

Case Report

Bullous Mastocytosis in an Infant: A Case Report

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ABSTRACT

Bullous mastocytosis is a rare skin disease characterized by dermal infiltration with mast cells and clinically by tendency for blister formation. It is more commonly seen in early infancy and childhood. Generally, it has good prognosis with tendency to spontaneous resolution. In this report, we describe an infant with recurring itchy widespread bullous skin disease. No systemic

involvement was detected. Skin biopsy specimen revealed dermal mast cell infiltration and subepidermal blister formation confirming the diagnosis of bullous mastocytosis. The patient showed a good response to both H₁ and H₂ selective antihistamines as well as potent topical steroid.

KEYWORDS: bullous dermatoses of infancy, mast cell, mastocytosis

INTRODUCTION

Mastocytosis encompasses a range of disorders characterized by over-proliferation and accumulation of tissue mast cells. Mast cell disease is most commonly seen in the skin, but other tissues of the body such as the skeleton, gastro-intestinal tract, bone marrow and central nervous system may also be involved^[1].

Mastocytosis is classified into three major clinical types: cutaneous, systemic and malignant forms^[2]. Cutaneous mastocytosis (CM) presents a very wide clinical spectrum ranging from an isolated cutaneous lesion to the generalized involvement of the skin. Accordingly, there are different clinical variants of CM: maculopapular (*urticaria pigmentosa*), mastocytoma, diffuse and/or erythrodermic as well as *telangiectasia macularis eruptiva perstans*. Irrespective of these clinical types of CM, the histopathological appearances are uniform. The common feature is an accumulation of increased number of normal looking mast cells in the dermis. Blistering can occur in all these clinical forms but bullous mastocytosis as a sole manifestation is rare and is seen primarily in infants and therefore, needs to be differentiated from other bullous eruptions of infancy^[3-6].

We report a case of an infant with recurring itchy extensive bullous lesions that were confirmed to be mastocytosis on histopathology and showed a favorable response to the treatment.

CASE REPORT

An 11-month-old Indian boy presented with a six month history of recurrent episodes of severely

pruritic generalized vesiculobullous skin lesions. Each episode of the skin eruption resolved within weeks. The bullae had initially been treated by a pediatrician with topical and systemic antibiotics for presumed staphylococcal scalded skin syndrome (SSSS) without beneficial response. There was no family history of a similar condition and his drug history was irrelevant. No symptoms of systemic involvement regarding the gastrointestinal tract (GIT) or the chest could be detected.

Skin examination showed widespread tense vesiculobullous lesions mainly on the trunk and neck (Fig. 1). Areas of erosions and scalded skin were also noted on the back (Fig. 2). Patches of hyperpigmentation and urticarial plaques were observed on the trunk and groin. Mucous membranes and nails were clinically free.

The various laboratory investigations including complete blood count (CBC), blood film, urinalysis, histamine in urine, serum chemistry profile and bone marrow aspiration were within the normal range or negative. No abnormal data could be seen in his abdominal and pelvic sonography. Swab and culture sensitivity from the blister floor showed no growth after 48 hours of incubation.

The skin biopsy was taken from an intact blister and adjacent apparently normal skin and was divided in two parts. The specimen from blister site was stained with hematoxylin-eosin (H&E), Giemsa and toluidine blue stains. Direct immunofluorescent studies for Ig G, Ig A and Ig M were performed on the specimen from the adjacent skin.

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Fig. 1: An 11-month-old infant with widespread skin lesions mainly on the trunk and neck



Fig. 2: A close-up view showing vesiculobullous lesions with areas of erosions and scalded skin on the trunk

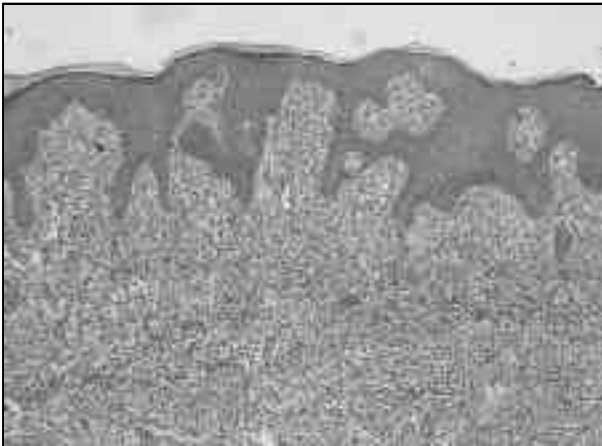


Fig. 3: Biopsy specimen showing sheets of uniform mononucleated cells with abundant cytoplasm below an intact epidermis (hematoxylin and eosin; original magnification X 10)

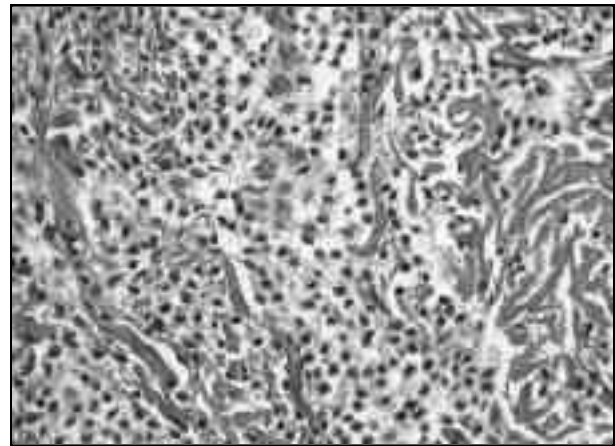


Fig. 4: The infiltrate is composed of round-oval cells with amphophilic-basophilic cytoplasm (H&E stain; original magnification X 40)

On histopathology examination using H&E stain, sheets of uniform mononucleated cells with abundant cytoplasm below an intact epidermis were seen (Fig. 3). This infiltrate was composed of monomorphic cuboidal cells (Fig. 4). Higher magnification demonstrated round-oval cells with amphophilic-basophilic cytoplasm. Few eosinophils were also seen within the infiltrate.

In addition, there was severe dermal edema with subepidermal bullae that contained eosinophils (Fig. 5). Both toluidine blue and Giemsa stains showed the characteristic metachromatic intracellular granules of mast cells (Fig. 6). Direct immunofluorescence from the adjacent skin revealed negative results.

Based on these findings, our diagnosis of **bullous mastocytosis** was confirmed. The child received both H₁ and H₂ selective antihistamines as well as topical potent steroid with good response. Complete clearance of his skin lesions was achieved by the end of two weeks. No recurrence was observed on follow-up 12 months later (Fig. 7).

DISCUSSION

Several skin diseases are characterized by the presence of blisters. In some of these diseases, the blister may be the only morphological clue to diagnosis; whereas in others, the blisters may account for a small minority of the total lesions^[7]. In an infant, the differential diagnosis of bullous eruptions includes epidermolysis bullosa, autoimmune bullous diseases, Steven-Johnson's syndrome, staphylococcal scalded skin syndrome (SSSS), and bullous mastocytosis. Bullae in children may occur in any form of mastocytosis but widespread bullous eruptions or bullae as a sole manifestation is rare and can be differentiated from other diseases by histopathology and immunofluorescence studies^[8].

If bullae are the predominant clinical features, as in the present case, the term bullous mastocytosis is applied. In such cases, blisters tend to be tense and usually heal without scar formation. It is important to differentiate such conditions from other clinical variants of CM which deteriorate with blister formation due to drugs that release histamine^[8-10].

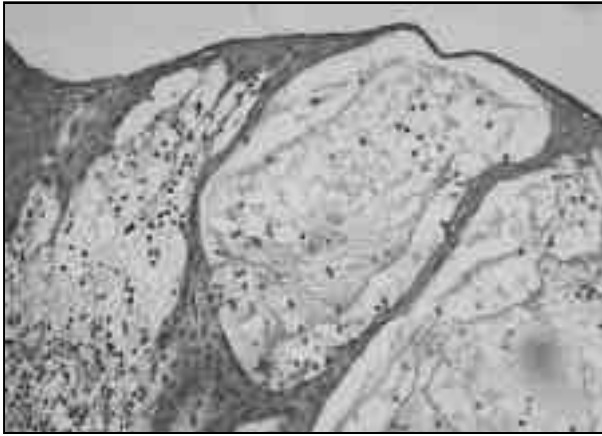


Fig. 5: Marked papillary dermal edema resulting in a subepidermal blister (H&E stain; original magnification X 20)

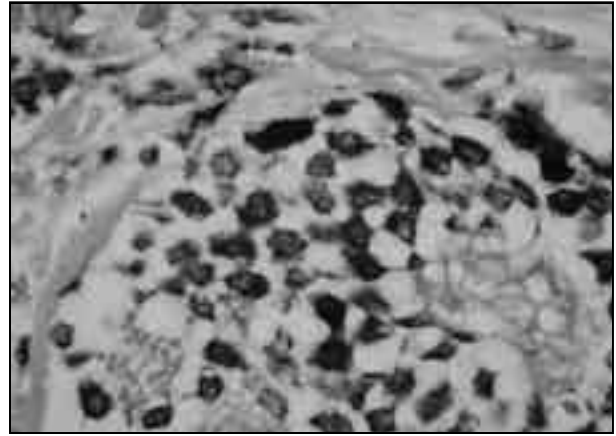


Fig. 6: Mast cells stained metachromatically with toluidine blue stain (Toluidine blue stain; original magnification X 40)



Fig. 7: Complete clearance of skin lesions with residual hypo- and hyperpigmentation. No recurrence was observed over a 12 month follow-up period.

Bullous mastocytosis may mimic the clinical appearance of SSSS^[11]. In the present case, the diagnosis was earlier missed for SSSS and was then settled by histopathology, negative immunofluorescence and bacteriology studies.

CM appears to occur sporadically, as in our patient. However, familial cases have also been reported occasionally^[12].

The consequences of mast cell accumulation in the skin include spontaneous or induced release of mast cell mediators, especially histamine, leading to pruritus, urtication, and dermal edema. This inflammatory dermal edema may be so severe as to result in a sub-epidermal blister formation^[13]. Because of regeneration of the epidermis at the base of the bulla, older bullae may be located intra-

epidermally^[14]. In our case, the blisters were located both in the intra- and sub-epidermal area.

The hyper-pigmented patches in CM could be explained by increased level of melanin pigment in the lower layer of the epidermis, probably due to the presence of a receptor for c-Kit ligand not only on the mast cell but even on the surface of melanocytes^[15].

The term systemic mastocytosis usually indicates the proliferation of mastocytes in more than one organ^[16]. In this case, there were no symptoms or signs of systemic involvement.

Generally, the long term prognosis of CM in infancy and childhood is good with a tendency to spontaneous resolution. However, a subset of children with congenital bullous mastocytosis is at a higher risk of fatal outcome and therefore, needs a more guarded long-term follow-up^[9,17]. Treatment options for bullous mastocytosis include the combined use of H₁ and H₂ selective antihistamines, and oral intake of mast cell stabilizing agent such as disodium cromoglycate. Also, topical potent corticosteroid therapy can improve cutaneous mast cell lesions^[10].

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