

Erythema elevatum diutinum associated with scleritis

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ABSTRACT

Erythema elevatum diutinum (EED) is a chronic and rare variant of leukocytoclastic vasculitis that is being reported nowadays frequently in association with HIV infection. It clinically manifests as asymptomatic to tender erythematous papules, plaques and nodules, usually with acral distribution and is rarely accompanied by systemic complaints other than arthralgia. The reported associations include preceding upper respiratory infections, hematological malignancies, lymphomas and monoclonal gammopathies. Here we report a 45 year old man with multiple joint pains, tender nodules over palms and soles and ocular pain and congestion, who was subsequently diagnosed as a case of EED and scleritis following histopathological examination. Skin as well as ocular complaints completely subsided with Dapsone monotherapy.

Key words: Dapsone, erythema elevatum diutinum, scleritis

INTRODUCTION

Erythema elevatum diutinum (EED) is a chronic and rare dermatosis that is considered to be a variant of leukocytoclastic vasculitis clinically manifesting as asymptomatic to painful erythematous papules, plaques or nodules. The lesions are usually distributed symmetrically on extensor surfaces of extremities. It is rarely accompanied by systemic features other than arthritis. Associated diseases include preceding streptococcal upper respiratory infections, Hepatitis B and HIV, lymphomas, hematological conditions and paraproteinemias. Here we report a case of sudden onset erythema elevatum diutinum in a 45 year old healthy man associated with severe arthritis and scleritis, with dramatic response to oral dapsone. Though arthritis is frequently reported with EED, so far, only one case of scleritis has been reported in literature.

CASE REPORT

A 45 year old man presented to us with multiple large joint pains of 8 days duration followed by development of multiple painful red raised lesions over both palms and back of both legs associated with redness of eyes.

There was no history of upper respiratory tract

infection, gastro-intestinal disturbances or fever. There was no past history of diabetes mellitus, hypertension or tuberculosis.

On examination multiple tender erythematous papules and firm nodules were present on both palms and both legs. [Figure 1] Bilateral scleral congestion was present. [Figure 2]. Both knee and ankle joints were swollen and tender with restricted movements.

There was no clinical evidence of Hansen's disease and no acid fast bacilli were detected by slit skin smear using modified Ziehl Nielsen technique.

Based on history and clinical examination a tentative diagnosis of Erythema elevatum diutinum (EED) was made which was substantiated further by a skin biopsy from a representative lesion showing a hyperkeratotic, acanthotic epidermis overlying dermis containing perivascular mixed inflammatory infiltrate comprising lymphocytes, neutrophils along with nuclear dust and fibrinoid necrosis of the vessel wall and extravasation of RBCs. There was presence of focal areas of basophilic collagen [Figures 3 and 4].

All hematological and biochemical tests were within normal limits except raised total leukocyte

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Figure 1: Erythematous nodules and plaques over both palms



Figure 2: Ocular congestion showing scleral vessels, more prominent over right sclera

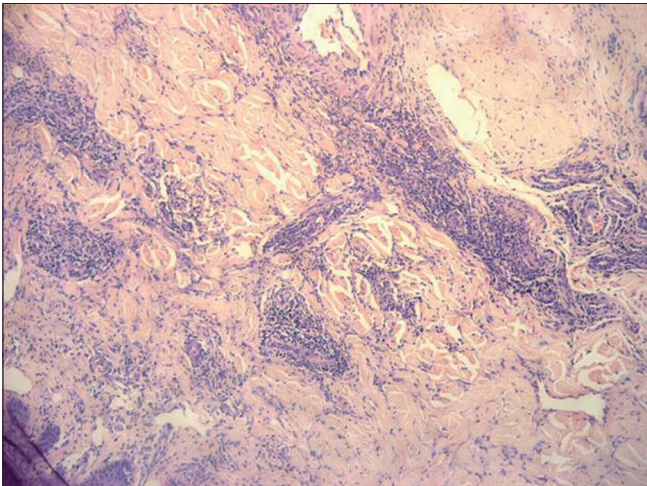


Figure 3: Photomicrograph showing hyperkeratotic, acanthotic epidermis overlying dermis containing perivascular mixed inflammatory infiltrate comprising lymphocytes, neutrophils along with nuclear dust and fibrinoid necrosis of the vessel wall and extravasation of RBCs with focal areas of basophilic collagen (H and E, $\times 10$)

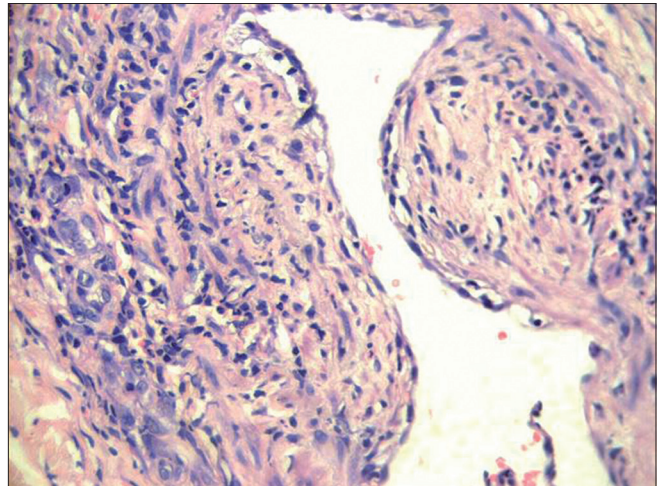


Figure 4: Photomicrograph focusing on a dilated blood vessel with edematous wall, surrounded by inflammatory infiltrate containing lymphocytes, neutrophils and nuclear dust (H and E, $\times 40$)

count and ESR. Other immunological and serological tests were negative. Ophthalmological opinion was sought and the clinical diagnosis of scleritis was confirmed.

Patient was started on 100 mg dapsone per day along with NSAIDs, to which there was a dramatic response with clearance of cutaneous, ocular and joint lesions within a couple of weeks' time and the patient was lost to follow-up after 3 months, by which time his lesions had considerably cleared up and he was advised to stop Dapsone.

DISCUSSION

Erythema elevatum diutinum (EED) is a rare chronic cutaneous eruption, currently classified under vasculitis, that is most

commonly seen in adults and is characterized by fibrosing red-violaceous, red-brown or yellowish papules, plaques or nodules which most commonly appear in a symmetrical fashion over the dorsa of the hands, the knees, buttocks and Achilles tendons.^[1]

It is a fairly uncommon condition; the only major case series reported consist of 13 cases from a large tertiary referral centre.^[2] While entertaining a diagnosis of EED other clinically similar diseases like Sweets syndrome, erythema multiforme and multicentric reticulohistiocytosis have to be ruled out. A skin biopsy is the most conclusive diagnostic measure for EED and exhibits features of small vessel vasculitis with predominant neutrophils and nuclear dust along with perivascular fibrinoid degeneration in the early lesions, whereas characteristic granulation tissue and fibrosis is seen in later lesions.

Extra cellular cholesterosis (due to liberation of cholesterol esters from damaged tissue which later get deposited in the surrounding), if observed in the fibrotic tissue, is another pointer towards EED.^[3]

EED is postulated to be caused by immune complex deposition in small vessel walls which induce inflammatory cascade, activation of cytokines like interleukin 8 which cause selective recruitment of leukocytes to blood vessels, leading to repetitive damage to blood vessels resulting in fibrosis and appearance of cholesterol crystals and myelin figures.^[4,5] EED has been associated with hematological diseases, recurrent streptococcal bacterial infections, hepatitis B and HIV, rheumatological disease, collagen vascular diseases, monoclonal gammopathies and lymphomas. The most common association is joint disease, though there are isolated reports of ocular abnormalities like nodular scleritis, panuveitis, autoimmune keratolysis and peripheral keratitis.^[6-8] Immunocomplexes may induce uveitis in EED.

Sulfones like dapsone and sulfapyridine are very effective in the treatment; other agents used include other immunosuppressants like cyclophosphamide, especially if associated with hematological abnormalities, SLE, paraprotenemias or leukemias. Niacinamide may also help as it acts as an anti-inflammatory agent and vasodilator. Intralesional corticosteroids may be tried in isolated plaques.^[1]

To conclude, our patient had a sudden onset of multiple erythematous plaques and nodules on flexor aspects of legs and on palms along with features of scleritis preceded by multiple large joint involvement. We could not elicit history of any preceding infection or illness. All the routine hematological and immunological tests were normal. He was diagnosed based on clinicopathological findings and was started on tab Dapsone 100 mg orally once at night. Interestingly, he showed a dramatic response with resolution of joint, eye and skin lesions within

a couple of days' time. To the best of our knowledge, there is only one more case report of scleritis associated with EED, but which did not respond well to oral Dapsone therapy and necessitated oral steroids.^[8]

Our report establishes that EED should be in the differential diagnosis of nodular lesions associated with joint and eye involvement, along with other classical causes like erythema nodosum leprosum, erythema nodosum, nodular vasculitis and panniculitis.

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