



PAEDIATRIC DERMATOLOGY

UNPROVOKED LOSS OF CONSCIOUSNESS; A CASE OF PEDIATRIC BLISTERING URTICARIA PIGMENTOSA

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Background: Cutaneous mastocytosis is characterized by pathologic accumulation of mast cells, resulting in clinical manifestations secondary to the inappropriate release of their mediators. The three major subtypes are Solitary Mastocytoma, Diffuse Cutaneous Mastocytosis and Urticaria Pigmentosa (UP), the most common subtype in children. Urticaria Pigmentosa presents as maculopapular lesions that may be extensive, confluent, and develop into blisters. Initially serum tryptase may be elevated and decrease over time. We present a case of blistering UP in association with gastrointestinal and neurologic symptoms.

Observations: A 12-month-old boy presented to hospital following two unprovoked episodes of loss of consciousness and emesis. He was previously diagnosed with mild scalp eczema and had transient “boil-like” lesions on his trunk and scalp since 4 months old. Prior to presentation, he developed diffuse erythema, irritability, and pruritus leading to hypotonia and loss of consciousness and rapidly developing scalp vesicles and bullae that were prone to rupture. He was afebrile with no mucous membrane involvement. Examination demonstrated tense, crusted, serous bullae and vesicles with background erythema on his scalp. Flaccid linearly arranged bullae and vesicles were noted on his trunk and back. Biopsies were negative for direct immunofluorescence to IgA, IgM, Fibrinogen, and C3; and positive for Toluidine Blue, Alcain Blue, and Calretinin suggesting a diagnosis of mastocytosis. Initially, serum tryptase was elevated at 17.1 ug/L.

Key Messages: Urticaria Pigmentosa is not commonly on the differential for pediatric bullous diseases. However, in the setting of histamine mediated symptoms such as vasodilation, gastrointestinal distress, and hemodynamic instability, physicians should consider a pathologic mast cell process. Early diagnosis is paramount for proper treatment of UP and prevention of severe life-threatening complications such as anaphylaxis and profound hypotension. As such, physicians should be aware of the association of UP with the gastrointestinal and neurologic events to aid early recognition.

