SHORT COMMUNICATION

POROKERATOSIS
(REPORT OF THREE UNUSUAL CASES)

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Three unusual cases of porokeratosis - linear, punctate and warty porokeratoses are reported with histopathological features. These three types are rarely seen in clinical practice.

Key word : Porokeratosis

Introduction

Porokeratosis is an autosomal dominantly inherited keratoatrophoderma with a wide variety of manifestations. It is characterised by a distinct peripheral keratotic ridge that corresponds histologically to the coronoid lamella. Five different forms can be distinguished: [1]-1) The plaque type classical porokeratosis of Mibelli, 2) Disseminated superficial actinic porokeratosis (DSAP), 3) Linear porokeratosis, 4) Porokeratosis plantaris, palmaris et disseminata and 5) Punctate porokeratosis. Only a few cases of porokeratosis have been reported from India.[2-5] Three different cases of linear, punctate and warty porokeratosis are reported.

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Case Reports

Case 1

A 9- year- old boy was brought for a single large hyperpigmented, linear lesion over the left upper limb of three years duration .The linear lesion measured 15" x 1" and extended from the styloid process of the radius to acromion process of scapula on the ventrolateral aspect of left forearm and ventral aspect of the arm , with central atrophy and hyperkeratotic, hyperpigmented margins. The typical furrow or crater was noticed near the elbow, (Fig.1).

Fig. 1: Linear porokeratosis with prominent crater over the elbow

No other family member had similar lesions. Histopathological study of biopsy specimen showed hyperkeratosis and typical coronoid lamella filled with parakeratotic cells (Fig.2). Mild lymphocytic infiltration was present in the der-

Fig. 2 : High magnification of coronoid lamella, showing parakeratotic cells (H & E 10 x 40).

mis. He was asked to apply topical 0.05% tretinoin cream daily at bed time for 3 weeks. There was not much improvement.

Case 2

A 50 - year - old man was seen for multiple asymptomatic, discrete, hard papular lesions on both palms and on the palmar aspect of all fingers of 10 years duration. Examination revealed numerous hyperkeratotic punctate pits on both palms and

Fig.3 : Punctate porokeratosis lesions over the palmar creases.

prominently present along the creases of both palms (Fig.3). There were no lesions on the soles and elsewhere over the body. All the routine investigations were within normal limits. No other family member had similar lesions. Histological study revealed features of porokeratosis. A diagnosis of punctate porokeratosis was made.

Case 3

A 45- year -old man presented with a large atrophic plaque with a warty and ulcerated growth at one end of the lesion over left leg, of 2 years duration. Patient was able to walk freely in spite of the growth over the left leg. No other family member was having similar lesion. All the investigations were normal. X-ray of the left leg showed no bony lesion. Examination revealed a large atrophic plaque of 20 cm x 10 cms in size with peripheral hyperkeratotic crater and a large warty mass with ulceration at the lower end of the atrophic plaque on the left leg on the lateral aspect (Fig.4). Clinically, porokeratosis of Mibelli with squamous cell carcinoma was suspected and two biopsies were taken separately from the atrophic plaque near

Fig 4: Large atrophic plaque with a warty mass at the lower margin.

the hyperkeratotic crater and other from the warty growth. Histopathology of the first one was consistent with the porokeratosis. The second one from the warty lesion showed hyperkeratotic finger-like projections with acanthosis, papillomatosis with central fibrovascular core in the epidermis. Histopathologically it was diagnosed as squamous cell papilloma without any evidence of malignant change. The excisional biopsy of the growth also showed the same histological feature and there was no evidence of any malignancy.

Discussion

Porokeratosis of Mibelli is a well known genodermatosis, but its variants and other types are rare. Linear porokeratosis is a distinct, rare entity whose nosological status remains unclear, occurring predominantly in children and young adults with a predilection for extremities. These lesions clinically resemble those of linear verrucous epidermal naevus. In our case, the boy had a single long linear lesion, involving the entire left upper limb from wrist to the acromion process without any lesions over the palms and elsewhere over the body, resembling linear verrucous epidermal naevus; But there was a clear hyperkeratotic crater near the antecubital fossae and histopathological examination revealed a coronoid lamella. Punctate porokeratosis is a rare variety of porokeratosis. It is characterized by seed - like punctate keratoses and/or pits on palms and soles. Sometimes they may be associated with linear porokeratosis. Clinically these lesions may be confused with punctate palmoplantar keratoderma, warts, arsenical keratosis, pitted keratolysis and a variety of other keratinising disorders. However, finding of a coronoid lamella seen as a parakeratotic column arising from a furrow in the irregularly acanthotic epidermis without an intervening granular layer clinched the diagnosis in our case. The premalignant potential of classical porokeratosis and disseminated superficial actinic porokeratosis and disseminated superficial actinic porokeratosis is well known.[6,7] But benign warty lesions clinically resembling squamous cell carcinoma can also occasionally be seen in porokeratosis of Mibelli.[5] Such cases are rarely reported. Our third case was a classical porokeratosis of Mibelli with warty ulcerated lesion. Histopathologically no evidence of malignancy, only benign squamous cell papillomatous changes were noticed.

Though porokeratosis is a known autosomal dominant genodermatosis, sporadic cases also can occur. All the three cases reported here are such sporadic ones without any family background.

References

1. Lever WF, Lever SG. Histopathology of the Skin, 7th Edn, JB Lippincott co, Philadelphia 1990; 70-72.

2.Singh G, Nigam PK, Kumar M. Linear porokeratosis. Indian J Dermatol Venereol Leprol 1987;53: 41 -42.

3.Gharpuray MB, Deshpande SG, Kulkarni V, et al. Porokeratosis (five cases), Indian J Dermatol Venereol Leprol 1988; 54: 255-257.

4.Ghorpade A, Ramam C. Punctate porokeratosis. Indian J Dermatol Venereol Leprol 1995; 61:123-124.

5. Rajendran N . Warty porokeratosis. Indian J Dermatol Venereol Leprol ,1989; 55: 59-60.

6. Gautam PK. Squamous cell carcinoma arising in porokeratosis of Mibelli. Indian J Dermatol Venereol Leprol 1991; 57 : 148 -149.

7. Idam K. Squamous cell carcinoma in lesions of disseminated superficial actinic porokeratosis. Arch Dermatol 1986; 22 : 853.