

picture: The epidermis showed hyperkeratosis, papillomatosis, acanthosis, elongation and branching of the rete ridges. The epidermis was diffusely infiltrated by cells which were characterized by a hyperchromatic, atypical nucleus. Some of the nuclei showed a crenellated periphery. The chorium was oedematous and the upper layers were moderately infiltrated by an admixture of cells consisting mainly of lymphocytes and histiocytes. The histological picture was consistent with the "Pagetoid" or "Bowenoid" type of Mycosis fungoides (5), (Fig. 3).

Biopsy material taken from non-verrucous lesions showed acanthosis, with elongation and branching of the rete ridges. The epidermis was infiltrated by "mycosis cells" which in some areas formed Pautrier microabscesses (Fig. 4). The upper layers of the chorium were densely infiltrated by an admixture of cells which consisted of lymphocytes, histiocytes and a considerable number of "mycosis cells".

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Disseminated Superficial "Actinic" Porokeratosis

A.-L. Kariniemi, M.-L. Kotovirta
and S. Stubb

*Department of Dermatology, University Central
Hospital, SF-00170 Helsinki 17, Finland*

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Abstract. An 81-year-old Finnish female had a 10-month history of a very pruritic eruption. In the clinical examina-

tion porokeratosis was suspected and histologically verified with the typical cornoid lamellae. The eruption involved also the unexposed areas of the skin. The patient had always avoided sunshine because it made her feel uncomfortable. The patient's sister, too, had a solitary lesion of porokeratosis. The pathomechanism of DSAP is discussed.

Key words: Porokeratosis; Cornoid lamella

Porokeratosis is a genodermatosis with autosomal dominant inheritance. The classic plaque type of the disease was first described by Majocchi in 1887 (6). In 1889 Mibelli termed the disease "porokeratosis", thinking that it would represent a disturbance in the keratinization of eccrine sweat ducts (5). Subsequently, different clinical types of porokeratosis have been described, all showing the typical histopathological finding, a parakeratotic cornoid lamella. Besides the classic localized porokeratosis of Mibelli, disseminated variants of the disease are also known (1, 10). In 1966 Chernosky reported on 12 patients with symmetrical lesions on the sun-exposed areas of the skin (2). This variant showed exacerbation following sun exposure. In 1967 Chernosky called this form disseminated superficial actinic porokeratosis (DSAP) (4). Unlike other types of porokeratosis DSAP usually begins after the third decade of life and the number of the lesions increases with age (3, 7).

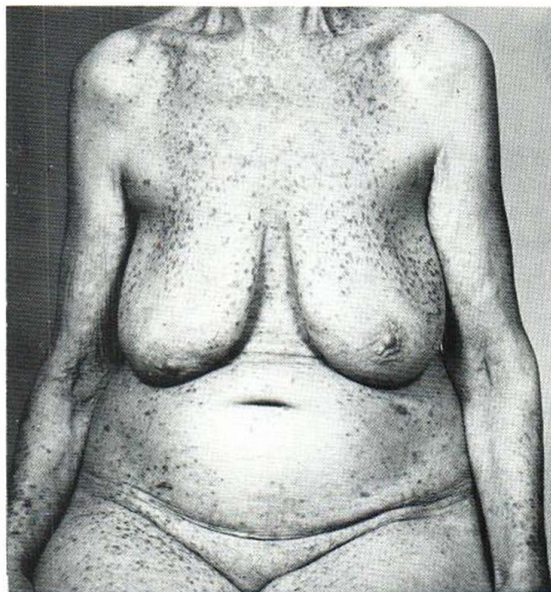


Fig. 1. The distribution of the lesions on the trunk.

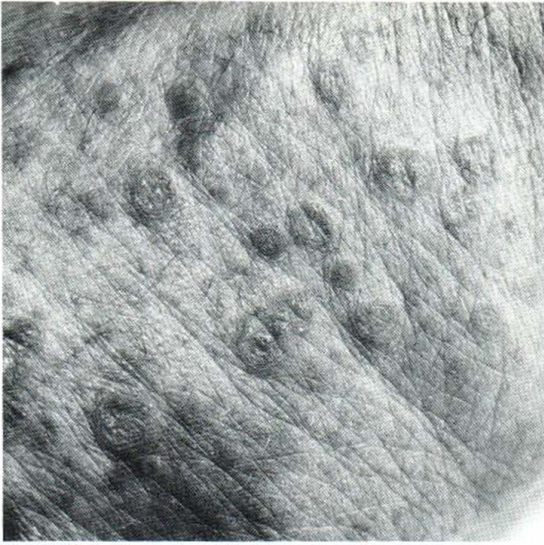


Fig. 2. Close-up of lesions with the thread-like border.

We present here a case of disseminated superficial porokeratosis with unusually late onset and wide distribution—mostly on non-actinic skin areas.

CASE REPORT

The patient is a Finnish woman, born in 1895, who had previously been quite healthy. During the last few years she had been suffering from compensated congestive heart failure, and in 1974 she had got thoracic herpes zoster, from which she had recovered well. In the spring of 1976 she began to suffer from a very pruritic dermatitis, first on her right arm, then also on the trunk, the other arm and the

thighs. In January 1977 she was examined at the Department of Dermatology, Helsinki University Central Hospital. At the clinical examination a symmetrical eruption was seen on the chest, back and thighs and both on the flexor and extensor sides of the arms. The eruption consisted of small red papules and circular brown plaques, 2–10 mm in diameter (Fig. 1). The plaques were surrounded by a thread-like scaly collar (Fig. 2). Most of them were seen on the clothes-covered areas of the skin. The patient had not noticed the deleterious effect of the sun, though she had never been an eager sun-worshipper. Routine laboratory studies and chest X-ray films gave findings within normal limits.

The first biopsy taken from a red papule was not diagnostic, but showed a peculiar intra-epidermal abscess composed of eosinophil granulocytes. The second biopsy taken from a brown keratotic plaque on the upper arm revealed the typical cornoid lamellae at the edges of the plaque (Fig. 3). The cornoid lamella was composed of a parakeratotic column embedded in a groove in the epidermis. The stratum granulosum beneath the cornoid lamella was missing (Fig. 4). The dermis was markedly thinned and there were remarkable solar elastosis, dilated vessels and infiltration of lymphocytes in the upper dermis.

During the 16 months we have followed the patient the eruption has got worse and the itching more severe. We have tried to control the itching locally with steroid and carbamid creams and systemically with antihistamines and sedatives without any notable result. Even PUVA was tried, carefully on one arm, but the treated area became erythematous and more pruritic.

DISCUSSION

In 1970 Reed & Leone suggested that porokeratosis might be due rather to hereditary mutant cell clones in the epidermis than to the disturbance of the eccrine sweat pores (9). In DSAP the abnormal cell

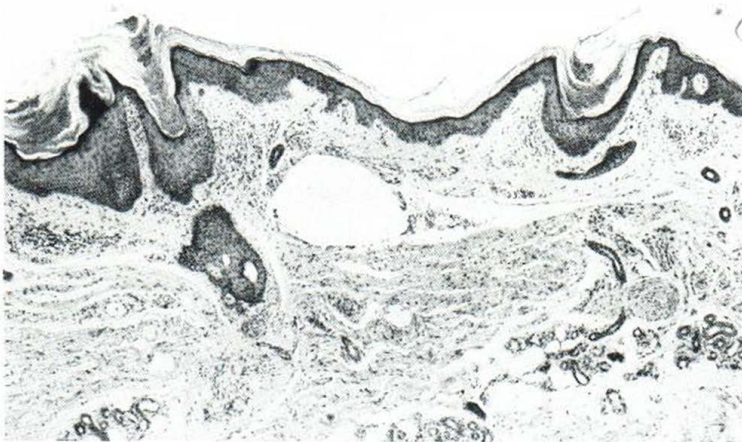


Fig. 3. The cornoid lamella at the edges of a keratotic plaque. Note the thinned dermis, solar elastosis, dilated vessels and lymphocytic infiltration in the upper dermis.



Fig. 4. The cornoid lamella with a parakeratotic column. The stratum granulosum is absent.

clones are more sensitive to ultraviolet light than are normal epidermal cells. The degree of penetration and the number of lesions correlate with the amount of sun exposure (3).

The disseminated superficial actinic form of porokeratosis is very rare in Finland, possibly owing to the small amount of sunshine in our country. In 1971 Niemi described a 38-year-old woman with superficial disseminated porokeratosis on the localized non-actinic skin areas, on the buttocks and the right wrist (7). The effect of sunshine on the eruption is not known.

The clinical and histological findings in our patient are suggestive primarily of DSAP, which is more usual in women and begins in middle-age. The unusually late onset of the disease in this case could be explained in terms of the low yearly dosis of sunshine in Finland.

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Epipodophyllotoxin (VP-16-213) in Mycosis Fungoides: A report from the Scandinavian Mycosis Fungoides Study Group

Lars Molin,³ Kristian Thomsen,¹ Gunnar Volden,²
Annika Bergqvist-Karlsson,⁴ Olle Hallberg⁶
and Lennart Hellbe⁵

Departments of Dermatology, ¹Finsen Institute, Copenhagen, Denmark, ²Rikshospitalet, Oslo, Norway, ³University Hospital, Linköping, ⁴University Hospital, Uppsala, ⁵County Hospital, Örebro and the ⁶Department of Oncology, County Hospital, Örebro, Sweden

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Abstract. Epipodophyllotoxin (VP-16-213) was administered to 9 patients with mycosis fungoides in various stages, most of them in the advanced tumour stage. In 4 of the patients VP-16 was combined with cyclophosphamide. VP-16 alone or in combination with cyclophosphamide was capable of inducing remission initially in all cases, complete in 2, partial in 3 and improvement to a