AUTOIMMUNE BULLOUS DISEASES

DIFFUSE CUTANEOUS BULLOUS MASTOCYTOSIS IN A FILIPINO GIRL : A CASE REPORT

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Introduction: Cutaneous Mastocytosis is characterized by a pathological increase and activation of mast cells in various tissues, particularly in the skin. It is the most common type of mastocytosis occurring in children. Bullous mastocytosis is a very rare variant with onset usually in the first year of life. It has a reserved prognosis with risk of systemic involvement, shock, and sudden death.

Case report: This is a case of a 1-year old female who presented with multiple vesicles and blisters on the scalp, face, neck and trunk. Patient sought consult with several dermatologist and diagnosed as autoimmune blistering disease. Biopsy was done and showed numerous mast cells predominantly and few lymphocytes in the dermis. Giemsa highlighted the mast cells and metachromatic granules. CD 117 showed membranous staining of the dermal infiltrate, which is compatible with mastocytosis. Serum tryptase was elevated. Patient was prescribed ketotifen syrup and topical glucocorticosteroid several weeks with noted decreasing and flattening of the lesions.

Conclusion: The diagnosis of diffuse cutaneous mastocytosis is difficult and is based on clinical manifestations, histopathology and immunohistochemistry. Direct Immunofluoresence is required to exclude autoimmune bullous diseases. Most of the cutaneous mastocytosis in children follow a benign course and undergo spontaneous involution during puberty. Patient education together with the parents and caregiver is very important.

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