Superficial Actinic Porokeratosis and Cystic Fibrosis

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A 24-year-old woman, presenting with cystic fibrosis, developed superficial actinic porokeratosis. Immunosuppression due to cystic fibrosis may be either the cause of or the exacerbating factor in superficial actinic porokeratosis in our patient.

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Porokeratosis is a distinctive skin eruption characterized by hyperkeratotic plaques with elevated borders and atrophic centres. In addition to the original plaque type described by Mibelli, several other varieties have been reported, including a superficial disseminated form, disseminated superficial actinic porokeratosis (DSAP), punctate porokeratosis of the palms and soles, and a linear type. Although each form represents a distinct clinical entity, they share similar histological changes, i.e. a characteristic column of parakeratotic cells (cornoid lamella).

Recently several authors reported the association of porokeratosis with immunosuppression. To the best of our knowledge, this is the first case report of superficial actinic porokeratosis occurring in a patient with cystic fibrosis.

CASE REPORT

A 24-year-old woman suffering from multiple hyperkeratotic lesions on both legs, was referred to the Department of Dermatology at Hadassah Medical Center. The lesions, with a diameter of 0.5–1.0 cm, had an elevated border and were asymptomatic (Fig. 1). There was no family history of porokeratosis of Mibelli. Cystic fibrosis, which was diagnosed in her infancy, was complicated by liver cirrhosis at the age of 10 years. In 1975 an urgent porto-caval shunt was performed following acute hemorrhage of varicose veins in the esophagus. Since 1981 she has been treated with glibenclamide for diabetes mellitus. During the past 2 years she had twelve episodes of pulmonary infections, most of them

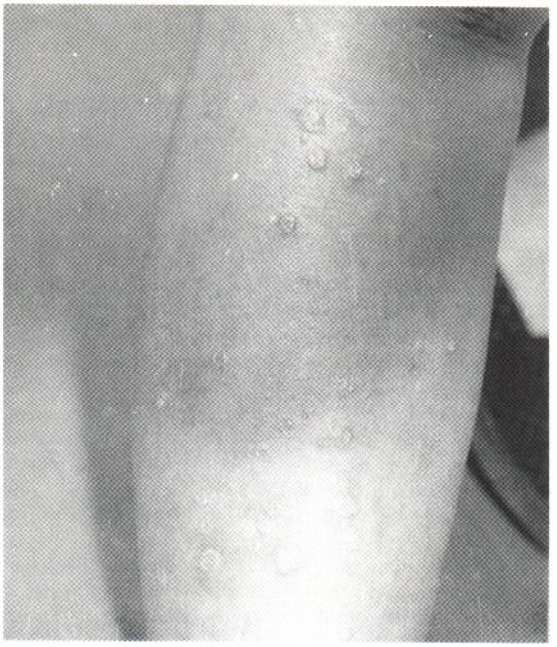

Fig. 1. Skin, showing keratotic papules with atrophic centres and well-demarcated, elevated, hyperkeratotic border.

due to *Pseudomonas aeruginosa*. A skin biopsy taken from a lesion on her legs disclosed the characteristic cornoid lamella (Fig. 2). Laboratory examinations revealed a decrease in the number of T_4 (helper) cells and a slight decline in the T_4 to T_8 ratio, which was 0.5. The T cell response to mitogens, phytohemagglutinin and concanavalin A, as well as lymphocyte response to pokeweed mitogen was slightly lowered.

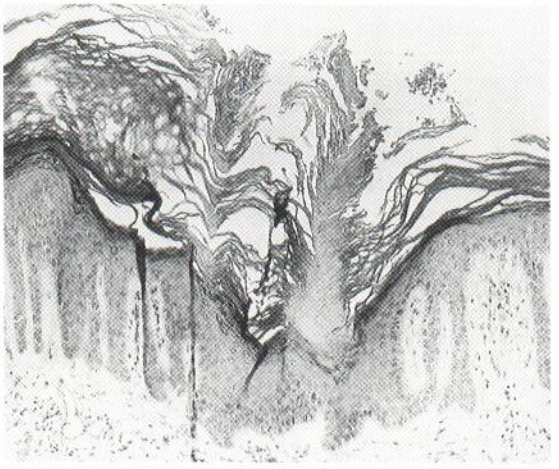

Fig. 2. Histopathology, showing the typical cornoid lamella.

The lesions were treated with liquid nitrogen, though with minimal response.

DISCUSSION

Recently, an association of porokeratosis of Mibelli with disseminated superficial actinic porokeratosis (DSAP) was reported in several immunosuppressed patients. The immunosuppression in these patients was due to renal transplantation (1, 2), malignancy and chemotherapy (3), mycosis fungoides (4), plasma exchange therapy for primary biliary cirrhosis (4), PUVA therapy for psoriasis (5) and immunosuppressive treatment for pemphigus foliaceus (6).

Read & Leone (7) have proposed that porokeratosis is a disease of the epidermis in which a mutant clone of epidermal cells is responsible for the formation of the parakeratotic column.

In our patient, aggravation of the pulmonary state seemed to correlate with the appearance of the superficial acting porokeratosis. It is known that overstimulation of the immune system in cystic fibrosis, due to secondary bacterial infections, may result in immunosuppression (8). This could account for the porokeratosis of Mibelli observed in our patient.

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