

POROKERATOSIS OF MIBELLI

A case report

A. V. SOLER

M.D., Dip. Derm.(Lond.), M.R.C:P: (U:K:)

Consultant Dermatologist

St. Luke's Hospital

Porokeratosis of Mibelli is characterised by keratotic plaques, sharply delineated from adjacent normal skin by a raised, horny ridge composed of a keratotic column of cells with a characteristic histology — the cornoid lamella. Other porokeratoses include that of Mantoux (1903), confined to palms and soles, and the recent more common type of disseminated superficial actinic porokeratosis of Chernosky (1967).

Case Report

The patient was a 72 year old male with a forty year history of a plaque over dorsum of left hand. There was slow increase in size peripherally with central clearing, but no bleeding or pain was present at any time. The patient was only worried about the cosmetic effect of the hard, scaly rim.

On examination, there was an eczematous-looking, oval shaped lesion, measuring 5 cm. along its greatest diameter, over dorsum of first interdigital space of left hand (fig. 1). The plaque had a characteristic hard, horny, gutter-like border with a shiny atrophic centre. No malignant changes were noted.

Biopsy confirmed the diagnosis of porokeratosis of Mibelli, showing the pathognomonic cornoid lamella. This consists of a tier of parakeratotic marginal material, sharply delineated and sandwiched by the normal hyperkeratosis on one side and the almost non-keratotic, atrophic epidermis centrifugally. The granular layer is absent beneath the lamella, which is cuffed in turn by a lymphoid cell infiltration in the dermis. There is slight dyskeratosis and premature keratinization of the Malpighian



Figure 1.

cells immediately below the parakeratotic column. In the section, the cornoid lamella has partly become detached, leaving a gutter filled up with blood during the biopsy procedure. (fig 2, 3).

Emulsifying ointment bathing followed by keratolytic agents like Whitfield's cream and 10% salicylic acid cream were given a trial, alternating them with diluted fluorinated steroids (bethamethasone 17-valerate). The hard rim persisted and carbon dioxide slush is now being applied. Excision is not considered due to the site and size of the lesion.

Discussion

Mibelli described the condition in 1893, and since then, fewer than 200 case reports have appeared in the literature (Butterworth & Streat, 1962). Mibelli believed the lesions to develop from a distur-

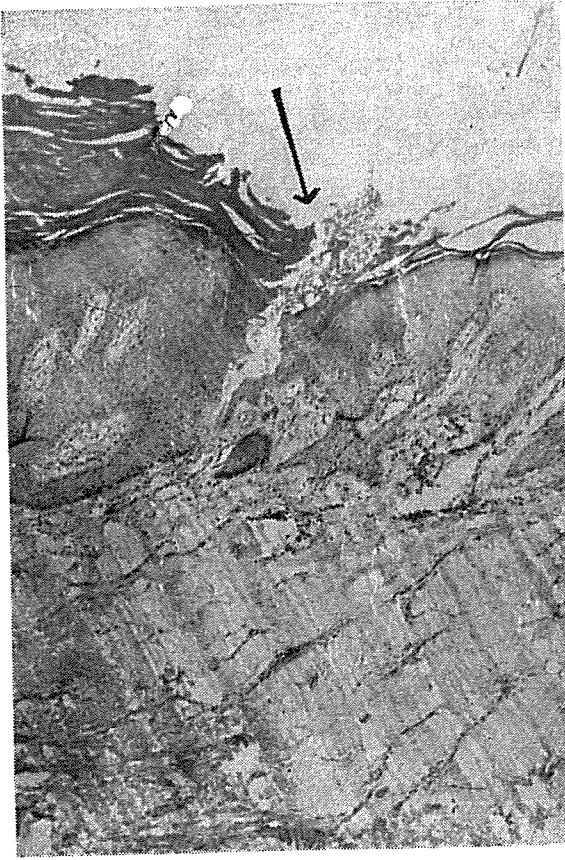


Figure 2.



Figure 3.

bance in keratinization of the acrosyringial portion of the eccrine (sweat) ducts, hence the name prokeratosis, but others (Montgomery & Lever 1961) have found that the cornoid lamella may be related to hair follicles or even independent of any cutaneous appendages. Further, it can occur on oral mucous membranes, glans penis, cornea, and other areas devoid of eccrine ducts, though the sites of predilection are the limbs, especially hands and feet, the neck, shoulders and face including scalp. With lesions of the posterior nailfold, the nails may be thickened, opaque or ridged.

The disorder usually appears in childhood, between 5 — 10 years, though lesions may be present at birth or appear first at puberty or early adult life. It persists indefinitely, remaining unchanged for many years, or extending slowly with long inter-

vals of quiescence. Males are effected two to three times more frequently. It is usually inherited as an autosomal dominant trait, though sporadic cases have been reported.

The initial lesion is a crateriform, horny papule which extends gradually to form a plaque of circinate, geographical or irregular contour, from a few mm. to several cm. in diameter. The peripheral zone of the plaque is raised and horny, and is surrounded by a furrow from which a thin wafer of keratin projects. The central zone is usually atrophic with loss of epidermal appendages; new lesions may however appear within it.

The lesions are asymptomatic except for the unsightly appearance. Frequently only one or a few lesions develop, but sometimes the number is large. A linear

type of porokeratosis resembling linear naevi has been described, usually unilateral and appearing in sites of previous trauma (Prince 1958, Savage 1953). Case reports of a disseminated superficial type with minimal changes have been described during early life and distributed over many areas without any predilection for sun exposed regions (Freund 1934, Andrews 1937), in contradistinction to the superficial actinic variety described by Chernosky. The classical lesions are not related to trauma, infection or systemic disease. Squamous cell carcinoma has developed occasionally in the atrophic skin usually in old age.

Histology reveals the honeycomb edge of the lesion to consist of hyperkeratosis and irregular acanthosis, which in a fortunate section is penetrated by a furrow filled with a column of parakeratotic cells in its centre, the so-called cornoid lamella, corresponding to the sharp rim of the lesion. The stratum granulosum is lacking below the lamella, and a few dyskeratotic and vacuolated cells are present within the Malpighian layer. In the dermis below there is usually a patch of lymphocytes and histiocytes sometimes with a few dilated capillaries. The centre consists of an atrophic Malpighian layer with flattened rete ridges and a chronic cellular infiltrate in the dermis.

Chernosky & Freeman (1967) and Braun-Falco & Balsa (1969), in an exhaustive histochemical study, found the basophilic granules in the cornoid lamella to contain RNA, DNA, and PAS-positive, diastase-resistant material, identical to those of parakeratotic nuclei, and excluded any histochemical similarity with eccrine sweat ducts. Many hold that the label of "parakeratosis centrifuga atrophicans", as suggested by Miescher (1941), is more appropriate than porokeratosis.

Ultra structural studies with the electron microscope (Mann *et al.* 1974) demonstrated the basophilic granular material in the cornoid lamella to consist of whole degenerate cells with pyknotic nuclei and dying cytoplasm. The dermis showed

clumps of fine fibrillar material resulting from definite foci of collagen breakdown and many fibroblasts with degenerative changes. The presence of large numbers of melanin granules in dermal fibroblasts and macrophages indicates a breakdown of basal lamina in some areas and damage to some melanocytes.

There is no effective treatment. Small lesions may be excised, or destroyed by deep electrodesiccation or cryosurgery.

Acknowledgements

My thanks are due to Prof. G. Xuereb of the Department of Pathology for providing the histology material.

References

- ANDREWS, G.C. (1937) *Arch. Derm. Syph.* 36, 1111.
- BRAUN-FALCO, O. & BALSA, R.E. (1969) *Hautarzt*, 20, 543.
- BUTTERWORTH, T., & STREAN, L.P. (1962) *Clinical Genodermatology*, Baltimore (Williams & Wilkins Co.) 26, 28.
- CHERNOSKY, M.E., & FREEMAN, R.G. (1967) *Arch. Derm.* 96, 611 — 624.
- FREUND, E. (1934) *Arch. Derm. Syph.* 170, 2.
- MANN, P.R., CORT, D.F., FAIRBURN, E.A., & ABDEL-AZIZ, A. (1974) *British Journal of Dermatology* 90, 607.
- MANTOUX, C. (1903) *Annals Derm. Syph.* 4, 15.
- MIBELLI, V. (1893) *Contributo allo studio delle ipercheratosi dei canali sudoriferi (porokeratosis)* *G. Ital. Mal. Veneree e Pelle* 28, 313 — 355.
- MIESCHER, G. (1941) *Year Book of Dermatology & Syphilology*, 198.
- MONTGOMERY, H., & LEVER, W.F. (1961) *Arch. Derm.* 84, 980 — 988.
- PRICE, H., (1958) *Arch. Derm.* 77, 477.
- ROOK, A., WILKINSON, D.S., EBLING, F.J.G., (1972) *Textbook of Dermatology*, Vol. 2, 1188.
- SAVAGE, J., (1953) *Proceedings of the Tenth International Congress of Dermatology*, London.