Case report

POROKERATOSIS HYPERTROPHICA ET DISSEMINATA

J. Miljković, M. Berčič and R. Kavalar

ABSTRACT

We report on a 54-year-old man with an unusual variant of porokeratosis that has been confirmed histologically. Nearly the whole skin surface was involved. On the heels and on the low extremities there were lesions of porokeratosis Mibelli of hypertrophic type with bizarre polycyclic configuration. On the upper extremities and on the trunk the skin lesions resembled an exaggerated variant of disseminated actinic porokeratosis. The lesions on the face were typical of the common type of the latter. Patient's father and elder brother have the same lesions but less pronounced. A systemic etretinate therapy during a period or 5 months was relatively successful. Because of the unusual clinical pattern the possibility of a new variant of porokeratosis is discussed.

KEY WORDS

porokeratosis Mibelli, hypertrophic and disseminated variant, systemic, etretinate therapy

INTRODUCTION

Porokeratosis was first described by Mibelli in 1893 (1). It is an inherited disorder of the skin, characterized by solitary or multiple lesions with hypertrophic border and an atrophic centre, most commonly located on the extremities.

The cause of the disease is unknown. Reed and Leone in 1970 suggested that abnormal clones of keratinocytes are responsible for the development of porokeratotic lesions (2).

Porokeratosis is probably an autosomal dominant condition with variable penetrance, where the predisposition to it is inherited but the predisposition to the type of porokeratosic lesions may be not. What determines the type of morphologic expression of the disease is not yet clarified (2).

A wide variety of clinical manifestations include small ring-like lesions, hypertrophic verrucous lesions, then lesions of the superficial disseminated type, lesions in zosteriform distribution, lesions occurring over the buccal mucosa and finally linear lesions that resemble linear verrucous epidermal nevus (3,4,5,6,). Development of squamous cell carcinoma or of Bowen's disease within features of porokeratosis had been reported in patients with solitary, disse-

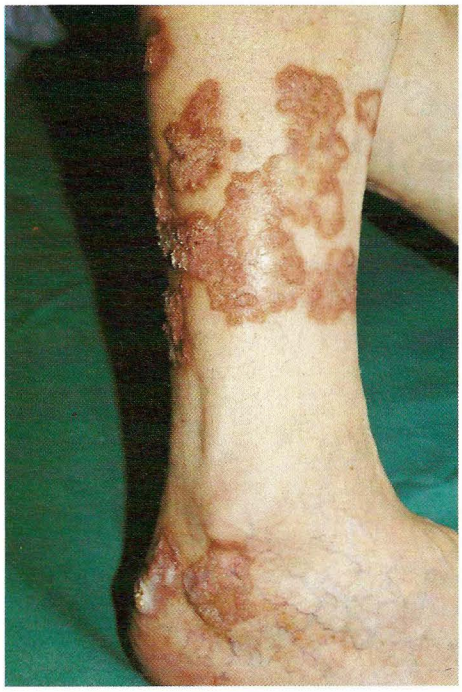


Fig. 1. Porokeratosis Mibelli. Verrucous hypertrophic reddish-brown plaques on the calf.

minated and linear lesions (7,3,9,10).

Cornoid lamellae formation is a consistent histopathologic finding of any type of porokeratosis (11).

CASE REPORT

A 54-year-old man was admitted to our department for evaluation of skin lesions that appeared at the age of 12. The first lesion developed on the left heel representing a verrucous papule that gradually transformed into a thick hyperkeratotic plaque of some centimeters in diameter. In subsequent years new similar lesions constantly developed on the other heel, on the feet, on the extensor sides of the extremities and later on also on the trunk and the face. Some of the lesions were large firm plaques, others were arranged as grouped flat verrucous papules. Changes once manifested never disappeared.

Because of hyperkeratotic plaques on the heels the patient had difficulties walking, but no other subjective symptoms. He therefore never applied any ointments or other therapy.

Patient's father and elder brother have

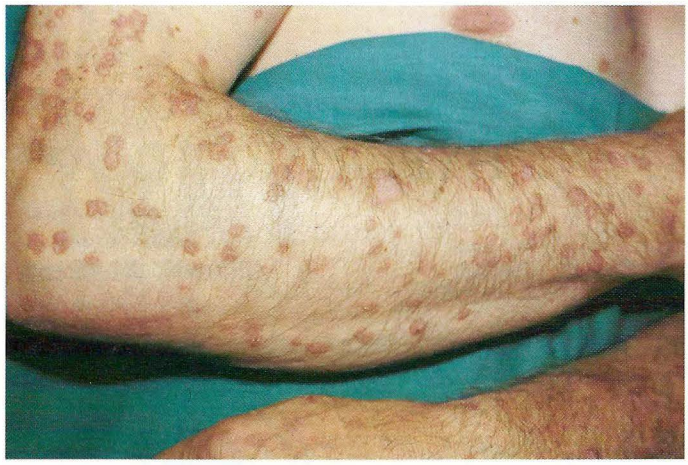


Fig. 2. Atrophic macules with red, slightly elevated keratotic rim, resembling disseminated superficial actinic porokeratosis.

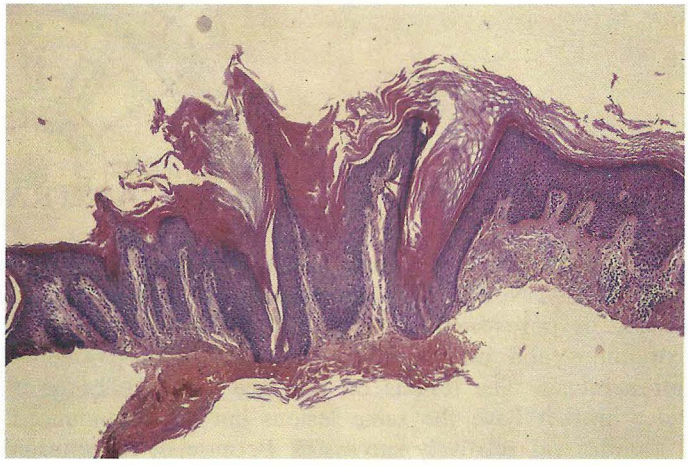


Fig. 3. Acanthotic epidermis with several parakeratotic plugs in epidermal invaginations. HE 10x4

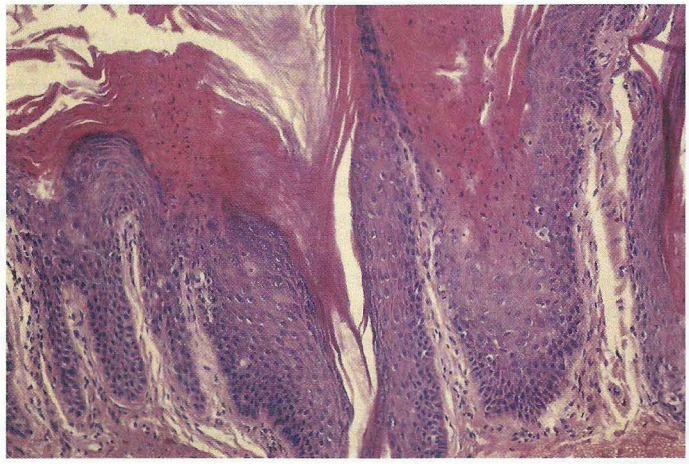
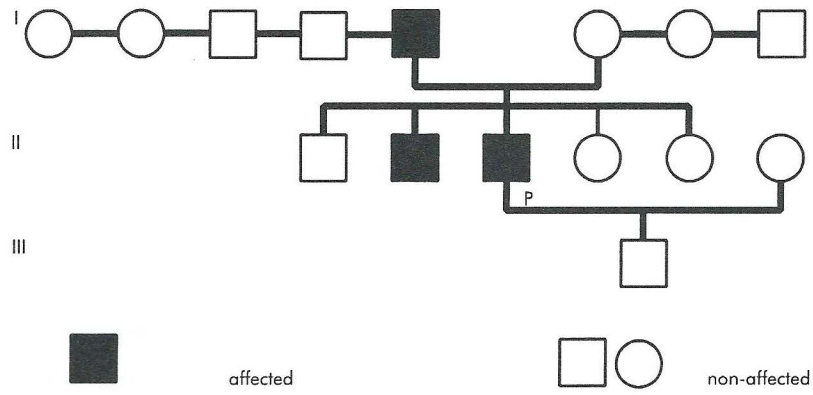


Fig. 4. Higher magnification of features in fig. 3 showing the cornoid lamella. HE 10x16

Table I. Pedigree of the patient with porokeratosis



similar, but less pronounced skin lesions. The 32-year-old patient's son has no skin lesions (Pedigree - Tab. I).

Except for skin lesions the patient had tuberculosis of the left hip-joint at the age of 7. Since then he is limping. Later on he got diabetes.

CLINICAL SYMPTOMS

On admission in June 1994 skin lesions of an extraordinary variegated pattern were found. Nearly the whole skin surface was involved. On both heels well demarcated verrucous lesions with several centimeters thick, crateriform horny masses were present. On the dorsa of the feet and on the extensor sides of the lower extremities there where numerous reddish-brown plaques of different size from 1 to 10 centimeters in diameter and more (Fig. 1) The borders of the plaques were elevated and hyperkeratotic, the centre of the lesions was slightly atrophic. A confluence or some plaques into large areas with polycyclic shape was evident. On both thighs and on the extensor sides on the upper extremities there were also numerous such lesions, but less verrucous. These lesions showed slight atrophic macules with a bright-red, only slightly elevated keratotic rim (Fig. 2).

Similar lesions were also present on the trunk. On the face, in the frontal regions and on the cheeks there were very flat atrophic macules resembling disseminated superficial actinic porokeratosis. Palms and soles were not involved.

Histopathology showed an irregular acanthotic epidermis with enormous orthokeratosis intermingled with parakeratosis. In some deep invaginations of the epidermis there were typical cornoid lamellae. Beneath the cornoid lamellae the stratum granulosum was absent. In the papillary dermis a slight perivascular lymphohistiocytic infiltrate was noted (Fig. 3 and 4). Routine laboratory finding were normal.

A systemic etretinate therapy and keratolytics were administered. The dose of 75 mg etretinate daily was given for one month; then the dose was reduced to 50 mg daily during the next 5 months. The effect of this therapy was relatively good; all lesions flattened.

DISCUSSION

In recent years several clinical manifestations of porokeratosis have been described and some classification schemes have been suggested (12,13,14). The most recent classification was proposed by Kopera et al in 1992. They systemized all described clinical variants of porokeratosis, mainly characterized by distribution of the lesions (15), (Tab. II).

The coexistence of two variants of porokeratosis has been reported in a few cases (16,17). In our case we have observed typical lesions of porokeratosis Mibelli namely hypertrophic verrucous plaques and also lesions of superficial actinic porokeratosis. Our case obviously does not fit into any of the proposed classifications. Concerning the interpretation of this case there are therefore two possibilities. Either the case is presenting a coexistence of two different variants of porokeratosis or it should be considered a new variant of the disease.

In our opinion, the coexistence of different variants of the same disease is more probable. So we support the view, that many areas of the skin can be affected by porokeratosis and that these areas may express different clinical variants (17).

Tab. 2. Types of porokeratosis and their features; cit in (15).

AUTHOR	TYPE	CLINICAL FEATURES	LOCALIZATION
Mibelli 1893		single or few annular lesions with hyperkeratotic border and atrophic centre	limbs, face, genitalia
Freund 1934	linear	linear configurated small papules	unilateral
Rhabari 1977	punctate	pin-point sized follicular keratotic papules	disseminated
Chernosky Freeman 1967	disseminated superficial actinic raised porokeratosis	multiple uniformly sized sun (5 mm) lesions with slightly border in symmetrical distribution	exposed areas, especially extremities
Guss et al. 1971	porokeratosis palmaris plantaris et disseminata	multiple annular lesions with hyperkeratotic border and central atrophy	palmoplantar
Coldner 1971	zosteriform	multiple annular lesions with hyperkeratotic border and central atrophy	corresponding to a nervous territory
McMillan 1976	linear with giant cornoid lamella	small circular plaques, sharply rising edges, central keratinous horn	
Schramm Bork 1982	neviform	(=syn. for zosteriform)	
Strani et al. 1983	zoniform	(=syn. for zosteriform)	

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AUTHORS' ADDRESSES

Jovan Miljković MD, dermatologist, Dpt. of Dermatology,
General Hospital Maribor, Ljubljanska 5, 62000 Maribor, Slovenia
Marija Berčič MD, PhD, dermatologist, same address
Rajko Kavalar MD, pathologist, Dpt. of Pathology, same address