PAEDIATRIC DERMATOLOGY

A RARE CASE REPORT OF ADULT ONSET ERYTHROKERATODERMA VARIABILIS PROGRESSIVA WITH FLEXURAL INVOLVEMENT.

Priyanka Karagaiah [1] - Leelavathy Budamakuntla [1] - Asha G S [1]

Bangalore Medical College And Research Centre, Dept Of Dermatology , Venerology And Leprosy, Bangalore, India [1]

INTRODUCTION: Progressive symmetric erythro-keratoderma (PSEK) is inherited as autosomal dominant trait in 50% cases while others have spontaneous mutations (sporadic) or autosomal recessive transmission. However, the pathomechanism involved remains poorly understood as the disorder also shows genetic heterogeneity. The lesions in PSEK are nonmigratory, well-demarcated, polycyclic, geographic shaped, erythematous, hyperkeratotic and mildly scaly plaques with striking symmetrical distribution over elbows, knees, dorsal hands and feet, buttocks and rarely face but typically spares the trunk. The plaques are slowly progressive and increase in number and size until puberty when they tend to stabilize or resolve spontaneously.

OBSERVATION: A 25 year old female patient with previous history of ichthyosis , presented with asymptomatic erythematous scaly plaques on the extensors and flexures of bilateral upper-limbs and axillae.Provisional diagnosis of Progressive symmetric erythro-keratoderma was made and biopsy was consistent with the finding seen in PSEK.

KEY MESSAGE : Flexures are often spared in PSEK and in 50 % cases it autosomal dominant. In this case the lesions are found on the flexural aspect and with adult onset. It lacks the migratory erythema seen in Erythrokeratoderma variabilis but considered to be of the same spectrum. Treatment is often difficult and benefit has been variable with systemic retinoids or topical emollients, keratolytics ointments (urea, salicylic acid, propylene glycol, lactic acid, alpha hydroxy acid), coal tar, retinoids, tacrolimus, and Vitamin D analogues with or without corticosteroid.