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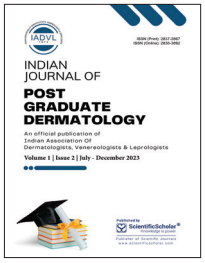

Clinicopathologic Challenge

Erythematous Plaques on the Extensors of the Extremities in a Middle-aged Female

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CLINICAL AND HISTOPATHOLOGICAL FINDINGS

A 62-year-old female presented with asymptomatic erythematous to brownish plaques on elbows, knees and dorsa of feet for 4 months. The lesions first appeared on her elbows and slowly progressed to involve the other sites. No history of joint pain, morning stiffness, bloody stools, diabetes, significant weight loss, prolonged diarrhoea or fever was elicited. On examination, multiple well-defined erythematous to brownish fibrotic plaques were present on the elbows, knees and dorsa of feet [Figure 1a and b]. A biopsy was performed from the plaque on the knee, and histological examination showed an unremarkable epidermis, mild dermal oedema with moderate to dense perivascular and interstitial pandermal infiltrate composed of predominantly neutrophils with fewer eosinophils, lymphohistiocytes and plasma cells. There was small vessel vasculitis with leukocytoclasia, endothelial cell swelling and fibrin deposits within and around the vessel walls [Figure 2a-c]. Direct immunofluorescence was negative. Rheumatoid factor, antinuclear antibody, fasting and postprandial blood sugar, human immunodeficiency virus (HIV), Venereal Disease Research Laboratory, hepatitis B surface antigen and anti-hepatitis C virus serologies were negative. Complete blood count, serum and urine protein electrophoresis did not reveal any abnormality.

DIAGNOSIS

Erythema elevatum diutinum (EED)

DISCUSSION

EED is a chronic and rare dermatosis which is a variant of leukocytoclastic vasculitis. It was first described by Hutchinson and Bury in the 1880s, and the condition was later named in 1894 by Radcliffe-Crocker and Williams.[1]

It presents with red to brown or yellowish papules, plaques or nodules distributed symmetrically over the extensor aspects of both upper and lower extremities, particularly involving the skin overlying the joints especially the elbows and knees.[1] Rarely, lesions may also occur on the face and ears. Initially, the lesions are soft, but eventually they fibrose and leave behind atrophic scars.[2]

EED has been associated with autoimmune diseases such as rheumatoid arthritis, coeliac disease, inflammatory bowel disease, type 1 diabetes and relapsing polychondritis. Associations with

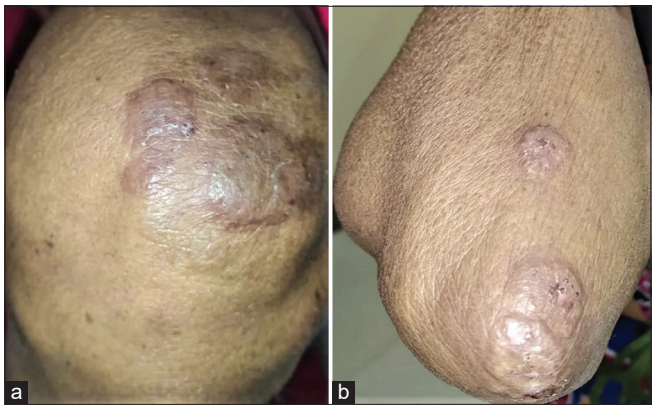

Figure 1: (a) Erythematous to brownish plaques present on the elbows, (b) Erythematous to brownish plaques present on the knees.

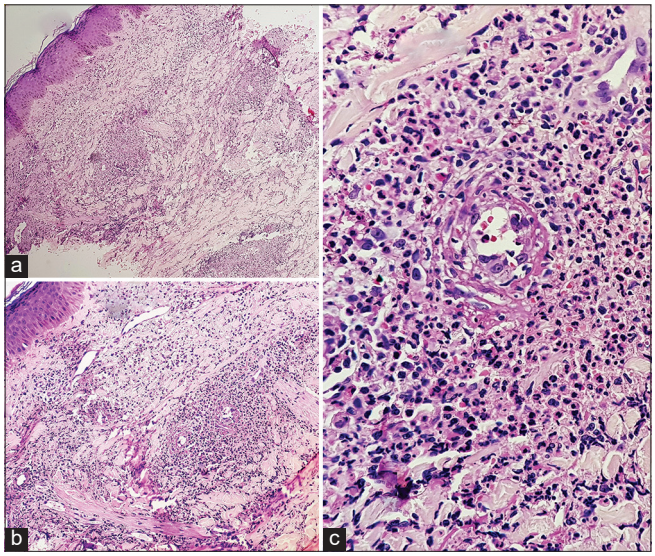

Figure 2: (a) Photomicrograph showing mild dermal oedema with moderate to dense perivascular and interstitial pandermal infiltrate (H and E, ×100). (b) Photomicrograph showing moderate to dense perivascular infiltrate composed of predominantly neutrophils with fewer eosinophils, lymphohistiocytes and plasma cells (H and E, ×200). (c) Photomicrograph showing moderate to dense perivascular infiltrate composed of predominantly neutrophils with fewer eosinophils, lymphohistiocytes and plasma cells. There is small vessel vasculitis with leukocytoclasia, endothelial cell swelling and fibrin deposits within and around the vessel walls (H and E, ×400).

infections, including *Streptococcus*, hepatitis, syphilis and HIV as well as malignancies such as hypergammaglobulinemia, IgA monoclonal gammopathy and myelodysplasia have also been suggested.[3]

Acute lesions of EED are characterised by leukocytoclastic vasculitis, without fibrin deposition. Eosinophils may also be present in the upper and mid dermis. Chronic lesions develop angiocentric eosinophilic fibrosis, capillary proliferation and infiltration of macrophages, plasma cells and lymphocytes. Cholesterol deposits in histiocytes and in the extracellular tissue, called 'extracellular cholesterolosis' may be present in older lesions, which is responsible for the yellow look of the lesions.[2]

Clinical differentials of EED include Sweet's syndrome which presents as asymmetrical tender lesions located on the arms, face and neck. On the other hand, EED lesions are chronic, symmetrical and classically located over the dorsa of the hands and knees, buttocks and Achilles tendons. Granuloma annulare is another differential which presents as erythematous annular plaques over the extensors of upper extremities. Histopathological differentials of EED include granuloma faciale as it also shows small vessel vasculitis, but shows a mixed inflammatory infiltrate, with mainly eosinophils and plasma cells as well a grenz zone. It presents as soft reddish brown nodules or plaques, with prominent follicular orifices and telangiectasias on the face.[3]

Treatment should first focus on treating the associated disorder, which itself may lead to resolution of the lesions. The first-line therapy is dapsone, with most lesions responding to the therapy. Second-line therapies include niacinamide, topical and intralesional steroids along with topical dapsone gel.[3]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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