

KYRLE'S DISEASE

Hyperkeratosis Follicularis et Parafollicularis in Cutem Penetrans

Reino Pajarre and Martti Alavaikko

From the Departments of Dermatology and Pathology, University of Oulu, Finland

Abstract. Kyrle's disease is a rare skin disorder, with about 20 well-documented cases reported. In the present case, the first to be reported in Finland, the clinical and histological findings are described and the differential diagnosis and the morphogenesis of the perforation are discussed.

In their review in 1968 Constantine & Carter (1) discussed previously published cases, 45 in all, from which they accepted 12 certain cases, in addition to five of their own. They drew the conclusion that the disorder begins about the age of 30. A clinically typical feature was symmetry. The lower extremities were affected most often. The formation of plaques correlated with the general density of lesions. Generally speaking, lesions caused little trouble and the early stages were seldom encountered. No "cured" case is on record. Neither has any effective therapy been discovered.

CASE REPORT

The patient is a former forestry worker, aged 73, with no history of skin eruption. During the last few years he has suffered from coronary disease with heart failure and poor circulation in the lower extremities. About a year before being admitted to hospital the patient developed a reddish itching skin eruption, beginning on the anal region and becoming widespread in 4 months. Concomitant with the appearance of small red papules, the diffuse reddening of the skin began to decrease. The extremely itchy small papules gradually developed into bean-sized hard papules, and formed several verrucous plaques when confluent. The lesions grew to maturity at about 6 months of age, after which they lost their plugs, gradually sank, and formed a scar at the skin level.

Clinical findings

The skin was generally pale, with livedo reticular appearance in some places and acrocyanosis in the feet.

On the extensor surfaces of the upper extremities and on the skin of thighs and legs there was a symmetrical distribution of umbilicated papules of pin-head size. The umbilicus consisted of a hard plug. In some papules, the plug was seen only after the removal of covering scales. On the lower extremities there were verrucous plaques formed by confluent large papules, measuring up to 3 cm in diameter (Fig. 1).

Histological findings

The histological examination was performed on five punch-biopsy specimens. The specimens were fixed in formaldehyde solution, embedded in paraffin and cut in serial sections. In addition to hematoxylin-cosin staining, acid-orcein-Giemsä staining and Verhoeff's staining were performed to visualize the elastic tissue.

In all five biopsies, characteristic keratin plugs were seen, situated in sharply delimited epidermal invaginations (Fig. 2). Each plug consisted of keratinous lamellae which filled the invagination, and parakeratosis was often seen at its bottom. Fine granulated basophilic material was seen between the lamellae. Hair follicles could not be demonstrated in most of these invaginations. In the epidermis under the keratin plug, str. Malpighi grew thinner and keratinizing spinal cells were seen instead. Cell vacuolization, nuclear pyknosis and dyskeratosis of separate cells was seen. At the bottom of a few invaginations, the cellular tip was keratinized. Around this keratinous mass was seen a dense lymphohistiocytic cell infiltration, having sharply-defined borders with the surrounding dermis. A rich proliferation of capillaries was noticed, too. Acanthotic areas with a broad str. Malpighi were seen in the epidermis bordering the epidermal invaginations. A clear perforation was seen in only one biopsy specimen.

COMMENT

In the histological differential diagnosis of Kyrle's disease (4) elastosis perforans serpigiosa and reactive perforating collagenosis (2) must be considered. In the former the basic change is the marked increase of the elastic tissue which

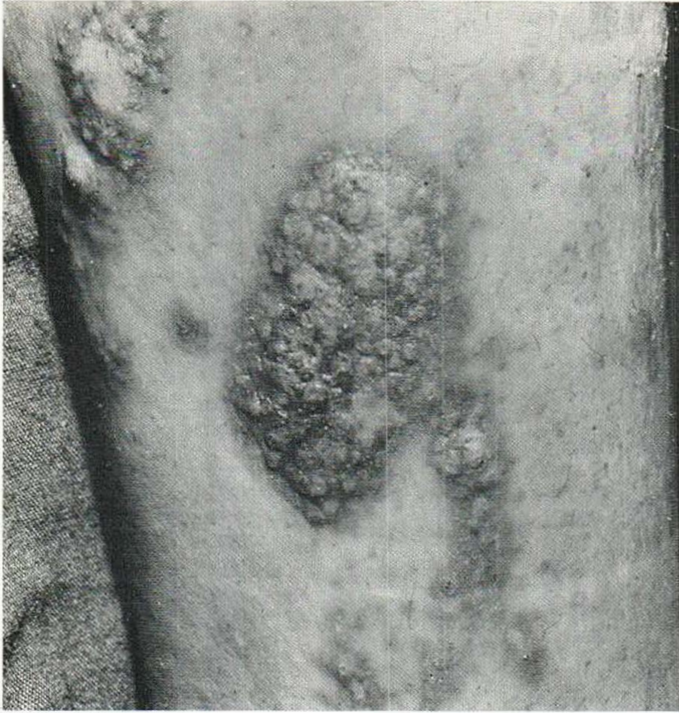


Fig. 1. Skin lesions on the lateral surface of the lower shin.

is such a typical feature that the name *elastosis perforans serpiginosa*, proposed by Dammert & Putkonen (3), is nowadays generally accepted. In our case no increase in elastic tissue was observed. Neither could we discover anything to give us cause to consider the degeneration of collagen to be a primary change. Mehregan & Coskey (5) paid attention to the fact that perforating folliculitis both clinically and histologically may resemble Kyrle's disease. In the former the keratin plug is always situated in the hair follicle. In our case most lesions were seen parafollicularly.

The penetration is emphasized in Kyrle's original study as well as in many other reported cases of Kyrle's disease. Constantine & Carter (2) considered the stressing of this feature somewhat misleading, because the term "penetration" implies that the epidermis remains passive. They pointed out that there is in fact abnormal keratinization of the whole epidermis. In our case such premature keratinization through the epidermis could be seen in many specimens.

The disintegration of epidermis that has lost its vitality and also the inflammatory reaction are natural changes and the absence of real perforation in the specimen does not exclude Kyrle's

disease if the disorder in other clinical and histological features is typical. Some cases of this kind are described (6).

REFERENCES

1. Constantine, V. S. & Carter, V. H.: Kyrle's disease. I. Clinical findings in five cases and review of literature. *Arch Derm (Chicago)* 97: 624, 1968.
2. — Kyrle's disease. II. Histopathologic findings in five cases and review of the literature. *Arch Derm (Chicago)* 97: 633, 1968.
3. Dammert, K. & Putkonen, T.: Keratosis follicularis serpiginosa Lutz (*Elastoma intrapapillare perforans verruciforme Miescher*). *Dermatologica* 116: 143, 1958.
4. Kyrle, J.: Über einen ungewöhnlichen Fall von universeller follikulärer und parafollikulärer Hyperkeratose (*Hyperkeratosis follicularis et parafollicularis in cutem penetrans*). *Arch Derm Syph (Berlin)* 123: 466, 1916.
5. Mehregan, A. H. & Coskey, R. J.: Perforating folliculitis. *Arch Derm (Chicago)* 97: 394, 1968.
6. Peterson, W. C., Goltz, R. W. & Hult, A.-M.: Hyperkeratosis penetrans (Kyrle's disease). *Arch Derm (Chicago)* 88: 210, 1963.

Received September 22, 1972

Reino Pajarre, M.D.
Department of Dermatology
University of Oulu
Oulu
Finland

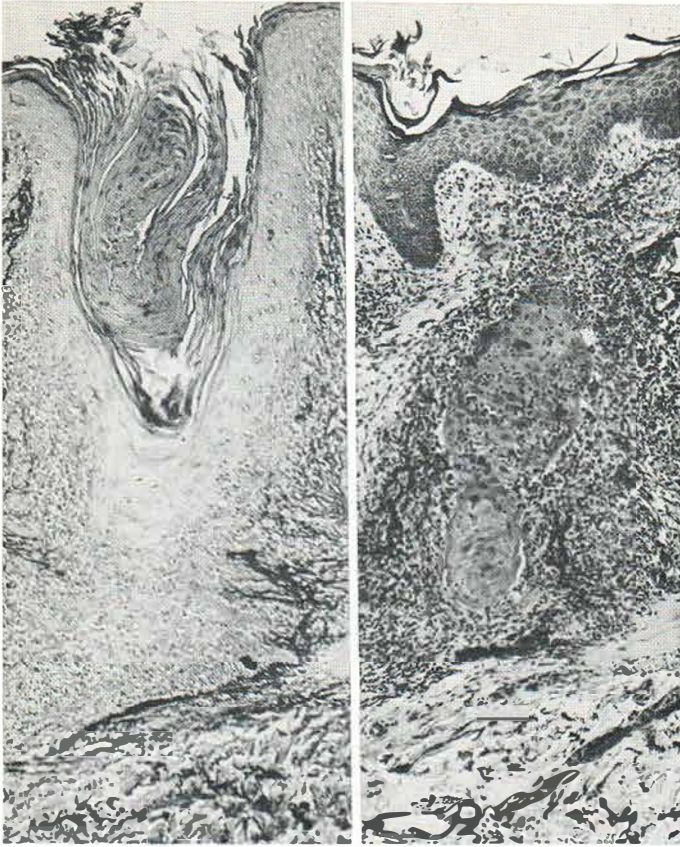


Fig. 2. Deep keratinizing epidermal downgrowth. A strong inflammatory reaction surrounding the epidermal ingrowth is well demonstrated. Acid-orcein-Giemsa, $\times 70$.