

A 9-month-old Child with Diffuse Cutaneous Bullous Erythema: A Quiz

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A 9-month-old boy presented to our outpatient clinic for a recurrent diffuse cutaneous transparent bullous rash accompanied by itching 4 months earlier. Erythema with erosion formed after the blisters ruptured, and gradually became dry and scabby. On physical examination of the involuting phase, there was generalized thickening of the skin and dark, geographic erythema with erosion or scabbing was present on the trunk (Fig. 1). Vital signs were normal. There was no evidence of systemic involvement (bone marrow, spleen, liver, gastrointestinal tract, etc.). Laboratory test

results, including complete blood cell count, liver and renal function tests were within the normal range.

What is your diagnosis?

- 1: Bullous diffuse cutaneous mastocytosis
- 2: Scalded skin
- 3: Toxic epidermal necrolysis (TEN)
- 4: Pemphigoid

See next page for answer.

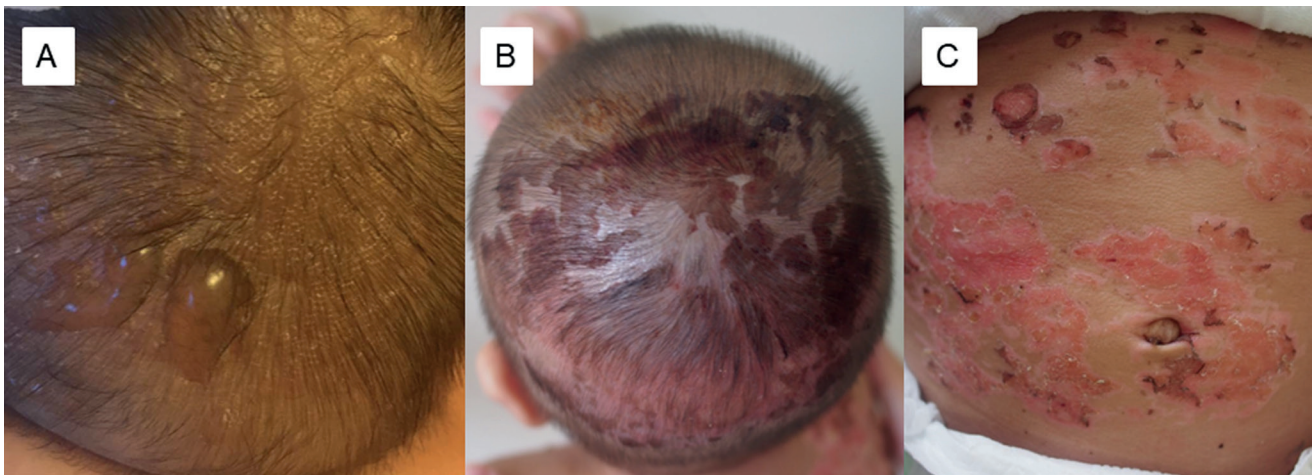


Fig. 1. Physical examination findings. (A) Bullous eruption on the scalp (acute-outbreak phase, photo from the parents). (B) Erythema, erosion, and scabbing (involuting phase) on the scalp and (C) trunk, with generalized thickened and dark skin.

ANSWERS TO QUIZ

A 9-month-old Child with Diffuse Cutaneous Bullous Erythema over Generalized Thickened and Dark Skin: A Commentary

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Diagnosis: Bullous diffuse cutaneous mastocytosis

Darier's sign was positive (after stroking the lesion, transitory reactions of erythema, and wheal formation). Skin biopsy and histopathological examination were undertaken. Haematoxylin-eosin staining showed a diffuse mononuclear cell infiltration in the upper dermis; moreover, the results of Giemsa and toluidine blue staining were positive (Fig. 2). A diagnosis of bullous diffuse cutaneous mastocytosis (DCM) was based on the presence of bullous cutaneous features, a positive Darier's sign, and the pathological findings. After topical use of mometasone furoate cream, the lesions significantly improved. During 4-year-follow up, blister recurrence gradually subsided, leaving residual pigmentation.

Mastocytosis occurs in 2 forms, cutaneous mastocytosis (CM) affecting only the skin, and systemic mastocytosis, affecting various internal organs, mainly the bone marrow and gastrointestinal tract (1). DCM is the most severe subtype of CM, applying only to patients with generalized thickened and dark skin. Infants with DCM often present initially with large blisters, generalized erythema, and pruritus. Other signs include flushing, urticaria, and a reddish-brown tinge to the skin, which appears thickened (2). Darier's sign as an initial clue of mast-cell infiltration was a consequence of elevated histamine release by the proliferated mast cells after mechanical irritation. The tendency to blister in children with DCM usually improves within 3 to 4 years, and most

cutaneous lesions spontaneously resolve by adolescence. However, a small number of DCM patients with a family history may have systemic organ involvement and a chronic course. We also need to consider the extra-cutaneous complications that may occur in children with DCM, such as lymphadenopathy, hepatomegaly, splenomegaly, diarrhoea, hypotension, and vasodilatory shock (2). The mainstay of medication treatment of DCM is antihistamines. External stimuli such as friction, heat, physical activity, certain foods, emotional stress, and certain medications should be avoided in daily life.

This report highlights that DCM should be considered in children with diffuse bullous eruption over generalized thickened and dark skin, especially with wheal formation after mechanical irritation. However, by adolescence, regression of the lesions is frequent in childhood-onset DCM.

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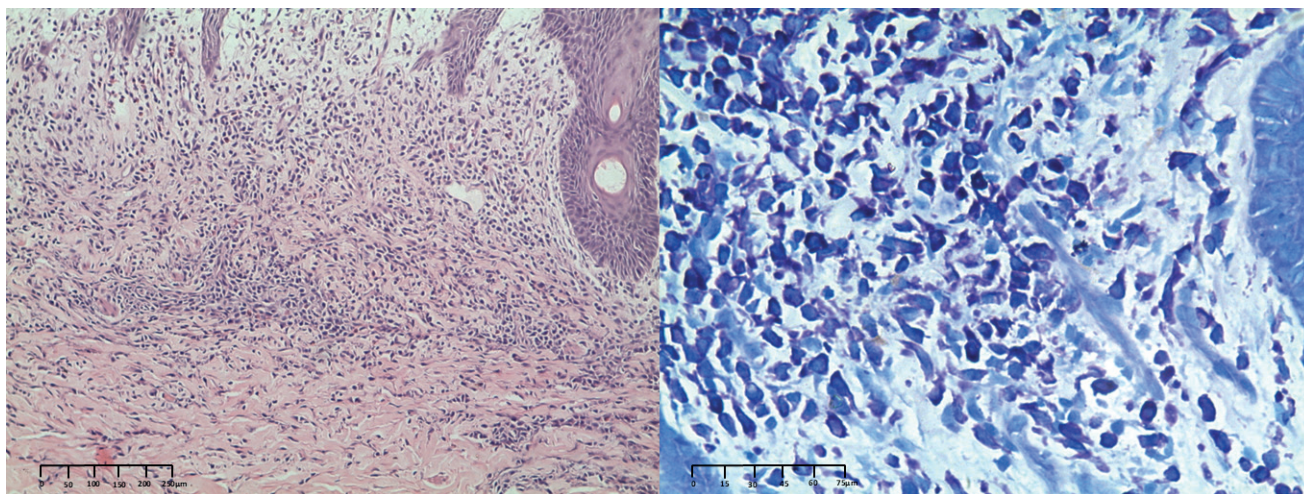


Fig 2. Haematoxylin-eosin (a, x100) and Giemsa (b, x400) staining showed mast cell infiltration and degranulation.