Paediatric erythema multiforme: not every bullous rash is chickenpox

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DESCRIPTION

A previously healthy 3-year-old girl presented to the emergency department with a 6-day-worsening pruriginous maculopapulovesicular rash with cephalocaudal progression predominantly in her face and upper limbs.

Two weeks before she had been admitted for a suppurative otitis media and was treated with systemic cefotaxime, fluconazole and topic clotrimazole and gentamicin.

At admission she was feverish and presented symmetrical distributed lesions on her face, upper and lower extensor surface of her limbs and scarce lesions on her trunk. Lesions evolved to round atypical target lesions with two different zones: a raised oedematous lesion with a central blister or necrotic centre. The lesions were at the same stage of evolution and affected her palms but not the scalp. Each individual lesion lasted more than 24 hours (figure 1).

Chickenpox followed by *Staphylococcus aureus* superinfection as bullous impetigo was considered and she was treated with flucloxacillin and clindamycin. There was a progressive worsening with oral mucositis with erythema and painful erosions and a superficial skin detachment of the lesions with a positive Nikolsky sign and 2% of body surface area epidermolysis (figure 2).

Stevens-Johnson syndrome (SJS) or an immune-mediated skin reaction as erythema multiforme (EM) was then considered. Skin biopsy was performed, systemic and topical steroids were started and antibiotics were changed to linezolid, in order to avoid beta-lactams. Laboratory investigation was extended. PCR skin swabs were negative to Herpes simplex virus (HSV) and Varicella zoster virus (VZV). Serological tests to HSV 1/2, VZV, HIV, Coxsackievirus, Cytomegalovirus and *Mycoplasma pneumoniae*, often associated with EM and

Figure 2 Atypical target lesion with a central bullae (black arrow). Malar superficial skin detachment is seen (yellow arrow, on the left). Severe oral mucositis is present on the right.

MIRM (*Mycoplasma*-induced rash and mucositis) were negative to acute infection. Blood and urine cultures were negative.

She was started on oral prednisolone 1 mg/kg/day for 7 days with progressive healing of the lesions and was discharged after 10 days. Besides residual skin hyperpigmentation, no complications associated with EM major were seen.

She has been followed up in the allergy outpatient clinic and was submitted to drug allergy tests. Specific immunoglobulin E to penicillin G and V, amoxicillin, ampicillin and cefaclor were negative. Skin prick tests and intradermal tests to paracetamol were negative. Epicutaneous tests to paracetamol, amoxicillin, flucloxacillin, ceftriaxone, ibuprofen, gentamicin, fluconazole and cefotaxime tested negative at 48 hours. Oral drug challenge tested negative to paracetamol. It was considered safe to use paracetamol, ibuprofen and amoxicillin on this child.

The atypical non-confluent raised target lesions with predominant acral distribution and centripetal spread made EM the most probable diagnosis supported by histological findings. Severe oral mucosa involvement classifies it as EM major.

Paediatric EM is rare and sometimes misdiagnosed. It was crucial to investigate differential diagnoses as life-threatening conditions such as SJS and possible causative agents such as infections, a well-known trigger in EM.¹⁻³ Although considered different clinical entities, both SJS and EM may be secondary to drugs, so it was prudent to stop possible causative drugs such as beta-lactams. Although EM is a self-limited condition, corticotherapy is suggested in severe cases, decreasing the days and severity of the rash.^{4,5}

Figure 1 Atypical target lesions: a round lesion with a raised oedematous border and a central blister or necrotic centre (black arrows).

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Learning points

- In contrast with chickenpox, erythema multiforme is characterised by lesions that evolve to typical or atypical raised target lesions with predominantly acral distribution; also, skin detachment is present, affecting less than 10% of the body surface area. PCR skin swabs allow to identify Varicella zoster virus in vesicles in chickenpox.
- Paediatric erythema multiforme is a rare condition, often misdiagnosed. It is a clinical diagnosis but skin biopsy may be needed to confirm it and exclude other conditions. Drugs are known precipitating factors. Differential diagnosis should include life-threatening conditions such as Stevens-Johnson syndrome, so it is important for the paediatrician to be aware of this condition.

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Contributors All authors were involved in the management of the patient to whom the case report refers to. AL and MB performed clinical history and collected the data and images. AL and SP wrote the manuscript, prepared the images and the learning points. All authors had a significant contribution, have revised and approved the manuscript for publication.

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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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REFERENCES

- 1 Zoghaib S, Kechichian E, Souaid K, *et al*. Triggers, clinical manifestations, and management of pediatric erythema multiforme: a systematic review. *J Am Acad Dermatol* 2019;81:813–22.
- 2 Read J, Keijzers GB. Pediatric Erythema Multiforme in the Emergency Department: More than "Just a Rash". *Pediatr Emer Care* 2017;33:320–4.
- 3 Siedner-Weintraub Y, Gross I, David A, *et al*. Paediatric erythema multiforme: epidemiological, clinical and laboratory characteristics. *Acta Derm Venereol* 2017;97:489–92.
- 4 Chan M, Goldman RD. Erythema multiforme in children: the steroid debate. *Can Fam Physician* 2013;59:635–6.
- 5 Kakourou T, Klontza D, Soteropoulou F, *et al*. Corticosteroid treatment of erythema multiforme major (Stevens-Johnson syndrome) in children. *Eur J Pediatr* 1997;156:90–3.

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