Generalized Benign Acanthosis Nigricans

A 4-year-old girl, born of a non-consanguineous marriage with normal growth and developmental milestones, presented with asymptomatic generalized hyperpigmentation. It started spontaneously at the age of 2 years, around the neck and axilla, and spread insidiously to involve the other parts of body. The skin gradually became thickened and rugose. There was no history of drug intake, polyuria, polydypsia, loss of appetite, excess weight gain or loss. Cutaneous examination revealed generalized hyperpigmentation with thickening of skin, which was accentuated in the back and sides of the neck, axillae, groins, dorsal hands and flexural areas of knees and elbows. The characteristic velvety plaque with corrugated surface could be appreciated in the flexures (***Fig*.1**). Mucous membranes, palms, soles, hair, and nails were unremarkable. Routine investigations, thyroid profile, insulin level, lipid profile, complete liver and renal function tests, and oral glucose tolerance test were within normal limits. Histopathological examination revealed marked hyperkeratosis, acanthosis and papillomatosis and was consistent with diagnosis of acanthosis nigricans. Epidermolytic hyperkeratosis was excluded on the basis of absence of bullous lesions and histopathological findings. Absence of mucosal and systemic involvement, and velvety texture of lesions in our patient were against the diagnoses of hemochromatosis and Addison's disease.

Generalized acanthosis nigricans, as seen here is rare and is most commonly seen in adults with an underlying malignancy. Age of onset more than 40 years, symptomatic (generalized pruritus), rapid progression and involvement of atypical sites such as mucosa (tongue and lips), Palm ("Tripe palm") and soles are clinical indicators of underlying malignancy. These findings were absent in our patient. The treatment is directed towards treating the underlying cause that includes, either weight reduction, discontinuation of offending drugs, correction of endocrinological abnormality or underlying malignancy. Emollients, keratolytics (topical retinoids, salicylic acid, lactic acid, urea), calcipotriol, systemic retinoids, CO2 laser ablation, and long-pulsed alexandrite laser may improve appearance.

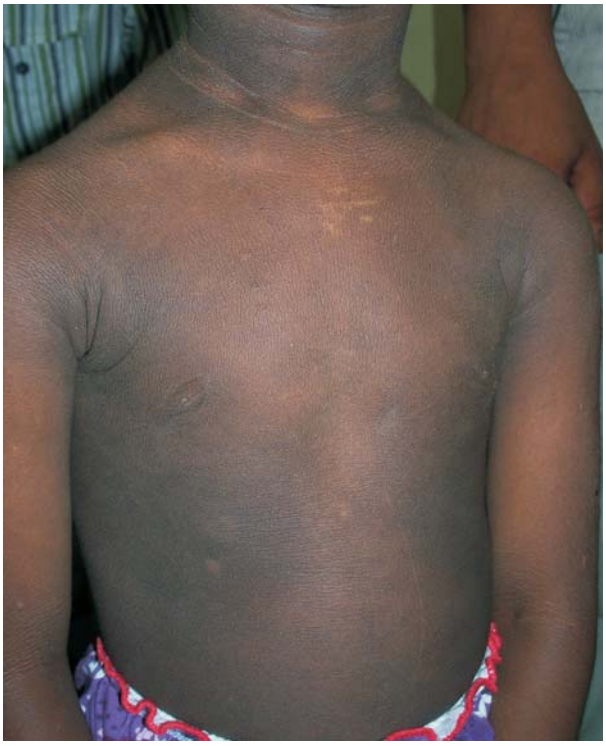

FIG.1 *Generalized hyperpigmentation associated with velvety skin thickening with corrugated surface.*

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An Infant with Skin Rash

An 8-month-old female with a history of eye discharge, presented with complaints of pustules on red tender skin, which ruptured to lead to erosions and peeling since 3 days. On examination, skin was tender with diffuse erythema. Whitish crusting and fissuring was seen in the perioral area and the neck (**Fig. 1**), with sparing of the mucosa. Flaccid pustules and blisters, few having ruptured to lead to erosions were seen on trunk, inner thighs and neck. Nikolsky sign was positive. Wrinkling of skin along with exfoliation was seen in the axillae. There was leucocytosis. Lesional pus for smear and culture sensitivity and blood culture were negative for *Staphylococcus.* Histopathology revealed focal loss of

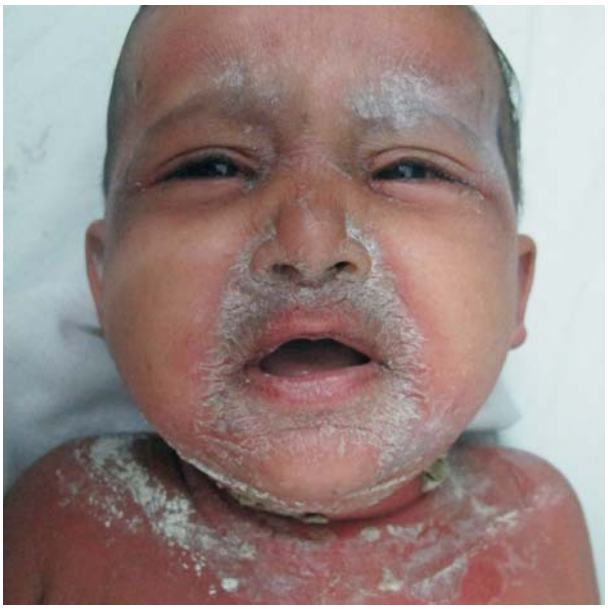

FIG.1 *Whitish crusting and fissuring in the perioral and perinasal areas with flaccid blisters in neck folds.*

upper epidermis and presence of acantholytic cells in the subcorneal layer. A diagnosis of staphylococcal scalded skin syndrome (SSSS) was made. Patient had a complete recovery with peeling within 10 days on treatment with antistaphylococcal antibiotics.

SSSS, caused by *Staphylococcus aureus* exfoliative toxins (ET) A and B, generally affects neonates, infants, and children less than 5 years of age, due to lack of protective antitoxin antibodies and immature renal function. Left untreated, large sheets of epidermis slough off to leave extensive areas of raw denuded skin that is sensitive and painful. The toxin is usually produced at a site distant from the lesions. ET acts as an atypical glutamate-specific serine protease that binds and cleaves desmoglein-1(found in the upper epidermis, absent in the mucosa) which explains the specific site of action in the superficial epidermis and the absence of mucous membranes affection in SSSS. Cultures from the skin lesions are negative for staphylococcus in almost all cases. It is important to send swabs from other areas such as the umbilicus, nasopharynx and conjunctivae. Anti-staphylococcal antibiotics, temperature regulation, maintaining fluid and electrolyte balance, nutritional management and skin care form the basis of treatment. The main differential diagnosis remains drug-induced toxic epidermal necrolysis (TEN) the differentiating factors in TEN being-adult onset, spared areas of the skin, mucosal involvement, presense of nikolsky sign only in involved skin (and not diffusely) and absence of perioral/ perinasal crusting.

It is important to recognize this often dramatic looking skin disorder early, especially in nurseries, with the help of the above-mentioned classical features.

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Rowell Syndrome

A 13-year-old female child presented with multiple erythematous papules and plaques with some of them showing targetoid appearance (***Fig.1a***), along with a single large 8 cm x 10 cm sized polycyclic, erythematous plaque, involving right side of face, upper neck and external ear area (***Fig.1b***). Oral mucosa showed erythematous plaque involving right side hard palate with overlying multiple superficial erosions (***Fig.1c***). She had history of low-grade fever, photosensitivity, malar rash, Raynaud's phenomenon, chilblains and recurrent oral ulceration since 6 months. There was no history suggestive of recent infection or drug intake prior to onset of lesions.

Laboratory studies showed positive antinuclear antibodies (ANA) in a speckled pattern at 1:160 dilutions, ESR 25 mm/I/h, and positive rheumatoid

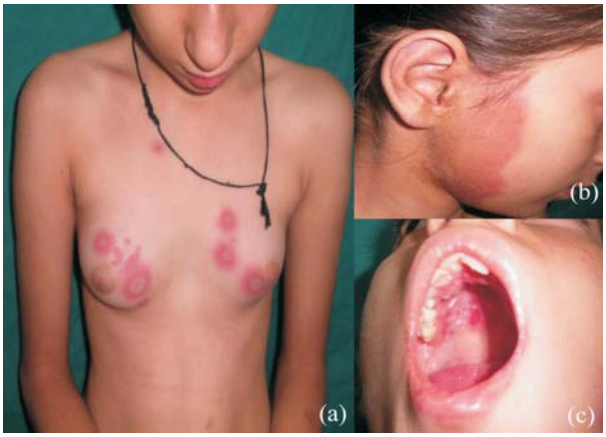

FIG.1 *Rowell syndrome*