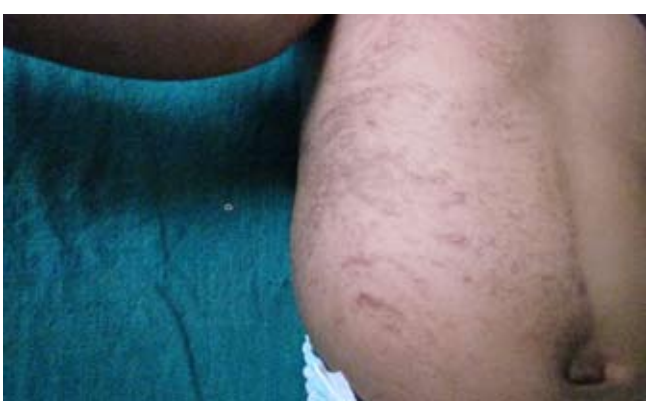
SPOT DIAGNOSIS (IMAGE GALLERY)



NEONATAL SKIN LESIONS

K Jagadish Kumar*, Veerana, Surya Prakash*, Vinay*, Vinutha**.**
**Department Of Pediatrics, **Department Of Dermatology, Venereology And Leprosy, JSS Medical College, JSS University, Mysore, India*

Address for Correspondence: Dr. K. Jagadish Kumar, Professor of Pediatrics, JSS Medical College, JSS University, Mysore, Karnataka, India.
Email: jagdishmandya@gmail.com

A 5 months old developmentally normal female child born to a 2nd degree consanguineous married couple presented with lesions on the trunk and legs since birth which were increasing in size. There were no similar complaints in the family members. On local

examination hyperpigmented linear, mosaic type of lesions were present on both legs and on the trunk. There was no alopecia, squint or obvious skeletal deformity. Other systems examintion was unremarkable. Skin biopsy showed skin with epidermis displaying orthokeratosis, thick irregular acanthosis with intra epidermal keratinisation consisting of whorls of keratinocytes. Masson-Fontana stain shows marked reduction in the melanin pigment. Dermis shows sparse perivascular lymphocytic infiltrate.

What is your diagnosis?

Features are consistent with Incontinentia pigmenti. Incontinentia pigmenti (IP) is a multisystem ectodermal disorder accompanied by dermatological, dental and ocular features and in a minority of cases may be associated with neurological deficit (1). The typical phenotype is a result of functional mosaicism` a phenomenon which occurs in X web addressed dominant disorders because of lyonisation (1). Classically the dermatological features are described in four stages. Stage 1: erythema, vesicles, pustules` Stage 2: papules, verrucous lesions, hyperkeratosis` Stage 3: hyperpigmentation` Stage 4: pallor, atrophy and scarring (2,3). The frequency of nail dystrophy may be as high as 40 percent but it is usually mild (1). Alopecia especially at the vertex and usually after blistering at this site is common but in most cases it is partial and goes unnoticed (2). Squints occur in over one-third of patients, often in association with refractive errors. The hallmark of ocular IP involves abnormalities of the developing retinal vessels and the underlying pigmented cells, and is present in over 40 percent of patients (1). The dental features of IP occur in over 80 percent of cases (1). Typical features include hypodontia, delayed eruption, impaction and malformation of the crowns, especially conical forms and accessory cusps. Breast hypoplasia and supernumerary is seen in 1 percent of cases (2). CNS involvement seen in one-third of patients includes motor and cognitive developmental retardation, seizure, microcephaly, spasticity, and paralysis (1). Skeletal abnormality includes somatic assymetry, hemivertebrae, scoliosis, spina bifida (1). The diagnosis of IP is founded on the clinical features. The classical florid rash of IP is diagnostic but unusual presentations can occur when skin biopsy may be necessary.

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References:

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