
Images in Clinical Practice

Urticaria Pigmentosa (Mastocytosis)

A one-year-old child was brought to us with multiple brown, well defined macular and papular lesions, symmetrically distributed mainly over trunk (*Fig. 1*) with scattered lesions over the face (*Fig. 2*) and extremities appearing at the age of 1 month. There were episodes of severe itching with urtication of lesions and occasional blister formation specially on stroking the lesions vigorously or following toweling of the child after bath. Firm stroking of the lesion on examination produced a linear wheal more pronounced within the lesion and a surrounding erythematous flare, the Darier's sign (*Fig. 3*). Systemic symptoms

such as flushing episodes, tachycardia, hypotension, fainting, diarrhea, vomiting with peripheral eosinophilia and hepatosplenomegaly should make one suspect systemic involvement in mastocytosis (bone, liver, spleen and gastrointestinal tract).

The parents were reassured and explained that the child is likely to become asymptomatic by 5-6 years of age and the lesions likely to spontaneously clear during adolescence. Histamine releasing drugs like aspirin, opiates (specially in cough syrups containing codeine) alcohol and some pre-anesthetic medications should be avoided. Vigorous rubbing of skin during oil massage and after bath is prohibited. Hydroxyzine hydrochloride, 2 mg/kg in 4 divided doses, was prescribed in our



Fig. 1. Brown hyperpigmented macules and papules on the trunk.



Fig. 2. Brown hyperpigmented lesions of the face.

patient to control symptoms. A combination of H₁ and H₂ receptor blockers sometimes with mast cell stabilizing agent like disodium cromoglycate may be needed to control severe systemic symptoms. Severe blistering of the lesions may necessitate use of corticosteroids systemically.

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Fig. 3. Darier's sign showing prominent whealing on the lesion after stroking.