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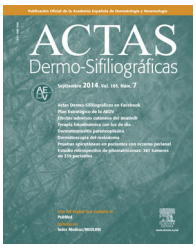

RESIDENT'S FORUM

Neutrophilic Dermatoses Associated With Autoimmunity☆

FR - Dermatosis neutrofílicas asociadas a autoinmunidad

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The neutrophilic dermatoses are a group of diseases characterized by skin lesions with variable degrees of infiltration by polymorphonuclear cells and leukocytoclasia without vasculitis.[1] Reports in recent years have described neutrophilic dermatosis lesions in patients with systemic lupus erythematosus (SLE) and other autoimmune diseases, such as rheumatoid arthritis,[2] either as skin involvement in the course of a previously diagnosed autoimmune disease or as the initial manifestation of such a disease. We believe it is important to recognize this type of lesion and its relationship with autoimmune diseases, as these lesions can be the first manifestation of such diseases in up to a third of cases.

In 1972, Fryha et al.[3] described the first case of neutrophilic dermatosis in a patient with renal glomerular disease and malar erythema; however, the patient did not have antinuclear antibodies and therefore did not strictly satisfy SLE criteria at that time. More than 50 cases of autoimmune diseases associated with neutrophilic dermatosis have since been reported. The majority of patients have been young women (mean age, 40 years), and skin involvement was the initial symptom of the disease in a third of cases. The lesions consist of erythematous, violaceous, or pink macules or papules that are sometimes painful. The lesions affect the limbs almost always, the trunk in more than half of cases, and the head and neck in almost a quarter. From a histopathological point of view, the degree of neutrophil infiltration can vary from paucicellular to dense infiltrates.[4] Other histologic changes (thickening of the basement membrane, presence of dermal mucin, or changes at the interface) can be seen in cases with a long-standing diagnosis of lupus, but are much rarer in patients in whom skin involvement is the initial symptom. The degree of leukocytoclasia also is variable, though signs of vasculitis area always absent. In half of the cases in which immunofluorescence studies are performed, deposits of immunoglobulin (Ig) G, IgM, IgA, or C3, almost always of granular appearance, are seen at the dermoepidermal junction.

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The treatment of these lesions does not differ from that of the underlying disease, and the lesions usually respond to systemic corticosteroid therapy (intravenous prednisone, 0.5-1 mg/kg) and to other immunosuppressants such as azathioprine or mycophenolate mofetil.

Thus, although long-term studies are required to determine the prognostic significance of these lesions on the course of the autoimmune disease, it would appear logical that, when investigating a patient with clinical and histologic features compatible with neutrophilic dermatosis, we should take a detailed medical history and perform a physical examination to look for other signs of connective tissue disease, as well as laboratory tests that include parameters for autoimmunity.

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