

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 Shaumburg-Lever,¹ 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 al² 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 3×5cm 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

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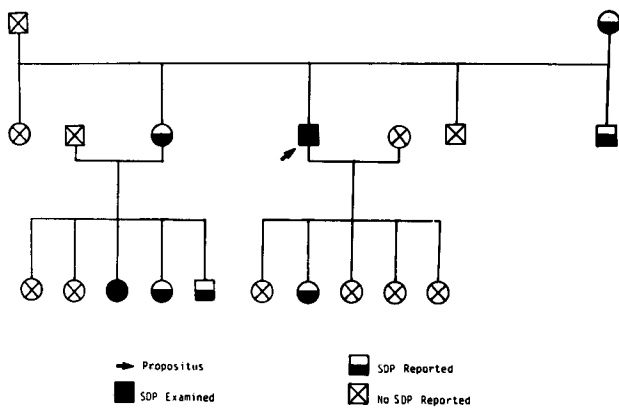


Fig. 1. Family tree. SDP indicates superficial disseminated porokeratosis.



Fig. 2. Porokeratotic lesions on the neck, trunk, and extremities.

blood cell count with differential was within normal ranges, erythrocyte sedimentation rate was 8mm/hr, routine urinalysis, stool examination for occult blood, roentgenogram of the chest were normal. The pattern of serum protein electrophoresis, the values of liver function test, blood levels of urea nitrogen and creatinine, 24-hour urine creatinine clearance, fasting blood sugar level, serum concentration of immunoglobulins, ASO titer, VDRL, peripheral T and B cell counts, T4/T8 ratio, and bone scan with radioisotope were all within normal limits or negative. The serum total hemolytic complement (CH50), C3, and C4 concentrations were within normal ranges. Delayed hypersensitivity reaction to 7 standardized recall antigens (Multitest® MERIEUX; Tetanus, Diphtheria, Streptococcus, Tuberculin, Candidin, Trichophyte, Proteus) were not disturbed.

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

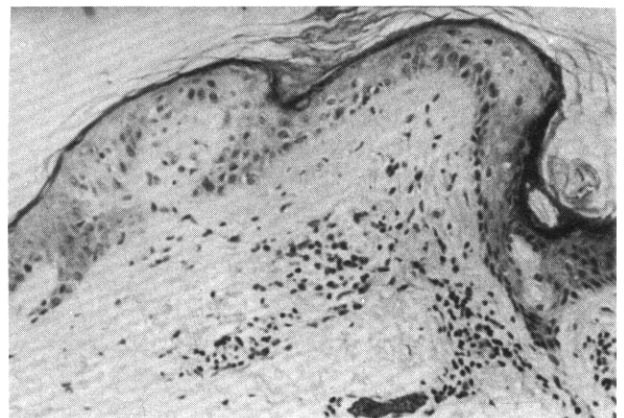


Fig. 3. Biopsy specimen taken from anterior chest showing cornoid lamella. Note the absence of a granular layer underlying the parakeratotic column (H & E, $\times 100$)

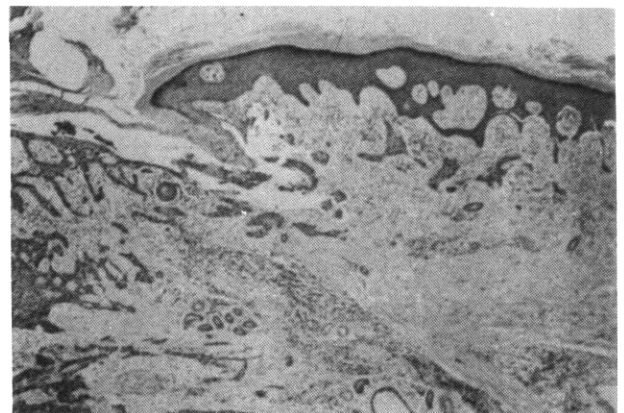


Fig. 4. Biopsy specimen taken from the dorsum of right thumb showing moderately differentiated squamous cell carcinoma (H & E, $\times 40$)

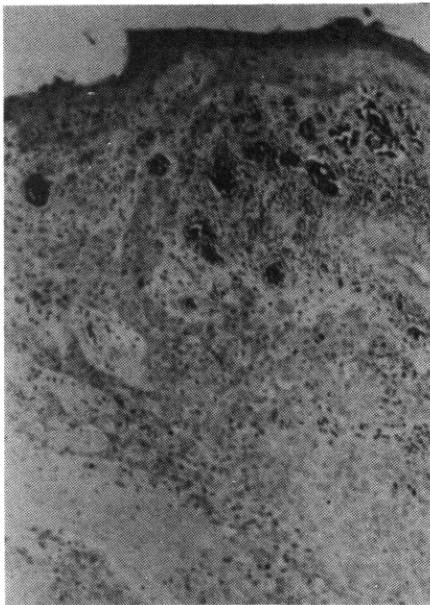


Fig. 5. Higher magnification of portion of moderately differentiated squamous cell carcinoma (H & E, $\times 400$).

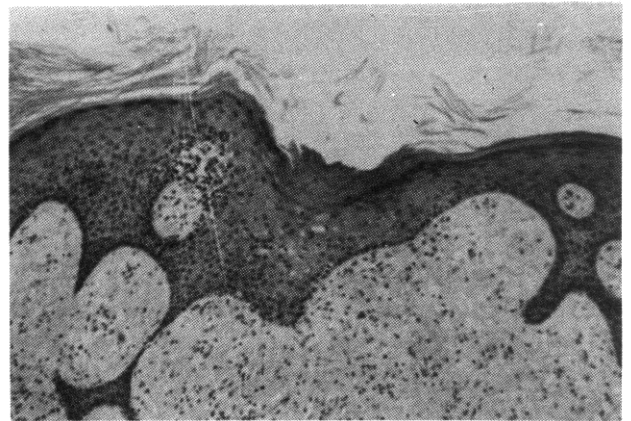


Fig. 7. Higher magnification of parakeratotic column of Fig. 6 (H&E, 100).

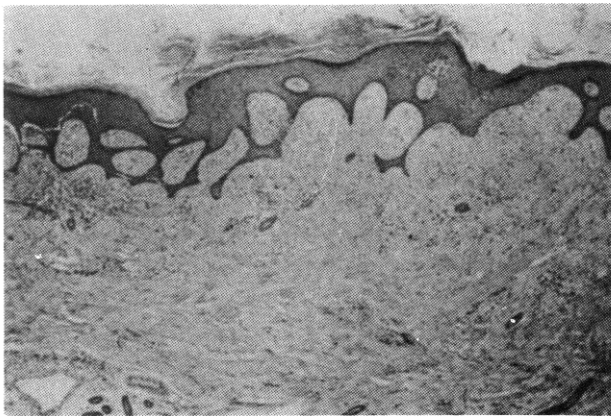


Fig. 6. Near the margin of the squamous cell carcinoma there is parakeratotic column (H & E $\times 40$).

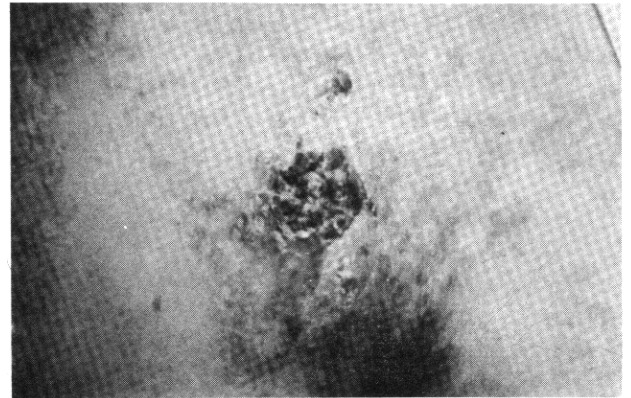


Fig. 8. Verrucous brownish tumors developed in porokeratotic lesions on the right upper arm.

evaluation of several verrucous, erythematous to brownish nodules and tumors, 1.0-3.0cm in diameter, on his right upper arm and his right shoulder of four months duration (Fig. 8).

Histologic sections revealed an irregularly acanthotic epidermis with diffuse anaplasia characteristic of Bowen's disease (Fig. 9, 10). These tumors were treated by excision.

DISCUSSION

Epidermal malignancies can develop in all types of porokeratosis. The development of squamous

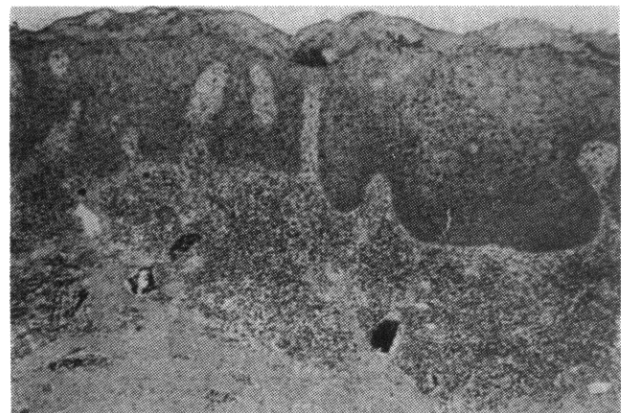


Fig. 9. Biopsy specimen taken from a verrucous tumor on the right upper arm shows anaplasia of Bowen's disease (H & E, $\times 40$).

cell carcinoma, Bowen's disease, and basal cell carcinoma in the lesions of porokeratosis of Mibelli have been observed.⁷⁻⁹ In addition, squamous cell

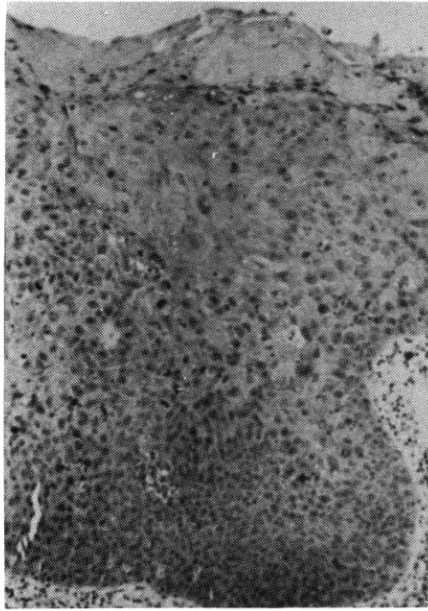


Fig. 10. Higher magnification of portion of Fig. 9 (H&E, $\times 200$).

carcinoma is now known to occur within lesions of porokeratosis palmris, plantaris, et disseminata,⁶ DSAP,⁶ and linear porokeratosis.¹¹

Also squamous cell carcinoma with metastasis have been reported¹¹⁻¹² in porokeratosis.

Reed and Leone¹³ have hypothesized that porokeratosis evolves from a clone of abnormal epidermal cells at the base of the cornoid lamella. Shrum *et al*⁶ concluded, from a study of the histopathologic features in their case, that the carcinoma might have developed from foci of epidermal dysplasia that also appeared to give rise to cornoid lamellae.

Most of the neoplasms reported have been on the distal areas of the extremities, where trauma as well as ultraviolet light might affect the skin. Also, immunosuppressive therapy and irradiation with x-ray or ultraviolet light might predispose one to the development of malignant changes.^{3, 11, 14} Our patient has developed a squamous cell carcinoma and Bowen's disease on sun-exposed areas. There has been a 2 year history of repeated local injuries on the dorsum of his right thumb where the squamous cell carcinoma developed. Perhaps the combination of exposure to sunlight in summer and repeated injuries might be the cause of the development of these cancerous lesions.

Numerous treatments have been attempted for

all types of porokeratosis, but the results generally have been poor.¹⁵ Small, solitary lesion can be excised or destroyed. Other modalities include liquid nitrogen, electrodesiccation, topical 5-fluorouracil, oral retinoids, and various topical keratolytic agents.¹⁶⁻¹⁷ Close observation, if lesions persist, is most important for early detection of malignant transformation.

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