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Visual Diagnosis

Nature's Canvas: An Infant With Stripes and Whorls

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This three-month-old girl developed skin lesions on day six of life. Initially, she had multiple vesicles on an erythematous background affecting her limbs, back, and abdomen. These lesions subsequently developed into brown-black linear patches. At age two months she developed warty, thick lesions on the extremities. There was no history of delayed development, seizures, or focal neurological deficit. On examination, she was microcephalic (OFC: 34.5 cm) and exhibited hyperpigmented linear and reticular lesions along the lines of Blaschko with facial sparing (Fig). The diagnosis was incontinentia pigmenti (IP).

IP is a rare neurocutaneous disorder with X-linked dominant inheritance or more frequently sporadic. IP is also known as Bloch-Sulzberger disease and is caused by a mutation in the *IKBKG* gene. It is usually lethal in males and thus occurs almost exclusively in females (92% to 95%).[1] The diagnosis is based on diagnostic criteria proposed by Landy and Donnai.[2] These criteria include dermatological, dental, retinal abnormalities, and recurrent miscarriage of male fetuses. The characteristic skin changes along the line of Blaschko appear as early as birth or within the first few weeks of life and as late as adolescence. The skin changes evolve in four sequential stages. Stage 1 (vesiculobullous) begins within a few weeks of life and is characterized by erythema and blistering that generally disappears by 15 months of age. Stage 2 (verrucous) appears within a few months of life and consists of a hypertrophic rash that disappears after a few months. Stage 3 is characterized by skin hyperpigmentation and usually persists untill adulthood. Stage 4 is not present in every patient and is characterized by atrophic scars, hypopigmented skin patches, and alopecia. Hair changes include alopecia; lusterless, wiry, and woolly hairs; madarosis; and sparse eye lashes. Other clinical findings include nail pitting, nail dystrophy, supernumerary nipple, hypodontia, retinal telangiectasia, retinal detachment, epilepsy, intellectual disability, and microcephaly.[3] Hypomelanosis of Ito is among the differential diagnoses of IP, and is characterized by hyper- and hypopigmentation without inflammation and verrucous lesions.[4]

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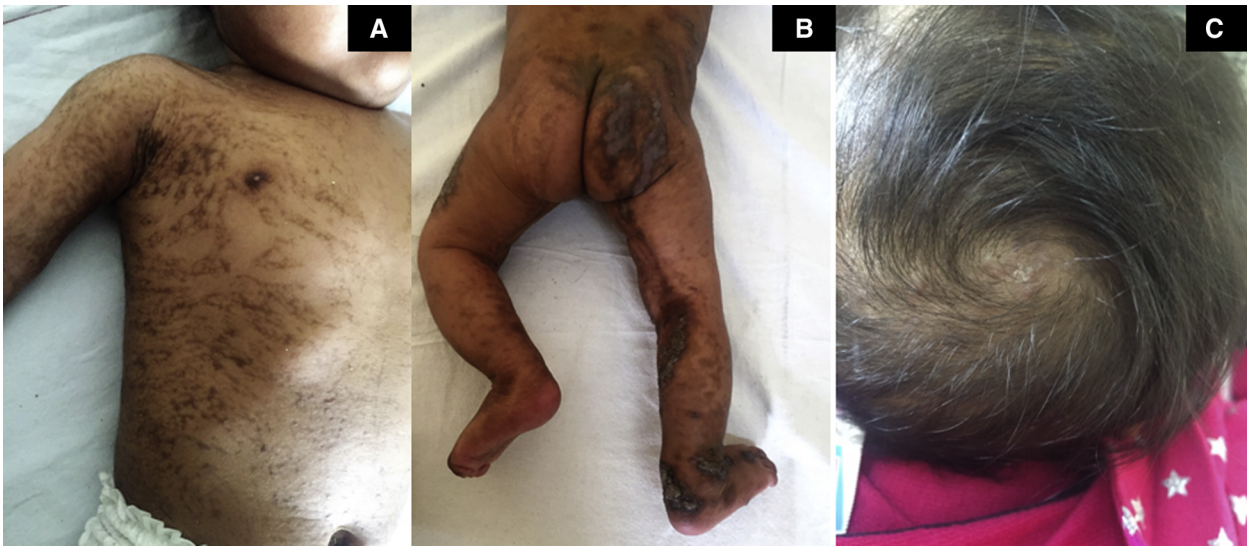

FIGURE. Multiple hyperpigmented linear and reticular lesions along the lines of Blaschko with normal intervening skin on trunk and extremities. Hypertrophic verrucous lesions on buttock, thigh, leg, and dorsum of foot (A and B). Thin sparse, dry, and lusterless scalp hairs with localized alopecia (C). The color version of this figure is available in the online edition.