International Journal of Research in Dermatology
Rijhwani M et al. Int J Res Dermatol. 2021 Jul;7(4):593-595
http://www.ijord.com

DOI: https://dx.doi.org/10.18203/issn.2455-4529.IntJResDermatol20212562

Case Report

Extensive erythema elevatum diutinum associated with IgA monoclonal gammopathy: a rare case report

Manish Rijhwani, Divya Yadav*, Manisha Nijhawan, Arvind Verma

Department of Dermatology, Mahatma Gandhi Medical College, Jaipur, Rajasthan, India

Received: 28 May 2021
Accepted: 15 June 2021

***Correspondence:**
Dr. Divya Yadav,
E-mail: rockin.diva3@gmail.com

ABSTRACT

Erythema elevatum diutinum (EED) is a rare form of leukocytoclastic vasculitis with an unclear pathogenesis. Almost 250 cases of EED have been reported in the literature, associated with several diseases and presented with various clinical features. Term 'diutinum' means chronic, describes one of the main characteristic features. The duration is very long, varying between one to more than 39 years. We report a rare presentation of EED in a 58-years-old man who presented with extensive and symmetrical, persistent, erythematous to violaceous plaques over trunk and limbs and popular-nodular lesions present over pinna and dorsum of hands with burning sensation and itching over few lesions, in association with IgA monoclonal gammopathy.

Keywords: Erythema elevatum diutinum, Extensive, IgA monoclonal gammopathy

INTRODUCTION

Erythema elevatum diutinum (EED) is rare, chronic leukocytoclastic vasculitis characterized by erythematous papules and nodules, occurring symmetrically on extensor surface. EED generally affects middle-aged population. First described by Hutchinson and Bury (1888), and named by Radcliffe-Crocker and William (1894).[1] Association with many infections, hematological disorders and inflammatory disorders is seen.[2] Etiology has been poorly understood.

CASE REPORT

58-years-old man presented with extensive, persistent, erythematous to violaceous bilaterally symmetrical plaque over face, trunk, limbs since 15 years (Figure 1). Erythematous plaques studded with firm papulo-nodules were present over the palms and sole, dorsum of hand and external ear since six months. Patient complained of itching and burning sensation in few lesions. Prominent onychorrhexis and onychodystrophy of few nails was noticed (Figure 2). Lesions initially started as erythematous papules over upper limbs. There were recurrent episodes of healing with hyperpigmentation and atrophy. No history of systemic illness. Patient was taking treatment from various doctors and was being treated as a case of psoriasis. We did biopsy keeping the differentials as mycosis fungoides, lichen planus, lupus erythematosus and EED and the histopathological report was suggestive of EED.

Routine laboratory investigations were normal except absolute eosinophil count (2.13 10^3/µl). Antistreptolysin-O titer was 144.5 IU/ml. VDRL, retroviral and hepatotropic-viral serologies were non-reactive. Chest X-ray, electrocardiogram were normal and Antinuclear-antibody titers, Mantoux test and slit skin smear were negative. Two biopsy samples were taken, one from a papulo-nodule and other from violaceous plaque. On histopathology, both showed diffuse infiltration and nuclear dust in reticular dermis with mild fibroplasia (Figure 3A). The neutrophilic and histiocytic infiltrate surrounded small thickened vessels. Histiocytes showed

pale vacuolated cytoplasm. Serum protein electrophoresis showed M-protein band in beta-zone (Figure 3B). Serum immune-electrophoresis was suggestive of IgA lambda monoclonal gammopathy (Figure 3C).

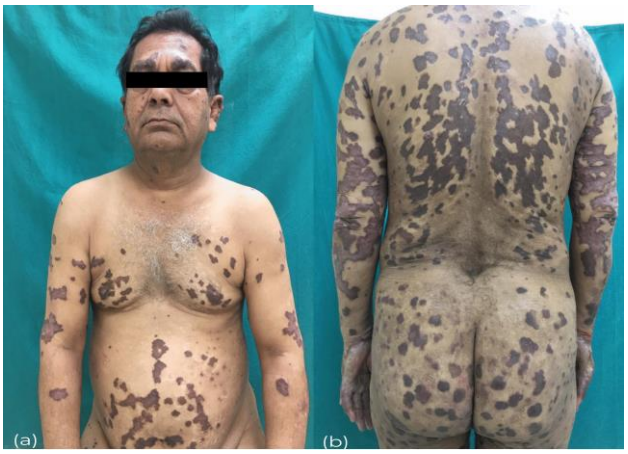

Figure 1: Multiple erythematous to violaceous plaques and few healed hyperpigmentation and atrophic patches over trunk and limbs.

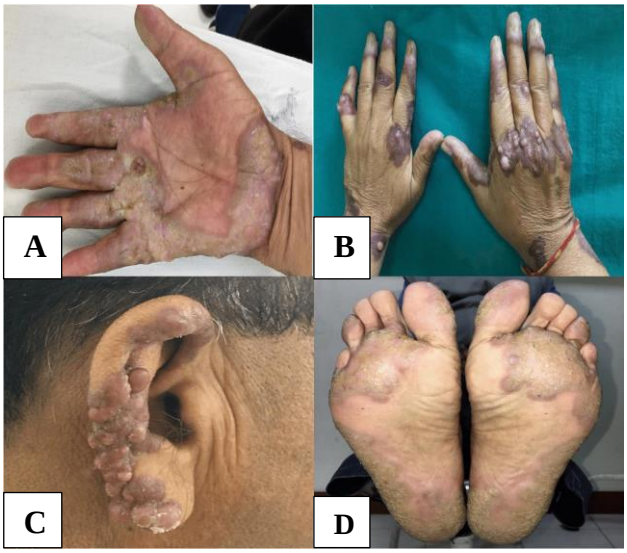

Figure 2: (A, B) Multiple erythematous to violaceous papules, nodules and annular plaques over hand; (C) multiple violaceous to red papule nodules over earlobe; and (D) soft and redundant skin with few nodules coalescing into plaques on the sole of both feet

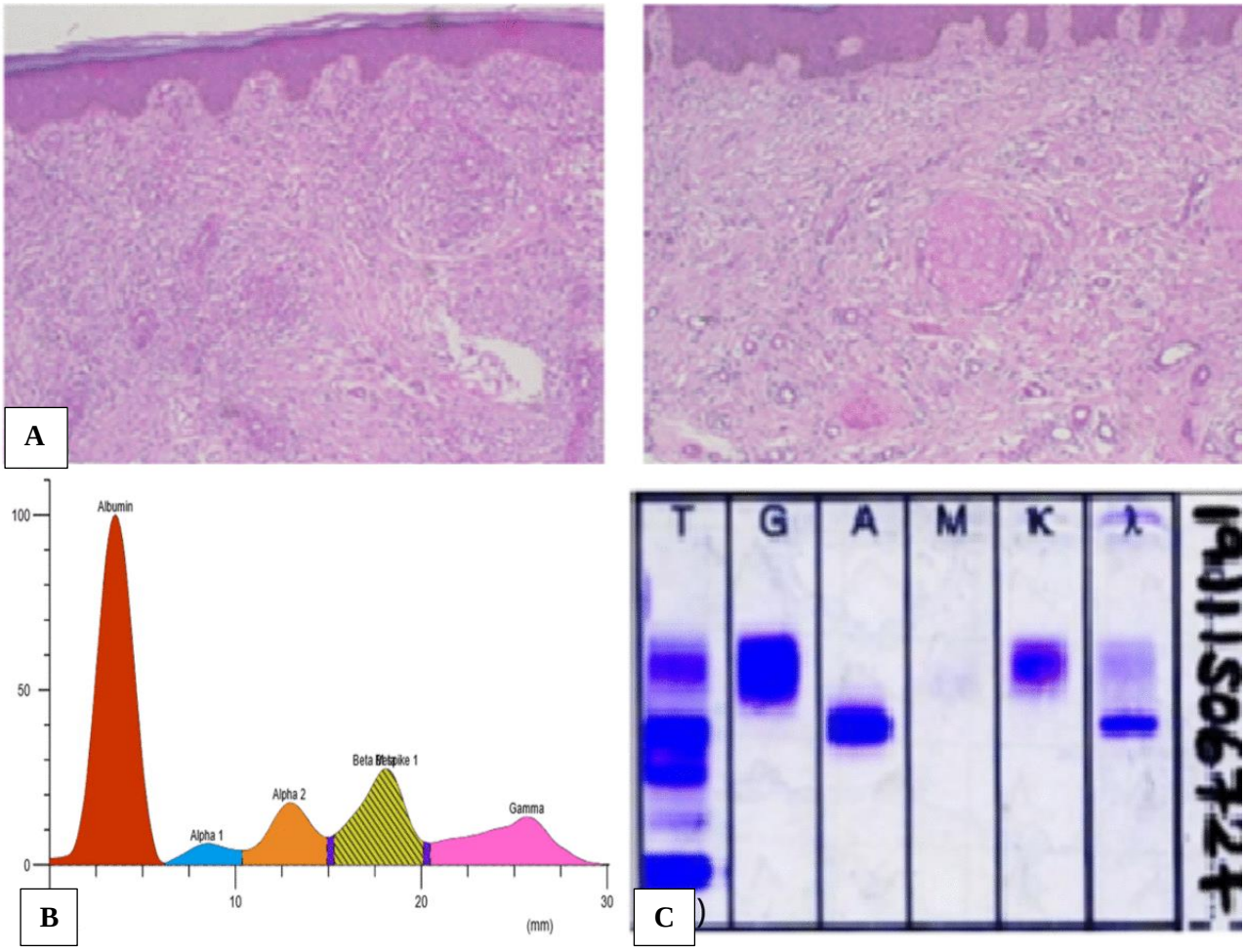

Figure 3: (A) EED at 100X magnification, H/E stain. Both the papule and plaque showed perivascular infiltrate containing neutrophils, with leukocytoclasis and some perivascular fibrin deposition; (B) serum electrophoresis: abnormal band in beta 2 zones in the test serum; and (C) IgA lambda monoclonal protein detected. (10.7 g/l).

Treatment was started with tablet dapsone 100 mg/day orally. Good response was seen within 15 days and lesions healed with hyperpigmentation. After 4 months of treatment, few similar lesions reappeared over shins and tablet colchicine 0.6 mg twice daily was added and improvement was seen. Intralesional triamcinolone 40 mg/ml is being given in papulonodular lesions of hands every three weeks, with good response.

DISCUSSION

EED is a rare form of cutaneous small vessel vasculitis, characterized by symmetrical, asymptomatic, persistent, violaceous plaques and nodules over the extensor surfaces of the extremities, with a predilection for the skin overlying joints, as well as the buttocks and the achilles tendon. Atypical sites like palm, sole, retroauricular and truncal area has also been reported.[3]

Pathogenesis of EED is not well understood. The widely accepted concept is that the immune complexes gets deposited in the post-capillary venules which activates the complement cascade and initiate leukocytoclastic vasculitis.[4,5] Langerhans'cell plays an important role in pathogenesis.[6] Ayoub et al suggested ANCA as marker of disease activity.[7] Association with various diseases like streptococcal infection, HIV, hepatitis B and syphilis, Wegener granulomatosis, rheumatoid arthritis is found.[8]

Eminent association of EED is with paraproteinemia, mainly monoclonal IgA gammopathy and IgA myeloma was reported in a study published by Arehimanditis et al (1977).[9] In Yiannias et al about 13 patients with EED, 6 had a hematologic abnormality, IgA gammopathy was most frequent.[10]

Therapeutic options include dapsone, tetracyclines, colchicine, nicotinamide, cyclophosphamide and corticosteroid injections.[11] First-line therapy includes treatment of underlying condition and dapsone.[12] Surgical management is done in severe and chronic cases.[13] Intermittent plasma exchange cleared flares of EED in patients with IgA paraproteinemia.[14]

CONCLUSION

To the best of our knowledge this case is a rare presentation of EED with extensive lesions. Patient is responding well to combination of dapsone and colchicines and intralesional therapy over papulonodules with decrement in severity of disease.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Rijhwani M, Yadav D, Nijhawan M, Verma A. Extensive erythema elevatum diutinum associated with IgA monoclonal gammopathy: a rare case report. Int J Res Dermatol 2021;7:593-5.