



PAEDIATRIC DERMATOLOGY

A BUBBLY BABY: A CASE OF DIFFUSE CUTANEOUS MASTOCYTOSIS IN A 1-YEAR OLD FILIPINO BABY GIRL

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BACKGROUND: Mastocytosis is characterized by an increased number in mast cells with abnormal growth and accumulation in one or more organs. Mastocytosis may be localized, or may have systemic involvement. Diffuse cutaneous mastocytosis presenting as bullae is a rare variant, with no known incidence and having only several case reports published.

CASE SUMMARY: A one-year old girl presented for multiple tense fluid-filled bullae on the trunk and extremities of one-year duration. On the ninth day of life, a solitary tense fluid-filled bulla was noted on the dorsum of the patient's left hand. The bulla spontaneously ruptured, but similar-looking bullae appeared on the face, scalp, trunk and extremities. These were associated with extreme pruritus, easy irritability, and flushing. Patient was observed to continuously scratch her skin, resulting to more bullae. Persistence of symptoms prompted consult. On physical examination, the patient presented with multiple tense fluid-filled bullae on the trunk and lower extremities, with multiple hyperpigmented macules and patches, and multiple erythematous blanching wheals. A leathery feel was appreciated on the entire skin surface. Darier's sign was positive. Histopathologic examination of a bulla on the trunk revealed a subepidermal split with neutrophils, and dense diffuse superficial-to-mid dermal infiltrates of numerous mast cells and occasional eosinophils, consistent with mastocytosis. The patient was evaluated for possible systemic involvement, but blood tests, holoabdominal ultrasound and a skeletal survey turned out negative. Round-the-clock antihistamine (cetirizine 0.2 mg/kg/day) was started, along with liberal use of emollients, and patient education.

CONCLUSIONS: Diffuse cutaneous mastocytosis appearing as multiple bullae is a rare presentation. A thorough history, a comprehensive physical examination, and histopathologic analysis lead to a definitive diagnosis. Supportive treatment with round-the-clock antihistamines, vigorous use of emollients, and avoidance of triggers to mast cell degranulation are the cornerstones of management. Spontaneous resolution by adolescence is the expected course.

