

IMAGES

melanin incontinence; usually mild as compared to classical lichen planus) were excluded based on their distinguishing features. Universal acquired melanosis is a diagnosis of exclusion. It is a progressive condition; long term prognosis and treatment is not established.

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FIG. 1 Generalized dark-black hyperpigmentation, mimicking negroid skin color. Note palm involvement.

Bullous Congenital Ichthyosiform Erythroderma

An 8 years-old girl presented with intermittent bullous lesions and rough thick skin since birth. At birth, her skin was reddish in color and was notable for spontaneous peeling. Soon after birth, she developed a bulla over leg, which healed spontaneously in 2 weeks without any scarring or pigmentation. However, she kept developing bullae intermittently. At around 5 months of age, she developed gradual thickening of skin. On examination, skin was dry and scaly. “Corrugated cardboard” like thickened skin was noted around joints, involving both extensor and flexor surfaces. Single erosion was found on right shin (**Fig. 1**). Systemic examination was non-contributory. Based on clinical presentation, she was diagnosed with bullous congenital ichthyosiform erythroderma (bullous CIE). Histopathology from the erosion showed marked hyperkeratosis, a thick granular layer, and vacuolar degeneration of the upper epidermis. These findings were consistent with the diagnosis of bullous CIE.

Bullous CIE is a rare autosomal dominant genodermatosis caused by mutation in epidermal keratins 1 and 10. It presents as erythroderma (involvement of more than 90% of skin with erythema, scaling with/without edema) and blistering in newborns, followed by a lifelong ichthyotic condition. As patients age, the scaling becomes thicker and the propensity to blister decreases. Palms and soles may be involved. This condition should be differentiated from non-bullous CIE (absence of history of bullae, presence of erythroderma) and epidermolysis bullosa (bulla formation at trauma prone areas, variable scarring absence of scaling or hyperkeratosis). The diagnosis is usually clinical; histopathology findings help



FIG. 1 Note “Corrugated cardboard” like hyperkeratosis in popliteal fossa (A), wrist and knee (B), and cubital fossa (C). Erosion over right shin (D).

in making a diagnosis. The term “epidermolytic hyperkeratosis” is often used as synonym for bullous CIE. Treatment in early period is directed towards treating secondary complications of erosions (sepsis, electrolyte imbalance etc). Later in life, emollients, urea 10%, topical and systemic retinoids are helpful.

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