GENETICS AND GENODERMATOSES

A CASE REPORT ON A RARE VARIANT OF ERYTHROKERATODERMA

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Background: Erythrokeratoderma is a group of rare inherited kertanisation disorders characterised by variable erythema and hyperkeratotic plaques. It is clinically categorised as the variable form, Erthrokeratoderma variabiilis (EKV) which is more common and the fixed type, Progressive symmetric erythrokeratoderma (PSEK). EKV can be inherited in either autosomal dominant or autosomal recessive fashion, although the former is the most common mode of inheritance.

Observation: An eight year old girl presented to Dermatology outpatient department for reddish brown lesions over trunk and limbs. The lesions were present since one month after birth and were asymptomatic. There was history of waxing and waning of few red coloured lesions which would also change in size and position. The child was born out of consanguineous marriage with uneventful birth history. Her developmental milestones were normal. Family history was insignificant. On cutaneous examination, there were well demarcated reddish brown scaly plaques over neck, chest, abdomen, back, buttocks, thighs , both extensor and flexor aspects of upper limbs and knees. The lesions over thighs and knees were bilaterally symmetrical. There was relative sparing of face, palms and soles. The lesions over chest were gyrate. Systemic examination was normal. KOH examination and culture was normal. Histopathological examination of the plaque over back revealed acanthosis, variable hyperkeratosis and perivascular lymphocytic infiltrate. The clinical and histopathological findings were consistent with Erythrokeratoderma variabilis(EKV) Cram Mevorah.

Key message: Several variants of EKV have been described among which EKV Cram Mevorah is one of the rarely described entity. The differential diagnosis include Erythrokeratodermia variabilis Mendes da Costa, Erythrokeratoderma en cocardes, acute or subacute lupus erythematosus, Netherton Syndrome.