

Urticaria Pigmentosa



An otherwise healthy 4-month-old boy presented with persistent pruritic skin eruptions that had been present since the first week of life. His mother reported that the lesions repeatedly swelled up following oil massages. Cutaneous examination revealed multiple discrete tan-brown waxy papules and nodules ranging from 5 to 15 mm in diameter, involving the trunk (**Figure, A**). His face, acral areas, and mucosae were uninvolved. On firm stroking of a lesion, localized urticaria surrounded by an erythematous flare was elicited (positive Darier sign) (**Figure, B**). Systemic examination and routine laboratory analysis were essentially unremarkable. The presence of abundant mast cell infiltrates on histopathology confirmed the clinical diagnosis of urticaria pigmentosa. Antihistamines were advised for symptomatic relief.

Cutaneous mastocytosis usually manifests as solitary mastocytomas, urticaria pigmentosa, and diffuse cutaneous mastocytosis; rarely, blistering, and xanthelasmoid variants are seen.¹ Although systemic involvement may be associated, malignant transformation is exceedingly rare. Histamine release from degranulation of an increased number of functionally normal mast cells in the dermis accounts for the Darier sign. Urticaria pigmentosa, caused by several

activating mutations in the *KIT* gene, is the most common cutaneous mastocytosis in children with infantile onset. It follows a benign course and frequently resolves by adolescence.² The differential diagnosis includes urticaria, nodular scabies, juvenile xanthogranuloma, arthropod stings, and autoimmune bullous disorders. Treatment is directed at controlling symptoms and avoiding triggers (physical, environmental, and drugs). ■

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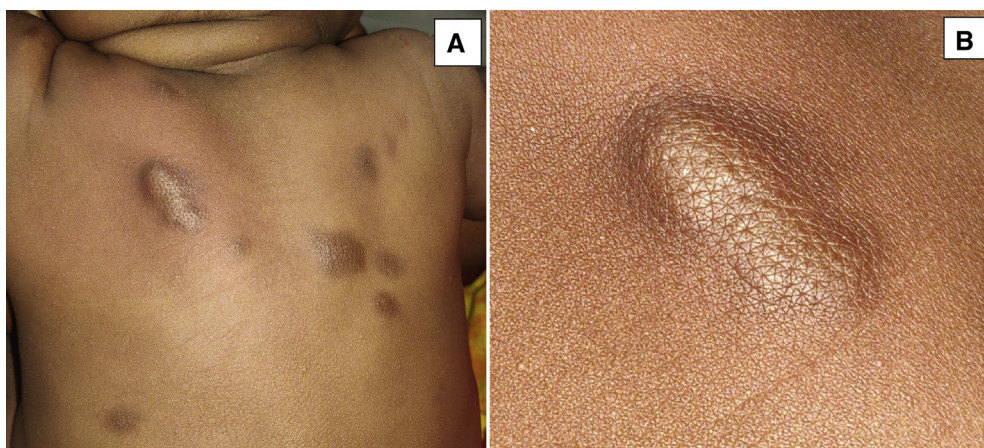


Figure. **A**, Multiple discrete tan-brown papulonodules on the patient's back. **B**, Positive Darier sign showing urtication with surrounding erythema on stroking of a lesion.