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REACTIVE PERFORATING COLLAGENOSIS (MEHREGAN)

Report of Two New Cases and Review of Literature

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Abstract. We report two cases of reactive perforating collagenosis (RPC) observed in Denmark. This skin disorder becomes manifest in childhood and is characterized by the occurrence of numerous pinhead-size keratotic papules over areas subject to trauma. The papular lesions increase in size to approximately 5 to 10 mm in diameter and become centrally umbilicated. This is followed by a period of regression in which the umbilicated lesions flatten out and eventually disappear completely. The life span of a lesion is between 6 to 8 weeks. Histologic examination of an early lesion reveals necrobiosis and bluish staining of the papillary connective tissue followed by a sequence of epithelial reaction and transepithelial elimination of the necrobiotic connective tissue.

Reactive perforating collagenosis (r.p.c.) was originally described by Mehregan and co-workers in 1967. A case was presented exhibiting an unusual reaction of the skin to superficial trauma. In response to minor trauma, certain changes took place within the papillary layer of the corium characterized by affinity of the connective tissue for hematoxylin. Secondary changes of the overlying epidermis eventually led to complete excoriation of the altered connective tissue. In the meantime 2 additional reports of cases (1, 8) have been published from USA.

CASE REPORTS

Case 1

A 3½-year-old white boy was first seen in July of 1971 in the outpatient clinic of the Finsen Institute for a skin eruption involving his face and extremities.

The child was born of a normal delivery. His mother suffered from a mild ichthyosis and his father had bronchial asthma. He was previously examined at an orthopedic clinic and was found to have bilateral coxa vara, pedes plani, and genua vara. The skin eruption was noticed before the age of 1 month. It consisted of discrete papules and pustular lesions first appearing on the trunk and later confined to the face and extremities. The eruption persisted for several years, with new lesions developing in crops while the older lesions regressed completely. His mother noted that the skin lesions developed particularly when the child had been playing in areas with coniferous trees. In July 1972, the child's skin eruption was limited to the dorsa of hands, face and the extensor surface of the lower extremities. Some papular lesions were arranged in a linear fashion suggesting Koebner phenomenon (Fig. 1). The earliest lesion was a pinhead-

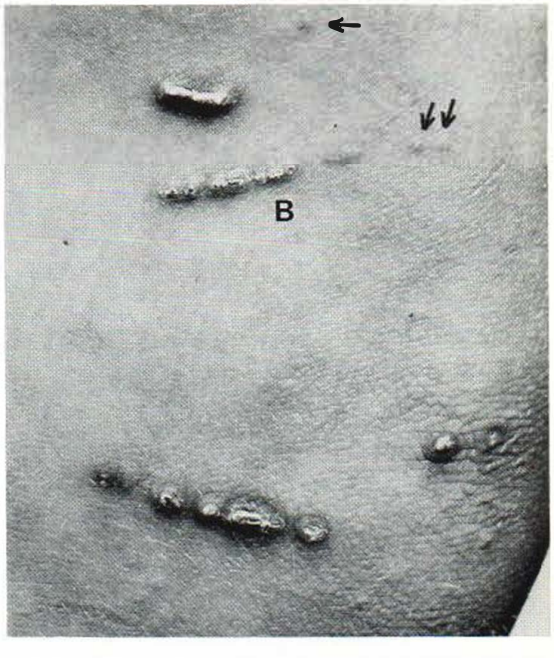

Fig. 1. Several lesions located on the medial side of the left knee. There is pronounced Koebner phenomenon. The earliest lesion is a pinhead-sized skin-colored papule (*arrows*). Two confluent lesions are becoming umbilicated (*B*). The other papules are well developed, umbilicated lesions.

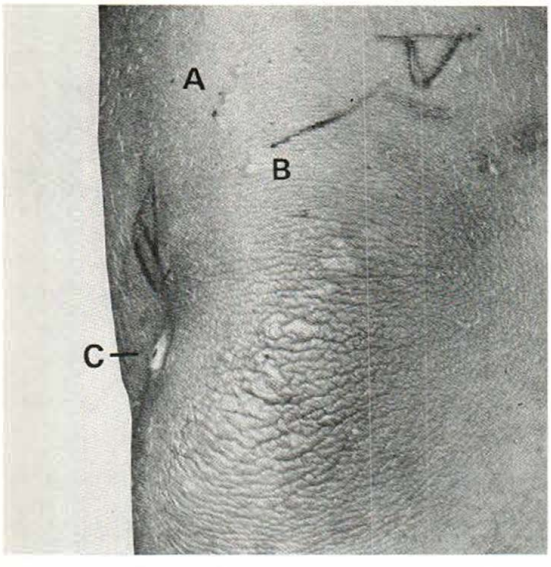

Fig. 2. Left elbow. Three early, non-umbilicated papules in linear formation (*A*). Below these is seen a lesion which is beginning to become umbilicated (*B*). (*C*) A well umbilicated papule in the stage of regression.

sized skin-coloured papule. Older lesions were 5 to 10 mm in diameter and showed a central area of umbilication containing brownish keratotic material. The central plug was adherent and could not be removed without causing bleeding. A fully developed umbilicated lesion usually reached its maximum diameter after a 3 to 4 week period. This period was then followed by a stage of regression in which the individual lesions flattened out and eventually completely disappeared. A complete cycle of the eruption occurred over 6 to 8 weeks.

Various laboratory studies such as erythrocyte sedimentation rate, haemoglobin, immunoglobulins (IgG, IgM and IgA), protoporphyrin in erythrocytes, ANF and AGKT gave results which were within the normal limits.

Case 2

A 7-year-old white boy was seen first at the outpatient clinic of the Finsen Institute in April 1967 for evaluation of a skin condition of 2 years' duration. A physical examination revealed numerous discrete 2–8 mm diameter erythematous papules, some covered with an adherent crust over the dorsa of the hands and on the face. A clinical diagnosis of molluscum contagiosum was made and the eruption was treated by application of 5% lapis solution. The patient returned 1 month later with a larger number of lesions, some showing a central area of umbilication and keratotic plugging. A biopsy specimen taken from a lesion on the dorsum of the hand was reported as epidermal ulceration and chronic inflammation.

The patient was seen again 5 years later in August 1972. The eruption had persisted since the first visit. There were, however, periods of remission, usually from late autumn until early in the spring. Physical examination revealed extensive involvement of the dorsa of the hands and feet, extensor surface of the arms, lower legs and face by numerous discrete papules and umbilicated lesions (Fig. 2). There were also a few whitish superficial scars.

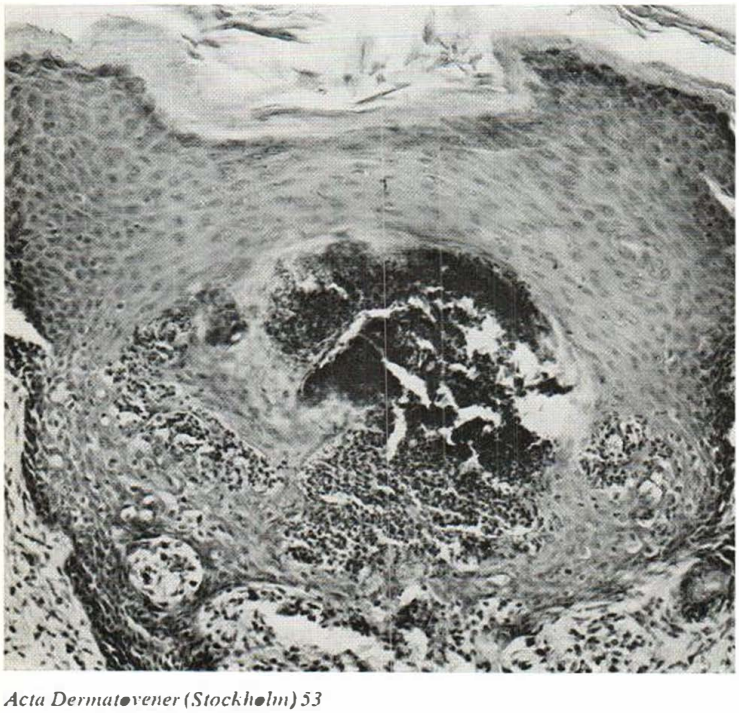

Fig. 3. Early non-umbilicated papule with connective tissue showing affinity for hematoxylin on top of a dermal papilla. (Hematoxylin and eosin, × 50.)

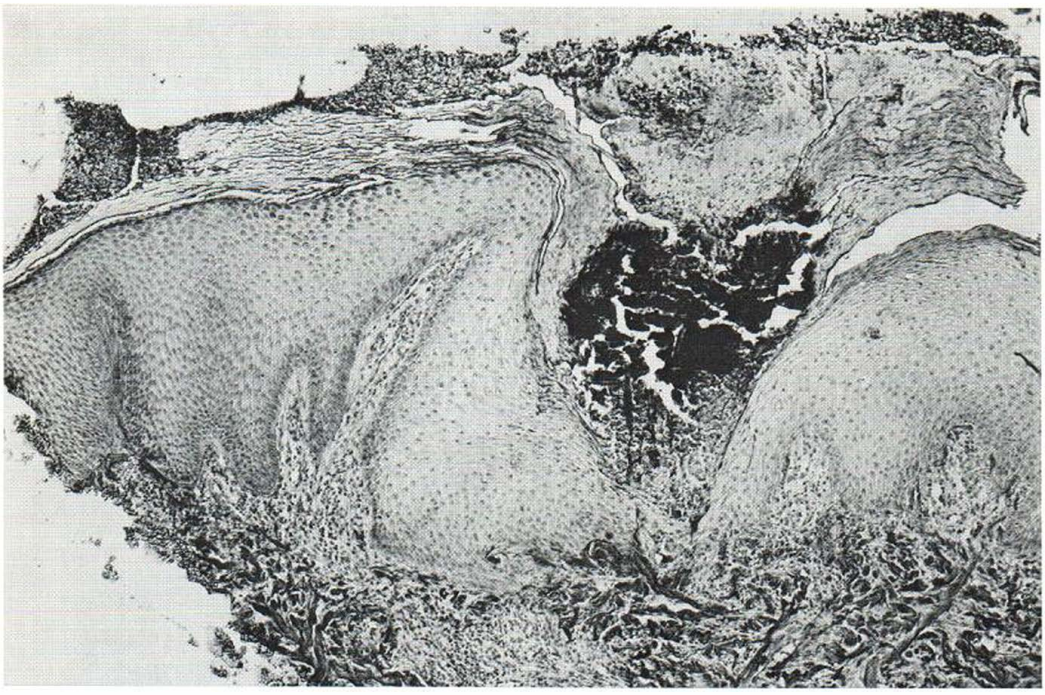

Fig. 4. Central parakeratotic material inside the area of epidermal depression of an umbilicated lesion. The central plug consists of parakeratotic keratin, masses of necrobiotic connective tissue containing numerous degenerating nuclei of inflammatory cells and thin, extruded collagen bundles. (Van Gieson stain, × 65.)

HISTOPATHOLOGY

A total of ten specimens were taken from the 2 patients for histologic examination. The paraffin-fixed tissue sections were stained by hematoxylin-eosin, acid orcein-Giemsa, Van Gieson and aldehyde fuchsin stains and also by a combination of alcian blue with PAS reaction. A section from an early non-umbilicated lesion from case 1 (Fig. 3) shows the epidermis to be acanthotic. In the centre of the lesion is a widened dermal papilla containing connective tissue, which stains a bluish colour with hematoxylin and eosin. However, most lesions biopsied were from the umbilicated stage and from the stage of regression. These lesions varied in size between 4 and 8 mm in diameter and showed a central area of umbilication plugged with a solid mass of brownish material. The histologic sections showed the central plug (Fig. 4) to consist of a mixture of parakeratotic material and thin layers of inflammatory cells. The underlying epidermis was irregularly atrophic and showed multiple foci of epidermal disruption. Through these areas was extruded necrobiotic connective tissue including thin collagen bundles. The collagen bundles were easily recognized within the central parakeratotic plug by their vertical direction (Fig. 5), especially in the sections stained by Van Gieson technique. The sections stained by acid orcein-Giemsa and by aldehyde fuchsin methods showed no evidence of dermal elastosis or elimination of the elastic fibres in the central keratotic plug.

COMMENTS AND REPORT ON THE LITERATURE

Reactive perforating collagenosis was first described in 1967 (2) as a definitive form of cutaneous reaction to superficial trauma. Uniformity in clinical manifestation and histologic findings in several additional cases was observed and those reported in the dermatologic literature have confirmed this hypothesis and have established this condition as a distinct clinico-pathologic entity (1, 3, 8). Relationship of the eruption to superficial trauma suggested by clinical data and by linear development of the eruption along the scratch marks (Koebner phenomenon) has been confirmed experimentally. Occurrence of this skin

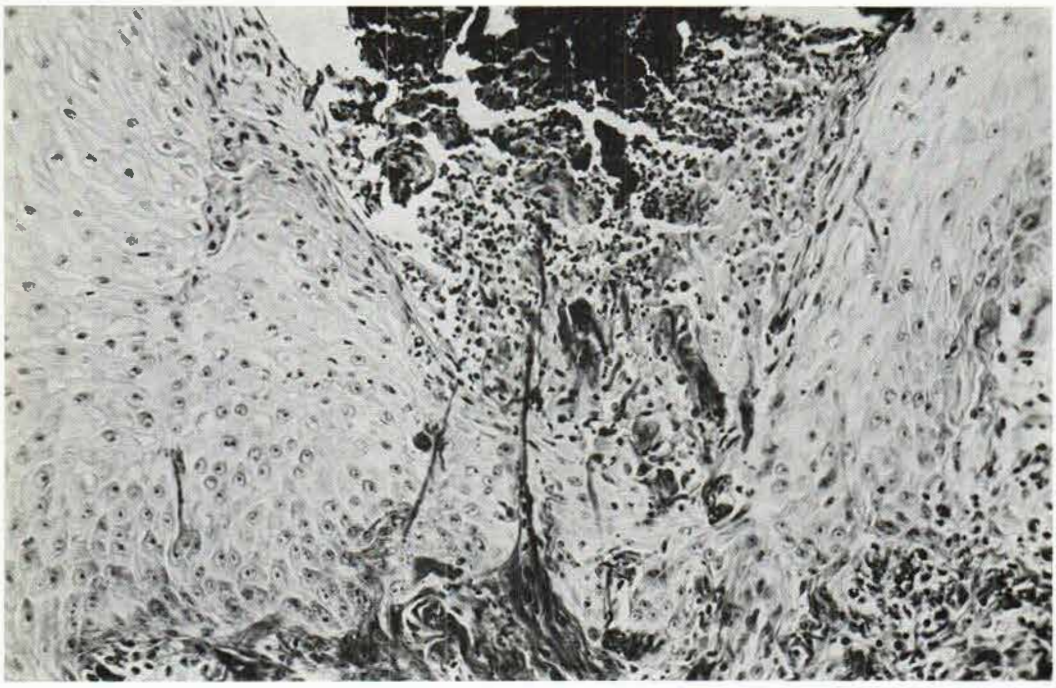

Fig. 5. Base of a central plug in an early stage of regression and repair. The epidermis at the floor of the crater is under regeneration but is still being perforated like a strainer by extruded collagen bundles. They are moving upwards in a direction perpendicular to the surface of the epidermis. (Van Gieson, × 195.)

disorder in several pairs of siblings suggests a hereditary type of connective tissue abnormality (3, 8). From the histologic examination of multiple lesions in various stages of development, the life cycle of the eruption has been reconstructed as follows: In response to superficial trauma to the skin of patients with RPC, multiple foci of necrobiosis occur, with bluish staining of the collagen bundles lying within the papillary layer of the corium. The suprapapillary epidermis becomes thin and shows multiple foci of disruption through which necrobiotic collagenous tissue and some inflammatory cells enter into the keratin layer. As more dermal material is eliminated a mixture of parakeratotic crust and collagen bundles is formed at the surface which now sinks into an area of epidermal depression. The process of transepithelial elimination continues until the supply of necrobiotic connective tissue is completely exhausted. This period then is replaced by a stage of repair in which epithelial regeneration closes all the areas of epidermal disruption. The keratotic plug at the surface of the lesion is flattened out and the lesion eventually disappears with a minimal or no residual scarring.

The process of transepithelial elimination explains the complexity of histologic changes observed in RPC and in elastosis perforans serpiginosa (4). This process is also observed in other dermatologic disorders such as in perforating folliculitis (5), nodular calcinosis cutis, perforating granuloma annulare (7) and in the so-called black heel (6).

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