A 5-years-old girl born to non-consanguineous parents presented with progressive generalized hyperpigmentation since 3 months of age. The darkening of skin was first noticed over the feet and face at the age of 3 months and gradually progressed to involve the entire body. There was no history of prior drug intake or other skin lesions, no history of discoloration of urine or photosensitivity. On examination, baby had generalized dark-black hyperpigmentation, oral mucosa was involved; genitalia were spared. Face showed patchy areas of normal skin. [Fig.1] The skin texture and sweat secretion were normal.

The skin biopsy revealed excessive melanin pigmentation of the epidermis with few melanophages in the dermis. A diagnosis of universal acquired melanosis (carbon baby syndrome) was made.

Addison disease (low blood pressure, hyponatremia, hyperkalemia, low serum cortisol level), Cushing disease (obese patient, cushingoid habitus, hyperglycemia, hypokalemia, elevated serum cortisol level), hemochromatosis (high transferrin saturation and ferritin level, genetic tests for the C282Y and H63D), and lichen planus pigmentosus (usually macular lesions, rarely diffuse hyperpigmentation, characteristic histopathology findings-lichenoid infiltration, basal vacuolar change and prominent...
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 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.

Ashim Kumar Mondal, Piush Kumar and Avijit Mondal
Department of Dermatology, Medical College and Hospital, 88, College street, Kolkata 700 073, West Bengal. docpiyush@gmail.com

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

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