POROKERATOSIS PALMARIS ET PLANTARIS DISSEMINATA

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A 30-year-old woman had porokeratosis palmaris et plantaris disseminata with unusual features. These included the sex of the patient, absence of family history of similar disease, onset of lesions on the shins and a significant furrow. There was also associated diabetes mellitus.

It is suggested that porokeratosis palmaris et plantaris disseminata falls in the middle of the spectrum of porokeratoses.

Key Word : Porokeratosis

Introduction

Porokeratoses are a group of disorders of keratinization which are inherited as autosomal dominant. Porokeratoses have been classified into various types which include porokeratosis of Mibelli, superficial disseminated porokeratosis, disseminated superficial actinic porokeratosis (DSAP), linear porokeratosis, punctate porokeratotic keratoderma and porokeratosis palmaris et plantaris disseminata (PPPD).

We describe a case affected by the rare variant of porokeratosis, PPPD.

Case Report

A 30-year-old Hindu female, in whom at age 20 years, numerous small, intensely itchy, papular lesions developed on both shins. These papules increased gradually in number involving practically the whole cutaneous surface. The sequence of site involvement was shins, followed by arms, palms and soles, face and lastly the trunk. The lesions enlarged in size becoming annular. There was no seasonal variation. No other family member had similar complaints.

Physical examination revealed innumerable hyperpigmented, hyperkeratotic annular lesions of size varying from 2 to 10 mm present symmetrically all over the body. (Fig. 1) These annular

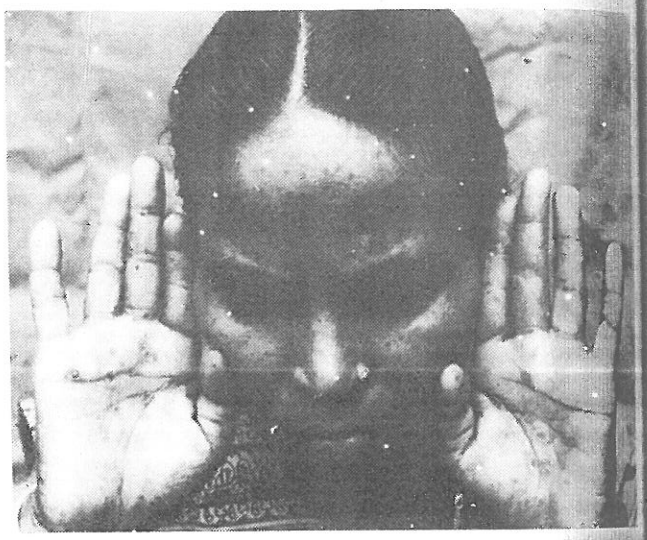

Fig. 1 : Numerous annular lesions with elevated border and deep furrow.

lesions had an elevated border with a furrow on the inner side, which was fairly deep in some of the lesions and central atrophy. There was no involvement of buccal mucosa. No other abnormality was detected on physical examination.

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Blood chemistry analysis showed hyperglycemia as the only abnormality with a fasting blood sugar of 160 mg%.

Histology revealed characteristic changes of porokeratosis. There were cornoid lamellae overlying a groove in the epidermis. Granular layer beneath the cornoid lamellae was absent.

Discussion

Guss et al in 1971,[1] first described a type of porokeratosis, different from that of Mibelli's and DSAP of Chernosky's. The condition has been named as porokeratosis palmaris et plantaris disseminata (PPPD). The authors have described this entity with features shared between these two earlier recognised conditions. PPPD appears in late teens and early twenties, in contrast to porokeratosis of Mibelli which starts in less than 10 years age group[2] and Chernosky's DSAP which starts in the late 30s or 40s.[3]

PPPD is inherited as autosomal dominant mainly affecting males.

The lesions have been described as beginning characteristically from the palms and soles. They have less developed cornoid lamellae, are bilaterally symmetrical with the size of lesions varying from 5 to 10 mm and absent furrow which makes it resemble DSAP closely.

The present case is the first case in Indian literature. Besides being a rare entity, this case had certain unusual features. It was associated with diabetes mellitus. Our patient was a female with no family history of similar disease. It was probably a mutation. The lesions began as itchy papules on the shins followed by lesions on the arms, in contrast to the characteristic onset of PPPD in the palms and soles. The cases reported had shown the absence of forrow but it was significantly present in our case, a feature of porokeratosis of Mibelli.

Some authors have observed existance of more than one type of porokeratosis in members of the same family.[4,5] We believe that PPPD is an entity in the middle of the spectrum of porokeratoses.

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