Case Report

Rheumatoid arthritis and pseudo-vesicular skin plaques: rheumatoid neutrophilic dermatosis*

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Abstract: A 54 year-old woman with a 3-year history of rheumatoid arthritis (RA) consulted us because of weight loss, fever and skin eruption. On physical examination, erythematous plaques with a pseudo-vesicular appearance were seen on the back of both shoulders. Histological examination was consistent with rheumatoid neutrophilic dermatosis (RND). After 3 days of prednisone treatment, the skin eruption resolved. RND is a rare cutaneous manifestation of seropositive RA, characterized by asymptomatic, symmetrical erythematous plaques with a pseudo-vesicular appearance. Histology characteristically reveals a dense, neutrophilic infiltrate with leucocitoclasis but without other signs of vasculitis. Lesions may resolve spontaneously or with RA treatment. This case illustrates an uncommon skin manifestation of active rheumatoid arthritis.
Keywords: Arthritis; Arthritis, rheumatoid; Rheumatoid factor.

INTRODUCTION

Rheumatoid arthritis (RA) is an autoimmune, chronic, systemic disease that affects 1% of the Western population, particularly women, during the fifth decade of life.[1] Clinically, it presents as a chronic, symmetric and destructive polyarthritis. Extra-articular manifestations of RA (ExRA) implicate a severe and active disease, and are associated with increased morbidity and premature mortality+.[2] It is believed that the same cytokines involved in the synovial pathology are responsible for ExRA, producing damage in multiple systems: musculoskeletal regions, skin, eye, lungs, heart, kidneys, salivary glands, blood vessels, central nervous system and bone marrow.

We describe a patient with severe and active RA who presented pseudo-vesicular skin eruption on her shoulders.

CASE REPORT

A 54 year-old female with a 3-year history of RA consulted us because of an asymptomatic, pseudo-vesicular skin eruption. She had not been on any treatment over the preceding 3 years, and was hospitalized because of a six-month history of weight loss, morning stiffness in her hands, fever and generalized arthralgia. On physical examination, the patient presented two symmetrically-distributed, erythematous plaques with a pseudo-vesicular appearance on the back of both shoulders (Figure 1). Laboratory exams revealed: microcytic anemia (hemoglobin: 10.1g/dl with MCV of 79.8fl), normal reticulocyte count; leucopenia (3,300 cells/uL) with normal neutrophil count; lymphopenia (729 cells/ul), high sedimentation rate (120mm/hr); hypoalbuminemia (2.9g/dl); and positive rheumatoid factor. A skin biopsy showed epidermal parakeratosis, irregular ac-

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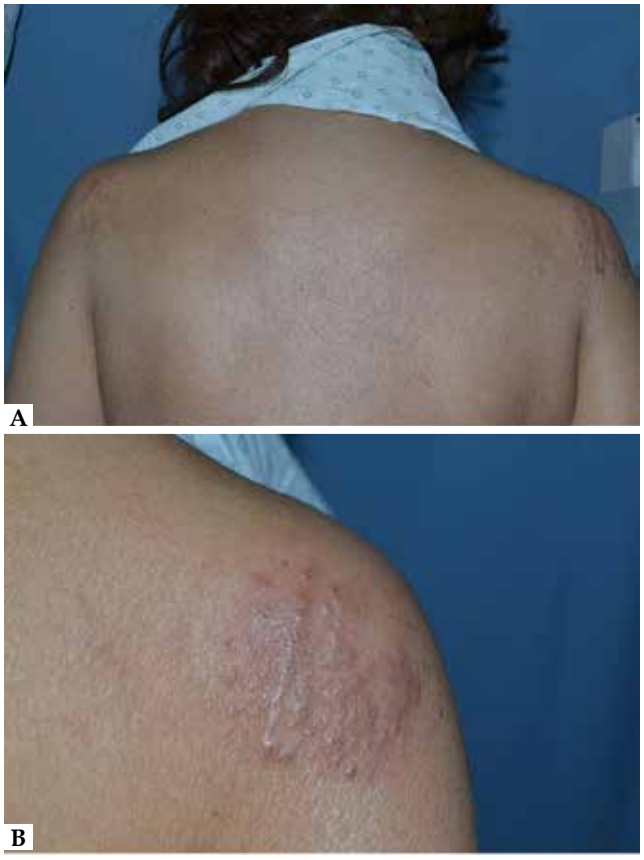

Figure 1: (a) Slightly erythematous plaques symmetrically located on the back of both shoulders. **(b)** Upon closer observation, the skin over the lesions was remarkable because of its pseudo-vesicular appearance

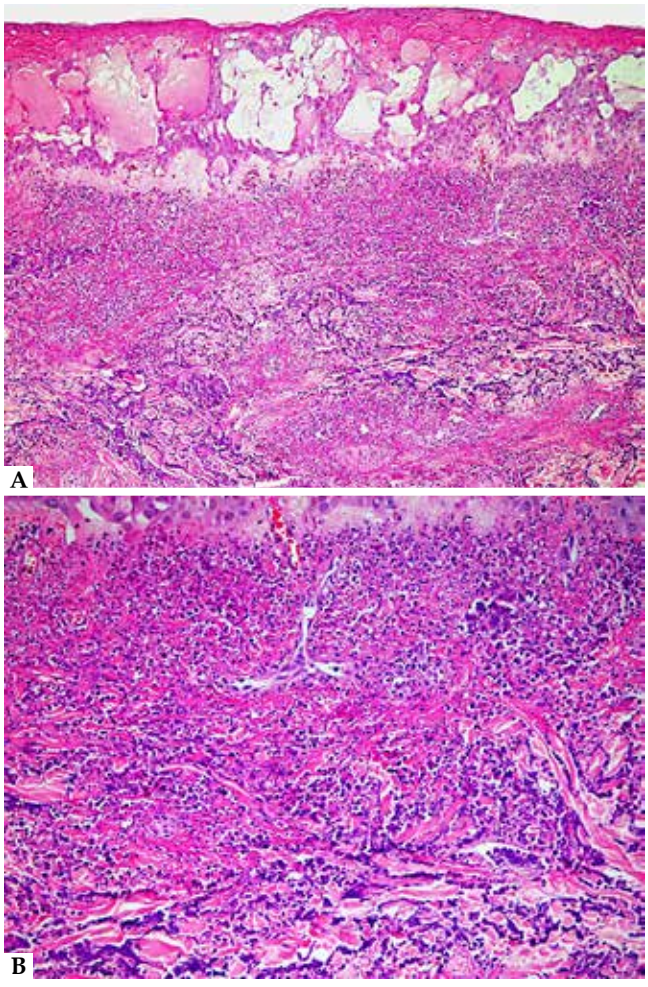

Figure 2: (a-b, Haematoxylin-eosin, X40 and X100, respectively.) Intense spongiosis and spongiotic microvesicles in the epidermis, with dense and diffuse infiltration of neutrophils at the dermis. No microorganisms were identified

anthosis, spongiosis with intraepidermal spongiotic blistering, and focal lymphocytes exocytosis. An accentuated papillary edema was noted on the upper dermis, followed by: an intense, predominately neutrophilic infiltration with necrosis in the deep reticular dermis; some vessels with a neutrophilic infiltration; as well as leucocitoclasis, but without any other signs of vasculitis (Figure 2). Histological findings were consistent with a diffuse, neutrophilic dermatosis, suggesting rheumatoid neutrophilic dermatosis.

Because of active and severe RA, a course of 60mg/d of prednisone was initiated, with rapid regression of skin eruption after the third day of therapy.

DISCUSSION

It is estimated that 40% of RA patients will develop an extra-articular manifestation.[3] Risk factors for ExRA include older age, presence of rheumatoid factor and/or antinuclear antibodies, early disability and smoking.[4]

Skin manifestations are the most frequent ExRA manifestations, the most common of which are rheumatoid nodules.[5] Other cutaneous ExRA include: skin ulcers, Raynaud phenomenon, vasculitis and pyoderma gangrenosum. Less frequent ExRA manifestations include palisading granulomatous dermatitis, neutrophilic lobular paniculitis, and neutrophilic dermatoses such as Sweet syndrome and rheumatoid neutrophilic dermatosis.[6]

Rheumatoid neutrophilic dermatosis (RND) was first described by Ackerman in 1978.[7] The clinical manifestations of RND include erythematous to yellow-colored papules, plaques or nodules, with an urticariform appearance. On rarer occasions, annular, ulcerative, crusted lesions may be seen, or sometimes lesions with a pseudo-vesicular surface, as in this case.[6] Lesions are symmetrically distributed above the joints, extensor surfaces of the extremities, or the trunk. [6,8]

Histologically, RND is characterized by papillary edema and a dense, neutrophilic infiltrate, which may extend into the subcutaneous fat, with some leukocytoclasis in the deep dermis. [9,10] The infiltrate may be admixed with eosinophils, lymphocytes and hystiocytes. Epidermal changes in RND are less prominent but, due to the dermal edema, intraepidermal blistering may be seen, as in the case here described, which correlates clinically with the pseuso-vesicular surface of the lesions.[9]

The main differential diagnoses of RND include: Sweet syndrome – which may be identical to RND clinically and histologically-, erythema elevatum diutinium, neutrophilic urticaria, urticarial vasculitis, pyoderma gangrenosum, bowel–associated dermatosis–arthritis syndrome and Behçet's disease.[6] Since RND and Sweet syndrome can be identical, RND may be regarded as Sweet syndrome in patients with active seropositive RA.

Lesions tend to resolve spontaneously or with improvement in the RA, as in this case. However, recurrence is common with exacerbation of RA. Lesions usually heal with no scarring.[6] Therefore, the first line treatment is RA therapy with systemic corticosteroids. Furthermore, anti-neutrophilic therapies – such as dapsone or colchicine- may lead to rapid resolution of RND.[6] ❑

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