HYPERKERATOSIS FOLLICULARIS AND PARAFOLLICULARIS IN CUTEM PENETRANS (KYRLE'S DISEASE) IN A CHILD

A Case Report

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This paper describes a case of hyperkeratosis follicularis and parafollicularis in cutem penetrans which appears to be the first case reported in this country.

Case Report

This eleven year old white female was first seen at Hacettepe Children's Hospital on April 13, 1964, because of an eruption on her extremities which was said to have been present since she was three years of age. The lesions first appeared on her hands and gradually spread to her arms and legs. The eruptions would flare up and subside from time to time, but had never cleared up in the course of eight years. The condition was variously diagnosed and treated as allergic dermatitis, pyoderma, papular urticaria, by several clinicians, without any cure. There were no subjective symptoms, such as itching, burning sensation or pain accompanying the eruptions. Any kind of trauma, especially cat scratch, caused new lesions at the site of the trauma within two or three days (Koebner's phenomenon). In addition to these lesions, the patient developed urticarial lesions and water blisters after eating several kinds of fruit. These lesions would disappear within a few days.

The patient is the first child of the family. The pregnancy and delivery were uneventful, and her development was normal. She started walking at the age of 16 months, and shortly thereafter her parents noted an abnormality in her gait. She was hospitalized several times since 1959 because of the walking difficulty and bowing of her legs. A diagnosis of vitamin D resistant rickets was made and bowing was corrected in 1961 by stabling procedure. Since then she has taken vitamin D_2.

The patient's paternal grandfather has hypertension (type unknown), and her maternal grandfather has diabetes mellitus. Her 36

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year old father had rheumatic heart condition. Her mother (36 years old) and her two siblings are in good health.

Physical examination revealed a well-developed, well - nourished white girl, appearing her stated age. Examination of the skin showed hard, discrete, slightly elevated, flat or cone - shaped, greyish or red - brown papules with a central crust, one to six mm in diameter, located on the dorsum of the hands, flexor and extensor surfaces of the forearms, legs, thighs and dorsum of the feet (Figures 1, 2 and 3). In addition to these discrete papules, the dorsum of the right hand exhibited papules in linear distribution (the patient had been scratched by a cat on this site a month prior to this examination). Several small, scattered, slightly pigmented depressions were also present.

Figure 1. Hyperkeratotic papules on the thigh.

Removal of the crusts revealed a cone-shaped depression. No pain was felt on palpation and the crusts came off without difficulty. Histological sections of the lesion showed a dense, concave, lamellar parakerototic plug, located in the follicular orifice (Figure 4). Although the epidermis was acanthotic, the stratum malpighii was thinned along the edges of the plug, and in some sections disappeared at the bottom of the plug. The plug itself penetrated into the corium (Figure 5). An inflammatory infiltrate made up of lymphocytes, histiocytes, epithelioid cells and a few giant cells was present in the upper corium (Figure 6). Various staining techniques (periodic

Figure 2. Early and mature papules, showing the effect of Koebner's phenomenon.

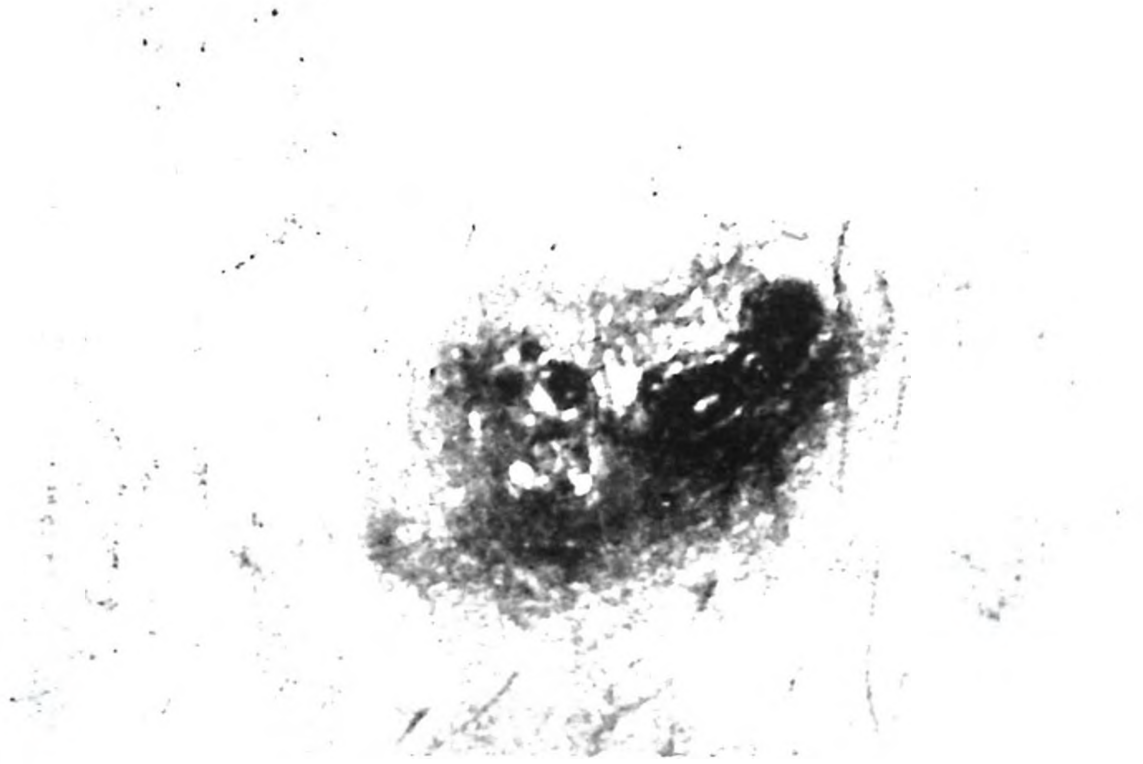

Figure 3. Close-up of the papules, indicating heavy keratotic plugs.

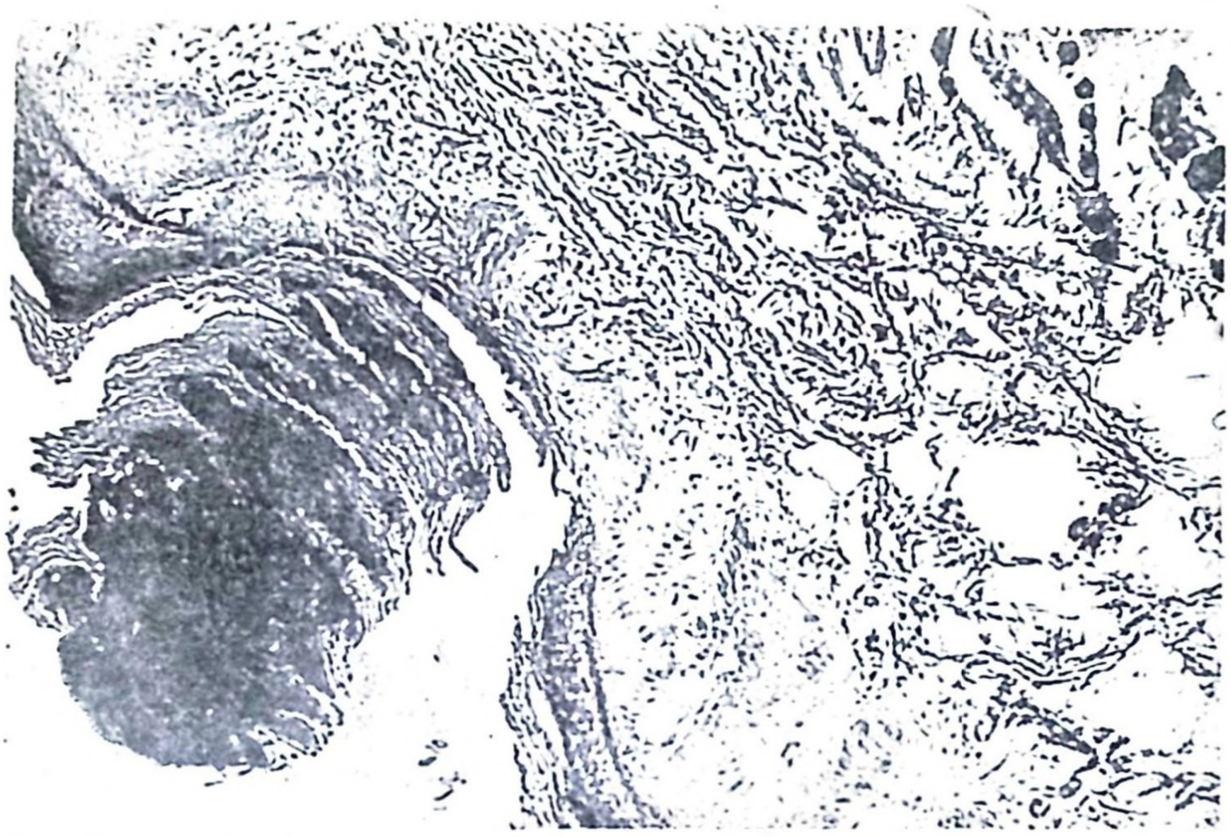

Figure 4. Low power magnification showing keratotic plug and thinning of the epidermis.

Figure 5. The plug penetrating the epidermis.

acid-Schiff, acid fast, Gram, Giemsa, von Kossa, Masson's trichrome and Verhoff's) were not diagnostic. Elastic fibers looked normal (Figure 7).

Laboratory Examination : Red blood count varied between 3,500,000 and 4,500,000/cmm; white blood count between 6,000 and 9,000/cmm with normal differential; hemoglobin between 11.3 and 14.5 gm/100 ml; platelets were within normal limits. Urinalysis : specific gravity, 1.014; pH, acid; glucose, negative; amino acids, ne-

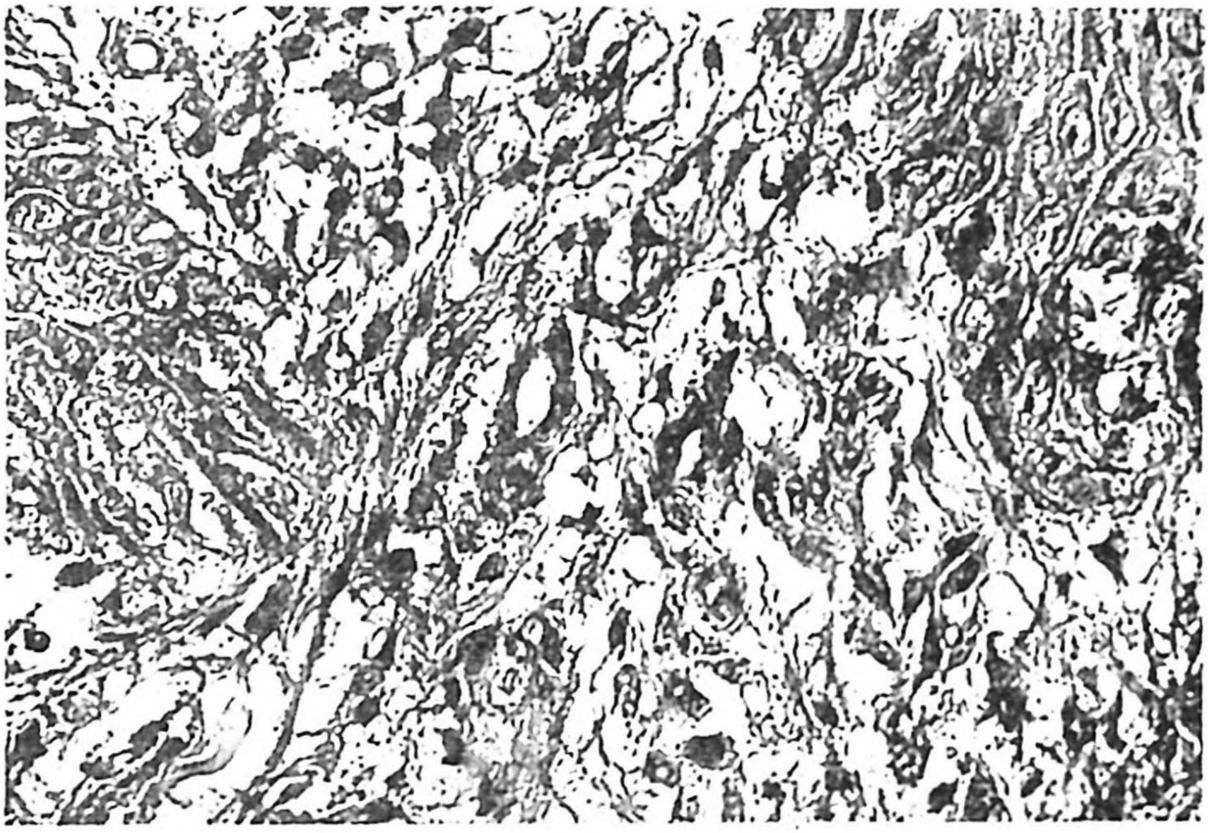

Figure 6. Granulomatous infiltrate.

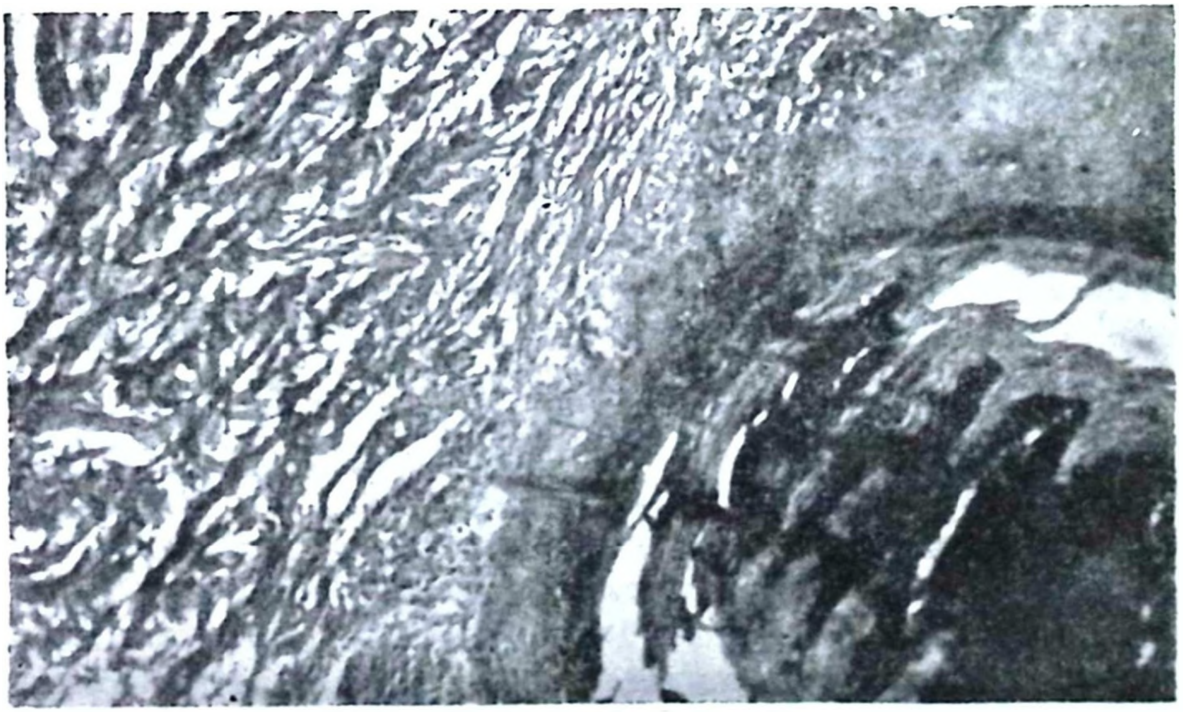

Figure 7. Normal elastic fibers.

gative. During vitamin D_2 treatment, urine Sulkowitch test became one plus to 4 plus. Fishberg test showed a urine specific gravity of 1.001 - 1.004 and 1.017 - 1.026. Serum Cl, K, Na, CO_2, NPN, blood

sugar and serum proteins (albumin and globulin) were all within normal limits. Alkaline phosphatase was normal. Blood cultures and tuberculin tests were negative. Serum phosphorus was 7.2 to 8.5 mg percent, and dropped to 5.2 to 6.5 mg percent with vitamin D_2 treatment. Serum calcium was within normal limits but became elevated to 13.8 mg percent with vitamin D_2 treatment. X - rays of the chest and an intravenous pyelogram were normal. Initial x-rays of the bones were reported to show active rickets, but follow-up pictures indicated a gradual improvement of this disease.

Discussion

Hyperkeratosis follicularis and parafollicularis in cutem penetrans was described by Kyrle in 1961.[1] About 30 cases have since been reported, but of these, de Graciansky et al,[2] have accepted only 12 as being authentic examples of this disorder.

Clinically the disease is characterized by horny papules at or near the follicular orifices anywhere except on the palms, soles, and mucosae. The eruptions may be localized or generalized. The primary lesion is a cone-shaped, slightly elevated, gray, brown or normal - colored papulae. These papules have a central adherent scale, or cornified cone, and some of the papules may be surrounded by an inflammatory halo. Starting as small as one to two mm in diameter, they may enlarge to 15 mm, and may coalesce to from polycyclic plaques with heavy crusts. The lesions are essentially asymptomatic and at all stages can be torn off with little difficulty, revealing a cone-shaped depression. Healing of the lesions after removal of the plugs may result in a slight, pigmented depression.

Histological examination is diagnostic. A large mass of keratin, either in the orifice of a hair follicle or independent of the follicle, is present. This mass is lamellar, may or may not be parakeratotic, and has been compared to a pile of saucers. Beneath this plug the epidermis may be acanthotic or atrophic, with flattening of the rete pegs. In some cases the keratin plug penetrates into the corium.[1, 2] This penetrating plug provokes a granulomatous inflammatory response with foreign body giant cells. When there is no penetration of the corium by keratin there is a slight, nonspecific infiltration. This penetration of the corium by keratin is a striking event but does not occur in all lesions and deGraciansky et al,[2] did not regard its occurence as essential to the diagnosis of this disease, accepting as authentic some cases which appeared to be typical but microsco-

pically showed only the presence of a hyperkeratotic mass above an intact epidermis.

A differential diagnosis must take into account the following :
elastosis perforans serpiginosa
porokeratosis Mibelli
lichen scrofulosorum
lichen spinolosus
keratosis pilaris
papulonecrotic tuberculide
keratosis follicularis
pityriasis rubra pilaris
hyperkeratotic epithelial nevi.

Except for the first, the conditions listed above do not cause complications in the clinical and histological diagnosis. Although elastosis perforans serpiginosa may be confused in some cases with Kyrle's disease in the clinical diagnosis, the histological findings are in fact quite different in that there are obvious abnormalities of the elastic fibers in elastosis perforans serpiginosa.[6]

Summary

The case of an eleven year old girl with Kyrle's disease is presented. Of especial interest in this case was that the patient also had vitamin D resistant rickets and Koebner's phenomenon.

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