Atypical presentation of erythema elevatum diutinum: A case report

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ABSTRACT:

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Erythema elevatum diutinum is a rare primary cutaneous vasculitis of unknown etiology. The most common site of involvement is extensor surface of the extremities with a predilection for the skin overlying joints. However, there are some atypical presentations that involve palms and soles.

We report a case of 47-year-old Thai male presented with multiple erythematous to brownish plaques on his back and right hand, discrete erythematous to brownish nodules on right elbows and painful hyperkeratotic plaques on soles. The histopathology revealed focal leukocytoclastic vasculitis with areas of variable concentric fibrosis around vessels. Direct immunofluorescence study showed positive for IgA and C3 at superficial blood vessels. He was diagnosed as erythema elevatum diutinum and was treated with topical corticosteroid, colchicine and dapsone**.** The patient showed good result in clinical during follow up.

Key words: Erythema elevatum diutinum, atypical presentation

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Introduction

Erythema elevatum diutinum (EED) is an uncommon chronic cutaneous vasculitis with unprecedented cause. Characteristically, patients present with symmetrical erythematous plaques and nodules on the extensor surfaces of extremities[2]. There are various co-occurring diseases with EED include paraproteinemia, lymphoproliferative disorders, autoimmune disease, chronic infection especially hepatitis B, Streptococcal and HIV infections[1-4].

Case report

A 47-year-old Thai male presented with painful hyperkeratotic plaques on soles for 2 years. He had burning sensation on lesions of his feet when he was excited. When he walked, he felt pain on his feet. Six months later, he developed asymptomatic erythematous to brownish nodules and plaques at dorsum of right hand, elbows and back. The patient has no known underlying disease.

Dermatological examination revealed multiple erythematous to brownish plaques on his back and right hand, discrete erythematous to brownish nodules on right elbows and painful hyperkeratotic plaques on soles with burning sensation (Figure 1). Superficial lymph node was not palpable. Skin biopsy taken from his right hand and left sole showed superficial and deep mixed inflammatory infiltrate throughout the dermis consisting of mainly neutrophils and nuclear dust. There are areas of variable concentric fibrosis around vessels (Figure 2). Direct immunofluorescence study from right hand lesionshowed positive for IgM and C3 at superficial blood vessels and positive for IgA and C3 at superficial blood vessels from left sole lesion. Complete blood count, blood chemistry, liver function test, hepatitis profile, anti-HIV and prostate-specific antigen were unremarkable. The serum protein electrophoresis revealed increasing of beta globulin, consistent with hyperlipoproteinemia without detection of monoclonal spike.

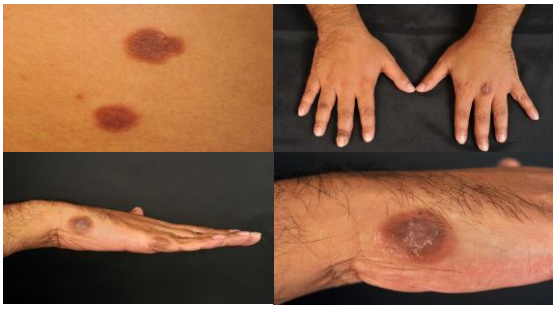

Figure 1 Clinical picture of patient. Multiple erythematous to brownish plaques on lower back and dorsum of hands.

He was diagnosed as erythema elevatum diutinum and was treated with dapsone, colchicine and topical corticosteroids with improvement during follow up period.

Erythema elevatum diutinum (EED) is an extraordinary form of primary cutaneous small

vessel vasculitis[1-3] with chronic relapsing course and spontaneous resolution[4]. It was initially described by Hutchinson in 1878. The name EED was later termed by Radcliff-Crocker and Williams in 1892[3]. Male are predominant than female, typical between the fourth and sixth decades[3]. The lesions are characterized by asymptomatic, red–brown papules, plaques and nodules on extensor surfaces of extremities. However, lesions on atypical sites such as palms and soles were also reported[1]. Typically, most lesions are asymptomatic but the patients may experience pain or a burning sensation after new lesions developed. Extracutaneous symptoms included arthralgia, fever or other constitutional symptoms.[3,5]

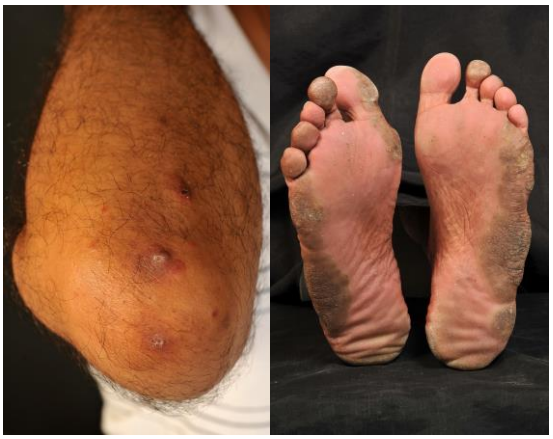

Figure 2 Clinical picture of patient. Erythematous to brownish nodules on right elbows. Multiple hyperkeratotic plaques on plantar surface of both feet.

Discussion

The pathogenesis of EED is still unknown, may be from deposition of immune complex in dermal blood vessels, which results in complement fixation and subsequent inflammation[3]. IgA anti-neutrophilic centromere antibody (ANCA) antibodies may be useful to be a clinical marker.[5]

There are various associated disease reported such as malignancy (multiple myeloma, B-cell lymphoma, breast cancer), infection (HIV, Streptococcus, hepatitis B), autoimmune disease (rheumatoid arthritis, diabetes mellitus, Crohn's disease, celiac disease), drugs (anti-tuberculosis drugs, cisplatin, erythropoietin) and paraproteinemia[2,3,4].

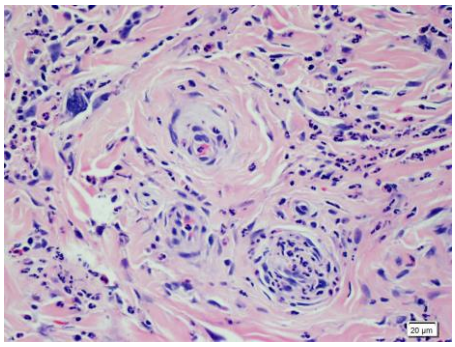

Figure 3 Histopathology shows the infiltrate consisting of mainly neutrophils, nuclear dust and lesser numbers of eosinophils and lymphocytes. There are areas of variable concentric fibrosis around vessels. Focal leukocytoclastic vasculitis is present. (Hematoxylin and eosin stain; original magnification ×40)

Dapsone is the mainstay treatment by impairing neutrophil function. If dapsone is not appropriate for the patients' condition, other less noxious agents such as colchicine, clofazimine, corticosteroids can be selected instead.[5]

Conclusion

We report an unusual case of erythema elevatum diutinum that response well to dapsone and colchicine. Histopathological and immunofluorescence studies were compatible with the disease. The clinical interest of this case is the history of burning sensation with hyperkeratotic plaques involving the soles. In this case, investigations were carried out to evaluate for underlying infection or malignancy and the results were within normal.

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