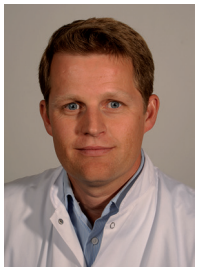
Recurrent UVA Photosensitive Erythema Multiforme



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Recurrent erythema multiforme (EM) causes substantial morbidity. Diagnosis and treatment are often prolonged and challenging. EM is most often related to herpes simplex virus (HSV), but other infections or systemic drugs can be implicated (1). Although the classic "target lesion" is an essential criterion for diagnosis, an atypical form, presenting with irregular-shaped blisters, has been described (2). This case report describes the clinical presentation, diagnosis and treatment of recurrent photosensitive erythema multiforme (PEM) presenting with typical "targets" and irregular blisters.

A 32-year-old woman presented at the acute referral unit of the Department of Dermatology, Gentofte University Hospital, Copenhagen, in late May 2014. She had had large bullae for 1 month, which had developed after a 1-week holiday in Egypt, where she had been exposed to the sun. After her return she had been very cautious concerning sun protection but new bullae were still developing. She did not take any prescription medication or over-the-counter drugs. Her medical history revealed no other complaints.

On clinical examination classical target lesions with central crusts were identified on the medial aspect of her left orbital area without involvement of sclera or conjunctiva. There was bilateral involvement of the nasal mucous membrane with oedema, inflammation and crusts. The right arm presented with a firm, large, fluid-filled bulla and a target lesion (Fig. 1A). The dorsal side of the hands and feet showed target lesions with central erosions (Fig. 1B). Culture of a throat swab was negative for haemolytic streptococci, expectorate was negative for Mycoplasma pneumoniae DNA, and bullae fluid was negative for herpes simplex DNA. Laboratory tests showed mild leucocytosis (10.4×10^9/l), in which neutrophils were predominant (8.13×10^9/l) and C reactive protein (CRP) 56 mg/l. Liver function tests were normal.

The woman had had similar rashes since she was a teenager, and a previous skin biopsy had shown subepidermal bulla with lymphocytes and epidermal apoptoses (Fig. 2). Over the years numerous swabs for herpes simplex had all tested negative and prophylactic treatment with acyclovir did not prevent new attacks. A photo provocation test induced oedema and erythema "en cocarde" inside and outside areas exposed to ultraviolet A (UVA) (3 standard erythema doses (SED)), but not to ultraviolet B (UVB) (2 SED).

Prednisolone, 37.5 mg/daily, was administered for 10 days and the blisters were lanced. The patient was advised to avoid sun exposure and to use sunscreen with a high UVA filter. The lesions healed without scarring and no new lesions appeared.

Recurrent vesiculobullous EM is a form of EM major that is most often triggered by HSV, and more seldom M. pneumonia or hepatitis C virus. Unfortunately in most cases the inciting factor is not identified (1).

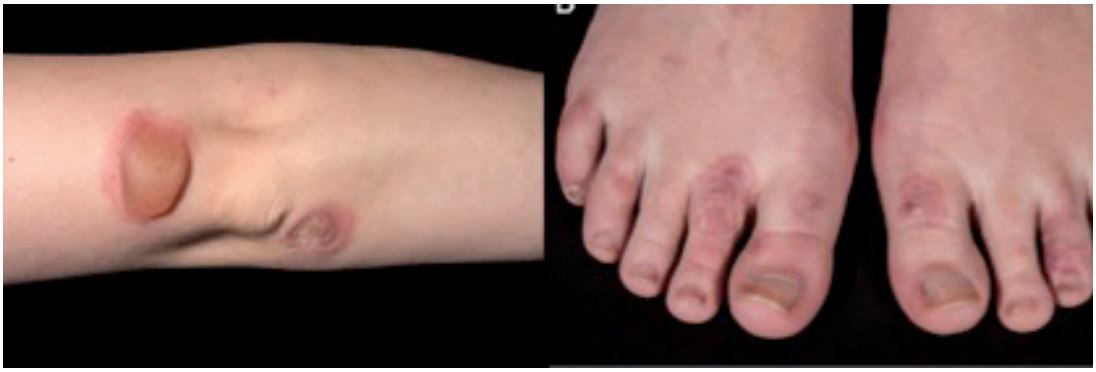

Fig. 1. Bullous and target lesions on: (A) the dorsal right arm, and (B) the feet in a woman with ultraviolet A (UVA)-induced erythema multiforme.

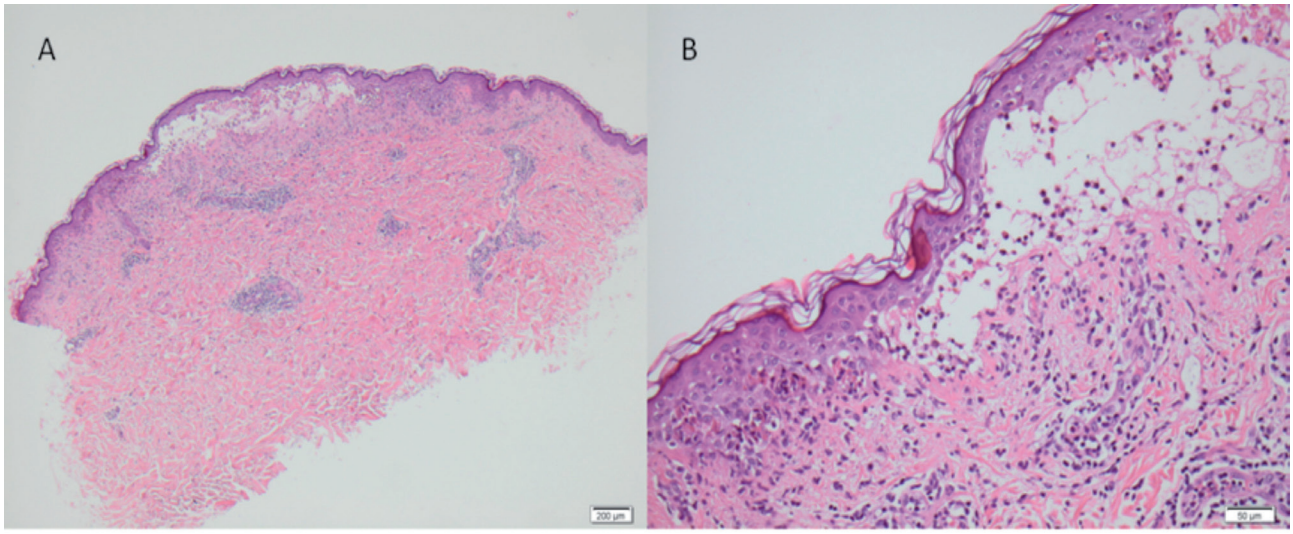

Fig. 2. Histopathology showing: (A) a central subepidermal bulla, with (B) dermal lymphocytic infiltrate and many apoptotic cells in the lower epidermis.

In this case recurrent EM was provoked by UVA. The distinction between PEM and an EM variant of polymorphous light eruption has been described previously (3), and recurrent EM following sun-induced PLE has also been reported (4, 5). In these 3 cases there was a delay of 2 weeks between initial PLE eruption and development of EM.

The involvement of mucosa and non-sun-exposed areas, a confirmatory biopsy, a positive UVA provocation test without delay, and the absence of pruritus and HSV infection substantiate the diagnosis of recurrent PEM. Drug-photo-induced EM and UVB-induced EM have been reported previously, but, to our knowledge, recurrent vesiculobullous EM induced by UVA has not been described previously (6–9). Phototesting might be indicated in recurrent PEM and appropriate sun behaviour alone may be sufficient to prevent attacks.

References

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