CASE REPORTS

KYRLE'S DISEASE IN A PATIENT WITH CHRONIC RENAL FAILURE

A J Kanwar, S Kaur, Arti Nanda, Rashmi Arora, Rajeev Sharma and B D Radotra

A 53-year-old female undergoing dialysis for chronic renal failure was seen with multiple discrete hyperpigmented papules of variable sizes each with a keratotic plug over the buttocks, back of thighs and knees of 2 months' duration. The lesions were mildly itchy. A diagnosis of Kyrle's disease was confirmed on histopathology. The lesions showed improvement with topical tretinoin.

Key words : Kyrle's disease, Chronic renal failure.

Kyrle's disease (KD) is a peculiar perforating disorder of the skin characterized by papules with a central keratotic plug. It was initially described by Kyrle in 1916,[1] and until recently, it has been considered a rare disorder.[2] However, reports of occurrence of this disorder in patients with chronic renal failure have revived interest in this entity.[3-6] The present communication describes KD in a patient undergoing haemodialysis for chronic renal failure.

Case Report

A 53-year-old female was seen for an eruption over the buttocks, back of thighs and knees of 2 months' duration. It was mildly itchy. The patient, a known diabetic had been on dialysis for the past 6 months for chronic renal failure. There was no family history of similar lesions and there was no clinical or laboratory evidence of any hepatic disorder in the patient. The lesions were multiple hyperpigmented papules of variable sizes each with a central keratotic plug (Fig. 1). Most of the lesions were discrete; a few were grouped to form small plaques. It was possible to remove the central horny plug from a few of the lesions with the help of a curette.

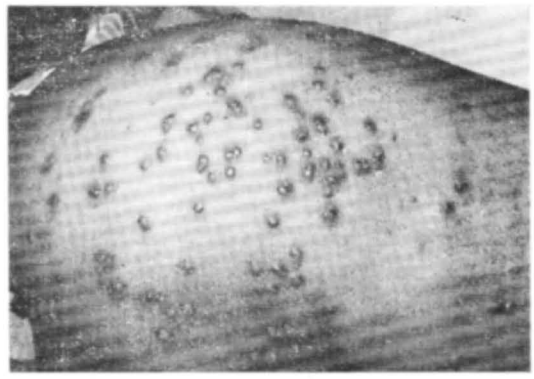

Fig. 1. Multiple hyperpigmented papules and nodules each with a central pale plug.

Histopathological examination of one of the papules on the thigh revealed a keratotic plug, partially parakeratotic, with basophilic material present in an invagination of the epidermis. There was disruption of epidermal cells at places and a round cell infiltrate in the upper dermis (Fig. 2). A Verhoeff-von Gieson stain for elastic fibres and a Masson trichrome stain for collagen fibres revealed no extrusion of these fibres into the keratotic plug. The patient was advised topical tretinoin (Retino A) with which

From the Departments of Dermatology and Pathology, Post-graduate Institute of Medical Education and Research, Chandigarh-160 012, India.

Address correspondence to : Dr A J Kanwar.

Fig. 2. Typical Kyrle's lesion with epidermal invagination filled with keratin and debris. Thinning of stratum malphigi and flattening of rete ridges is also seen. (H and E, X 140).

the lesions flattened and the pruritus decreased over a month.

Comments

Kyrle's disease is an exceedingly rare dermatologic disorder with distinct clinical and histopathologic findings.[7,8] The earliest lesion is a small papule with a silvery scale which gradually enlarges. The fully developed papules are reddish-brown or hyperpigmented with a central, cone-shaped plug that can be removed with a curette. The papules may be follicular or extra-follicular in location and may coalesce into verrucous plaques.[9] The extensor surfaces of extremities are the most common site of involvement. Though any part of the body may be affected, face, mucous membranes, palms and soles tend to be spared.[3] In our patient, the lesions were confined to the lower extremities only. Though some patients exhibit Koebner's phenomenon,[7] it was absent in our patient. Clinically, the differential diagnoses considered were prurigo nodularis, keratoacanthomas, lichen planus hypertrophicus and perforating folliculitis. However, these were excluded on histopathology.

Several patients of Kyrle's disease have associated diabetes mellitus, so much so that the disorder has been considered to be an unusual manifestation of the diabetic syndrome.[10] Other associations have included clinical or laboratory evidence of hepatic abnormalities[6,7] or congestive cardiac failure[6,7] and hypothyroidism.[3] However, the association of KD with chronic renal failure is interesting. Hood et al[3] observed 9 cases of KD among 200 patients receiving haemodialysis for chronic renal failure which is an extremely high incidence.[9] All these patients were black, though in cases reported in the literature, the disease is three times more prevalent among whites than blacks.[3] In 7 of these, the renal failure was related to insulin-dependent diabetes mellitus. In our patient also, diabetic nephropathy contributed to renal failure.

The primary pathogenetic event in KD is a disturbance of epidermal keratinization which takes place at the level of the basal cell layer, inciting dermal inflammation and resulting in the classic histopathology of KD.[2] It is possible that metabolic derangements associated with diabetes mellitus and renal failure are somehow responsible for abnormal keratinization or connective tissue changes, but precise mechanisms are at present unknown.[2] A variety of topical keratolytic agents have been used for the treatment of KD.[2] We observed flattening of the lesions and decreased pruritus with twice daily applications of tretinoin.

References

1. Kyrle J : Hyperkeratosis follicularis et parafollicularis in cutem penetrans, Arch Dermatol Syphilol, 1916; 123 : 466-493.
2. Patterson JW : The perforating disorders, J Amer Acad Dermatol, 1984; 10 : 561-581.
3. Hood AF, Hardegan GL, Zarate AR et al : Kyrle's disease in patients with chronic renal failure, Arch Dermatol, 1982; 118 : 85-88.
4. Aram H, Szymanski FJ and Bailey WE : Kyrle's disease, Arch Dermatol, 1969; 100 : 453.
5. Ephraim AJ and Barbanti BJ : Kyrle's disease, Arch Dermatol, 1971; 101 : 704.

6. Moss HV : Kyrle's disease, Cutis, 1979; 23 : 463-466.
7. Carter VH and Constantine VS : Kyrle's disease. I. Clinical findings in five cases and review of literature, Arch Dermatol, 1968; 97 : 624-632.
8. Constantine VS and Carter VH : Kyrle's disease. II. Histologic findings in five cases and review of literature, Arch Dermatol, 1968; 97 : 633-639.
9. Lever WF and Schaumburg-Lever G : Kyrle's disease : Degenerative diseases, in : Histopathology of the Skin, JB Lippincott, Philadelphia, 1983; p 273-275.
10. Moschella SL : Diseases of nutrition and metabolism, in : Dermatology, Vol 2, Second ed, Editors, Moschella SL and Hurley HJ : WB Saunders, Philadelphia, 1985; p 1928-1929.