Porokeratosis: Clinical Observation of 29 Cases

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Background **: Porokeratosis is a disorder of epidermal keratinization which is characterized by the presence of cornoid lamellae. Since the original description of the plaque form of Mibelli, various types of porokeratosis have been reported.**

Objectives **: The purposes of this study were aimed at evaluating the clinical and biological features of porokeraosis.**

Methods **: The hospital charts and slides of 29 patients with porokeratosis were reviewed. Only the cases showing characteristic cornoid lamellae by histopathological examinations were included.**

Results **: Female patients outnumbered the male ones (M:F = 1:2.2). Ages of the onset of porokeratosis varied from infancy to 68 years. Disseminated superficial actinic porokeratosis (DSAP) was the predominant type (22 cases). The Mibelli type was seen in 3 cases and the linear type in 4 cases. Only one case had a family history of porokeratosis. Two patients had complained of pruritus in the lesions, which were the DSAP and linear type, respectively. In that linear type, squamous cell carcinoma developed. Eight patients had other associated diseases. Although 6 patients were treated with CO_2 laser, topical isotretinoin or cryotherapy, the lesions tended to recur, persist or even progress.**

Conclusions **: Porokeratosis showed various clinical features. DSAP was the predominant type. Two patients complained of itching of the lesions, which were the DSAP and linear types, respectively. Within that linear type, squamous cell carcinoma developed. We speculate that pruritus might be an ominous sign of malignant transformation of the porokeratosis. We suggest that the Mibelli type and linear type of porokeratosis should be either excised or destroyed, or in widespread cases close follow-up should be mandatory.**

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Key Words : Porokeratosis

Porokeratosis is a hereditary disorder of keratinization characterized by an extending keratotic lesion with an atrophic center and a prominent peripheral ridge with a fissure at its summit. Cornoid lamella, a parakeratotic column, is its hallmark and an essential pathognomonic feature for diagnosis[1-6]. Since the original description of the plaque form of Mibelli[7], various types of porokeratosis have been reported. The clinical presentation of porokeratosis of Mibelli has been greatly expanded, and seven variants have been clinically distinguished: the plaque type, giant porokeratosis, disseminated superficial porokeratosis (DSP), disseminated superficial actinic porokeratosis (DSAP), linear porokeratosis, porokeratosis plantaris palmaris et disseminata (PPPD), and punctate porokeratosis[10]. Among them, the classification of 5 types were prefered; porokeratosis of Mibelli, the

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linear porokeratosis, DSAP, PPPD, and punctate porokeratosis.

MATERIALS AND METHODS

We reviewed the hospital charts and slides of 29 patients with porokeratosis. Only the cases proven by histopathological evaluations were included among the patients who visited our department from April 1990 to December 1997. Clinical evaluations were performed regarding the age, sex, age of onset, distribution, and associated diseases. We classified the subtypes according to the clinical presentations. Subjective symptoms and family histories of skin lesions were also recorded by chart review.

RESULTS

1. Twenty female patients and nine male patients were included (M:F = 1:2.2).
2. Ages of the onset of porokeratosis varied from

Table 1. Clinical data of porokeratosis

Patients	Age/Sex	Onset.	Locations	Associated diseases
Mibelli type				
1	M/58	48y	shoulder, arm, leg	
2	F/37	37y	right cheek	
3	F/60	30y	right cheek	
Linear type				
1	F/10	3y	arm	
2[a]	M/57	37y	forearm and wrist	SCC within the lesion
3	M/14	9y	right palm (palmoplantar)/ external genitalia (linear)	
4	F/40	2m	Right inner thigh	Endometriosis
DSAP				
1	F/48	48y	face, anterior chest, forearm	Myoma uteri
2	F/50	48y	face	
3	F/55	53y	extremities	
4	M/57	42y	extremities	Bell's palsy
5	F/51	21y	whole body	Hypertension
6[a]	M/67	62y	trunk, extremities	
7	F/46	42y	face, forearm	
8	F/39	29y	face, extremities	
9	M/45	45y	extremities	Chronic urticaria
10	F/35	25y	face, extremities	
11	M/49	41y	trunk, extremities	
12	F/42	22y	face	
13	M/50	50y	trunk, forearms	
14	F/43	33y	face	
15	F/40	39y	face, extremities	
16	F/49	45y	face, trunk, extremities	
17	F/27	27y	face, neck, arms	
18[b]	F/55	45y	face, neck, forearm	
19	F/57	42y	whole body	
20	F/38	34y	face, forearm, dorsum of hand	Polymorphous light eruption
21	F/56	53y	whole body	
22[c]	M/78	68y	face, extremities	Diabetes mellitus, Parkinson's disease

[a] presence of pruritus, [b]presence of family history, [c]hyperkeratotic type

infancy to 68 years. The mean age of onset was 33.4 years.

3. Disseminated superficial actinic porokeratosis (DSAP) was the predominant type (22 cases) (Fig. 1). Among them, one case showed the hyperkeratotic type of DSAP (Fig. 2). The Mibelli type was seen in 3 cases (Fig. 3) and the linear type in 4 cases (Fig. 4). One case showed both

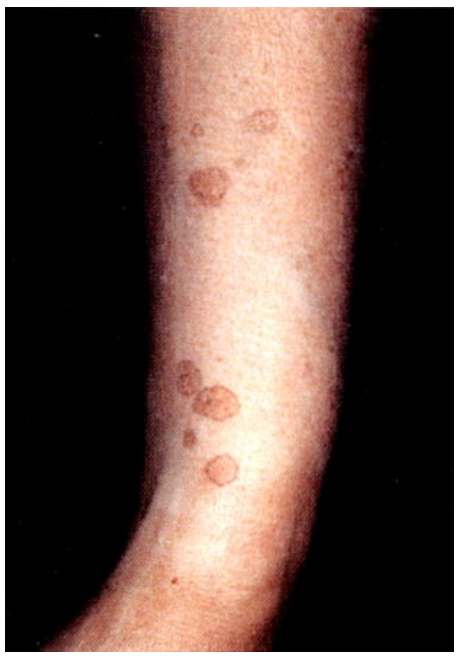

Fig. 1. Several brownish papules with peripheral keratotic castles showing characteristic findings of DSAP.

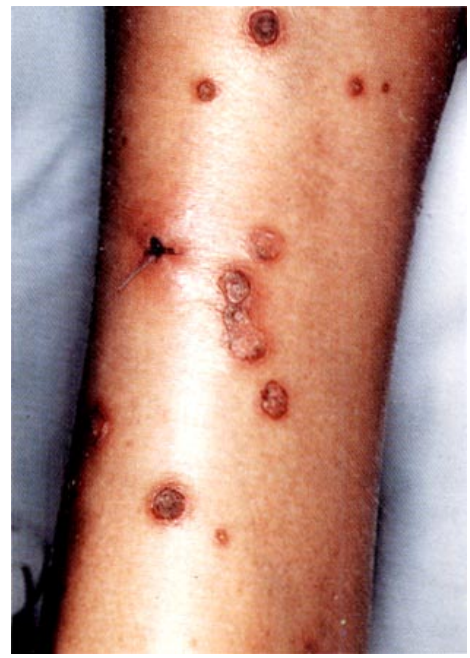

Fig. 2. Numerous brownish hyperkeratotic papules with peripheral ridges on the shin showing the hyperkeratotic variant of DSAP.

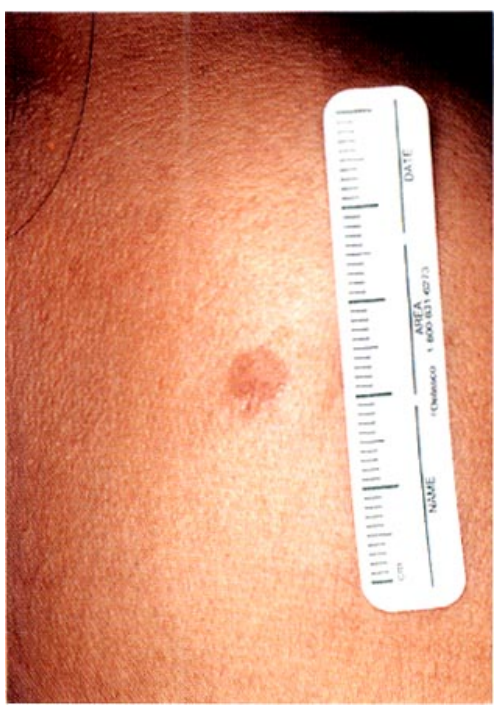

Fig. 3. Solitary discrete brownish papules on the cheek showing the Mibelli type.

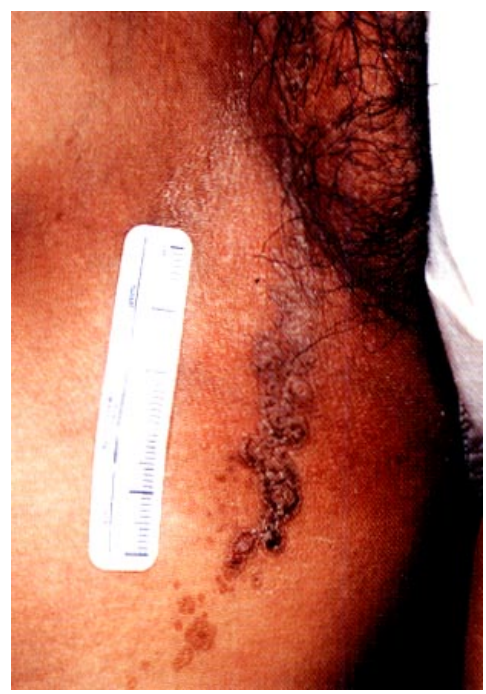

Fig. 4. Linear type of porokeratosis on the inner thigh.

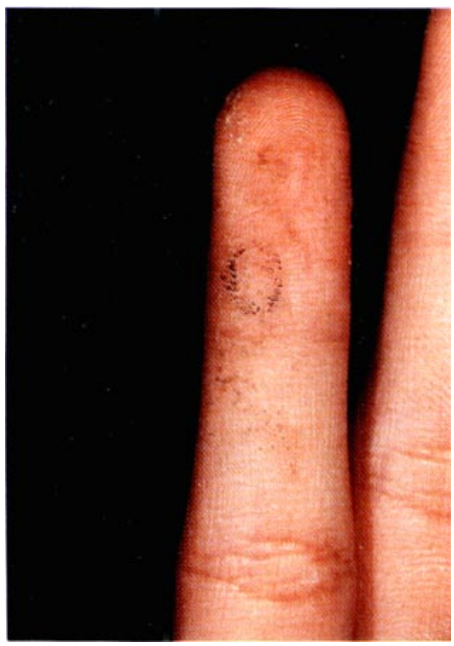

Fig. 5. Palmar porokeratosis.

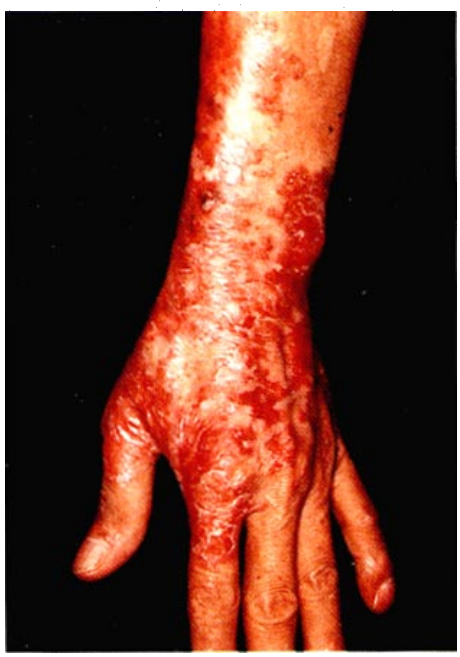

Fig. 6. The linear type of porokeratosis which developed squamous cell carcinoma within the lesion.

types of palmoplantar porokeratosis (Fig. 5) on the right palm and the linear one was present on the groin. Almost all the lesions were confined to the sun-exposed areas, such as the face, anterior chest and extremities. In 3 of 4 cases of the linear type, the porokeratosis had developed in the first decade on the covered areas.

4. Only one case with DSAP had the family history of porokeratosis. Two patients had complained of pruritus in the lesions, which were the types of DSAP and linear types, respectively. In that linear type (Fig. 6), squamous cell carcinoma developed . Eight patients had associated diseases including myoma uteri, Bell's palsy, hypertension, chronic urticaria, endometriosis, leiomyosarcoma in the small intestine, polymorphous light eruption, diabetes mellitus and Parkinson's disease.

5. Although 6 patients were treated with a CO_2 laser, topical isotretinoin or cryotherapy, the lesions tended to recur, persist or even progress except in 2 cases of the Mibelli type which were treated with a CO_2 laser.

DISCUSSION

In our cases, 20 female patients and 9 male patients were included (M:F = 1:2.2). The ages of the onset of porokeratosis varied from infancy to 68 years. The mean age of onset was 33.4 years. The Mibelli type was seen in 3 cases and the linear type in 4 cases. One case showed both types of palmoplantar porokeratosis (Fig. 5) on the right palm and the linear one was present on the groin. DSAP was the predominant type. In 22 patients with DSAP, 16 ones were females in the third and sixth decades. One male patient showed the hyperkeratotic type of DSAP. The hyperkeratotic variant of the porokeratosis has until now been described only 5 times in the literature, and all were the Mibelli type[8,9]. In our case, the skin lesions were distributed over the sun-exposed areas. Histopathological examinations revealed several hyperkeratotic and parakeratotic cornoid lamellae, multiple eosinophilic bodies, degeneration of basal cells, a mild lymphohistiocytic infiltrate around the blood vessels in the dermis, and actinic changes in the upper dermis. To our knowledge, this case is the first report of the hyperkeratotic variant of DSAP. We suggest that the hyperkeratotic variant of DSAP should be considered in multiple hyperkeratotic lesions over the sun-exposed areas.

The pathogenesis of porokeratosis is unclear. The presence of genes involved in the pathogenesis of porokeratosis has been postulated. The genes perhaps may express small keratotic papules, over both exposed and covered areas. Sunlight and/or

ultraviolet light then gradually plays an important role in evolving these papules into typical plaques over the sun-exposed areas[11]. However, other triggering events, such as immunosuppressive therapy after organ transplantation have also been discussed[12]. Immunosuppression might trigger the expression of a mutant clone of epidermal cells, either directly or by disrupting the growth dynamics of the epidermis, or by impairing the immune surveillance[13].

Almost all the lesions were confined to the sun-exposed areas, such as the face, anterior chest and extremities. Interestingly, in 3 of 4 cases of the linear type the porokeratosis developed in the first decade on the covered areas. Only one case with DSAP had a family history of porokeratosis. 2 patients had complained of pruritus in the lesions, which were the DSAP and linear types, respectively. In the linear type, squamous cell carcinoma developed. The lesions were treated with cryotherapy and under close follow-up. All the types of porokeratosis were described to have malignant potential such as squamous cell carcinoma as in our case, Bowen's disease, or basal cell carcinoma especially in immnunosuppressed individuals[12]. Although occurrence of skin cancers in porokeratotic lesions provides clinical evidence that porokeratosis is precancerous, the molecular mechanisms of the carcinogenesis remain unclear. Recently, frequent p53 overexpression has been reported in the Mibelli, DSAP, and linear types and this might be related to the carcinogenic potential of porokeratosis[10].

Eight patients had associated diseases including myoma uteri, Bell's palsy, hypertension, chronic urticaria, endometriosis, leiomyosarcoma in the small intestine, polymorphous light eruption, diabetes mellitus and Parkinson's disease.

The effective therapy is that of physical destruction by electrodesication, cryotherapy, or CO_2 laser, with their undesirable potential for scarring and depigmentation[12]. The more superficial lesions may respond temporarily to topically applied retinoids or 5-fluorouracil, or a combination of both[11]. Although 6 patients were treated with a CO_2 laser, topical isotretinoin or cryotherapy, the lesions tended to recur, persist or even progress except in 2 cases of the Mibelli type which were treated with a CO_2 laser.

In conclusion, porokeratosis showed various clinical features. DSAP was the predominant type. We speculate that pruritus might be an ominous sign of malignant transformation of the porokeratosis. Since porokeratosis is a premalignant lesion, the lesions should be excised or destroyed, or in widespread cases close follow-up should be mandatory.

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