Images in Clinical Practice

Hypomelanosis of ITO

A 12-year-old male boy presented with repeated episodes of generalized tonic clonic seizures since the age of 1 year. There was global development delay and mental retardation. On examination hypopigmented macules were present in bizarre configuration following lines of Baschko (*Fig. 1*). The palms, soles and mucous membranes were not involved. A diagnosis of Hypomelanosis of ito was made. CT scan brain showed diffuse gyral thickening in right fronto-parietal region (cortical dysplasia). EEG showed generalized epileptiform discharges, skin histopathology showed decreased number of melanocytes in the basal layer with no inflammatory cells.

Hypomelanosis of Ito is characterized by skin lesions which are generally present at birth but may be acquired within 2 years of life; these bizarre, patterned hypopigmented macules are arranged over the body surface in sharply demarcated whorls, streaks and patches that follows the lines of Blaschko; with sparing of palms, soles and mucous membranes. The hypopigmentation remain throughout childhood but fades during adulthood. There is associated mental retardation, seizures and microcephaly. The other associations include thoracic and



Fig. 1. Child with hypomelanosis of ITO.

strabismus, nystagmus and cardiac defects. The differential diagnosis includes stable leukoderma and Incontinentia pigmentia.

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