

CASE REPORT

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Unilateral solitary erythema elevatum diutinum

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Abstract

Background: Erythema elevatum diutinum (EED) is a rare disease with unknown etiology, various presentations, broad spectrum of underlying conditions, and unified pathology. So, every case of it merits further investigation for better understanding of its causes and pathophysiology.

Case presentation: I report the case of a 32-year-old otherwise healthy man with an 8-year history of a refractory solitary lesion on his right hand and prompt response to dapsone 100 mg BID within 2 weeks without any recurrence after 2 years of follow-up.

Conclusion: This case report aims to inform clinicians about EED in order to consider it in the differential diagnosis of any chronic refractory papule, nodule, or plaque on the relevant skin sites due to its protean manifestations, potential underlying diseases, and, finally, excellent response to dapsone.

Keywords: Vasculitis, Leukocytoclastic, Sulfone drugs

Background

EED is a rare distinctive leukocytoclastic vasculitis limited to the skin with characteristic distribution of skin lesions which are usually found symmetrically over the extensor surfaces of joints with a predilection for the skin overlying the small joints of the hands and knees and prompt response to sulfone medications [1]. It has been associated with various rheumatologic diseases, viral infections, celiac disease, and immunoglobulin A paraproteinemias [2]. Atypical presentations of the disease have also been reported in the literature such as involvement of the palms and soles and a solitary lesion in a patient with rheumatoid arthritis [3, 4].

The aim of this report is persuading clinicians to consider EED in the differential diagnosis of chronic refractory papules, nodules, or plaques on the characteristic skin sites. This suggestion is based on several reasons. First, EED usually has a protracted and insidious course

and mimics other conditions in its development. Second, the disease itself has atypical presentations in addition to its rarity which further complicate its timely diagnosis. Third, despite of the different unrelated conditions leading to EED, its pathological findings are constant and uniform which raise the question “is EED a distinct entity or an epiphenomenon?” which necessitates searching for underlying diseases. Finally, several studies have shown a good response to dapsone, therefore making it the treatment of choice.

Case report

A 32-year-old otherwise healthy man presented with an 8-year history of a tender skin lesion on his right hand. On examination, there was a well-defined firm red-brown plaque with some nodularity on its surface measuring about 2 × 2 cm just on the extensor surface of proximal interphalangeal joint of the third finger of right hand (Fig. 1A). The lesion had been started as a small red plaque that reached to its current size within 1 year. Other examinations were unremarkable. He was not smoker and reported no alcohol or illicit drug use. His family history included hypertension and a recently

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diagnosed polymyalgia rheumatica in his mother. Because of the failure of local and systemic treatments including corticosteroids, a biopsy had been taken 3 years ago which revealed pseudoepitheliomatous hyperplasia with mixed inflammation and no clear evidence of any specific diagnosis. He was reluctant to undergo another procedure. Inflammatory markers including erythrocyte

sedimentation rate and C-reactive protein reported normal. His rheumatologic profile, hepatitis B virus, hepatitis C virus, and human immunodeficiency virus serology were negative, and no monoclonal band was detected in plasma electrophoresis. Based on its location, appearance, refractoriness to common treatments, and a familial history of autoimmune disease, a clinical diagnosis of erythema elevatum diutinum (EED) was suspected, and a trial of dapsonsone 100 mg BID started with significant resolution of the lesion within 2 weeks which further supported the diagnosis (Fig. 1B, C).

EED is a rare distinctive leukocytoclastic vasculitis limited to the skin in which the post capillary venules of dermis is involved with intense neutrophilic infiltration [2]. The disorder is distinctive because of the characteristic distribution of skin lesion which usually found symmetrically over the extensor surfaces of joints with a predilection for the skin overlying the small joints of the hands and knees and prompt response to sulfone medications [1]. It has been associated with various rheumatologic diseases, viral infections, celiac disease, and immunoglobulin A paraproteinemias [2]. Atypical presentations of the disease have also been reported in the literature such as involvement of the palms and soles and a solitary lesion in a patient with rheumatoid arthritis [3, 4].

EED etiology is unknown, but deposition of immune complex and resultant complement activation have been hypothesized which subsequently lead to neutrophilic infiltration and fibrin deposition in and around small dermal vessels [2].

Conclusion

By considering of its rarity, atypical presentations, and multiple associated diseases, the question “is EED an epiphenomenon and therefore a final common pathway of various heterogeneous diseases or a distinct entity?” needs to be further investigated.

Awareness about the disease has important clinical implications because of the failure of common local and systemic treatments (e.g., corticosteroids), dramatic response to sulfone drugs such as dapsonsone or sulfapyridine, and requirement for screening of the potential underlying diseases and avoiding misdiagnosis.

Abbreviations

EED: Erythema elevatum diutinum.

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