Linear and Whorled Nevoid Hypermelanosis with Hemiatrophy

A two-years-old boy presented with abnormal body pigmentation and body asymmetry noted since birth. A preceding vesicular eruption was absent. He was developmentally normal. Family history was insignificant. Examination revealed linear and whorled hyperpigmented macules in a reticulate pattern along the Blaschko's lines over trunk and limbs with sparing of palms, soles and mucous membranes (***Fig.* 1**), suggestive of Linear and Whorled Nevoid Hypermelanosis (LWNH). There was left-sided hemi-atrophy. Systemic examination was unremarkable. Fundoscopy was normal. Skin punch biopsy revealed increased pigmentation of the basal layer without incontinence of the pigment. Karyotype, echocardiography and MRI brain were normal.

LWNH is a rare disorder and may be associated with developmental delay, autism, seizures, ocular, cardiac and skeletal abnormalities. Hemi-atrophy has been reported only rarely. It needs to be differentiated from other linear pigmentary disorders distributed along the Blaschko lines. Incontinentia pigmenti is an X-linked dominant disorder presenting in females with a vesicular phase preceding the pigmentary phase. Linear epidermal nevus has characteristic hyperkeratotic plaques. Hypomelanosis of Ito and segmental nevus depigmentosus present with hypopigmented lesions.

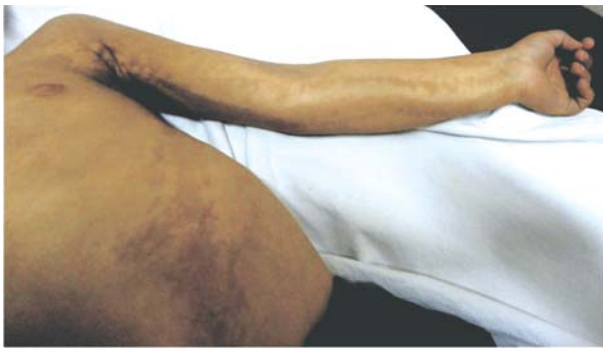

FIG. 1 *Linear and Whorled Nevoid Hypermelanosis: Linear and whorled hyperpigmented macules in a reticulate pattern along the Blaschko's lines over trunk and limbs.*

PUNEET JAIN, BISWAROOP CHAKRABARTY AND SHEFFALI GULATI
Division of Pediatric Neurology, Department of Pediatrics, All India Institute of Medical Sciences, New Delhi,110029 India.
sheffalig@yahoo.com

Pityriasis Lichenoides

A 2-year-old male child presented with multiple asymptomatic erythematous scaly papules and plaques with peripheral scales all over the face, scalp, trunk and extremities since 3 weeks (***Fig*.1**). Prior to this, there was a history of fluid filled lesions of 2 weeks duration which left behind Chicken pox like scars following symptomatic treatment. Skin biopsy showed typical features of Pityriasis lichenoides et varioliformis acuta (PLEVA). He was treated with narrow band UVB with good results.

Clinical spectrum of Pityriasis lichenoides ranges from acute papular lesions that rapidly evolve into papulovesicles, necrosis and varioliform scarring (PLEVA) to small, scaly, benign-appearing papules (pityriasis lichenoides chronica or PLC) both with a generalized distribution. This affects young adults and occasionally children. Though the etiology of this condition is unclear, infectious/drug related hypersensitivity and premycotic lymphoproliferative disorder are the mainstay theories. The diagnosis is by histopathology. In PLEVA, as there are polymorphic lesions of papules and vesicles appearing in crops which may heal with scars, a differential diagnosis of varicella has to be considered. Hemorrhagic necrosis and a course of waxing and waning are not common in varicella.

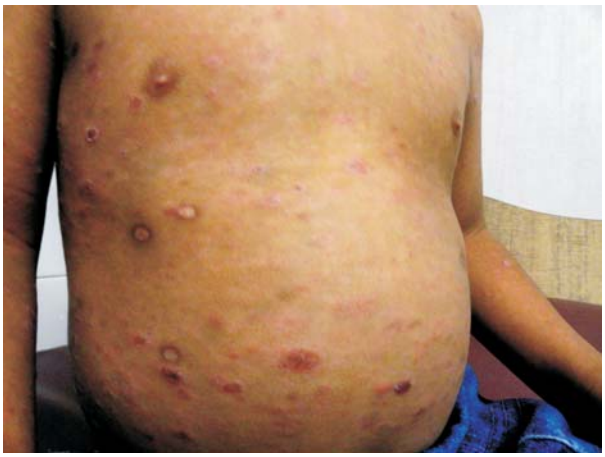

FIG. 1 *Erythematous scaly lesions and scars.*

As PLC presents as small papules with scaling, pityriasis rosea and psoriasis need to be ruled out. While typical pattern and a self-limiting benign course excludes pityriasis rosea; symmetry, distribution pattern and silvery scales point to the diagnosis of psoriasis. PLC may also have adherent 'mica like' scales, which, when detached, reveal a shiny brown surface. If seen, this is a distinctive diagnostic feature. The present case had features of both PLEVA and PLC.

Though the condition is self-limiting, as the course is unpredictable, it warrants therapy. Oral antibiotics, topical corticosteroids and phototherapy have been tried with variable success.

SHYLAJA SOMESHWAR AND SATISH UDARE
Department of Dermatology,
MGM Medical College, Navi Mumbai,
Maharashtra, India.
shaila38@yahoo.co.in