He was treated with dapsone 200 mg daily and the skin cleared.

A blood sample from each twin was examined at the Institute of Forensic Medicine in Copenhagen, and the HLA type for both was HLA-$A_1$9; B8; Dw3, which confirmed that they were monozygotic.

COMMENT

The monozygotic twins described here with GSE and DH, respectively, were HLA-B8 and Dw3, in agreement with previous studies. There is an increased frequency of B8 in GSE (80–90%) and in DH (80%) as compared with only 20–30% in normal controls (4). Recently, HLA-Dw3 has been shown to occur in GSE and DH in a higher frequency than HLA-B8 (4).

Twin cases tabulated to date demonstrate 67% concordance in ostensibly monozygotic twins and 33% in dizygotic twins (Table 1). This demonstrates that the theory that GSE is due to a single gene is untenable.

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Some Unusual Findings in Porokeratosis Mibelli

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Abstract. Light microscopy in a case of unilateral porokeratosis Mibelli revealed a cyst-like dilatation in the upper corium containing two cornoid lamella complexes. Among other findings, elastic globes as part of thickening and fragmentation of elastica, which in this case may be related to microangiopathia deserve to be mentioned.

The cornoid lamella in porokeratosis Mibelli has always been found to be located in funnel-like epidermal invaginations both in connection with epidermal adnexa and in regions free of adnexa. The parakeratotic nature of cornoid-lamella cells has been suggested on the basis of histochemical and ultramicroscopical findings (2, 6). This is consistent with a deficient granular layer, perinuclear vacuolation and nuclear pyknosis in the epidermis below the cornoid lamella. The occurrence of the cornoid lamella in places where no relations to adnexa could be found, was the reason for the term of "parakeratosis centrifuga atrophicans" suggested by Miescher (7) and basically accepted by Braun-Falco & Balsa (2).

A focal disturbance afflicting a clone of epidermal cells resulting in aberrant keratinization was an alternative suggestion to the theory of adnexal origin (4, 8).

The reason for the present publication is the unique detection of two cornoid lamella complexes in a cystic dilatation of an intradermally located hair follicle.

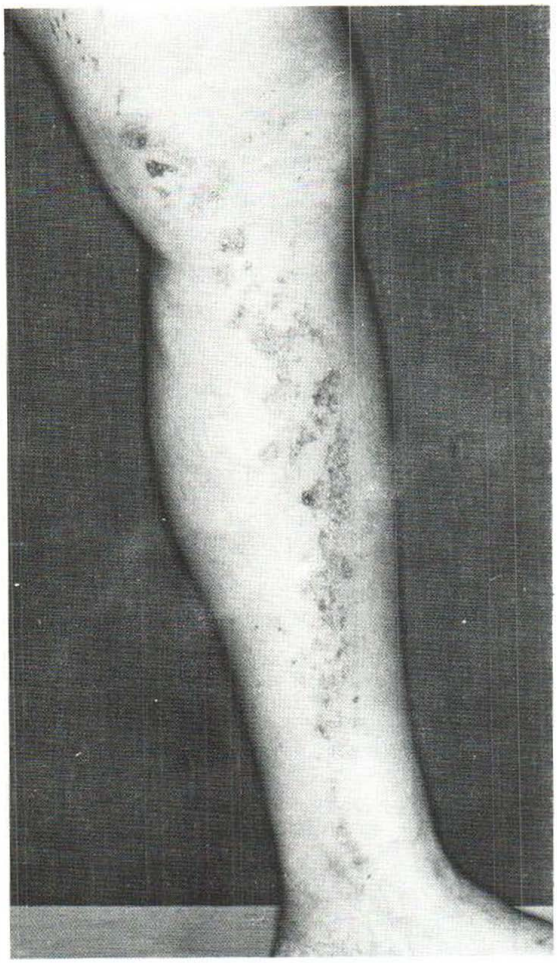


Fig. 1. Unilateral striated lesion of porokeratosis Mibelli.

CASE REPORT

A woman aged 38 years developed at the age of 10 years an eruption of non-irritant, asymptomatic, round, somewhat depressed lesions bordered by a tiny keratotic ridge. At first the eruption was on the inner side of the left knee. Over the years, new lesions arose coalescing to form striated plaques involving two-thirds of the inside of the left leg (Fig. 1) and isolated lesions 3–5 cm in size on the dorsum of the left hip, on the buttock and left arm. No lesions appeared on the right side of the body.

Biopsies were taken for light and immunofluorescence microscopy. Staining for light microscopy: Hematoxylin-eosin, van Gieson, PAS, PAS-diastase, Weigert-elastica, Toluidine blue and Congo red. The biopsies included the peripheral ridge.

HISTOPATHOLOGY

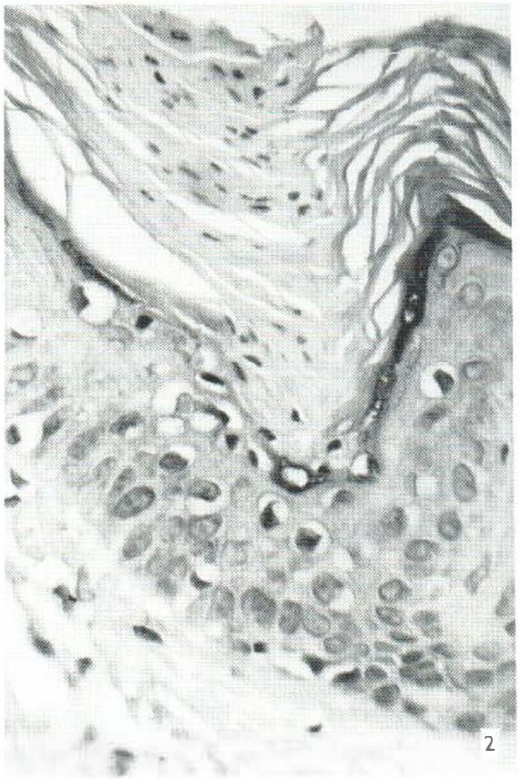
The major part of the excision is represented by an atrophic epidermis lacking rete ridges, lying on the inside of the cornoid lamella. Beneath the columns of parakeratotic cells in the cornoid lamella the stratum granulosum was almost totally absent and replaced partly by necrobiotic cells with effaced cell contours and vacuolated cells with excentric nuclei (Fig. 2). The most noteworthy change in the atrophic part of the epidermis is a varying degree of cell polymorphy as to size and tinctorial qualities, connected with loss of stratification somewhat resembling a precancerous epidermal condition. The basement membrane is thin, in some places interrupted and totally absent on the part corresponding to advanced necrobiotic changes in the epidermis in an area farthest from the cornoid lamella.

A hitherto unpublished finding was a cyst-like dilated hair follicle in the upper part of the corium containing two cornoid lamellae originating in the cyst wall, encircling an orthokeratotic core (Fig. 3). The cyst wall beneath the orthokeratotic mass contains 2–4 layers of granular cells. In contrast, their absence over a zone of vacuolated cells is directly involved in the formation of the cornoid lamella. The cyst was in all sections located in the vicinity of the epidermal cornoid lamella, suggesting an original connection between them, but no direct transition was found.

An atrophy of surrounding connective tissue may be the cause of a slight displacement of the cyst. In the deeper part of the corium in the same zone an empty follicular cyst was found in direct connection with a suggested atrophic sebaceous gland (Fig. 4).

The bulbus of this hair follicle could not be traced in further sections. The blood vessels exhibited marked regressive changes at all levels. Fig. 5 shows a thickening and splitting of their wall and in some places a markedly reduced lumen with only a few ghosts of endothelial cells at the lumen. The rest of them can be traced to be extruded into the vessel wall. The picture of microangiopathy is

Fig. 2. Columnar parakeratosis, lacking stratum granulosum, and degenerated and vacuolated granulocytes in the corresponding part of epidermis.
Fig. 3. Follicular, cyst-like dilatation with two cornoid lamellae (arrows) encircling an orthokeratotic core. The epidermal part of a cornoid lamella is to the right of the cyst-like dilatation.
Fig. 4. Empty follicular cyst-like dilatation with an adjacent atrophic sebaceous gland.
Fig. 5. Thickened and split vessel walls with defective endothelial and perithelial cells. Ghosts of endothelial cells within the vessel walls are shown.



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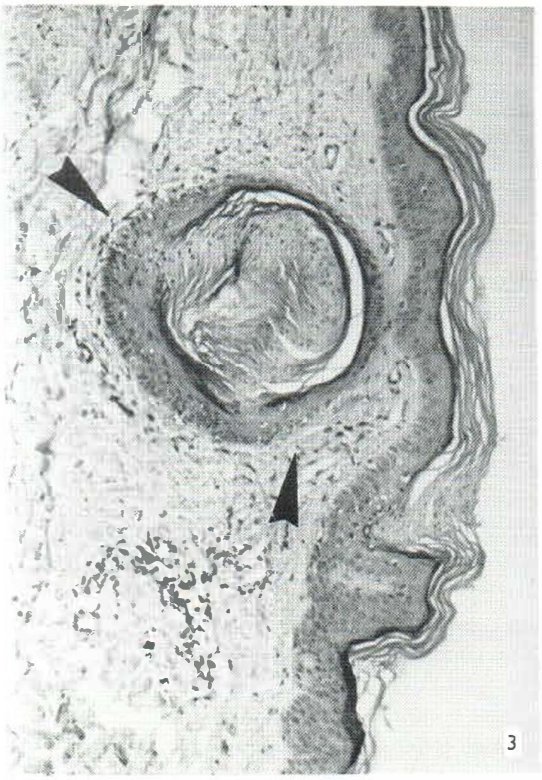

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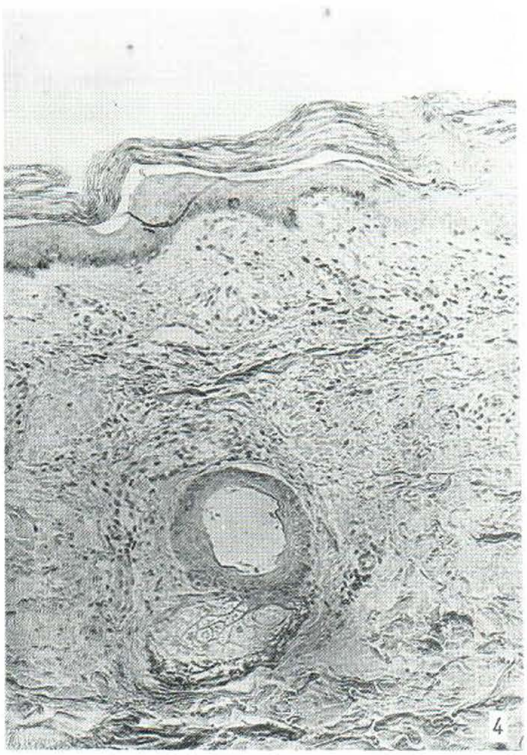

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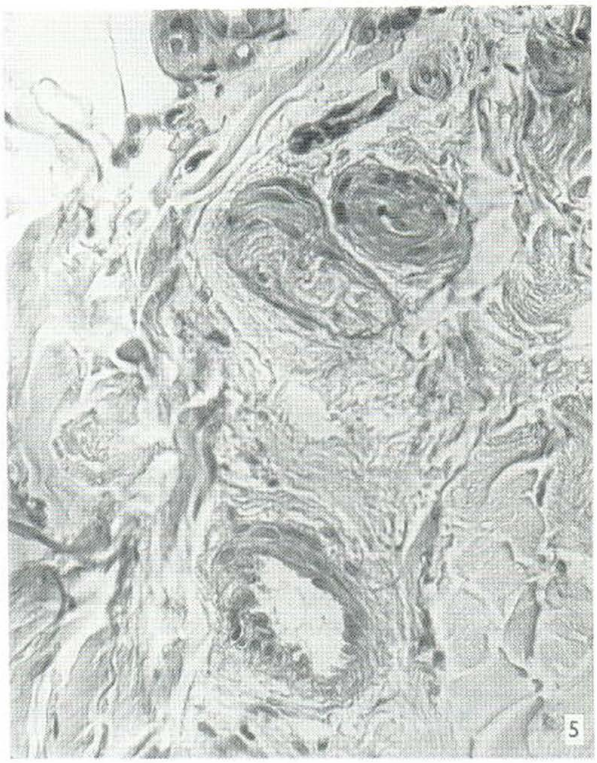

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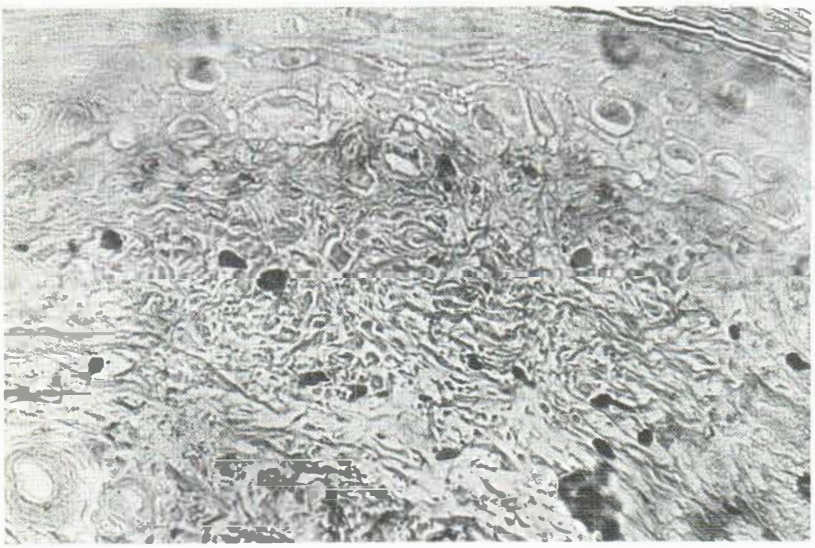

Fig. 6. Elastic globes, mainly in the upper part of the corium.

completed by a marked PAS positivity in the vessel wall and a comprehensive loss of pericytes.

Atrophic and degenerative changes in the sweat glands were seen in form of dilated lumina, effacement of cell contours, and vanishing nuclei. A fragmentation and thickening of elastica and many elastic globes were found mainly in the upper part of the corium (Fig. 6). A slight inflammatory infiltration was limited to the vicinity of the cornoid lamella and atrophic adnexa.

DISCUSSION

In recent years a suggestion has been put forward that the cornoid lamella can originate both in orifices of adnexa and in adnex-free epidermal areas (2, 4, 6). A double cornoid lamella in the infundibulum has previously been shown (1) which, together with our finding of two cornoid lamellae in a follicular cyst located in the vicinity of the epidermal part of the cornoid lamella, emphasizes a follicular predilection. Columnar parakeratosis was shown even in Darier's disease (1). This fact, in connection with findings of an admixture of dyskeratotic cells on the base of the cornoid lamella, suggests a relationship on the basis of a nevoid genesis. A degenerative and atrophic process on epidermal adnexa, repeatedly emphasized, even leading to their total destruction, may in many cases explain why no connections between the epidermal indentation containing the cornoid lamella and the deeper parts of adnexa could be found.

As a minor curiosity, we could also demonstrate elastic globes as part of the thickening and fragmentation of elastica. Degenerative changes on connective tissue have been found to be linked to microangiopathia in diabetics (5). It should be noted, however, that microangiopathic changes have been found even in 10.75 % non-diabetics (3), which may open up an interesting way to consider the pathogenesis of some hitherto unelucidated or problematic dermatologic phenomena.

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Perioral Dermatitis with Epithelioid Cell Granulomas in a Woman: A Possible New Etiology

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Abstract. Perioral dermatitis, a distinctive clinical entity yet of obscure etiology, is not uncommonly seen among Saudi middle-aged females. A case of a 30-year-old woman with a perioral lupoid micropapular erythematous eruption is described. Histopathological examination showed evidence of granulomatous sarcoidal reaction similar to that described by Gianotti et al. (3), Gianotti (2), and Georgouras & Koesard (1). Synthetic acrylic fibre meshes widely used by Arabian women to cover their faces, are considered in the etiology of this eruption.

Key words: Gianotti facial dermatitis; Granulomatous sarcoidal reaction; Acrylic fibres

CASE REPORT

A woman, aged 30 years, was examined in January 1978 because of an itchy facial eruption of 3 months' duration and resistant to topical treatment. The eruption had commenced around the mouth and covered the lower half of the face within 2 weeks. The forehead and ears were not involved. The patient stated that this eruption had appeared after the use of new black synthetic meshes to cover the lower half of her face. She did not use any cosmetic preparation.

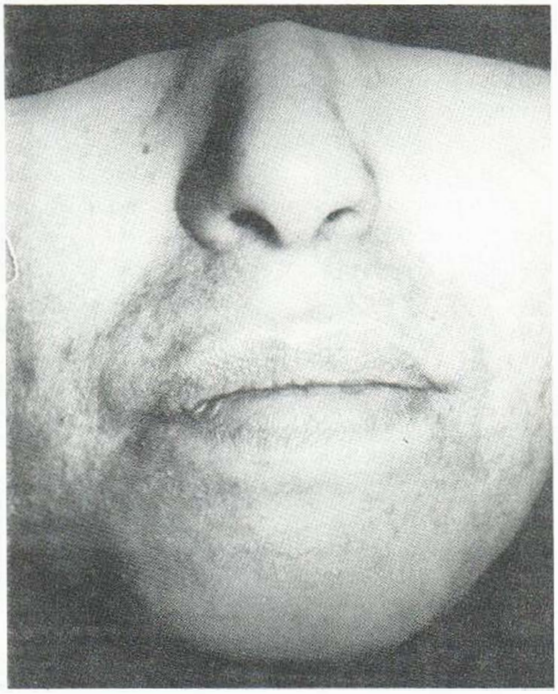

Fig. 1. Perioral dermatitis.

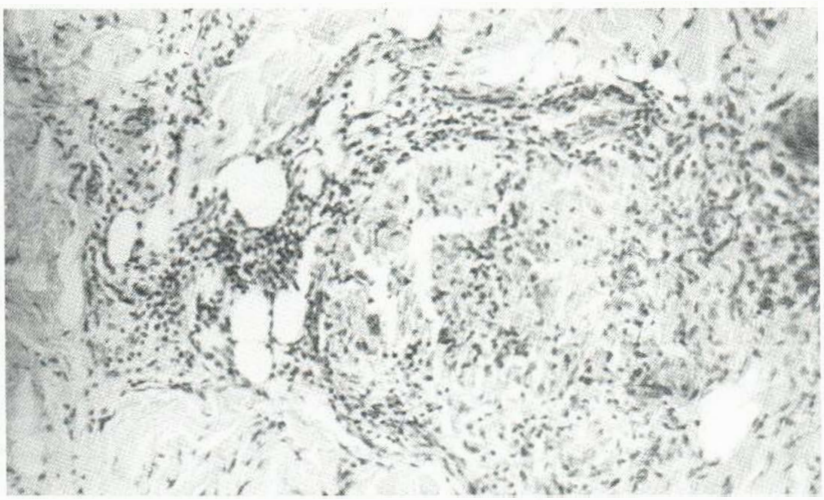

Fig. 2. Epithelioid cell granuloma.