CASE REPORTS

POROKERATOSIS
(Report of five cases)

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Five cases of porokeratosis are reported. Two of these were of the classical Mibelli type, in one case only one segment of the limb was involved in a linear fashion, in the fourth case a linear lesion involved the entire left side of the body, and the fifth case was of the disseminated superficial type. Typical histopathological features were observed in porokeratosis of Mibelli and also in the case of disseminated superficial type. In one case short contact dithranol and isotretinoic acid treatment was successful.

Key words : Porokeratosis.

Porokeratosis is a rare, chronic, progressive keratoatrophoderma inherited as a dominant character.[1] Only a few cases have been reported from our country.[1] Lever and Schaumberg-Lever[2] described five different types of porokeratosis. These are porokeratosis of Mibelli (classical type), superficial disseminated type, linear porokeratosis, disseminated superficial actinic porokeratosis (DSAP) and porokeratosis palmaris et plantaris disseminata (PPPD).

Two types of histopathological lesions are seen.[2] Firstly, the classical lesion with an invagination of keratin in the epidermis with the central cornoid lamella. This is seen in porokeratosis of Mibelli. The second type is the superficial craters filled with parakeratotic material seen in the other varieties.

No satisfactory treatment is available for porokeratosis. However recently, corticosteroids[3] and topical retinoids[4,5] have been tried successfully. We are reporting five cases of porokeratosis with different clinical features.

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Case Reports

Case 1

A 9-year-old girl was brought for reticulate hyperpigmented lesions over her legs, abdomen and both forearms. She was absolutely healthy about 2 years ago when the lesions started appearing on the legs and then gradually spread to involve the other parts of the body. The lesions were asymptomatic but posed a cosmetic problem. A shallow furrow was present at the margins of the lesions. Histopathological examination showed the characteristic keratotic invagination of the epidermis with central cornoid lamella. The granular layer was absent at this site. Generally such disseminated superficial porokeratosis lesions do not show a parakeratotic column rising from the groove but only a mass of parakeratotic cells on top of a flattened epidermis with intact granular layer.[2] This histopathologic picture was also seen alongside the one with invagination and cornoid lamella.

Case 2

A 13-year-old female reported with lesions over her body since the age of two months. She was a full-term normally delivered baby who was absolutely alright at the time of birth.

She was born to non-consanguinous parents and was the second sib in the family. Since the age of 2 months her parents noticed hyperpigmented lesions over the left great toe which gradually extended in a linear fashion to involve major portions of the left lower limb and the sole. In a similar fashion, the left upper limb was also involved and linear, hyperpigmented lesions were also seen on the left side of the face. To our knowledge his hemidistribution of lesions is not reported. No other member of the family had a similar problem. The lesions were non-pruritic, reticulate and hyperpigmented, with well-defined raised margins. The clinical diagnosis of linear type of porokeratosis was made and histopathology showed a superficial crater filled with parakeratotic cells.

Case 3

A seven-year-old girl had annular lesions restricted to the left thigh, and arranged in a linear fashion (Fig. 1). Morphology and histopathology in this case was similar to the one seen in case 2.

Case 4

A 14-year-old boy had a gradually progressive annular lesion over the left side of the upper lip involving the vermilion border. The lesion was atrophic in the centre and had a raised border with a central furrow. Histopathology confirmed the diagnosis of porokeratosis of Mibelli as there was keratin invagination into the epidermis with the absent granular layer and a central parakeratotic cornoid lamella.

Case 5

A 16-year-old female patient came for a gradually spreading thick lesion on the medial side of the left palm. The lesion started two years ago as a small papule and went on gradually spreading at the periphery. It had thick hyperpigmented scales which were very difficult to remove. The margin of the lesion showed a characteristic furrow (Fig. 2). The medial side of the hand and the little finger looked atrophic. The terminal phalanx of the left little finger had another similar keratotic lesion, and there was difficulty in the movement of the little finger. There was no family history of a similar disorder. We treated the case with short contact dithranol treatment before bath and 0.05% isotretinoic acid application at bed time. The entire lesion including the margins became much softer and the central portion

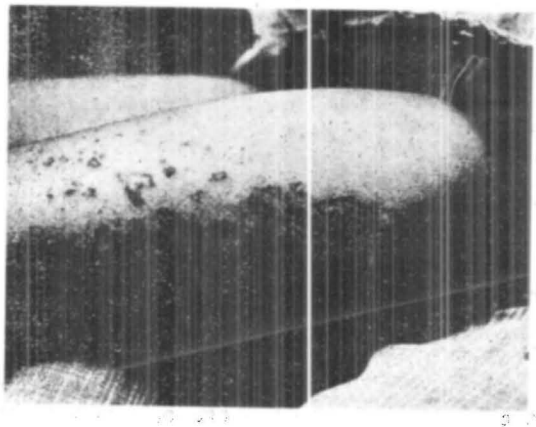

Fig. 1. Annular hyperpigmented lesions restricted to the left thigh.

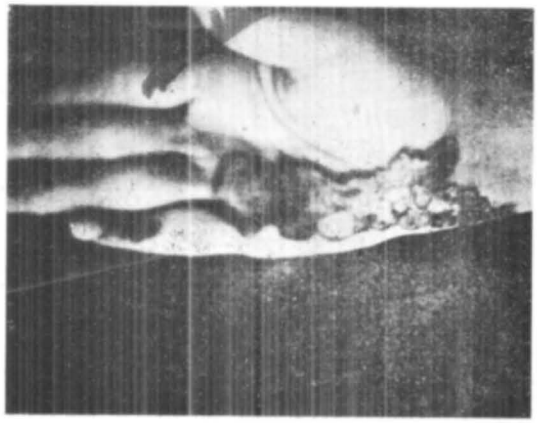

Fig. 2. Thick hyperkeratotic plaque on the medial side of the left plam and terminal phalanx of the little finger.

was devoid of scales, when last examined (Fig. 3). Porokeratosis generally has central atrophy. Our case had presented with thick scales probably because of various herbal medicines that she had used.

Comments

Porokeratosis is a rare disorder, though several cases remain unreported. Two of our cases were of the classical Mibelli type, one involved only one segment of the limb, another involved the entire left side of the body and one was of disseminated superficial type. Recent reports suggest the use of local and systemic corticosteroids[3] or retinoids.[4,5] In one case short contact dithranol and isotretinoic acid treatment was successful.

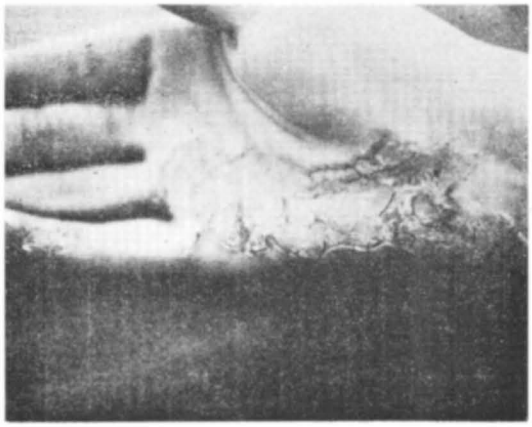

Fig. 3. Improvement seen in the lesion on the palm after treatment with short contact dithranol and isotretinoic acid.

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