Erythema elevatum diutinum in association with IgA monoclonal gammopathy: A rare case report

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ABSTRACT

Erythema elevatum diutinum (EED) is a rare form of vasculitis characterized clinically by red-violet brown papules, plaques, and nodules mainly involving the extensor surfaces; histologically by leukocytoclastic vasculitis in early lesions, and fibrosis and cholesterolosis in late lesions. EED has been associated with many systemic disorders including infections, autoimmune disorders, and both benign and malignant hematological disorders. As it is a rare form of vasculitis and only 250 cases reported till date, we report a case of EED in association with IgA monoclonal gammopathy with partial response to dapsone treatment.

Key words: Erythema elevatum diutinum, IgA monoclonal gammopathy, leukocytoclastic vasculitis

Access this article online

Website: www.idoj.in

DOI: 10.4103/2229-5178.185472

Quick Response Code:

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INTRODUCTION

Erythema elevatum diutinum (EED) is a chronic leukocytoclastic vasculitis, initially described in 1888 by Hutchinson. It can occur at any age but peaks in sixth decade with an equal sex ratio. EED has been associated with many infections, hematological disorders, connective tissue disorders, and inflammatory disorders. IgA monoclonal gammopathy is the commonest association of EED.

CASE REPORT

A 55-year-old woman presented with erythematous, violaceous papules, plaques, and annular lesions over her upper and lower limbs since 2 years. Lesions initially started as erythematous papules over both lower legs that resolved after taking oral prednisolone. There were recurrent episodes of healing with hyperpigmentation. There was no history of systemic symptoms.

On examination there were multiple erythematous to violaceous papules, plaques, and annular lesions, and purpuric lesions over both upper and lower limbs. Lesions were bilaterally symmetrical. Healed hyperpigmented macules and atrophic scars were present over the lower legs. There were dry necrotic ulcers over elbows and knees [Figures 1 and 2].

Routine laboratory investigations were normal except elevated erythrocyte sedimentation rate (40 mm 1st hr). Antinuclear antibody titers, retroviral and hepatotropic viral serologies were negative. The Venereal Disease Treatment Laboratory test was nonreactive. Antistreptolysin O titers were <200 mIU/mL. Chest radiograph, electrocardiogram, and Mantoux test were normal and slit skin smear was negative. Histopathology of violaceous plaque revealed leukocytoclastic vasculitis; dilated, thickened dermal blood vessels with prominent endothelial cells, neutrophilic infiltration within and around the dermal blood vessels, occasional eosinophilis, nuclear dust, and fibrin deposition around blood vessels [Figures 3 and 4]. Serum protein electrophoresis showed M-protein band

Cite this article as: Patnala GP, Sunandini AP, Rayavarapu R, Yandapalli PS. Erythema elevatum diutinum in association with IgA monoclonal gammopathy: A rare case report. Indian Dermatol Online J 2016;7:300-3.

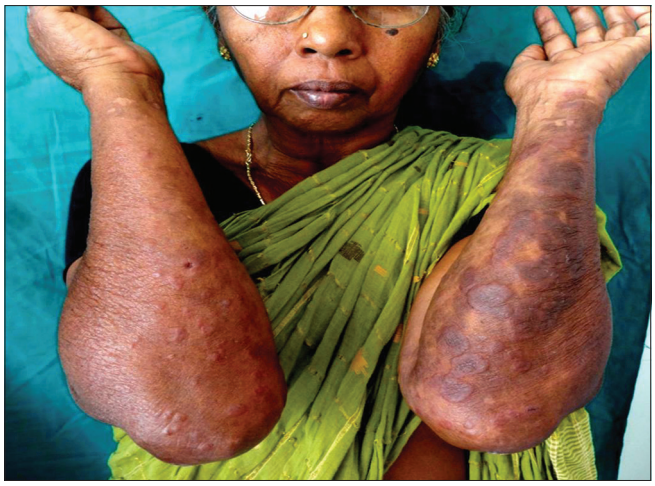

Figure 1: Multiple erythematous to violaceous papules, annular plaques over upper limbs

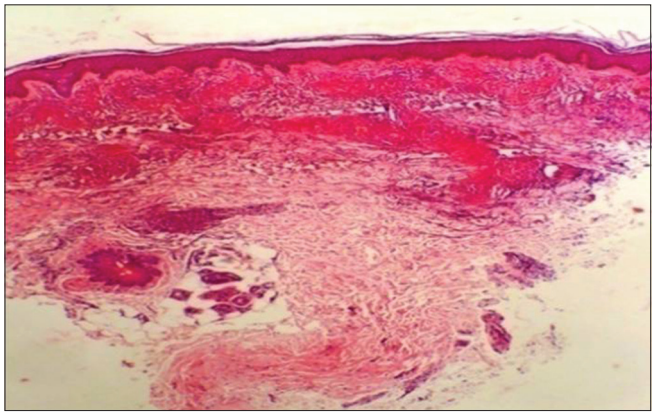

Figure 3: HPE (H and E stain, ×40): Leukocytoclastic vasculitis, deposition of fibrin and eosinophils

in beta-2 zone [Figure 5]. Serum immunoelectrophoresis was suggestive of IgA-lamda monoclonal gammopathy [Figure 6]. Urinary Bence–Jones proteins were negative. Bone marrow aspiration was normal. Axial skeletal survey showed no lytic lesions or osteoporosis. Serum calcium levels were normal. She was diagnosed as a case of EED associated with IgA monoclonal gammopathy. Treatment was started with dapsone 100 mg/day orally. Healing of lesions was observed after 48 h of treatment [Figures 7 and 8]. Lesions recurred after 2 weeks, and she was started on doxycycline 100 mg/day. Response was obtained with healing of old lesions and no new lesions.

DISCUSSION

EED is a rare, chronic dermatosis that can occur at any age, with an equal sex ratio[1] The condition however peaks in the sixth decade. Clinically, lesions present as firm, tender, brownish-red to purple, papules, plaques, or nodules. Extensor aspects of the extremities, usually near joints such as the fingers, hands, elbows, ankles, and knees are the

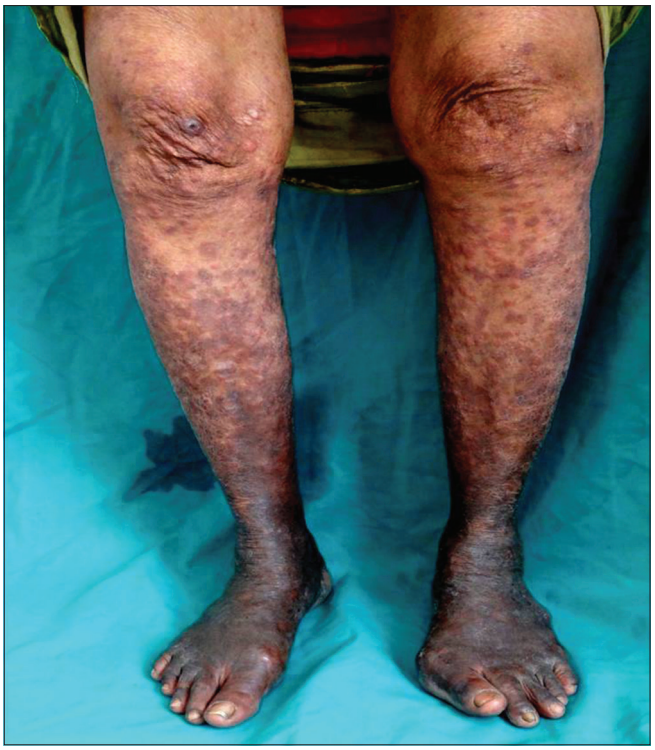

Figure 2: Multiple erythematous to violaceous papules, annular plaques and purpura, healed hyperpigmentation and atrophy over lower limbs

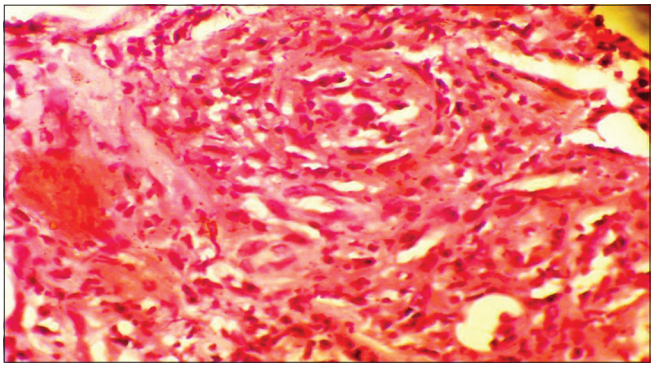

Figure 4: HPE (H and E stain, ×10): Leukocytoclasia with prominent endothelial cells

preferred locations. However, occurrences at atypical sites have been reported, including truncal, retroauricular, palmar, and plantar areas.[2] EED may also present as a solitary lesion.[3] The lesions of EED are usually asymptomatic, but pruritus, pain, and arthralgia of the involved joints have been reported.

Early lesions are characterized histologically by leukocytoclastic vasculitis,[1] with neutrophilic infiltrates around the blood vessels in the mid-dermis admixed with eosinophils, lymphocytes, plasma cells, and nuclear dust. Late lesions are characterized by mixed inflammatory cell infiltrate, fibrin deposition, cholesterol deposits in histiocytes, and extracellular tissue (extracellular cholesterolosis).

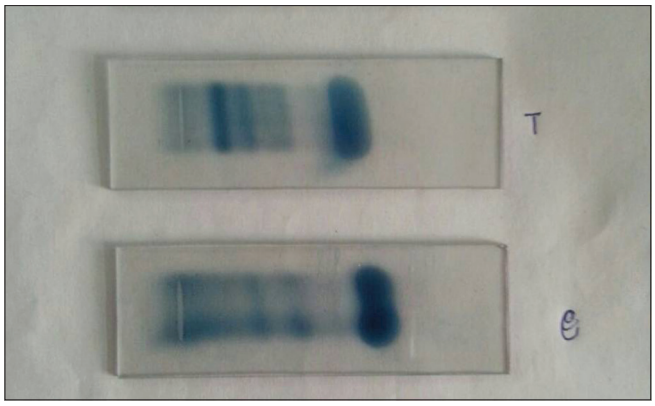

Figure 5: Serum electrophoresis: abnormal band in beta 2 zone in the test serum (T) upper slide, lower slide – control serum (C)

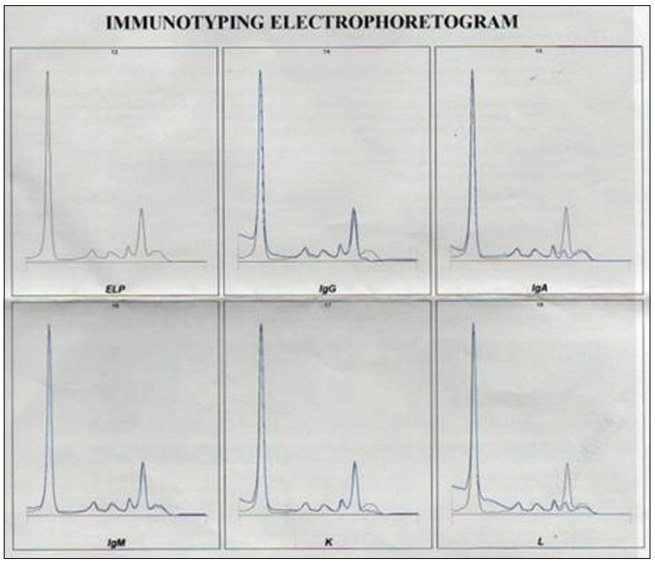

Figure 6: Serum immunoelectrophoresis: Blue graph represents patient's serum and black graph represents control serum

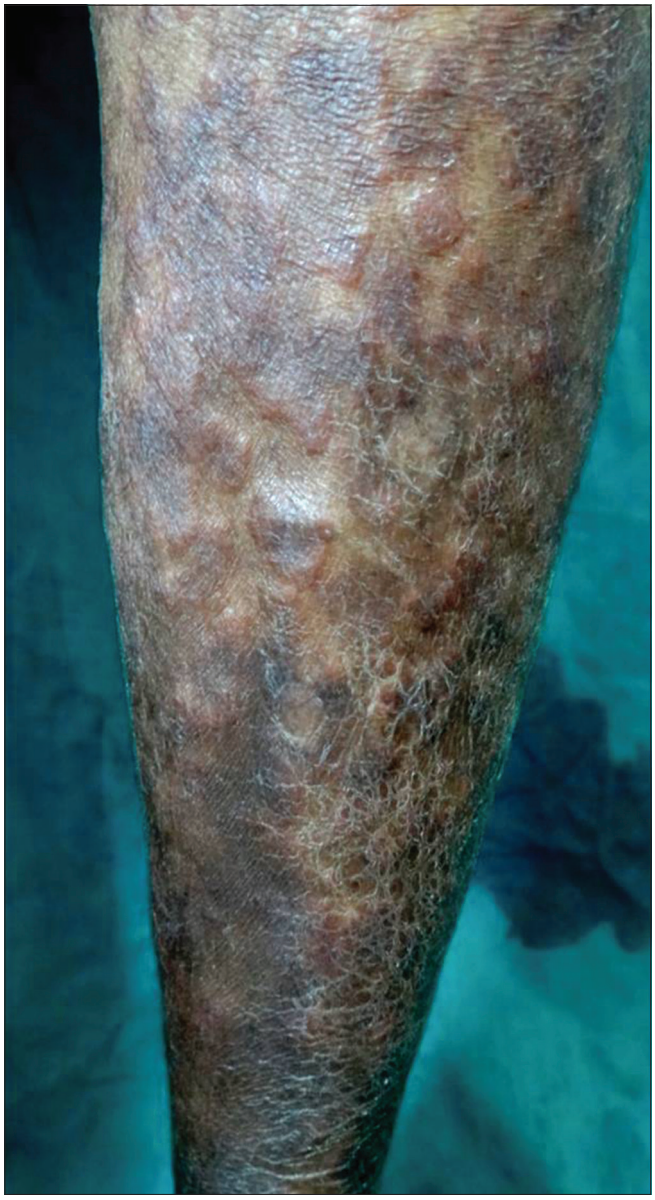

Figure 7: Before treatment with dapsone

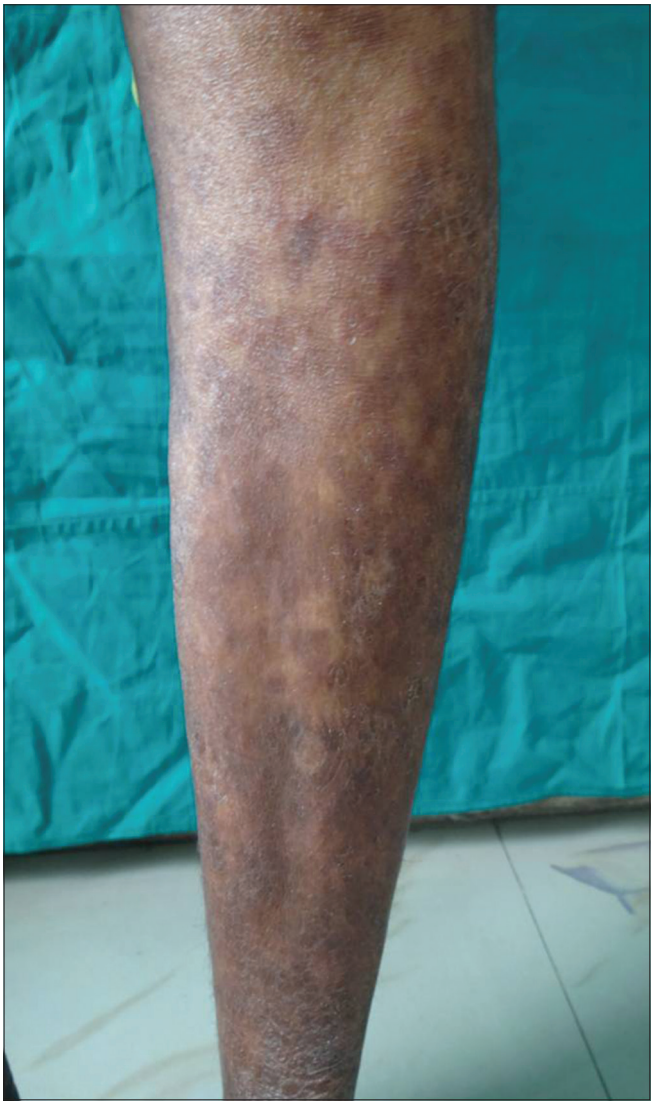

Figure 8: After treatment with dapsone on the third day

We report this rare form of cutaneous vasculitis characterized typically by red-brown to violaceous papules, plaques, purpuric

lesions, and necrotic lesions that were distributed over inner aspect of forearms and symmetrically over lower legs, healing with hyperpigmentation and atrophic scars. The case was diagnosed as EED on histopathology.

Most cases of EED have been described to be associated with a number of systemic diseases, including streptococcal infection, human immunodeficiency virus,[4] hepatitis B virus and syphilis, autoimmune diseases such as inflammatory bowel disease,[5,6] Wegener's granulomatosis, relapsing polychondritis, lupus erythematosus[7] and rheumatoid arthritis, hematological disorders such as plasma cell dyscrasias (multiple myeloma,[8] IgA monoclonal gammopathy[9]), lymphomas, and leukemias.[10]

The present case was associated with IgA monoclonal gammopathy. Multiple myeloma was ruled out as the patient was asymptomatic and there were no lytic lesions on axial skeletal survey. Bone marrow aspiration was normal. Serum total proteins, serum calcium, and alkaline phosphatase levels were in normal range.

Serum M-proteins are identified in 1% of asymptomatic healthy persons older than 50 years. This paraproteinemia without any signs and symptoms of multiple myeloma is called monoclonal gammopathy (IgA paraproteinemia). These patients are periodically followed up for multiple myeloma.

Association of EED with paraproteinemia especially monoclonal IgA gammopathy and IgA myeloma has been reported since 1977.[9]

In the review by Yiannias *et al*.[9] about 13 patients with EED, six patients had a hematologic abnormality, IgA gammopathy was the most frequent (four patients). EED preceded the myeloproliferative disorders by an average of 7.8 years in this study.

The present case was started on dapsone 100 mg/day, rapid resolution of lesions was observed after 48 h, recurrent lesions were managed by adding doxycycline, and the case was periodically followed for plasma cell dyscrasias.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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