BULLOUS MASTOCYTOSIS MIMICKING CONGENITAL EPIDERMOLYSIS BULLOSA

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INTRODUCTION

- Bullous mastocytosis is a rare and severe variant of mastocytosis presenting in the first year of life and has been linked to Glu-839-Lyc c-kit mutation (1).
- Bullous mastocytosis has a guarded prognosis with potential risk of shock and sudden death (1).
- It is misdiagnosed as staphylococcal scalded skin syndrome, erythema multiforme or epidermolysis bullosa due to its close resemblance with them, however, the histological findings clearly distinguishes it from others (1).

CASE

- A seven months old male child presented with history of severe itching & redness of skin followed by appearance of clear fluid filled bullae which would rupture on their own or settle leaving behind hyperpigmented velvety plaques.
- There was no history of vomiting, flushing, diarrhea associated with eruption of bullae.
- On examination there were multiple hyperpigmented velvety plaques, some having a peau'd orange appearance involving whole body, sparing palms and soles (Fig. 1 & 2). Darrier's sign was positive (Fig.3).
- Histopathology: Thinned out & atrophic epidermis with sheets of Mast cells having regular round to oval nuclei and dense cytoplasm in superficial & mid dermis (40X & 400X) (Fig. 4). Giemsa staining showed Metachromatic granules of Mast cells (40X & 400X) (Fig. 5).
- Bone marrow aspiration/biopsy or serum total Tryptase level could not be performed due to financial constraints.
- The patient was prescribed Syrup Hydroxyzine & Calamine lotion with advice to follow up regularly.

DISCUSSION

CLASSIFICATION
It is important to differentiate between cutaneous mastocytosis, systemic mastocytosis and localized mastocytomas as their clinical behaviors and long-term outcome are diverse (2).

BULLOUS CUTANEOUS LESIONS
Can be present in all forms of mastocytosis; however when it is predominant presentation, it is called bullous mastocytosis, with frequent systemic involvement (2).

CLINICAL FEATURES
Thickened skin, with an enhancement of cutaneous folds, hyperpigmentation in over half of the cases and positive Darier's sign (3).

HISTOPATHOLOGY
- Abnormal proliferation of dermal mast cells.
- Special stains used to detect mast cells:
1. Giemsa
2. Toluidine blue
3. Leder (3)

TREATMENT
Main goal is to control the signs and symptoms provoked by the release of mast cell mediators with systemic corticosteroids and anti-histaminics (3).

CONCLUSION

- Despite the relative rarity of bullous mastocytosis, its diagnosis is important, both because of the multiplicity of cutaneous manifestations and risk associated with consequent symptoms, illustrating the clinical complexity that may come with the disease.
- Hence knowledge and prevention of agents that trigger release of mast cells mediators with counselling and treatment is required.

REFERENCES

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3. Heide R, Tank B, Oranje AP. Mastocytosis in childhood. Pediatr Dermatol 2002;19:375-81.

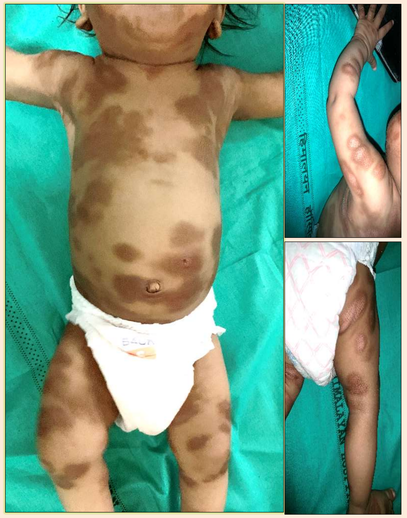

Fig. 1 & 2: Multiple velvety hyperpigmented plaques, some having a Peau'd orange appearance

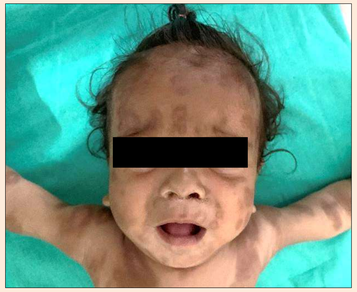

Fig. 2

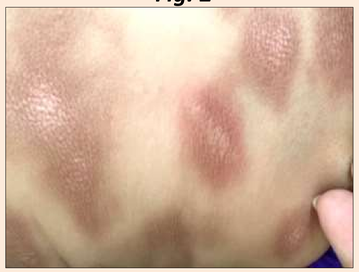

Fig. 3:Positive Darrier's sign

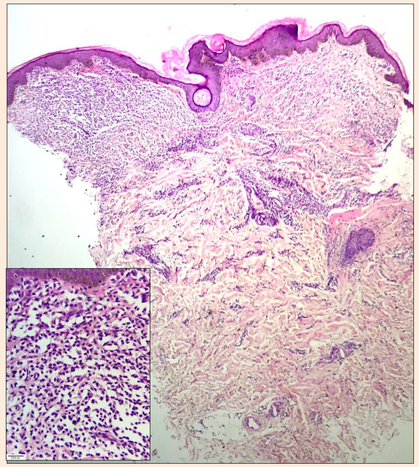

Fig. 4: Atrophic epidermis with sheets of mast cells having regular round to oval nuclei and dense cytoplasm in superficial & mid dermis (40X) Inset (400X).

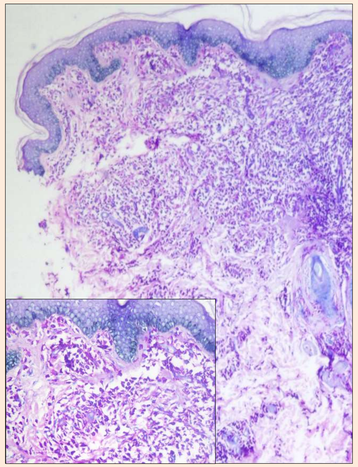

Fig. 5: Giemsa staining: Metachromatic granules of mast cells (40X) Inset (400X).