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Bullous eruptions in an infant - A rare case report

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Abstract
Background: Infantile vesiculobullous lesions are caused by infections, drugs, congenital and autoimmune disorders. Mastocytosis refers to over-proliferation and accumulation of tissue mast cells, encompassing a spectrum ranging from indolent cutaneous to malignant and systemic conditions. Bullous mastocytosis is a rare cutaneous form.
Case characteristics: A male infant with recurring bullous lesions was confirmed to be mastocytosis on routine histopathology and special stains. Our patient was managed with antihistaminics and close monitoring with strict avoidance of triggering factors.
Conclusion: Our patient responded very well to antihistaminics with complete subsidence of lesions without recurrence at 6 months follow-up. Hereby, we report this rare skin disorder encountered at our tertiary care centre. Bullous mastocytosis, though rare should be considered as one of the differentials in children with vesiculobullous lesions.

Keywords: Mastocytosis, bullous, child

1. Introduction
Mastocytosis, is characterized by abnormal growth of mast cells in skin and other organs like bones, gastrointestinal tract, liver and spleen. The term "mastocytosis" was coined by Sezary and Chauvillon in 1936 [1] According to a consensus revised classification based on clinical presentation, pathology and prognosis, mastocytosis is divided into four major forms- indolent, mastocytosis with hematological disorder, aggressive forms and mast cell leukemia [2] Cutaneous mastocytosis presents a wide clinical spectrum ranging from isolated cutaneous lesion (mastocytoma) to diffuse involvement (urticaria pigmentosa, telangiectasia macularis eruptive perstans). Diffuse cutaneous mastocytosis includes two forms- pseudoxanthomatous/xanthomatous and bullous. Although blistering can occur in any form, bullous mastocytosis as a sole manifestation is rare and predominantly encountered in infancy. Symptoms are attributable to release of mast cell mediators [3] Childhood cutaneous mastocytosis generally follows a benign course with spontaneous involution around puberty [4] However, bullous mastocytosis has been reported to have a variable prognosis warranting careful and long-term follow up [5]

2. Case report
An 11 month old child presented with spontaneous eruption of fluid filled lesions over body since three months of age. These episodes were associated with itching and redness of face but not preceded by triggering factors like drugs or infections nor were they accompanied by diarrhea, vomiting, abdominal pain or chronic productive cough. The child was a second product of non-consanguineous marriage born after a full term normal vaginal delivery. Family history was noncontributory and the child had an updated immunization status. The anthropometric and nutritional status were grossly normal. General and systemic examination were unremarkable except for pallor. Dermatological examination revealed multiple grouped coalescing vesicles and tense bullae with surrounding erythema over trunk, back and neck (Fig 1a). Palms, soles, mucosae were spared while nails and eyes were normal. Shearing of the epidermis from underlying dermis on tangential pressure (Nikolsky's sign) and extension of bulla (Bulla spread sign) were negative. Stroking of normal skin resulted in an urticarial plaque (Darrier's sign). Routine laboratory investigations were normal except for hemoglobin of 7.5 gm %.

Grams stain and culture sensitivity of vesicular fluid were non-productive. Tzanck smear did not reveal any acantholytic cells (ruled out pemphigus vulgaris) or multinucleated giant cells (varicella) but showed mast cells with Giemsa stain. Histopathological examination revealed subepidermal blister (Fig 1b) with moderate band like and perivascular infiltrate of cuboidal mast cells confirmed by demonstration of purple cytoplasmic granules on Toluidine blue stain (Fig 2).

A final diagnosis of bullous mastocytosis was made and the child started on syrup cyproheptadine (0.25 mg/kg/d in 2 divided doses) with syrup ranitidine (1 mg/kg twice a day). The parents reported remarkable improvement in terms of severity and frequency of episodes within two weeks with complete regression of lesions at six weeks. The child was called for regular follow up in view of the possibility of systemic manifestations and parents were counseled regarding course of disease and complete avoidance of triggering factors. Fortunately, no recurrence has been observed after six months.

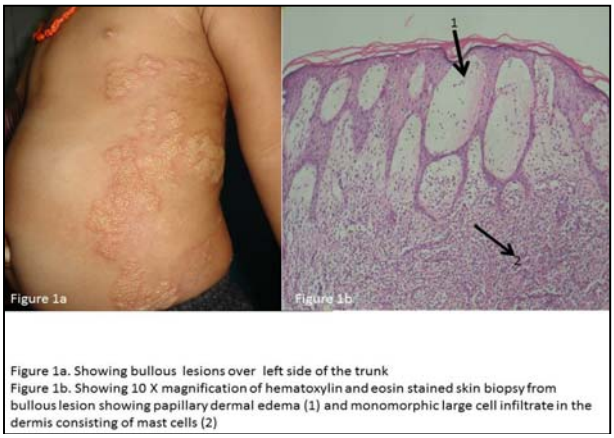

Figure 1a. Showing bullous lesions over left side of the trunk
Figure 1b. Showing 10 X magnification of hematoxylin and eosin stained skin biopsy from bullous lesion showing papillary dermal edema (1) and monomorphic large cell infiltrate in the dermis consisting of mast cells (2)

Fig 1a: Multiple grouped coalescing vesicles and tense bullae with surrounding erythema over the trunk

Fig 1b: Histopathological examination revealed sub-epidermal blister with moderate band like and perivascular infiltrate of cuboidal mast cells

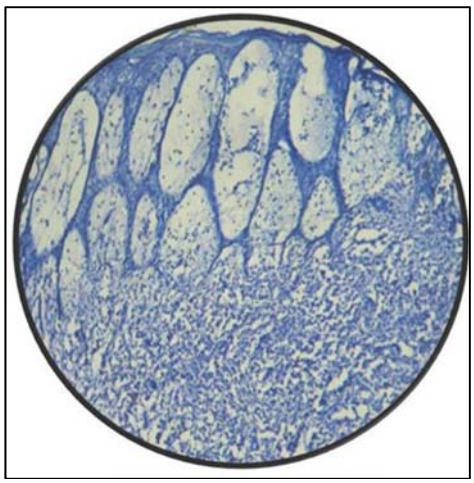

Fig 2: Histopathological examination with Toluidine blue stain showing plenty of mast cells infiltrate along with epidermal blister

3. Discussion

Bullous eruptions in infancy have a wide range of differential diagnosis. Several skin diseases are characterized by blisters - bullous impetigo, staphylococcal scalded skin syndrome, congenital syphilis, epidermolysis bullosa, autoimmune bullous disorders, drug eruptions and rarely mastocytosis.

Bullous mastocytosis is an extremely uncommon variant of cutaneous mastocytosis with indeterminate prevalence usually manifesting in the first year of life. Typical childhood disease is linked to Glu-839-lyc-skit mutation, whereas adult and systemic disease are associated with Asp-816-Val-ckit mutation [4]. Childhood mastocytosis differs in many respects from adult onset disease. Characteristic presentation of pediatric onset mastocytosis consists of cutaneous manifestations either solitary mastocytoma, urticaria pigmentosa or rarely diffuse cutaneous mastocytosis. Internal organ involvement like bone marrow or gastrointestinal is rare compared to adults [5]. Plasma histamine levels may be elevated. Symptoms and signs are both systemic as well as related to specific organ involved and occur as a result of generalized and local release of histamine, leukotrienes, prostaglandins. Massive release of these mediators produces flushing, tachycardia and rarely hypotension, syncope or shock [5].

Consequences of mast cell accumulation in skin include pruritus, urtication and dermal edema which may be so severe as to result in subepidermal blister formation [6] similar to our case. Blisters are tense initially with clear fluid, of variable size. Chronic disease is typically characterized by thickened skin [5]. Often, hyperpigmented patches may be observed due to increased melanin pigment in epidermis as a result of a common receptor for c-kit ligand on mast cells and melanocytes. Stroking or trauma of affected skin causes a wheal with flare or hypotension and flushing in case of large or extensive lesions. The demonstration of mast cell increase in superficial and mid dermis is considered gold standard for diagnosis of mastocytosis and was confirmatory in our case. Infrequently, an infant may exhibit hepato-splenomegaly or skeletal lesions due to mast cell hyperplasia. Fortunately our patient did not display any such involvement.

Many agents stimulate degranulation of mast cells such as bacterial toxins, physical stimuli (heat, cold, sunlight, friction), poisons (snake, hymenoptera), biological peptides (wasp, bees, lobster, ascaris) and drugs (aspirin, codeine, morphine, contrast media etc.) Hence, avoidance of aforementioned agents should be emphasized. Generally, childhood mastocytosis follows a favourable course but bullous subtype may occasionally have a reserved prognosis as observed by Murphy *et al* wherein the child met with a fatal outcome [7] Patients at high risk for shock or sudden death are those with extremely early onset disease (congenital/neonatal), extensive bullous lesions and/or vital organ involvement like cardiovascular or renal [8]. Our patient is expected to have a good prognosis in view of his minimal systemic involvement and encouraging response to treatment.

Treatment is currently limited to palliation of symptoms like pruritus and flushing. Preferred therapeutic options are first generation antihistaminics like hydroxyzine or cyproheptadine which have been shown to produce relief in pruritus, bullae and flushing [9]. The addition of H2 antihistaminics like ranitidine may ameliorate gastrointestinal manifestations like hyperacidity, diarrhea and abdominal pain [10]. Sodium cromoglycate and ketotifen are often reserved as second line agents for patients who fail

to respond satisfactorily to antihistamines. Rarely, infants may develop extensive bullous eruptions accompanied by shock like illness and gastrointestinal hemorrhage suggestive of massive mast cell mediator release. Such children require intensive care support with initiation of corticosteroids and epinephrine in addition to H1 and H2 inhibitors [10]. Generally bullae heal without scarring but local care and prevention of infection are mandatory and topical steroids are to be used with caution. We administered cyproheptadine due to its proven safety profile in small children and noted marked improvement within six weeks. The patient was kept under close observation with no recurrence over six months follow-up.

Thus bullous mastocytosis although rare, must be considered in the differential diagnosis of infants presenting with bullous eruptions.

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