LINEAR POROKERATOSIS OF MIBELLI

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An unusual case of linear porokeratosis manifesting unilaterally in a 25 year old male patient is reported for rarity and clinical interest.

Key Word : Porokeratosis

Introduction

Porokeratosis of Mibelli (PM) is an autosomal dominant genodermatosis with variable penetrance, characterised by a defective keratinization that histopathologically manifests as a stack of closely packed parakeratotic cells traversing the stratum corneum- the cornoid lamella.[1] Since the original description of this entity in 1893 by Mibelli, various other forms of this disease have been described. These are the disseminated superficial actinic prokeratosis of Chernosky and Freeman (DSAP);[2] porokeratosis plantaris palmaris et disseminata of Guss et al (PPPD);[3] porokeratosis punctata palmaris et plantaris of Brown (PPPP);[4] linear porokeratosis-Rhabari et al [5] and reticulate porokeratosis of Helfman et al.[6] Among these variants the linear porokeratosis is considered to be rare.

Case Report

A 25-year-old male patient presented with a $2^1/_2$ year history of slowly progressive asymptomatic violaceous skin lesions over the right half of the body. The lesions initially started over the right knee and gradually extended both upwards and downwards over the affected limb in a linear fashion and later to the abdomen, chest and upper limb on the right side. Cutaneous examination revealed multiple hyperpigmented violaceous keratotic papules and plaques arranged unilaterally over the lower limb, abdomen, chest and upper limb on the right side. Some of the papules were discrete while others had confluenced to from annular plaques. In majority of the lesions the border was raised and the center was atrophic and partially depressed (Fig. 1) Close examination of the lesions especially the annular ones revealed a characteristic groove traversing the margins. In addition a linear verrucous plaque 3 x 2 cm size was present over the dorsal aspect of the right great toe encroaching the nail plate (Fig. 2) Rest of the skin and systemic

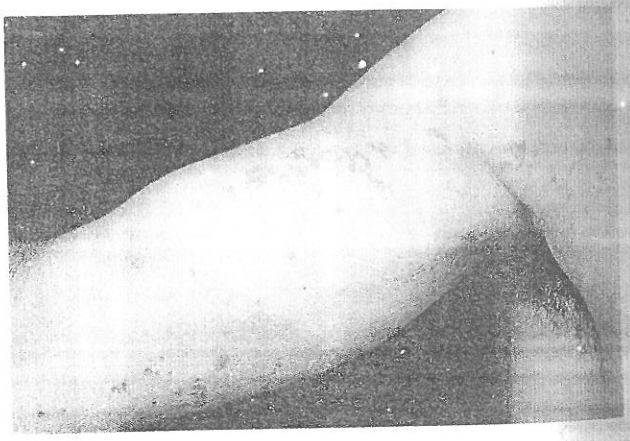

Fig. 1. Hyperkeratotic papules and plaques with raised borders and depressed centre over the upper limb in a linear pattern

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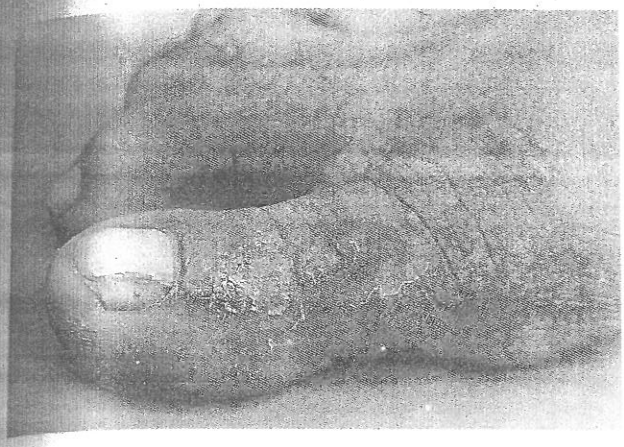

Fig.2. Linear verrucous plaque of porokeratosis over the right great toe simulating linear epidermus naevus

examination did not reveal any abnormality.

Histological examination of the biopsy taken from the lesions over the arm and over right great toe on H & E staining showed characteristic parakeratotic column of cells (cornoid lamella) traversing the stratum corneum, with underlying epidermis showing absence of granular layer and dissolution of basal cells. Lymphocytic infiltrate was seen in the upper dermis corresponding to the cornoid lamella.

Comments

A striking similarity was noted on clinical appearance of the lesions over the upper limb and abdomen to lichen planus and the verrucous plaque over the right great toe to linear epidermal naevus in our patient but the presence of cornoid lamella on histopathological examination left no doubt about the diagnous of PM.

PM is believed to be an autosomal dominant genodermatosis with varying degree of penetrability. However isolated cases may occur due to mutation similar to the patient described herein. Various theories about the pathogenesis of this disorder have been described in the literature. Reed and Leone [1] postulated that the lesions arise in a clone of epidermal cells which can be seen as vacuolated cells underlying the cornoid lamella. This clone of cells spreads laterally with a growth rate more rapid than the resisting epithelial cells so that a fold on groove is formed at the point of pressure as the lesions spreads through the epidermis.

The abnormalities in the dermis underlying cornoid lamella have been suggested to be important in the pathogenesis of PM. Chromosomal instability has been found in cultured fibroblasts from the lesional skin.[7] Actinic radiation, aging, trauma and infection are belived to trigger the disease in a genetically predisposed individual. Reddy et al [8] proposed an interesting theory of the role of female hormones in preventing the development of prokeratosis in famales of reproductive age group. While describing two cases of linear PM, Warren et al [9] reported that the linear lesions may represent a somatic gene mutation in embryonic life rather than Koebner's phenomenon.

In conclusion, it may be considered that linear PM originates from an abnormal clone of epidermal cells carrying the porokeratosis trait arranged along a dermatomal distribution early in embryonic life. The exact event which triggers the area to develop clinical lesions may not be obvious in all cases.

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