


Atypical presentation of erythema multiforme

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DESCRIPTION

We report a case of a previously healthy adolescent girl, with a history of alpha thalassemia minor, admitted to the emergency department due to painful, symmetrical, non-pruritic, violaceous flat macules with an inflammatory halo throughout the upper arms for four days. The patient reported sore throat the week prior to admission, for which she was prescribed a non-steroidal anti-inflammatory drug (NSAIDs) for three days, with resolution of her complaints. On the third day taking the prescribed medication, the patient noticed the appearance of painless red macules on the dorsum of the left hand progressing centripetally through the arm, becoming more conspicuous and painful during the following days (figure 1). On clinical examination, the patient showed stable vital signs, with painful violaceous flat macules with an inflammatory halo, on both upper arms up to the elbow, measuring 2–10 mm, without coalescence. There were no signs of mucositis. Blood tests revealed known mild anaemia, an erythrocyte sedimentation rate of 7 mm/hour and CRP (C-reactive protein) of 1.1 mg/L. *Mycoplasma pneumoniae* and Epstein-Barr viral serologies were negative and the remaining studies were unremarkable, except for elevation of antistreptolysin O (ASO) titre of 608 IU/mL (normal <200 IU/mL), with negative anti-DNase B. This was considered to be unspecific since elevation of ASO titres can be seen in immune-mediated diseases.¹ Considering the history of NSAID use, the atypical appearance and the progressive nature of the lesions, differential diagnosis with fixed-drug eruption (FDE) was proposed. Cutaneous biopsy showed hydropic degeneration of the epidermis' basal layer, with apoptotic keratinocytes and areas of confluent necrosis in the spinous layer, as well as a superficial perivascular lymphocytic infiltrate consistent with erythema multiforme (EM) (figure 2). The lesions regressed within four weeks, without the need for any medical treatment, with complete restitution of the skin. EM is an acute, self-limited immune-mediated dermatosis, with acral distribution and distinctive symmetrical lesions, mostly affecting the extensor surfaces of the body with or without mucosal involvement.² Common aetiological factors are herpes virus simplex 1 and 2, Epstein-Barr virus, *M. pneumoniae*, immune-mediated diseases, immunisations, and in <10% iatrogenic-related due to medications such as sulfonamides, NSAIDs and amoxicillin, among others.^{2,3} When lesions do not regress, further investigation may be necessary considering its association with organ cell malignancies such as renal cell carcinoma or gastric adenocarcinoma.⁴ Diagnosis is made by



Figure 1 Red macules on the dorsum of the left hand progressing centripetally through the left arm.

observation of typical target-like lesions and clinical history, dismissing cutaneous biopsy in most cases.⁵ However, as in our case, the differential diagnosis between EM and FDE is not always straightforward and cutaneous biopsy may be warranted. FDE is a hypersensitivity reaction characterised by well-demarcated, violaceous, circular patches, recurring in the same anatomical region with exposure to an offending drug. Although self-limited in most cases, repeated exposure can evolve to a life-threatening form of FDE and withdrawal with future avoidance of the drug is recommended.⁶ A conservative approach to treatment of EM is recommended by discontinuing the drug in lighter cases. In moderate to severe cases, topical to systemic corticosteroids may be prescribed. If an underlying infectious cause is suspected, it must include appropriate treatment.^{4,5}

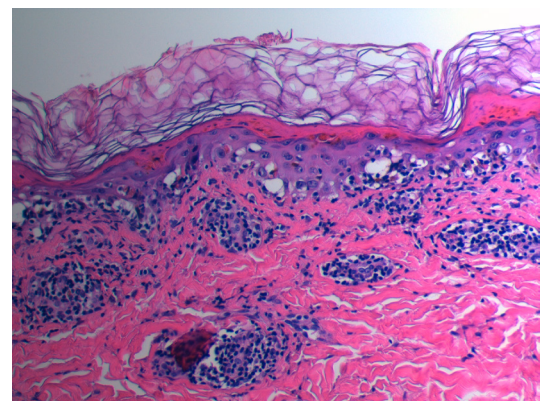


Figure 2 Vacuolar degeneration of the epidermis and keratinocyte apoptosis with lymphocytic superficial perivascular infiltrates (H&E ×100).



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Learning points

- ▶ Erythema multiforme (EM) is an acute, self-limited immune-mediated dermatosis, with acral distributed distinctive symmetrical lesions, mostly affecting the extensor surfaces of the body.
- ▶ Diagnosis is made through clinical history and observation, and in selected cases the differential diagnosis between EM and fixed-drug eruption may be difficult and a skin biopsy may be warranted.
- ▶ Most of the cases spontaneously resolve with no need for further treatment.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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