

WHAT IS YOUR DIAGNOSIS?

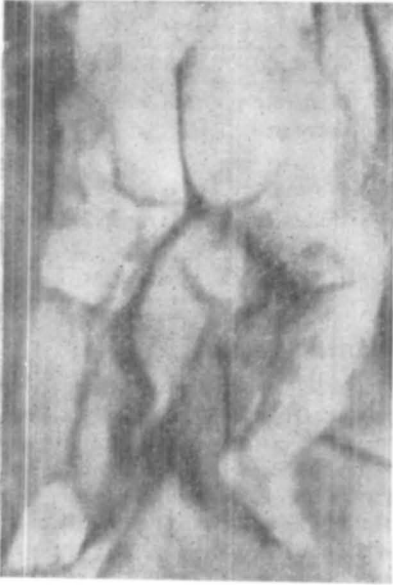


Fig. 1



Fig. 2

Two and a half months old female baby presented with pigmentation and blisters of her skin. The baby was born with pigmentation all over the body and a week after birth developed the blisters first on the feet and later on the hands and other parts. Examination revealed bizzare hyperpigmentation (Fig. 1) all over the body except face. In some areas there was erythema, with blistering (Fig. 2) and crusting. Few of the areas on the extremities showed early verrucous change. No other abnormality was detected in the child. There was no family history of similar problem.

Differential Diagnosis :

1. Drug reaction
2. Incontinentia Pigmenti
3. Urticaria pigmentosa.

(Diagnosis overleaf)

Biopsy showed multiloculated intraepidermal blister. The blister contained strands of differentiating cells moving upwards across the blister. The inflammatory cells in the blister were predominantly eosinophils. The epidermis in the blister area was acanthotic and the basal cells were damaged. There were clumps of free pigment in the upper dermis, which showed mild chronic inflammatory reaction. PAS stain revealed absence of basement membrane.

Final diagnosis :

Incontinentia pigmenti

Incontinentia pigmenti is a rare syndrome in which skin and other ectodermal structures are involved. Cutaneous manifestations are pigmentation, erythema with blistering and lichenification with verrucous changes. Pigmentation is bizarre and is usually seen at birth. One-third of the children develop erythema and vesiculation finally leading to pigmentation. Genetic transmissions of this disease is not obvious although some consider it as a genetic disorder — sex-linked to females.