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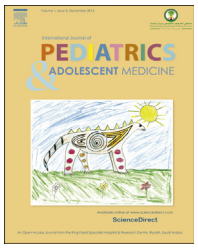

CASE REPORT

Bullous eruption in an infant, what's your diagnosis?

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1. Case presentation

A three-month-old male infant presented with generalized blisters that were first noted when he was two months old. A portion of blisters formed spontaneously and a portion formed when the patient was scratched or rubbed. The lesions were first observed on the hands and feet and spread to the scalp, face and trunk. The infant was otherwise healthy. The mother reported that there was no history of local or systemic symptoms, including flushing, pruritus, shortness of breath, hypotension and diarrhea. Furthermore, the family history was negative for any dermatologic, gastrointestinal or hematologic diseases. Cutaneous examination revealed multiple tense vesicles, bullae, and erosions on the face, scalp, trunk and extremities (Fig. 1). All of the patient's skin was erythematous and thickened. It had a leathery appearance and a doughy texture, and the skin markings were exaggerated. The patient was positive for Darier's sign. There was no evidence of organomegaly or lymphadenopathy. Bacterial blister culture results were negative. Direct immunofluorescence was also negative. Histological examination of a biopsy specimen taken from a tense blister revealed sub-epidermal bullae with a dense infiltration of mast cells and a number of eosinophils in the upper dermis (Figs. 2 and 3).

2. What's your diagnosis?

Diffuse cutaneous mastocytosis with generalized bullae.

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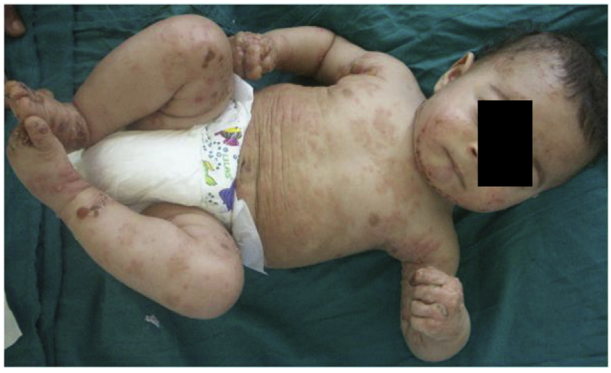

Figure 1 Multiple tense bullae and erosions on the trunk, extremities, palms and soles with exaggeration of skin markings.

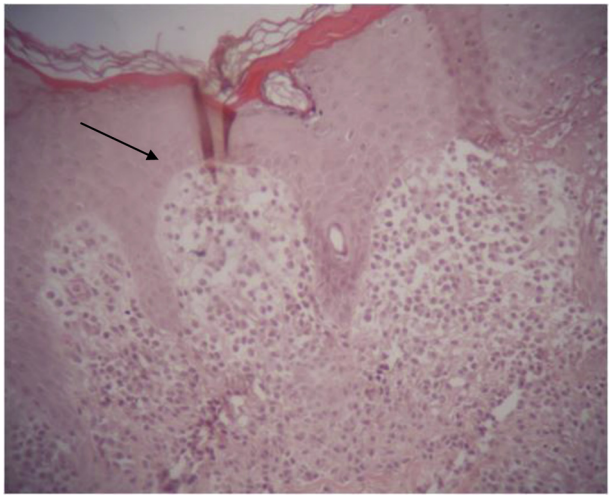

Figure 2 Sub-epidermal bullae with a dense cellular infiltration in the upper dermis (marked by an arrow).

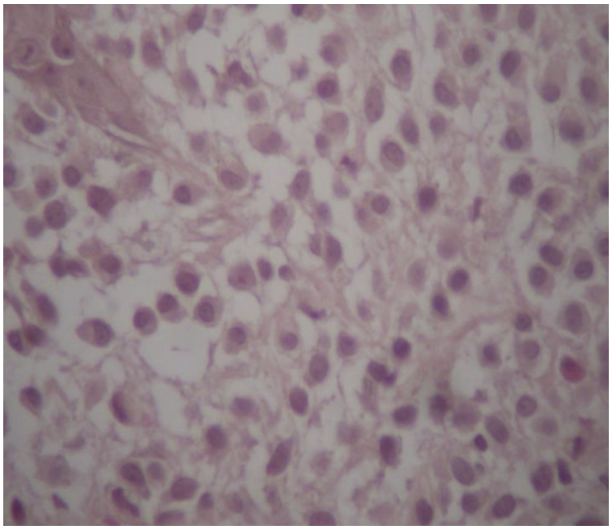

Figure 3 Dense mast cell infiltration with some eosinophils in the upper dermis.

3. Discussion

Cutaneous mastocytosis is a rare condition related to an abnormal proliferation of mast cells and their accumulation in the skin without any evidence of extracutaneous organ involvement [1]. There are 4 different clinical variants of cutaneous mastocytosis: urticaria pigmentosa, solitary mastocytoma, diffuse cutaneous mastocytosis (DCM) and telangiectasia macularis eruptive perstans. Among the mastocytoses that affect children, DCM is the rarest subtype and occurs predominantly in infants. It generally arises within the first 3 years of life [2]. DCM is a heterogeneous disease and presents with 3 different symptomologies: reddish skin with widespread large bullae (as in our patient); yellow-orange infiltration and minimal small blisters or pachydermia; and extremely folded skin [3,4]. Blistering may be present with varying degrees of erythroderma and pruritus and usually occurs during the early stages of life, as occurred in our patient [5,6]. The blisters present in a variety of sizes and initially contain clear fluid that may become hemorrhagic with time [1]. Histological findings in DCM include loosely arranged mast cells throughout the dermis with subepidermal edema leading to vesiculobullous lesions, especially in children. A variety of special stains, including toluidine blue, Giemsa and immunochemistry for CD117 (c-kit) may be used to highlight mast cells [7]. The close clinical differentials include autoimmune bullous dermatoses, epidermolysis bullosa, staphylococcal scalded skin syndrome, incontinentia pigmenti, epidermolytic hyperkeratosis and toxic epidermal necrolysis [8]. A crucial sign for the diagnosis of DCM is whealing in response to rubbing of affected skin areas (Darier's sign) [9]. A positive diagnosis of DCM is then confirmed based on clinical features and the results of histopathological analysis. A combination of antihistamines is the standard treatment for DCM. Cromolyn sodium, ketotifen or corticosteroids may also alleviate DCM symptoms [1]. Finally, DCM has an overall good prognosis and usually resolves spontaneously between the ages of 15 months and 5 years. A close follow-up is usually recommended for these children because of extensive involvement [5,10].

This case report highlights that DCM with generalized bullae should be considered in the differential diagnosis of infants presenting with generalized bullae and erosions.

Conflict of Interest

None.

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