GENETICS AND GENODERMATOSES

ERYTHROKERATODERMIA VARIABILIS: A REPORT OF A TUNISIAN CASE

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Background: Erythrokeratodermia variabilis and prgressiva (EKVP) is a rare genodermatosis, predominantly
inherited in an autosomal dominant mode. It is characterized by two morphologic clinical features: fixed hyperkeratotic plaques and transient erythematous patches. Literature data reported very few cases of EKVP in north Africa. We present herein a sporadic case of EKVP in a four-year-old tunisian boy.

Observation: A four-year-old boy with no past medical history was referred to our department for evaluation of
skin lesions. He was born to non-consanguineous unaffected parents. Lesions appeared four months ago and progressively enlarged. Dermatological examination revealed multiple, hyperkeratotic, brownish plaques, well demarcated, with geographic borders, variable in size and shape. These lesions were fixed and symmetrically distributed on elbows, the perineal region and the back of the thighs. There was not palmoplantar keratoderma. The clinical lesions were suggestive of EKVP. A cutaneous biopsy was performed. The histopathologic examination revealed orthokeratotic hyperkeratosis and papillomatosis, with a church spire configuration of the epidermis. The dermis was unchanged. The patient's skin had been normal at birth. The clinical and histopathological findings were consistent with the diagnosis of EKV. The patient showed a partial improvement under topical treatment with keratolytic agents.

Key message: EKVP is a rare inherited disorder of keratinization. The diagnosis is usually missed. Cutaneous lesions are heterogeneous and include migratory patches and fixed hyperkeratotic plaques. Treatment is only symptomatic. It usually involves the use of topical keratolytic agents and emollient. Low dose systemic retinoid may be beneficial in severe cases.