



PAEDIATRIC DERMATOLOGY

URTICARIA PIGMENTOSA: CASE REPORT

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Background: Mastocytosis is defined as an overproliferation and accumulation of tissue mast cells. Urticaria Pigmentosa is one of the forms of cutaneous mastocytosis and is clinically characterized by macules, papules and red-brown plaques. Most cases of Urticaria pigmentosa happens in children. Dermographism may be present in one-third of the patients and the Darier's sign in 90% of the patients. Systemic involvement is rare.

Observation: Eight-year-old patient with hyperchromic macules accompanied by severe pruritus. The parents report that the lesions started when the child was six years old and has been increasing in size and number. Dermatological examination shows several brownish maculae of varying shapes and sizes spread on the trunk, abdomen and upper and lower limbs. Positive Darier sign. A biopsy of the skin was performed for histopathology that demonstrated the presence of numerous mast cells in the reticular and deep dermis confirming the diagnosis of urticaria pigmentosa. Introduced antihistamines and ketotifen for control of the condition. Abdominal ultrasonography, chest x-ray, blood count, blood biochemistry, feces parasitology and urinalysis, all with negative results and / or within the normal range.

Key message: Diagnosis of Urticaria Pigmentosa can be made clinically. However, a definitive diagnosis requires a skin biopsy. The hallmark finding histologically for Urticaria Pigmentosa is having an increased amount of mast cells in the dermis. Mast cells have a cuboidal or rounded appearance. Clinical feature consist of reddish or brownish patches on the skin, with dissemination of variable density. Lesions are variable in size, round or oval. Darier's sign is the formation of a wheal upon stroking or rubbing skin lesions and suggests the diagnosis of cutaneous mastocytosis. The pathogenesis of mastocytosis is due to several mutations in the tyrosine kinase receptor KIT. Systemic symptoms can occur such as pruritus, flushing, abdominal pain, diarrhea, palpitations, dizziness, and syncope.

