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*Case Report*

# Erythema elevatum diutinum involving palms and soles: a case report and literature review

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**Abstract:** Erythema elevatum diutinum (EED) is a rare chronic inflammatory dermatosis and a part of the spectrum of cutaneous leukocytoclasticvasculitis. The most common site of involvement is extensor surface of the extremities, with a predilection for the skin overlying joints, particularly hands, feet, elbows and knees, as well as buttocks and Achilles tendons. Here we report a case of EED with atypical presentation involving palms and soles. The patient showed dramatic response to the treatment with prednisolone combined with *Tripterygium wilfordii glycoside* (TWP). The lesions improved significantly after three months of therapy. We will also review the atypical cases of EED that were previously published in English literature.

**Keywords:** Erythema elevatum diutinum, leukocytoclasticvasculitis, multiple myeloma

## Case report

A 51-year-old Chinese female presented with a bluish nodular lesions that first appeared on the lateral border of bilateral foot, gradually progressing to involve toes and soles. The lesion continued to grow and later involved fingers and palms within one-year duration (**Figure 1A-D**). The lesions were pruritic and painful enough to disrupt her in daily work. She was otherwise well, with no systemic symptoms. There was no relevant medical history, and she was not taking any regular medications. Clinical differential diagnoses were Sweet syndrome, palmoplantarkeratoderma, erythema multiforme, and bullous pemphigoid. Presentation at an atypical site could be the reason why EED was not suspected at a first glance. However, a biopsy was taken and the histological findings were consistent with EED (**Figure 2**). It made us to screen the patient for other conditions including malignancy. Laboratory investigations including complete blood count, renal, hepatic and thyroid function, serologies for syphilis, HIV and hepatitis, rheumatoid factor, antinuclear antibody, blood sugar, P-ANCA and C-ANCA, were all within the normal range. The only remarkable laboratory findings were: elevated M-protein in serum (35 g/l), elevated serum ß2 microglobulin (3.12 mg/l), increased ESR (78 mm/hour), increased kappa, decreased lamda, and kappa: lamda ratio = 6.04:1. Bone marrow aspiration showed clonal plasma cells > 13%. Based on these findings, the diagnosis of EED in association with asymptomatic multiple myeloma was made. The patient was referred to haematological examination. In agreement with the haematologist the following treatment regime was administered to the patient: oral prednisolone 15 mg once every day, oral TWP 20 mg three times a day, topical application of indomethacin solution and tacrolimus ointment once daily. On subsequent dermatological follow up visits, the rapid regression of the lesions was noticed. After two months of therapy, TWP was stopped and prednisolone was tapered. Topical medications remained the same. Treatment was discontinued after 3 months. The lesions after three months of treatment are shown in figure (**Figure 1E-G**).

## Discussion

Erythema elevatum diutinum is a rare and distinctive form of chronic cutaneous vasculitis clinically characterized by symmetrical distribu-

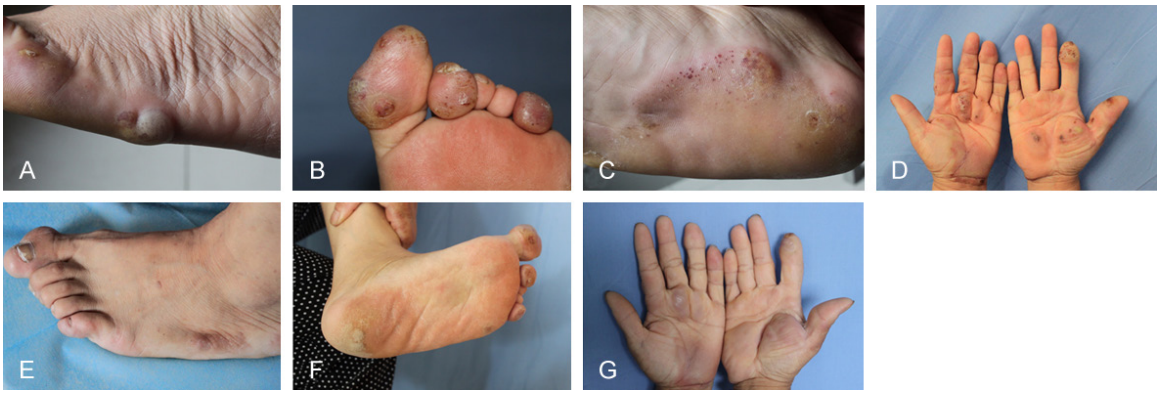


**Figure 1.** Clinical picture of patient before and after treatment. A: Voilaceous nodule on erythematous base on left lateral foot; B: Reddish papular lesions with scales involving multiple toes; C: Dry scaly plaque with multiple hemorrhagic spots of different size on right foot; D: Papulonodular lesion on palms. Some papules have coalesced to form plaque; E: 3 months after treatment lesion disappeared leaving slight hyperpigmentation; F: 3 months after treatment the lesion improved; G: 3 months after treatment decrease in the thickness of plaques and nodules.

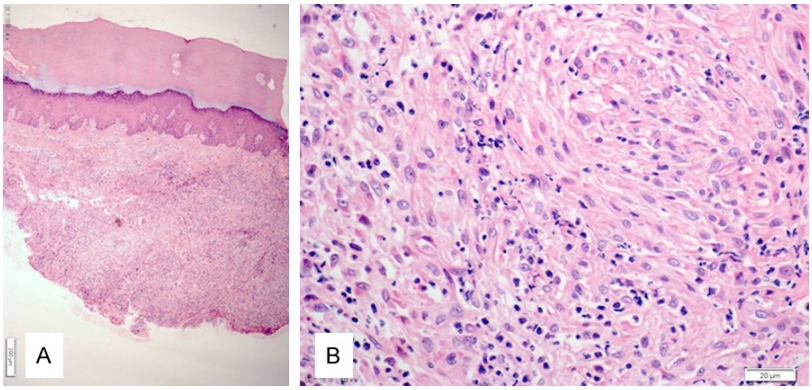


**Figure 2.** Histopathology. A: Histology shows diffuse neutrophil infiltration and fibroplasia in the dermis (Hematoxylin and eosin stain; original magnification ×4); B: Histology shows broken neutrophils and scanty eosinophils in the dermis (Hematoxylin and eosin stain; original magnification ×40).

tion of erythematous violaceous papules and nodules, isolated or confluent with hardened consistency over extensor surfaces of the extremities [1]. However, typical lesions at atypical sites or atypical lesions at typical sites have been reported (**Table 1**) [4, 6-10]. The etiology of EED is unknown, but it is presumed to be due to vascular deposition of immune complexes [2]. Although it is a chronic condition, most cases respond well to treatment with dapsone, which is considered to be the initial treatment of choice for EED [3]. Although in our case, dapsone was not given to the patient due to its' hematologic side effects. We planned to start prednisolone at a dose of 0.5 mg/kg, but the patient refused to take that amount of steroid because she was afraid she might have side effects. Therefore, we put her on minimum dose of prednisolone (15 mg) combined with TWP. As one of the extracts of *Tripterygium wilfordii* plants, TWP is a non-steroidal immune inhibitor with many pharmacological activities including anti-inflammatory and immune suppression [5, 11]. Our patient responded well to the given treatment regimen with decrease in the thickness of plaques and nodules and a regression of pruritus and pain within 3 months. TWP has been widely used to treat autoimmune and inflammatory conditions like rheumatoid arthritis, and psoriasis. In addition, its' intraperitoneal injection can inhibit graft versus host reaction, and also delayed-type hypersensitivity [12]. However, its' use in EED has not been previously reported. To our knowledge, our case is the only one, which was treated with prednisolone plus TWP, with a good treatment response. Therefore prednisolone + TWP can be a novel treatment for EED associated with multiple myeloma, but it has to be further evaluated.

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**Table 1.** Clinical features of atypical presentation of EED

| Reference | Patient age/sex | Duration of disease | Symptoms | Type of lesion | Site of involvement | Associated conditions | Treatment | Outcome |
|---|---|---|---|---|---|---|---|---|
| Futei et al. [4] | 62 y/M | 18 m | Pain & recurrent lesions | Verruca vulgaris like hyperkeratosis on soles, subungual hemorrhage, onycholysis, paronychia | Palms, soles, nails | Malignant B-cell lymphoma, benign monoclonal gammopathy | Cyclophosphamide, adriamycin, vincristine and prednisone | Eruption completely stopped reappearing |
| García-Meléndez et al. [6] | 45 y/M | 2 y | Chronic joint pain | Linear rope like plaques & voilaceous nodules | Palms, helixes | None | Denied by patient | Lost to follow up |
| Mohamadreza et al. [7] | 77 y/Fe | 2 y | Significant weight loss & low back pain | Verrucous plaques | Palms & soles | IgA gammopathy | Dapsone | Lesions began to soften and flatten within few days |
| Maruthappu et al. [8] | 55 y/Fe | 2 y | Swelling of fingers & joint pain | Hyperkeratoticvasculitic lesions with central necrosis, angular beaded plaque | Elbows, terminal digits, nape of neck | Bechet disease | Dapsone | Rapid regression of cutaneous lesions within 2 weeks |
| Dronda et al. [9] | 32 y/M | 1 m | None | Red nodules | Elbows, knees, soles, right wrist | HIV seropositive, CLD, infection with hepatitis B, C & D viruses | Dapsone | Skin lesions improved within 5 days and resolved completely over a 2-week period. |
| Ben-Zvi et al. [10] | 47 y/M | 2 y | Tender lesions | Asymmetrically scattered reddish-brown nodules | Upper back | None | ILCS | Flattening of lesions, which lasted during followup period of 1 year. |

Y: years, m: months, M: male, Fe: female, CLD: chronic liver disease, ILCS: intralesional corticosteroids.

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## Disclosure of conflict of interest

None.

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