Bullous Erythema Elevatum Diutinum Associated with Immunoglobulin a Monoclonal Gammopathy: An Atypical Variant

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Sir,

An atypical and very rare type of presentation of erythema elevatum diutinum (EED), an usual case of bullous EED not associated with human immunodeficiency virus or acquired immunodeficiency syndrome, is presented here.

We present a 60-year-old male patient with a personal history of diabetes, arterial hypertension, smoking, dyslipidemia, and avascular necrosis of the hip who presented 1 month after evolution of extensive, painful, and itchy skin lesions. He was afebrile and had a good overall condition. He had no history of trips to abroad, contact with animals, occupational exposure to substances, or unsafe sex.

Denuded and crusty red-brown plaques were found on the extensor surfaces of forearms, arms, buttocks, thighs, and legs [Figure 1]. They showed a symmetrical distribution and spared his trunk and face [Figure 2]. Some lesions were ulcerated.

Histopathological analysis of three of the plaques demonstrated an extensive neutrophilic vasculitis, leukocytoclasia, subepidermal blisters, and fibrinoid necrosis in the dermis [Figure 3]. Direct immunofluorescence was negative.

The results of blood analysis were 13,000 leukocytes/µL (8630 neutrophils/µL), hemoglobin 11.8, and 310,000 platelets/µL. Glucose, urea, liver enzymes, lipids, ions, albumin, proteins, immunoglobulin G (IgG), IgA, IgM, IgE, proteinograms, and tumor markers were normal. Antinuclear, p-anti-neutrophilic cytoplasmic, c-anti-neutrophilic cytoplasmic, and antiphospholipid antibodies were all negative. Human immunodeficiency virus, syphilis, hepatitis B and C viruses, and Toxoplasma spp. serologies were negative. The urine test was normal. A kappa IgA monoclonal paraprotein could be found in 24-h urine and blood using immunofixation. Bone marrow aspiration was negative for multiple myeloma.

Differential diagnoses included autoimmune blistering disorders (mainly dermatitis herpetiformis), bullous leukocytoclastic vasculitis, and bullous EED.

The possibility of dermatitis herpetiformis was discarded considering the histopathology, the negativity of the direct immunofluorescent, and the negative results of antitissue transglutaminase and antiendomysium antibodies.

A bullous leukocytoclastic vasculitis could be considered in this case; however, due to the clinical features, distribution and symmetry of the lesions, complete lack of response to high doses of corticosteroids, and good response to dapsone, a diagnosis of bullous EED was thought of.
After a month of treatment with oral dapsone 100 mg/day, the resolution of nearly all the lesions was noteworthy [Figure 4], only showing residual fibrotic nodules located on the knuckles [Figure 5].

EED is a rare type of chronic neutrophilic vasculitis located over the extensor surfaces of limbs, including knuckles, knees, elbows, and ankles. Usually, erythematous–violet nodules, papules, and plaques are found. Vesicles, pustules, or blisters are much less common. EED usually spares the trunk and has a typical symmetrical distribution.

Histopathologically, it is common to find neutrophilic infiltrate in the dermis, leukocytoclasia, vasculitis, and endothelial swelling. Perivascular fibrosis is seen in chronic lesions.[1]

EED has been associated with a vast list of diseases such as multiple myeloma, IgA monoclonal gammopathy,[2] leukemia, lymphoma, human immunodeficiency virus infection, syphilis, streptococcal infection, hepatitis B, celiac disease, inflammatory bowel disease, rheumatoid arthritis, cryoglobulinemia, systemic lupus erythematosus, solid tumors, and other neutrophilic dermatoses (pyoderma gangrenosum and Sweet’s syndrome).[3]

Dapsone is the first choice of treatment, usually showing a fast response. Other second-line therapies include steroids, tetracyclines, methotrexate, nicotinamide, and colchicine.

The erosive and bullous EED variant is very unusual and it has been reported most times in patients infected by human immunodeficiency virus.[4,5] Dapsone resistance is more common among these patients.

Bullous EED should be taken into account in differential diagnoses of bullous diseases.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.