A Case of Squamous Cell Carcinoma and Bowen's Disease Associated with Superficial Disseminated Porokeratosis

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We describe the clinical and pathologic observation of a 50-year-old man with superficial disseminated porokeratosis who developed a squamous cell carcinoma on the dorsum of his right thumb and Bowen's disease on his right upper arm. The tumors were surrounded by lesions of superficial disseminated porokeratosis and were thought to develop from the dysplastic epidermal cells located under the cornoid lamellae.
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Porokeratosis is a rare genodermatosis with an autosomal dominant mode of inheritance. It is characterized by one or many atrophic, keratotic patches surrounded by a distinct, and raised ridge showing the cornoid lamella. Several clinical forms of porokeratosis that have been described by Lever and Shaumberg-Lever, are as follows: (1) the plaque type, originally described by Mibelli, (2) the superficial disseminated form, (3) disseminated superficial actinic porokeratosis (DSAP), (4) the linear form, (5) the punctate form.

The superficial disseminated porokeratosis, which have been described by Guss et al in 1971 as porokeratosis palmaris, plantaris, et disseminata, often shows widely distributed lesions especially on the trunk, palms, and soles. It usually appears during adolescence and gradually spreads until virtually the entire body is involved.

Development of squamous cell carcinomas, basal cell carcinomas or Bowen's diseases within lesions of porokeratosis have been reported in patients with solitary lesions as well as in disseminated lesions.3-10

We describe a case where both squamous cell carcinoma and Bowen's disease developed in areas of superficial disseminated porokeratosis.

REPORT OF A CASE

A 50-year-old farmer presented to the dermatology clinic at Hanyang University Hospital in March 1988, with a 30-year-history of multiple, hyperkeratotic lesions on his face, neck, trunk and extremities. The lesions had developed initially on his trunk and spread to his arms, legs and face. A dark hyperkeratotic, verrucous tumor developed on the injured site of his right thumb 2 months previously. The tumor was surrounded by the superficial hyperkeratotic lesions. He was in good general health. There was no significant past history except for a history of repeated trauma to remove the spine on the dorsum of his right thumb for the past 2 years. At that time, he attested to similar conditions in three generations of his family, including his daughter (Fig. 1).

Physical examination on his first visit disclosed a 3x5cm hyperkeratotic verrucous tumor on the dorsum of his right thumb and hundreds of annular, dry, hyperkeratotic lesions, 0.3-1.0cm in diameter, on his face, neck, trunk, and extremities; except the palms, soles and mucous membranes were spared (Fig. 2).

Laboratory data were as follows: a complete
A biopsy specimen taken from a lesion on the anterior chest showed characteristic changes of porokeratosis. These included a cornoid lamella composed of a column of parakeratosis overlying a slight "dell" in the epidermis, beneath which the granular layer was thinned to absent, and a few surrounding dyskeratotic keratinocytes (Fig. 3). A skin biopsy taken from the tumor on the dorsum of his right thumb showed moderately differentiated squamous cell carcinoma which infiltrated into the deep dermis (Fig. 4, 5). Near the margin of the squamous cell carcinoma, there was a deep groove filled with a parakeratotic column (Fig. 6, 7). Subsequently, disarticulation of the right first metacarpocarpal joint was performed.

In September, 1988, the patient returned for