Recurrent anaphylaxis in an infant with Cutaneous Mastocytosis

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Introduction

Rashes during infancy are mostly secondary to infection or allergy but can be due to rare serious disorders with poor prognosis; as in this case.

Case Report:

A 10-mo-old boy presented with a rash since 5 mo of age. Vesicular eruptions appeared in the right axilla then spread to the trunk, neck and back, associated with irritability, colic and vomiting. The bullae became tense then confluent with sizes between 10-50 mm, with serous content and indurated erythematous base. The lesions spread further to the diaper area and extremities with positive Darier sign without involvement of mucus membranes.

Frequent flare-ups and anaphylaxis followed, and were related to hot climate, sun exposure, or bathing, recovered partially on treatment with parenteral dexamethasone. Food allergy was excluded. At age of 10 mo, he developed extensive hemorrhagic bullae (figure 1), protracted anaphylaxis and multi-organ failure, then transferred to the pediatric ICU for assisted ventilation, adrenaline IV drip, methylprednisolone.

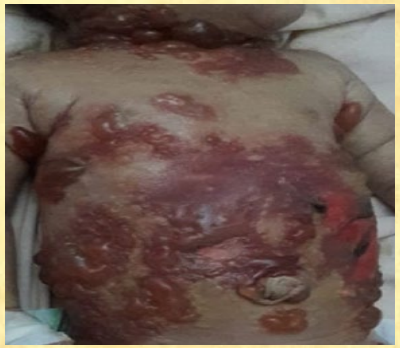

Figure 1: Skin lesions during the last severe exacerbation at 10 mo of age

Mastocytosis was suspected. Serum tryptase level was very high (120 ng/ml, Nl < 11). Skin biopsy revealed dermal and sub-epidermal vacuoles and dense mast cells infiltrate, compatible with bullous mastocytosis. Other investigations including bone marrow aspirate were normal supporting the diagnosis of diffuse cutaneous mastocytosis. C-Kit analysis did not reveal mutation.

Corticosteroids and tyrosine kinase inhibitor (imatinib mesylate) were administered. Anaphylaxis action plan, avoiding common triggers, skin care, tacrolimus ointment, ketotifen, were given. Prednisolone and imatinib were discontinued after 1 mo of improvement. Full recovery occurred by the age of 30 mo.

Conclusion:

Diffuse cutaneous bullous mastocytosis is a rare disease that typically presents during early infancy and the course is rather complicated with recurrent anaphylaxis. The course and prognosis are influenced by the density and distribution of the mast cells. Management depends on avoidance of triggers of mast cell activation, symptomatic therapies and in severe conditions, imatinib can be of value.