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WARTY POROKERATOSIS
(A clinical variant of porokeratosis of Mibelli)

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A 40-year-old male developed porokeratosis of Mibelli with an unusual warty encrustation on the left foot and hand. The diagnosis was confirmed histopathologically. The warty type of porokeratosis is a rare distinct clinical variant.

Key words : Porokeratosis Mibelli, Warty.

Porokeratosis of Mibelli is a rare clonal[1] disease with an abnormal type of keratinization. It is a progressive kerato-atrophoderma characterised by extending plaques of hyperkeratosis succeeded by atrophy. Faulty keratinization occurs in the advancing border with the loss of granular layer and formation of the histopathognomonic cornoid lamella.[2] It is inherited as a simple dominant character with partial limitation to the males.[3] According to Cockayne,[4] the proportion of affected males to females is about 2 : 1. It occurs at all ages. Various morphological forms of the disease have been described.

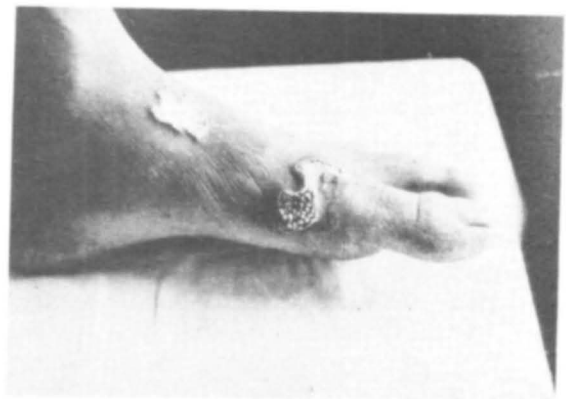

Fig. 1. Porokeratosis of Mibelli with verrucous appearance.

Case Report

A 40-year-old man was seen for asymptomatic, atrophic and hyperkeratotic plaques and papules over the left leg and left hand for the last 7 years. The initial lesion started as a small papule in the left leg and spread peripherally causing a depressed centre. This was followed by the appearance of the other lesions in the left hand. There was no positive family history. The lesions were asymmetrical and circinate with a verrucous centre and well-defined margins (Fig. 1). Atrophic areas were present at a few places. The nails, mucous membranes and hair were normal. Routine blood and urine investigations were within nromal limits. Skin biopsy taken from the periphery of the lesion was consistent with the diagnosis of porokeratosis of Mibelli.

Comments

The term porokeratosis was coined by Mibelli[5] because he considered that the histopathological cornoid lamella of parakeratotic column of cells originated from the sweat duct. However, subsequently, other workers found that the lamella developed not only in the sweat ducts but also in other areas free of any glandular structure. Thus the term porokeratosis is a misnomer. Following the report by Mibelli from Italy, various reports

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were added to the literature. Moncorps[6] recorded 95 cases. Bloom and Abramowitz[7] reviewed 24 reports dealing with 37 cases and reported a family in which three members were affected. According to Butterworth and Strean,[8] not more than 200 cases had been reported. To our knowledge, only 20 cases had been reported from India.[9-16]

Porokeratosis may assume different clinical forms. The lesions may be faint or prominant, small or extensive, atrophic or hyperplastic, hyperpigmented or hypopigmented, asymptomatic or pruritic.[17] Various clinical forms include classical plaque type, superficial disseminated eruptive form, disseminated superficial actinic porokeratosis (DSAP) of Chernosky,[18] hyperkeratotic warty type, hyperplastic form with an inflammatory response leading to ulceration and crusting, linear forms (Freunds forma minima), zosteriform type[19] and porokeratosis plantaris palmaris et disseminata (PPPD).[20] Verruciform lesions of porokeratosis Mibelli are considered as a rare distinct clinical variant. It is quite likely to be confused with diseases like hypertrophic lichen planus and verrucae. However, an accurate diagnosis is achieved by the identification of cornoid lamella in histopathological sections.

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