

# Coexistence of Porokeratosis of Mibelli with Linear Porokeratosis

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We report a case of porokeratosis of Mibelli coexistence with linear porokeratosis in a 28-year old male. He had multiple discrete reddish to brownish annular plaques with peripheral keratotic ridges on the left side of the neck and axilla, and such lesions extended in a linear fashion on the left side of the upper arm and posterior aspect of the lower extremity. The cutaneous lesions started on the left neck and axilla about 6 years prior to consultation. Two years after that, new lesions developed on the left side of the upper arm and lower extremity in a linear fashion. There was no family history of similar skin disorders. A histological examination showed cornoid lamellae in the horny layer. Hypogranulosis, dyskeratotic cells and perivascular mononuclear infiltrations were also seen beneath the cornoid lamella.

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Porokeratosis(PK) is an uncommon disorder of keratinization that is characterized histologically by the presence of a compact column of parakeratotic cells termed a cornoid lamella. Although histologically similar, clinically five forms of PK are recognized, including PK of Mibelli, disseminated superficial actinic PK (DSAP), porokeratosis palmaris, plantaris, et disseminata (PPPD), punctate PK, and linear PK<sup>1</sup>. Although there are a number of reports of the variants of PK, the coexistence of two or more of the variants is rare. Herein we report a case exhibiting the clinical features of PK of Mibelli and linear PK simultaneously in one patient; to our knowledge, this is the second description of those variants coexisting in Korea.

## CASE REPORT

A 28-year-old man visited our clinic because of groups of annular and linear skin eruptions on the left side of his neck, axilla, upper arm, buttock, and lower extremities. He first noticed scaly annular lesions on his left neck and axilla about 6 years previously. Two years later, linear streaky skin eruptions had developed and extended along his left forearm, buttock, and lower extremity.

Examination of the neck and axilla revealed groups of reddish to brownish annular rim-like, or gyrate plaques with keratotic peripheral ridges and atrophic centers (Figure 1A, B). Such lesions were arranged in a linear fashion on the left upper arm, buttock, and posterior surface of the lower extremities (Figure 2A, B). Though all of the lesions were cosmetically distressing, they were asymptomatic. His familial and past medical histories were not significant. There was no history of previous trauma or physical injury. On physical examination, specific findings were not seen except for the skin lesions. Routine laboratory studies were within normal limits or negative. Biopsy specimens from the peripheral ridges of both annular and linear lesions

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**Fig. 1A, B.** Reddish to brownish annular rim-like, or gyrate plaques with keratotic peripheral ridges and atrophic centers on the left neck and around the left axilla.

demonstrated columns of parakeratosis, termed cornoid lamellae, in the basket-weave cornified layer of the stratum corneum. Hypogranulosis, dyskeratotic cells and perivascular mononuclear infiltrations were also seen beneath the cornoid lamella (Figure 3). The clinical picture supports the diagnoses of both PK of Mibelli and linear PK. We tried to remove all the lesions with topical 0.025% tretinoin twice a day for 2 weeks but this was not effective.

## DISCUSSION

The incidence of the PK variants is not clear,

but Sasson reviewed 281 cases of PK, and of these, 116 were classified as DSAP (41.3%), 92 as Mibelli (32.7%), 42 as linear (14.9%), 21 as PPPD (7.5%), and 10 as punctate (3.6%)<sup>2</sup>. In Korea, of the 79 cases, 44 were classified as DSAP (56%), 22 as Mibelli (28%), 11 as linear (14%), and 2 as punctate (2%)<sup>3</sup>. The mean age of all patients with PK is 43.9 years<sup>2</sup>, but is 33.4 years in Korea<sup>3</sup>. Overall, males and females are equally represented, but the male-to-female ratio varies considerably among the different types<sup>2,3</sup>.

Reed and Leone suggested that PK results from mutant clones of epidermal cells, which extend peripherally<sup>4</sup>. This is supported by auto-transplantation experiments<sup>5</sup> and by detection of abnormal DNA ploidy<sup>6</sup>. Ito *et al* found that dyskeratotic cells beneath the cornoid lamella were lacking in SH groups and rich in SS linkages in a study using N-(7-dimethylamino-4-methyl-3-coumarinyl)maleimide (DACM) staining<sup>7</sup>. This suggests a tendency towards rapid or incomplete keratinization of the cornoid lamella forming keratinocytes. There is substantial evidence that in some cases, immunosuppression triggers the disease<sup>8</sup>. Alternatively, an infectious agent, perhaps a virus, might induce the disorder<sup>9</sup>.

PK of Mibelli is probably transmitted as an autosomal dominant trait with a distinct predilection for males<sup>2,3,10</sup>. The dermatosis has an early onset and gradually evolves into its characteristic morphology. The lesions may be single, few, or numerous, and are usually found on the face, trunk, and extremities. Our patient had had groups of annular rim-like, or gyrate, reddish to brownish plaques with keratotic peripheral ridges and atrophic centers on his left neck and axilla, since the age of 22, with no family history such lesions (Figure 1A, B). Occasionally, isomorphic/Koebner's phenomenon may be seen<sup>11</sup>. Hair, nails, and mucous membranes are usually spared. The lesions may simulate other skin diseases such as atrophic lichen planus, verruca vulgaris, actinic keratosis, squamous cell carcinoma, basal cell carcinoma, scarring, milia, and solitary inflammatory lesions<sup>12</sup>. Linear PK is clinically characterized by lesions following the lines of Blaschko<sup>13</sup>. The embryological interpretation of the Blaschko line is that it corresponds to the direction of growth of clones of cutaneous cells (epidermal and dermal)<sup>14</sup>. It usually appears in childhood and primarily involves the extremities<sup>13,15</sup>.

**Fig. 2.** A: Aggregation of small round plaques in a linear fashion on the left upper arm.  
B: A brownish red colored, irregular, hypertrophic, linear plaque extended from the buttock to the ankle.

**Fig. 3.** Skin biopsy, taken from the plaque of the left leg, shows a cornoid lamella in the basket-weave cornified layer of the stratum corneum. Hypogranulosis, dyskeratotic cells and perivascular mononuclear infiltrations are seen beneath the cornoid lamella (H&E,  $\times 200$ ).

The differential diagnosis of linear PK of childhood includes linear verrucous epidermal nevus, lichen striatus, linear psoriasis and linear lichen planus<sup>15</sup>. However, demonstration of a cornoid lamella in histological sections is the defining characteristic<sup>16</sup>. Our patient had confluent reddish to brownish hyperpigmented plaques with atrophic centers and peripheral keratotic ridges arranged in a linear fashion on his left upper arm, buttock, and posterior surface of the lower extremity (Figure 2A, B). Biopsy specimens from the peripheral ridges of both the annular and linear lesions demonstrated columns of parakeratosis, termed cornoid lamellae, in the basket-weave cornified layer of the stratum corneum. Hypogranulosis, dyskeratotic cells and perivascular mononuclear infiltrations were seen beneath the cornoid lamella (Figure 3). However, the depth of cornoid lamella may vary from full invagination to superficial lo-

**Table 1.** Summary of reported cases of coexistence of variants of porokeratosis in Korea

Authors	Sex/Age	Clinical types	Duration (years)	Inheritance	Treatment
Kim et al <sup>17</sup>	F/23	Linear	—	+	5-FU cream
		DSAP	—		
Park et al <sup>18</sup>	F/43	Linear	21	—	Cryotherapy
		DSAP	2		
Kim et al <sup>19</sup>	F/46	Linear	26	—	Cryotherapy
		DSAP	26		
Lee et al <sup>20</sup>	F/20	Linear	13	—	Chemical peeling with TCA
		Mibelli	13		
Present case	M/28	Mibelli	6	—	Tretinoin
		Linear	4		

calization in all clinical PK variants<sup>16</sup>. With the above findings, the diagnosis of the coexistence of PK of Mibelli with linear PK is strongly suggested.

The coexistence of several porokeratotic variants is rare. It has been described for DSAP with linear PK and for PK of Mibelli with linear PK in Korea<sup>17-20</sup> (Table 1). PK was formerly believed to be an autosomal dominant condition with variable penetrance, where the predisposition to PK is inherited but not the predisposition to the type of PK<sup>4,21</sup>. In view of the coexistence of different PK variants in one patient, it now seems likely that porokeratoses are genetically determined disorders that are carried on closely linked gene loci<sup>21</sup>. It is interesting that all of the cases of PK of Mibelli with linear PK in the literature<sup>13,20-22</sup>, including the present case, occurred in a unilateral fashion. Therefore, it strongly represents a mosaicism for two closely linked genes responsible for the Mibelli type and linear PK.

Malignant skin tumors such as squamous cell carcinoma, basal cell epithelioma, and Bowen's disease may affect 7% of porokeratotic patients<sup>23</sup>. These have been substantiated by the studies of Gray et al, who used several markers of keratinocyte maturation and differentiation, including involucrin, fillagrin, cytokeratin, and growth activation markers (psi-3), and reported that staining patterns of keratinocytes beneath the cornoid lamella was similar to that of squamous cell carcinoma<sup>24</sup>. Otsuka et al demonstrated abnormal DNA ploidy in lesional epidermal cells, a sensitive marker of premalignant conditions<sup>6</sup>. Malignant degeneration may occur in all types of porokeratosis, in particular, giant form and linear PK

are more susceptible to malignant transformation<sup>2,13</sup>, so our patient should be recommended for a careful follow-up. Older patients and those with long-standing disease are at increased risk for development of malignancy<sup>2</sup>.

Treatment of PK is empirical. It is, however, essential to consider the malignant potential of this dermatosis. Surgical excision has been practised for small lesions. Alternative therapies, including electro-cautery, curettage, liquid nitrogen, keratolytics, topical 5-fluorouracil, and intralesional steroids, have been used in the past with disappointing results. Retinoids have also been successful in inducing remissions in a few cases<sup>25</sup>, but results have been variable. We tried to remove all lesions with 0.025% tretinoin application twice a day for 2 weeks but could not gain a satisfactory effect. Dermabrasion and carbon dioxide laser excision have recently been proposed as alternatives that are cosmetically and functionally superior to conventional therapies<sup>26,27</sup>.

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