Erythema Multiforme

When targets lead to diagnosis

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Introduction

A 40-year-old Colombian patient, allergic to non-steroidal anti-inflammatory drugs and with no other relevant medical history, attended the emergency department because of a 4-day mucocutaneous eruption without associated fever. She had presented with a similar self-resolving clinical picture two years before, which was diagnosed as chickenpox. She did not take any medication and denied having had a herpes outbreak or suffered other infections in the previous few weeks.

On examination, she presented with multiple targetoid erythematous-violaceous circular plaques on the thorax, arms, and back of the hands, some with a central vesicle (Figure 1). The oro-genital mucosa showed painful hemorrhagic crusts and erosions (Figure 2).

Comment

Erythema multiforme (EM) was suspected clinically. A skin biopsy was performed and treatment was started with a tapered dosage of prednisone, topical methylprednisolone (1 mg/g), and ebastine (10 mg/12h). The biopsy showed vacuolar degeneration of the basal layer of the epidermis and spongiosis with isolated colloid bodies corresponding to an interface dermatitis. Direct immunofluorescence showed fibrinogen deposits within the basement membrane and IgG, IgA, IgM and fraction C3 within the
intraepidermal colloid bodies. Both results supported the diagnosis of EM. After two weeks, the lesions resolved, leaving residual hyperpigmentation.

EM is a sudden-onset hypersensitivity reaction which usually resolves within 2-4 weeks. It usually manifests in young individuals with target-like concentric erythematous-bullous skin lesions, predominantly affecting acral and facial areas. These lesions typically measure less than 3 cm in diameter, have a regular round shape with a central dusky zone and two concentric outer rings. EM targetoid lesions are often palpable and should be distinguished from the atypical “targets” that can be seen in Stevens-Johnson syndrome (SJS) or toxic epidermal necrolysis (TEN), which are round, asymmetric, non-palpable lesions with only two zones and a poorly defined border. The EM spectrum has been classified into major and minor forms, the latter associated with mild or absent mucosal affection and acral location of the skin lesions. Severe mucosal involvement is characteristic of major EM and usually consists in vesiculobullous lesions which rapidly develop into painful erosions that can give rise to serohemorrhagic crusts. General symptoms such as fever, asthenia and arthralgias usually precede and accompany the skin lesions.

Infections trigger a significant percentage of cases, the most common being herpes simplex virus and Mycoplasma pneumoniae. The administration of drugs such as sulfonamides, non-steroidal anti-inflammatory drugs and penicillines, account for less than 10% of cases.

EM requires differentiation from SJS/TEN as these conditions can be life threatening. Many other conditions can display targetoid vesiculobullous lesions or mucosal involvement and could be mistaken for EM, including urticaria, fixed drug eruptions, bullous pemphigoid, paraneoplastic pemphigus or Rowell syndrome. A skin biopsy could be helpful to distinguish these entities from EM. Possible findings on direct immunofluorescence include granular deposition of C3 and IgM at the dermo-epidermal junction. However, direct immunofluorescence is usually non-specific in EM and its purpose is to exclude other similar conditions.
Mild disease can be managed in most patients with topical steroids, analgesic agents and antihistamines. When identified, treatment of any underlying infection should be instituted. Patients with severe EM may require systemic steroids.

It is important to differentiate EM from other erythematous-bullous pathologies, with typical target-like lesions, a key diagnostic clue.

The patient in this manuscript has given written informed consent to publication of their case details.

References:
Figure 1: Clinical image: Classic lesions of erythema multiforme manifesting a targetoid appearance with quasi-vesiculation, distributed on the dorsal aspects of both hands.

Figure 2: Clinical image: Erosions and thick hemorrhagic crusting of the vermilion lips and the perioral region.