POROKERATOSIS OF MIBELLI : DISSEMINATED NON ACTINIC FORM

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Two cases of nonactinic - disseminated porokeratosis of Mibelli are reported. Autosomal dominant mode of inheritance was observed in one case.

Key Word : Porokeratosis of Mibelli

Introduction

The term "porokeratosis" was first coined by Vitterio Mibelli in 1893. Though a misnomer, it denotes a faulty form of keratinization. Reed and Leone[1] thought that this disorder was due to a benign, heritable derangement of some epidermal cells.

Clinically it is characterized by annular, sharply demarcated lesion with a raised hyperkeratotic border topped by a furrow. Histopathologically "coronoid lamella" is the hallmark. Six clinical variants have been described : (1) classical porokeratosis of Mibelli (3) linear porokeratosis (4) disseminated superficial porokeratosis (5) disseminated superficial actinic porokeratosis (6) and porokeratosis palmaris plantaris disseminata. (PPPD).

Associations with Bloom's syndrome, multiple cancers of the skin, lung and colon,[2] immunosuppression and organ transplantation,[3] and with dermatomyositis[4] have been reported. Family studies in most of the cases suggested an autosomal dominant mode of inheritance.[5]

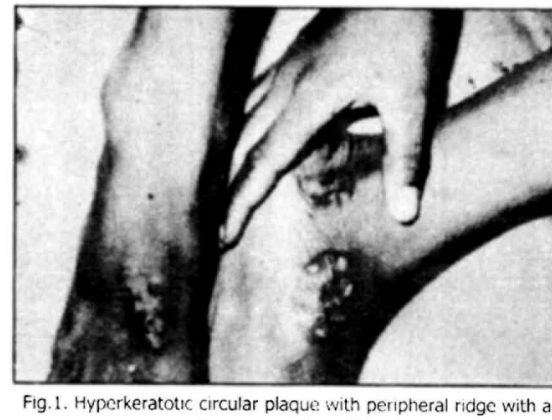

Fig.1. Hyperkeratotic circular plaque with peripheral ridge with a furrow over the ankle.

We report two cases of non-actinic disseminated form of porokeratosis of Mibelli and in one of them autosomal dominant mode of inheritance was

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observed.

Case Report

Case 1- A 11- year old boy presented with asymptomatic, multiple skin lesions of 2 years duration. There was history of similar lesions in his father also. Examination revealed multiple, greyish - black, circular, dry hyperkeratotic plaques of size ranging from 0.5 cm x 0.5 cm to 2 cm x 2 cm in diameter with central atrophy and bordered by a hyperkeratotic peripheral ridge with a furrow. The lesions were present mainly on the extensor aspects of the legs, arms, dorsum and the sides of the feet, soles (Fig.1), trunk with prominent involvement of the face, the upper right eyelid and lower lip. There was complete sparing of the palms, nails, scalp and mucous membranes. The father was also examined and was found to be having similar type of lesions. Histopathology of the lesions in the patient as well as in the father was consistent with the features of porokeratosis of Mibelli.

Case 2- A 22- year-old man presented with asymptomatic multiple skin lesions of 5-6 years duration. There was no history of similar lesions in the family. Examination revealed brownish-black, multiple, dry, circular plaques of varying sizes with central atrophy and peripheral keratotic ridge, distributed over the face, arms, legs and the trunk. Scalp, mucous membranes, palms, soles and nails were spared. Histopathology of the lesion was consistent with the features of porokeratosis of Mibelli.

Discussion

Porokeratosis of Mibelli affects males more often than females. In our cases also both the patients were males. The father of one of the patients had porokeratosis of Mibelli, which indicated autosomal dominant mode of inheritance. This mode of inheritance, has been observed in most of the cases of porokeratosis of Mibelli.[5]

Presence of multiple raised plaques with keratotic border and furrow involving exposed as well as covered areas in both the patients suggest non actinic disseminated form with classical lesions of porokeratosis of Mibelli. On the contrary very flat lesions have been reported in disseminated superficial porokeratosis.[6]

References

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