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Isolated unilateral erythema elevatum diutinum: a case report.

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Abstract

A 35 year old married female presented with asymptomatic swellings over lateral aspect of right foot since 1 year with occasional pain gradually increasing in size. There was no history of any other complaint. Rest of cutaneous and systemic examination including eye examination was within normal limits. Biopsy from one of the lesion was pathognomonic of erythema elevatum diutinum(EED). The patient was treated with oral dapsone with good response within 6 months.

Key words: Unilateral, erythema elevatum diutinum, dapsone

Introduction

Erythema elevatum diutinum (EED) is a rare, form of cutaneous leukocytoclastic vasculitis, characterized clinically by red-to-yellow-brown papules, nodules, and plaques, typically affecting extensor surfaces and the dorsal aspects of joints in a symmetric distribution.[1] EED may be associated with several immunological and infectious diseases, collagen vascular diseases, hematological disorders, inflammatory bowel disease, B-Cell lymphomas, Myeloproliferative disorders.[2] Unilateral EED is a rare occurrence.

We report a case of isolated and unilateral EED treated successfully with oral dapsone therapy.

Case report

A 35-year-old female presented with history of asymptomatic brownish raised lesions over the lateral aspect of right foot of one year duration. There was history of gradual increase in the number and the size of the lesions. The lesions were not associated with itching and burning sensation or pain. There was no history of preceding or associated upper respiratory tract infection, gastro-intestinal disturbances, fever and malaise. She was

not a known diabetic or hypertensive and had no past history of tuberculosis or contact with tuberculosis.

On examination her vital parameters were within normal limits. There were multiple unilateral brownish nodules on the lateral aspect of right foot (**Fig 1**). No similar lesions were seen anywhere else. The nodules were brownish, smooth surfaced, firm in consistency, non-tender and not fixed to the underlying tissues. There was no significant lymphadenopathy, hepatosplenomegaly or joint deformity or any eye complaint.



Fig 1: Multiple brownish nodules over right lateral foot.

The differential diagnosis of Keloid, fibromatosis and late stage Kaposi's sarcoma were thought and patient was investigated.

The complete hemogram, blood biochemistry, X-ray chest and electrocardiogram were normal. ASLO titer was normal and throat swab did not show any growth. The tests for rheumatoid factor, ANA, dsDNA as well ELISA for HIV were negative. Serum IgA, IgG and IgM levels were within normal limits. Urine examination for Bence-Jones proteins was negative. Eye examination revealed no abnormality.

Skin biopsy from a nodule revealed normal epidermis and a circumscribed dense dermal infiltrate of predominant neutrophils with few lymphocytes, eosinophils especially in the perivascular areas with fibroplasia and thickening of collagen (**Fig 2,3**). The blood vessels showed endothelial cell swelling, infiltration of walls with neutrophils, extravasation of erythrocytes and perivascular nuclear dust and perivascular fibrosis (**Fig 4**).

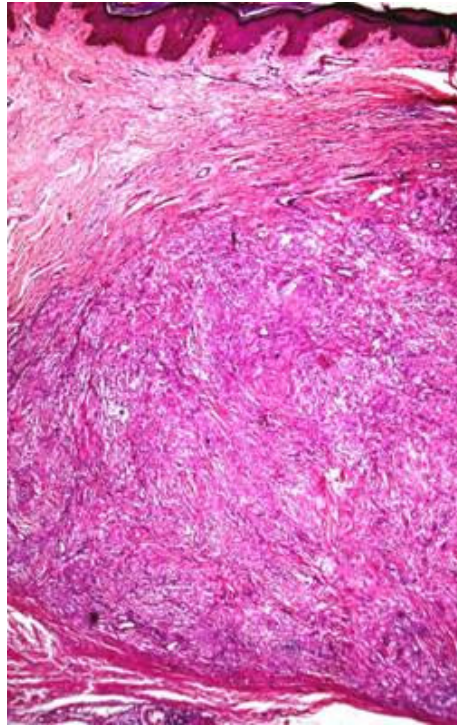


Fig 2: A circumscribed collection of inflammatory infiltrate in dermis with fibroplasia. (H&E, 25X).

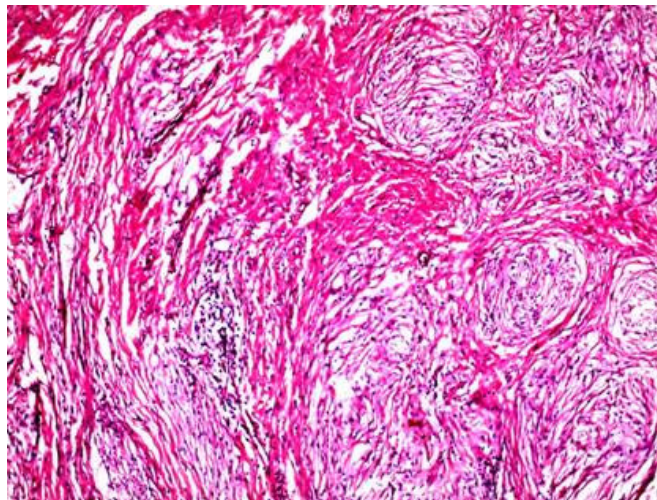


Fig 3: Dense mixed infiltrate with predominance of neutrophils, nuclear dust, perivascular fibrosis and vessel damage. (H&E, 100X)

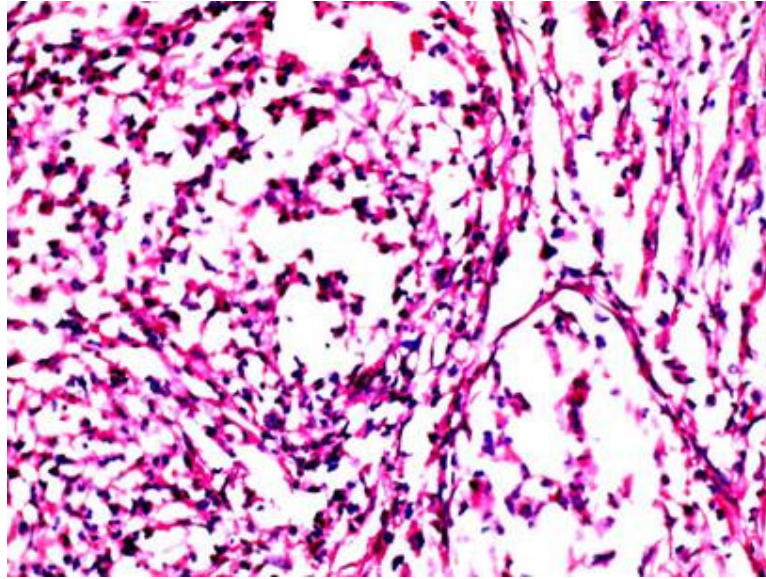


Fig 4: The infiltrate invading vessels shows neutrophils with nuclear dust. (H&E, 400X).

On clinico-pathological correlation the final diagnosis of unilateral isolated erythema elevatum diutinum (EED) was made. The patient was started on 100 mg of dapsone daily at bed time. A significant flattening of the lesions was noticed by the end of six months of therapy (**Fig 5**).



Fig 5: Nearly complete flattening of lesions after 6 months of dapsone therapy.

Discussion

It was first described by Hutchinson in 1888 and by Bury in 1889. [2] It is a rare form of cutaneous leukocytoclastic vasculitis, characterized clinically by red-to-yellow-brown papules, nodules, and plaques which typically occur over extensor surfaces and the dorsal aspects of joints in a symmetric distribution. The trunk and the mucosae are usually spared. Rare presentations include vesicular, bullous, and ulcerative lesions. Lesions are generally asymptomatic but occasionally may be tender, painful, or pruritic. Constitutional symptoms like arthralgias and fever may occur though systemic vasculitis is not present. It occurs usually in third to sixth decade but childhood cases especially with vesiculo-bullous lesions also occur. The disease follows chronic course with frequent relapses although it may spontaneously remission may occur rarely. Lesions tend to become firm and indurated over time and heal with pigmentary changes and atrophic changes with loss of collagen in the dermis. [1,2,3]

Exact pathogenesis of EED is not known but it is thought to be a consequence of an arthus like reaction to bacterial or viral antigen with formation of circulating antigen antibody immune complexes that get deposited in dermal vessels leading to activation of complement cascade and neutrophil activation with consequent vascular damage. [2,4,5]

EED is well known for its systemic associations like HIV infection, chronic/recurrent streptococcal infection, verrucous cutaneous carcinoma, immunologic disorders, collagen vascular diseases like rheumatoid arthritis and lupus erythematosus, hematological diseases like IgA monoclonal gammopathy, myeloproliferative disorders, inflammatory bowel disease, gout, scleritis and uveitis and a thorough examination and work up of such cases is warranted to find any systemic problem. [1,2,4,6]

It needs to be differentiated from similar conditions like granuloma faciale, granuloma annulare, reticulohistiocytosis, xanthomas, Kaposi's sarcoma, pyoderma gangrenosum, dermatitis herpetiformis, Sclerosing hemangioma and dermatofibroma. Histopathology is helpful to rule out many of these conditions. [1,2] Histologically early lesions show a dense, perivascular, predominantly neutrophilic infiltrate involving superficial and mid dermis with leukocytoclasia, fibrin deposition and vascular injury though eosinophils, plasma cells and histiocytes may be seen. Older lesions show perivascular fibrosis with onion-peel appearance, intracellular cholesterol deposition and capillary proliferation. [1] Thus skin biopsy is usually diagnostic and other investigations like direct immunofluorescence and electron microscopy may be done in doubtful cases. [2]

Dapsone is the treatment of choice with good therapeutic response as was seen in our case. Various other drugs like colchicine, sulfapyridine, niacinamide and tetracycline and steroids either topical, intralesional or oral have been used with variable success. Most important part of treatment is to unmask any serious systemic disease. Patients with IgA gammopathy may need intermittent plasma exchange in addition to above measures.

Our case showed good response to oral dapsone had no any systemic association and lesions were unilateral, which is rare in EED.

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