

Erythema Elevatum Diutinum (EED): A Distinctive Vasculitis with Acute-on-Chronic Features

Dear Editor,

A 53-year-old man presented at our institution in June 2012 for a painful rash on his hands and feet. He had been treated prior to hospitalisation with topical and oral antifungals and antibiotics without success. Microbiological stains and cultures done were also negative. His past medical history included hypertension, Type 2 diabetes mellitus and end-stage renal disease.

Physical Examination

There were multiple tender violaceous plaques and nodules on his feet, legs and hands with an extensor predilection (Fig. 1). Pustulation was noted on some lesions. Trunk and face were spared. There was no lymphadenopathy.

Progress

He was investigated thoroughly to exclude an underlying infective process. Repeated full blood counts were normal, C-reactive protein was not elevated and erythrocyte sedimentation rate was 27 mm/hr. Anti-nuclear antibody and human immunodeficiency virus (HIV) serology was negative. Tumour markers such as carcinoembryonic antigen (CEA), alpha-fetoprotein (AFP) and prostate-specific antigen (PSA) were not elevated. A chest radiograph did not reveal any abnormalities. X-rays of the left foot showed chronic osteomyelitis of the second metatarsal (MT) head.

Histopathology

A skin punch biopsy of the left foot nodule was taken. Histopathology revealed dermal infiltrates of histiocytes and neutrophils (Fig. 2). There was epidermal acanthosis and parakeratosis. These features were considered to be in keeping with Sweet syndrome and he was started on oral prednisolone 30 mg per day.

Six weeks later, he reported minimal improvement. An alternative diagnosis of erythema elevatum diutinum (EED) was considered. On histopathological review, prominent endothelial cell swelling was noted and the neutrophilic infiltrates were confirmed to be perivascular rather than diffusely distributed, features less typical of Sweet syndrome and more consistent with EED (Fig. 2).

The patient was started on dapsone in August 2012. However shortly after, he was re-admitted with a severe pneumonia and eventually succumbed to sepsis. A review of the lesions while the patient was in the intensive care unit about 5 weeks after dapsone therapy showed a remarkable clearance of the lesions.

Discussion

EED is a rare, cutaneous form of leukocytoclastic vasculitis, characterised by nodules and plaques that may become indurated and tender over time.¹ Histopathologically, EED typically shows a perivascular



Fig 1. Photo showing the presence of reddish brown plaques and nodules involving the extensor surfaces of the small joints of the patient's hands.

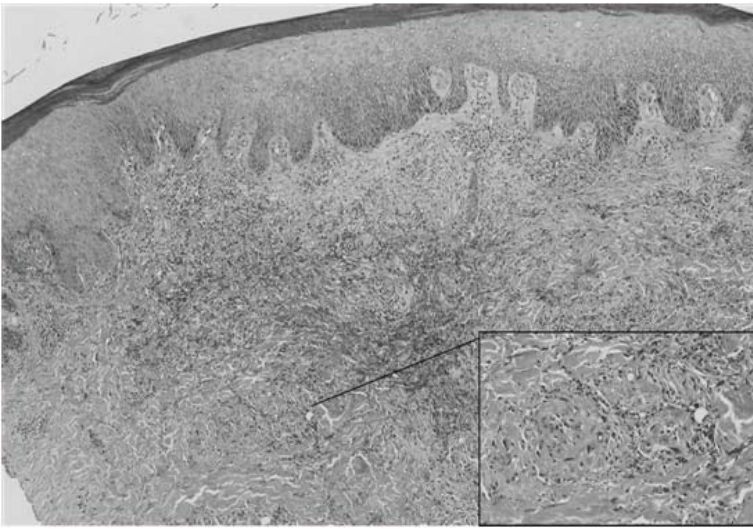


Fig. 2. A photomicrograph of the skin biopsy shows acanthotic epidermis with underlying perivascular and interstitial infiltrates of neutrophils, oedema and fibroplasia. Inset: Higher magnification view of the dermal blood vessels shows endothelial cell swelling with surrounding neutrophilic infiltrate (haematoxylin and eosin staining x40-main, x200-inset).

neutrophilic infiltrate that involves the superficial and mid dermis with fibrin deposition and endothelial swelling. Older lesions may demonstrate perivascular fibrosis and capillary proliferation.² The concomitant presence of the acute features of perivascular neutrophilic infiltrate and endothelial swelling and the more chronic features of dermal fibroplasia and epidermal acanthosis epitomises the histopathology of EED. Differential diagnoses of EED include urticarial vasculitis, neutrophilic dermatoses, Kaposi sarcoma, dermatitis herpetiformis, dermatofibroma, and granuloma annulare.³

Additionally, EED may occur in association with a variety of underlying disease entities such as malignancy, connective tissue and autoimmune disorders,⁴ immunocompromised states, and chronic infection.⁵ It was interesting to note that this patient had a concomitant chronic osteomyelitis of the second MT of his left foot. This patient had features consistent with EED but the diagnosis was unfortunately delayed due to a preoccupation to exclude infection and more common conditions like Sweet syndrome.

Although not usually fatal, EED can cause significant morbidity. Hence we hope to raise awareness of this rare condition so that prompt diagnosis and treatment can be instituted.

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