A Case of Basal Cell Carcinoma Arising in Linear Porokeratosis

Jung Sub Yeum, M.D., Jin Chun Suh, M.D., Dong Ju Shin, M.D.

Department of Dermatology, Fatima Hospital, Taegu, Korea

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 keratoderma 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 (PPP), 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 infiltrate 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 lymphs 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

Received December 13, 2002
Accepted for publication March 12, 2003
Reprint request to: Jung Sub Yeum, M.D., Department of Dermatology, Fatima Hospital, 302-1 Sinam-Dong, Don-Gu, Taegu, 701-600, Korea
Tel. (053)940-7380, Fax: (053)954-7417
E-mail. skinfriend@korea.com

*This case was presented at the 54th Annual Meeting of the Korean Dermatological Association on April 17, 2002.
Fig. 1. Unilateral widespread linear and whorled lesions of porokeratosis following Blaschko’s line.

Fig. 2. Curious linear distribution of lesion on the back. Note the pigmented plaque area on the left side of lower back, in which malignant transformation has taken place (arrow).

Fig. 3. Microscopic section through border of typical lesion of porokeratosis. Note cornoid lamella on right with plug of parakeratotic cell, loss of granular layer and perivascular mononuclear cell infiltration in the dermis (H & E, ×200).

Fig. 4. Histopathologic finding of plaque on the center of porokeratotic lesion showing irregular proliferation of basaloid tumor cell, peripheral palisading and cleft formation surrounding tumor (H & E, ×100).

parakeratotic column, so-called cornoid lamella (Fig.3), which represents the histopathologic hallmark common to all clinical variants of porokeratosis. The epidermis underlying this parakeratotic column was devoid of granular layer. The biopsy specimen taken from the pigmented plaque in the center of the linear lesion showed undifferentiated BCC (Fig.4).

Although the lesions do not follow the unilateral
Table 2. Table 1. Reported cases of BCC arising in porokeratosis

<table>
<thead>
<tr>
<th>Date</th>
<th>Author</th>
<th>Race</th>
<th>Sex</th>
<th>Age</th>
<th>Site of carcinoma</th>
<th>Histologic type</th>
<th>Type of porokeratosis</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1973</td>
<td>Sarkany</td>
<td>Caucasian</td>
<td>Male</td>
<td>55</td>
<td>Right thigh</td>
<td>BCC</td>
<td>Porokeratosis</td>
<td>No data</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>of Mibelli</td>
<td></td>
</tr>
<tr>
<td>1982</td>
<td>Glicman</td>
<td>Caucasian</td>
<td>Female</td>
<td>75</td>
<td>Right leg Neck</td>
<td>BCC</td>
<td>Porokeratosis</td>
<td>Radiotherapy</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>of Mibelli</td>
<td></td>
</tr>
<tr>
<td>1989</td>
<td>Cheong</td>
<td>Asian</td>
<td>Female</td>
<td>58</td>
<td>Back</td>
<td>BCC</td>
<td>PPPD</td>
<td>Surgical excision</td>
</tr>
<tr>
<td>2002</td>
<td>Presented case</td>
<td>Asian</td>
<td>Male</td>
<td>77</td>
<td></td>
<td>BCC</td>
<td>Linear porokeratosis</td>
<td>Surgical excision</td>
</tr>
</tbody>
</table>

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 porokeratosis.

Malignant degeneration occurring in porokeratosis was first noted in 1942 by Vigne. And then various malignancies have been previously described in all types of porokeratosis. To date, there have been reports of squamous cell carcinoma(SCC), BCC, Bowen’s disease 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 agents. 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.

REFERENCES