URTICARIA, ANGIOEDEMA

DIFFUSE CUTANEOUS MASTOCYTOSIS: STUDY OF 12 CASES

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Introduction: Diffuse cutaneous mastocytosis (DCM) is an extremely rare and severe form of cutaneous mastocytosis characterized by mast cell infiltration of the skin. DCM presents at birth or in infancy. Clinically it presents with widespread spontaneous blistering with erosions, erythroderma or thickening of the skin. It is often associated with flushing, abdominal pain, vomiting, hypotension and anaphylactic shock. Few cases have been described in literature.

Objective: The aim of the study is to characterize the clinical manifestations of DCM presenting at a tertiary care children hospital.

Materials and Methods: This study is a retrospective review of all children less than 18 years diagnosed with DCM presenting to the department of pediatric dermatology at our institute in the last 26 years. DCM was diagnosed based on the cutaneous, laboratory and histopathological examination. Serum tryptase levels were done in all children. Bone marrow biopsy was done in selected cases only. All the demographic details were included in a pro-forma.

Results: A total of 12 cases diagnosed with DCM were included in the study. The age of presentation ranged from birth to 11 months. Congenital onset of lesions was seen in 2 children and presented with erythroderma. Bullous lesions were the most common presentations in all the cases. Blistering was seen in all children and appeared periodically up to 2 to 4 years. Large hemorrhagic bullous variant and infiltrative small vesicular variant was seen in 50% cases. 3 children developed septicemia which was managed with antibiotics. Long term control of the disease was seen in all surviving children except in one child which died due to systemic complications.

Conclusions: Our study showed favorable prognosis in all children except in one child. DCM can progress to systemic mastocytosis and hence guarded prognosis should be given to parents.