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Case Report

Kyrle's Disease: A Case Report

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Abstract

Observations: Kyrle's disease was first described under the name hyperkeratosis follicularis et parafollicularis in cutem penetrans in 1916. This rare dermatologic entitiy is characterized by a chronic, more or less extensive, papular eruption over the upper body and the legs. We report a case of Kyrle's disease involving the face, neck, scalp and lower extremities. Based on laboratory and other findings, chronic renal failure was diagnosed. Histopathological analysis of skin lesions showed presence of keratotic, partly parakeratotic plug invaginating the epidermis. Kyrle's disease is classified among the perforating skin diseases, in which reactive perforating collagenosis, perforating folliculitis and elastosis perforans serpiginosa take place.These acquired perforating dermatoses usually have been associated with diabetes mellitus or chronic renal failure. Further investigations with more cases are needed to understand the underlying pathogenesis.

Introduction

Kyrle first described this disorder in 1916 under the name hyperkeratosis follicularis et parafollicularis in cutem penetrans [1]. Kyrle's disease, a rare skin disorder, is characterized by a chronic, more or less extensive, papular eruption over the upper body and the legs. These papules may be follicular or parafollicular and contain a central cone-shaped plug. Characteristically, the disease does not involve the mucous membranes and palmo-plantar surfaces [2]. Histopathologically, a keratotic plug fills an epithelial invagination. Parakeratosis is present in parts of the plug that sometimes penetrates in the dermis [3]. Most patients have association with systemic diseases [4].

Herein we report a case of *Kyrle*'s disease associated with chronic renal failure.

Case Report

We present a case of *Kyrle*'s disease involving the face, neck, scalp and lower extremities. The patient was a 46-year-old male with a one year history of pruritic papules with central keratotic plugs. He had chronic renal failure for 5 years and underwent hemodialysis three times a week since that time. Family history was noncontributory and there was no previous history of skin disease. Papular eruption had developed first on his lower extremities which progressed to involve his neck, face and scalp (**Figure 1** and **2**). The early skin lesions started as hyperkeratotic papules of 2-5 mm. In a few months these lesions enlarged to 5-8 mm in diameter. Laboratory tests during the period 1996-2001 showed the follow-

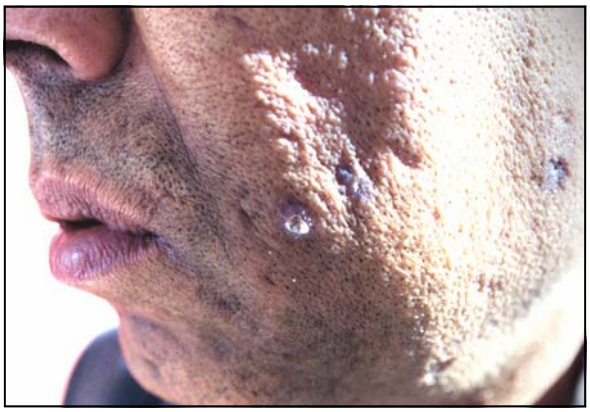

Figure 1. Keratotic papules on the face

ing values: blood urea ranged from 75-202 mg/dl and serum creatinine, 2.0-8.3.

Skin biopsy specimen revealed a keratotic, partly parakeratotic plug invaginating the epidermis. Epidermal perforation was absent (**Figure 3**). 0.1% retinoic acid cream was applied two times daily. After 6 months moderate flattening of the lesions was noted. Lesions recurred when topical treatment was discontinued.

Discussion

Kyrle's disease is one of the perforating skin diseases, in which reactive perforating collagenosis, perforating folliculitis and elastosis perforans serpiginosa take place. These acquired perforating dermatoses usually have been associated with diabetes mellitus or chronic renal failure. It has been also reported with other conditions including tuberculosis, pulmonary aspergillosis, scabies,atopic dermatitis, AIDS, neurodermatitis, malignant, hepatic and endocrinological disorders [**5, 6, 7**].

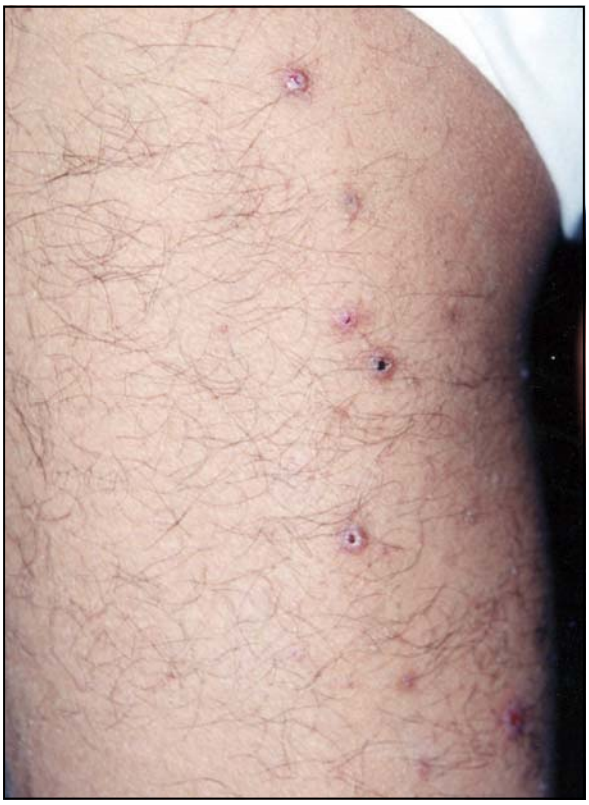

Figure 2. Lateral aspect of the left thigh

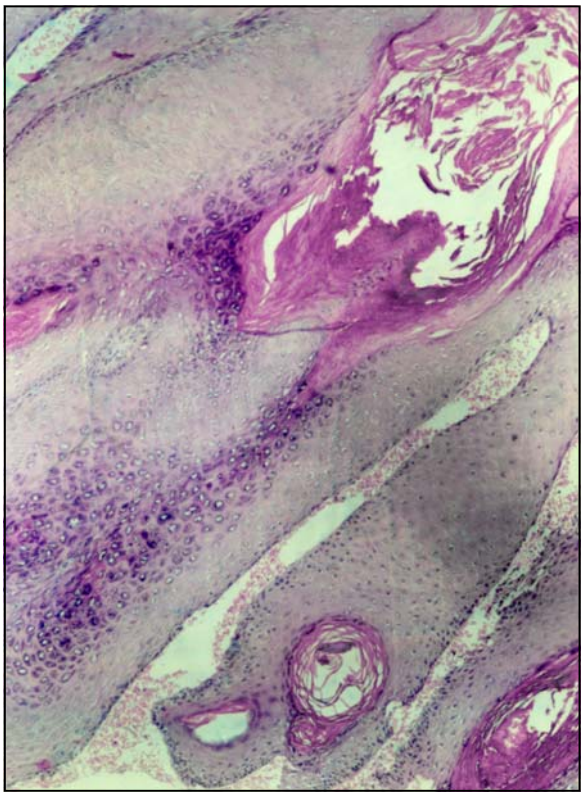

Figure 3. Keratotic plug invaginating the epidermis (HE x 40).

The etiology of *Kyrle*'s disease remains unknown. Although it is suggested to be an autosomal recessive genodermatosis, the mode of inheritance is not obscure [**8, 9**]. Hereditary association was not confirmed in our patient. This disease predominantly affects without a predilection of sex or race [**10**]. *Alyahya* et al. [**11**] suggest that it might also exist in children and also described the first conjunctival changes in a case of *Kyrle*'s disease. The basic pathogenic event seems to be a focal disturbance of epidermal cell proliferation and differentiation which leads to a keratotic plug. Progressive dislocation of the level of keratinization toward the dermal-epidermal junction is felt to be the reason for this event [**8**]. It is suggested that one of the extracellular

matrix protein, fibronectin could be involved in the pathophysiological mechanism in *Kyrle*'s disease as well as the other perforating dermatoses [12]. Regression of small lesions with clindamycin leads to proposals of other hypotheses that microorganisms may play a role in the pathogenesis of *Kyrle*'s disease at least in the initial stage of lesions [9]. Like our case with chronic renal failure, this disorder has been reported in association with diabetes mellitus, renal disease, hepatic insufficiency and congestive heart failure [4]. Nine cases of *Kyrle*'s disease among 200 patients were reported by *Hood* [13] who underwent hemodialysis beacuse of chronic renal failure. Although the pathogenesis of *Kyrle*'s disease is unknown, it has been treated with numerous agents. Some of these methods are: retinoic acid preparations, electrocautery, cryotherapy, CO_2 laser surgery, high-dose vitamin A, oral retinoids (isotretinoin and etretinate), ultraviolet irradiation after curetting the hyperkeratoses and a combination of oral retinoids and psoralen plus UVA [8, 14, 15]. Discontinuation of these treatments usually results in recurrence of the lesions [8]. Further investigations with more cases are needed to understand the underlying pathogenesis.

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