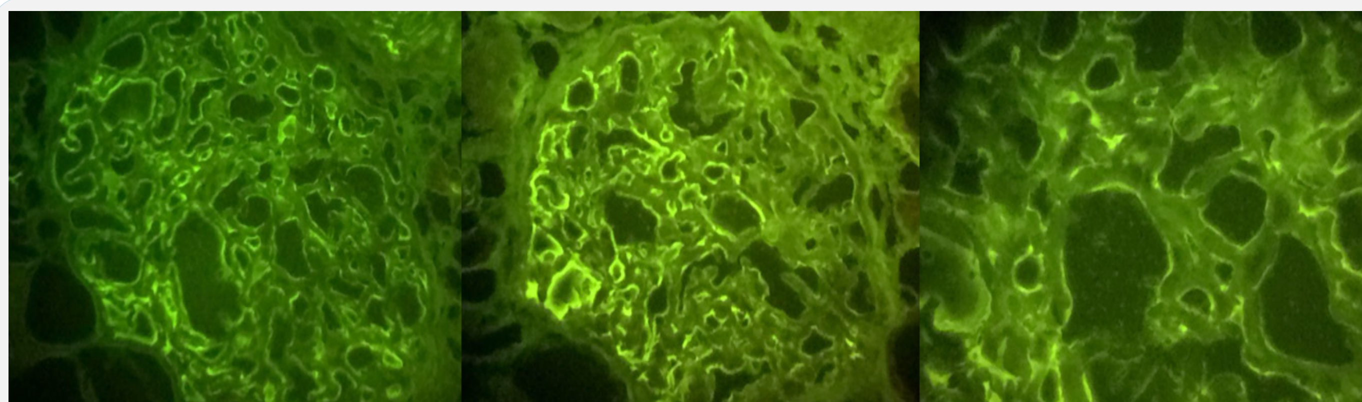

Figure 1: Subtypes of the "Full-house" pattern of direct immunofluorescence C3, IgG and IgM, respectively.

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Discussion

In a study with 13,080 SLE patients, 12,542 (95.9%) performed ANA by immunofluorescence indirect HEp-2 with positive results and several titers. For the 1:40, 1:80, 1:160, and 1:320 titers the sensitivity rates were 98.4%, 97.8%, 95.8% and 86.0%, respectively. In fact, with a titer of 1:160, less than 5% of patients with SLE had negative ANA. This negative examination in a context of high suspicion of SLE is a clinical challenge [2]. In recent studies, only 0.14% of the cases present this negative serological marker. In rare cases of lupus ANA negative with nephritis, renal biopsy is necessary because, in addition to establishing histological classification, it has therapeutic implications and excludes other diagnoses such as thrombotic microangiopathy. Thus, we report here a rare case of SLE ANA negative which diagnosis was made by renal biopsy, nephropathy with “Full-house” pattern being an important clue to the diagnosis [3,4].

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