

A Case of Basal Cell Carcinoma Arising in Linear Porokeratosis

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Porokeratosis are characterized by distinct clinical findings of a keratotic ridge that corresponds to the cornoid lamella on histology and has well defined potential for malignancy.

We report a case of basal cell carcinoma(BCC) arising in linear porokeratosis in a 77-year-old man. (Ann Dermatol 15(2) 78~81, 2003).

Key Words : Basal cell carcinoma, Linear porokeratosis

Porokeratosis of Mibelli is a familial chronic progressive keratoatrophoderma which demonstrates a characteristic clinical and histologic picture. Since Mibelli described three cases in 1893, several clinical variants have been identified. The others include disseminated superficial actinic porokeratosis(DSAP), porokeratosis plantaris palmaris et disseminata(PPPD), linear porokeratosis and punctate porokeratosis.

The unifying feature is the histologic presence of cornoid lamellae. This is characterized by a thin column of parakeratosis extending through the surrounding orthokeratotic stratum corneum, an absent or decreased stratum granulosum beneath the parakeratotic column, either vacuolated or dyskeratotic cells in the underlying stratum malpighii, and a perivascular lymphocytic infiltration in the papillary dermis.

Malignant degeneration has been observed and various malignancies have been previously described in the all types of porokeratosis but we

could not find a case of BCC arising in linear porokeratosis. We present herein a case of BCC arising in this type of porokeratosis in a 77-year-old man.

CASE REPORT

A 77-year-old man presented with diffuse but linear eruptions over large area of the body. The lesion had been growing slowly during childhood and was occasionally pruritic. During the course of disease, a pigmented papule developed in the center of the erythematous lesion on the back. Neither his parents nor his siblings have similar lesion.

Physical examination revealed a linear arrangement of discrete, brownish-gray, annular plaques on the back. It consists of numerous small oval papules with an atrophic, sometimes red brown center and a raised edge. It involved also forehead, left chest, shoulder and arm(Fig.1). In the lower back, a solitary dark pigmented, 2 x 3 cm sized, flat plaque developed on the center of porokeratotic lesions was noted(Fig.2). Regional lymph node were not palpable and the remainder of physical examination was not contributory. On laboratory tests including a complete blood cell count, urinalysis, liver function test, BUN/creatinine, the results were within normal limits or negative.

Biopsy specimens were obtained from the peripheral ridge of the patch and the central pigmented plaque. The specimen of the peripheral ridge showed atrophic epidermis bordered on side by

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Table 2. Table 1. Reported cases of BCC arising in porokeratosis

Date	Author	Race	Sex	Age	Site of carcinoma	Histologic type	Type of porokeratosis	Treatment
1973	Sarkany ⁵	Caucasian	Male	55	Right thigh	BCC	Porokeratosis of Mibelli	No data
1982	Glicman ⁶	Caucasian	Female	75	Right leg	BCC	Porokeratosis of Mibelli	Radiotherapy
1989	Cheong ⁷	Asian	Female	58	Neck	BCC	PPPD	Surgical excision
2002	Presented case	Asian	Male	77	Back	BCC	Linear porokeratosis	Surgical excision

lines of Blaschko on the back but the left chest and arm lesions occurred in an unilateral linear form resembling a linear verrucous epidermal nevus. A diagnosis of linear porokeratosis with associated BCC was made. Total excision for BCC and oral administration of retinoic acid for the treatment of linear porokeratosis and for prevention of cancer development was done.

DISCUSSION

The porokeratosis has well defined potential for malignancy but the exact relationship between porokeratosis and malignancy is not clear. Occasional reports of apparent malignancy in lesions of porokeratosis has stirred an uncertainty concerning the malignant potentials of the porokeratosis. However, the mutated clone theory has been used to explain the increased incidence of carcinomas associated with porokeratosis¹⁻².

Malignant degeneration occurring in porokeratosis was first noted in 1942 by Vigne³. And then various malignancies have been previously described in all types of porokeratosis. To date, there have been reports of squamous cell carcinoma (SCC)⁴, BCC⁵⁻⁷, Bowen's disease⁸ arising in porokeratosis.

Linear porokeratosis, also referred to as zosteriform porokeratosis, is characterized clinically by linear and whorled verrucous plaques that appear at birth or in childhood. Like other lesions of porokeratosis long-standing lesions of linear porokeratosis may transform into malignancy. But we could not find a case of BCC arising in this type of porokeratosis. In fact, there were only three cases of BCC arising in porokeratosis in English literature and then all reported cases of BCC had arisen in

Porokeratosis of Mibelli type and PPPD type.

A summary of the available facts in respect of previous reported cases of BCC arising in porokeratosis is presented in Table 1. and our own cases are included. Although there were cases with BCC arising from porokeratosis of Mibelli and PPPD, the fact that from linear porokeratosis, which has never been reported in the English literature, is very interesting.

Like other porokeratosis, a generally accepted mode of treatment does not exist for linear porokeratosis, variable therapeutic success has been achieved with keratolytic agents and other agents⁹. We tried to treat with retinoic acid, applied systemically, has favorable therapeutic effect on hyperkeratosis and has been used in different hyperkeratotic conditions including porokeratosis.

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