Multiple cutaneous mastocytomas

Sir,

Cutaneous mastocytosis is defined by abnormal infiltration of mast cells limited to the skin. Urticaria pigmentosa and mastocytoma are the most common clinical types while diffuse cutaneous mastocytosis is a rare entity. Mastocytoma is a solitary nodular lesion which may be present at birth or appears within the first 3 months of life and resolves spontaneously with age.[1]

A 3-month-old boy presented with skin-colored raised lesions, progressively increasing in number and scattered all over the body noted a few hours after birth. He had history of multiple episodes of blistering within these raised skin lesions. There were no systemic complaints. Antenatal, birth and developmental history were normal.

On cutaneous examination, there were 11 discrete, well-defined, nodulo-plaques over face, trunk, and extremities [Figure 1]. A few serous fluid-filled bullae were present on top of the lesions on the back. Darier's sign was positive [Figure 2a]. While the child was being evaluated as an in-patient, he started developing episodes of flushing, which lasted for 3-4 minutes [Figure 2b]. These were not associated with hypotension or tachycardia.The child also developed two new lesions over the scalp.

The histopathological examination of one of the nodules revealed sheets of mast cells in the dermis [Figure 3a]. On toluidine blue staining, intracellular metachromatic granules were seen [Figure 3b].

Hematological parameters were normal except for slightly low hemoglobin (11 gm/dl) and eosinophilia (absolute eosinophil count 500/microliter). His serum biochemistry was normal. Radiological investigations and bone marrow examination were normal.

The parents were advised regarding avoidance of potential triggers of mast cell degranulation. The child was treated with various oral H1 antihistamines (cetirizine, dexchlorpheniramine, promethazine, hydroxyzine). Fluticasone propionate 0.05% ointment application was advised over the nodules. No other drugs were started in view of the age of the child and the doubtful safety profile of mast cell stabilizers in children less than 2 years of age. The episodes of flushing became infrequent, no new lesions appeared, and there was some flattening of skin lesions after 3 months. Subsequently, the child was lost to follow-up.

A majority (65%) of patients with cutaneous mastocytosis present in childhood, among which, 55% do so before the age of 2. Mastocytomas are present in 10-35%. Most mastocytomas appear during the first year of life with around 42% presenting at birth. Vesiculation and bulla formation are observed in 33% of mastocytomas.[2] Our patient had multiple mastocytomas which is rare.[1]

Flushing occurs in 6.5% of patients and abdominal pain has also been described in cases with mastocytomas. Our patient experienced multiple episodes of flushing which were not associated with hypotension. Systemic symptoms are rare in pediatric cutaneous mastocytosis. These are seen more often in those with extensive skin involvement, episodes of flushing, early onset of disease and bullous lesions.

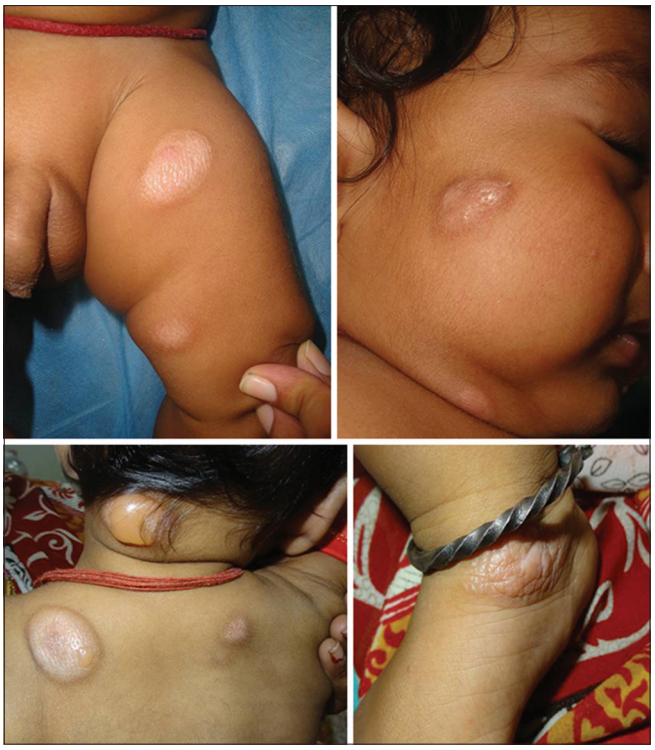

Figure 1: Multiple nodulo-plaques, also showing bullae, at various sites

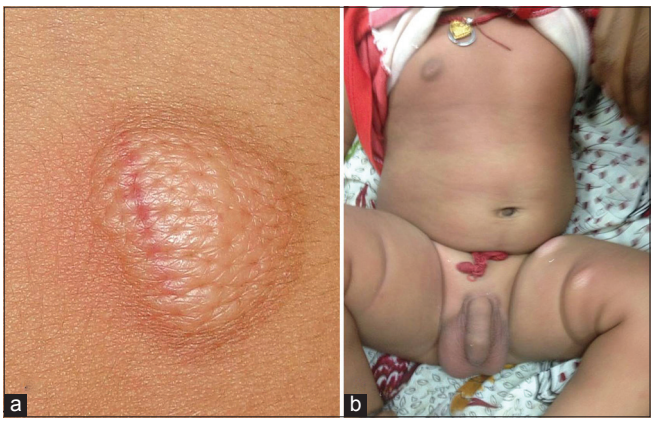

Figure 2: (a) Darier's sign. (b) Generalized erythema during an episode of flushing

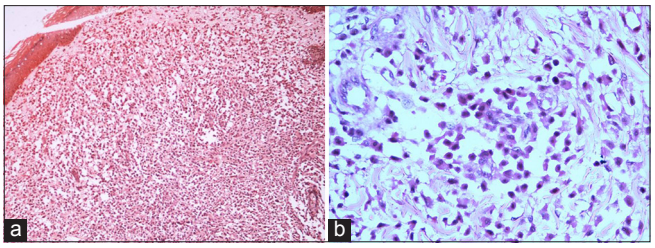

Figure 3: (a) Sheets of mast cells in dermis (H and E, ×10), (b) Toludine blue stain showing mast cells with granular cytoplasm (×40)

These patients have a potential risk for experiencing shock or sudden death.[3]

Plasma tryptase and histamine concentrations reflect the increased proliferation of mast cells and/or their activation.[4] Bone marrow biopsy in those with childhood-onset cutaneous disease is not recommended unless there is evidence of systemic disease such as unexplained peripheral blood abnormalities, hepatosplenomegaly, and lymphadenopathy.[5] It has been suggested that a serum tryptase level exceeding 100 ng/ml or more is an indication for bone marrow examination regardless of age.[2] Due to monetary constraints, the plasma tryptase levels could not be done in our patient. As he had frequent flushing episodes, appearance of new skin lesions and multiple mastocytomas, bone marrow examination was carried out, which turned out to be normal.

The therapy for mastocytosis is focused on providing symptomatic relief and avoidance of triggering factors. H1 antihistamines control pruritus, flushing and urticaria. In spite of receiving antihistamines, our patient had, an initial, increase in the duration of flushing episodes. We assume that this was because triggering factors like friction could not be avoided due to the multiplicity of lesions.The other reason could be an inadequate dose of antihistamines due to lack of sufficient data on dosage in the very young. Sometimes, flushing can be caused by chemical mediators other than histamine.To add to the problem, the safety profile of drugs with mast cell stabilizing properties like ketotifen and azelastine in infants and the very young has not been established.[6] Surgical excision of solitary mastocytoma, topical calcineurin inhibitors and topical corticosteroids are among the other modalities of treatment.

The course of the disease in children is often benign and there is usually no progression to systemic disease. The occurence of multiple mastocytomas with bullous lesions as a presenting feature is unusual. The control of flushing in such an instance may be difficult. There is need for more therapeutic options and guidelines for the treatment of pediatric mastocytosis, especially in very young infants.

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