

Incontinentia Pigmenti

A one-month-old girl was brought with multiple erythematous vesiculobullous skin lesions over distal part of limbs and trunk, since birth. The lesions were in the form of vesicles coalescing at places, arranged in linear configuration (*Fig. 1*). These lesions gradually subsided leaving behind linear streaks of blackish hyperpigmentation. The infant had no involvement of the central nervous, ocular or skeletal system. The infant was treated with emollients and topical antibiotics for secondary infections. At six months of age, the skin lesions were in pigmentary stage. The pigmentation was in the form of different whorls over abdomen (*Fig. 2*) and had a linear pattern over extremities.

Incontinentia pigmenti (IP) (syn: Bloch-Sulzberger disease) is a rare hyperpigmentary disorder with a X-linked dominant inheritance. It is characterized by four phases-vesicular, verrucous, pigmentary and hypopigmentary stage. In 80% cases other defects are seen in the form of dental, skeletal, nail anomalies, microcephaly, seizures, psychomotor retardation, strabismus, optic nerve atrophy, retinal detachment, cataracts etc. The skin lesions are benign and follow lines of Blaschko. These lesions resolve by the age of one year but pigmentary stage may last for years. Pigmentation fades away by adulthood, leaving no sequelae. Blood levels of IgE are raised with eosinophilia.

The blistering stage of the lesion needs to be differentiated from *Herpes simplex* and Bullous Impetigo. The lesions are linear and in clusters in classical Incontinentia Pigmenti



Fig. 1. Healing verrucous stage of incontinentia pigmenti.



Warty phase needs to be differentiated from linear epidermal birthmarks or warts. Hyperpigmentary stage needs to be differentiated from moles and other causes of hyperpigmentation. Hyperpigmentation of Incontinentia Pigmenti is classically in whorls. Skin lesions of Incontinentia pigmenti are self limiting. Treatment is usually symptomatic in the form of emollients and topical antibiotics if required for infection.

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Fig.2. Pigmentary stage of incontinentia pigmenti.
