KYRLE'S DISEASE

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Summary

Three cases of Kyrle's disease are reported. All the three cases showed lesions with typical morphology and histology. In one instance the disease was present in the earlier generation. An interesting feature is the observation of a Koebner – like phenomenon.

KEY WORDS : Kyrle's disease, Follicular papules, Koebner reaction

Kyrle's disease is a rare disorder of keratinization, first described as a specific entity by Kyrle[1] in 1916. It is characterized by papules situated at or adjacent to hair follicles. The papules may appear anywhere on the body except mucous membranes. As the papule ages a central keratotic plug forms which on removal leaves a crater like depression. The lesions come in crops, last for weeks to months and eventually heal with atrophic scars[2].

In the last three years and a half three cases of Kyrle's disease occurring in three different age groups were encountered. Since the disease is rare we considered these cases worth reporting. Besides, in case 1, there was a family history of similar illness occurring in the mother of the patient.

Report of Cases

Case No. 1

A 37 year old lady was seen in the out patient department with history of chronic skin lesions which started at the age of 10 years. The lesions were appearing in crops and fresh lesions appeared as the old ones kept subsiding. Most of the lesions healed with pitted scars. Examination revealed hyperpigmented, hyperkeratotic papules not more than 0.5 cm in diameter, mostly over the legs and feet, but few over the face and rest of the body. Many of these showed central keratotic plugs and a few central crater-like depressions. The lesions were discrete and linearly arranged over parts of the legs suggestive of Koebner phenomenon (Fig. 1). Pitted scars were also seen in these areas. Hair, nails, mucous membranes, palms and soles were normal. There was no evidence of diabetes mellitus. Pedigree pattern is depicted in Fig. 2.

Case No. 2

A 14 year old boy presented with asymptomatic papular lesions which were present for 9 years. These had first appeared on the face and then on the extremities and dorsa of the hands. The hyperkeratotic papules with surrounding erythema later acquired a central plug and healed with pitted atrophic scars which looked like pock marks especially on the face (Fig. 3). There was no family history fo similar skin disorder.

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Received for publication on 27—6—1981

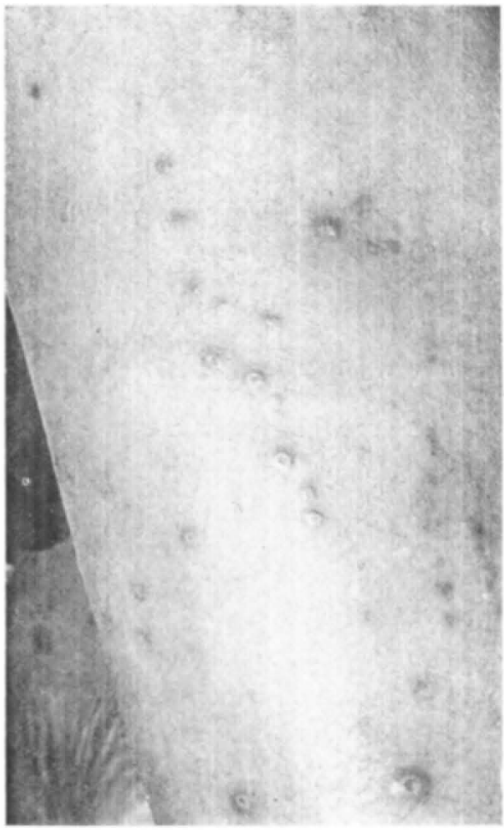

Fig. 1
Typical papular lesions of Kyrle's disease in linear arrangement (Koebner phenomenon)

Case No. 3

A 70 year old man with diabetes mellitus was referred to our skin out patient department with history of itchy skin lesions of two months' duration. The lesions were numerous over the abdomen, back and upper extremities. Each lesion had lasted for about three weeks before healing with atrophic pitted scars. The lesions and scars were seen arranged in lines transversely and vertically over the back and upper extremities, simulating Koebner phenomenon. Palms and soles were free. Mucous membrane was normal. There was family history of diabetes mellitus but not of any similar skin disorder.

Biopsy of the lesions from all three cases confirmed the diagnosis of Kyrle's disease. Histopathology in each case demonstrated hyperkeratotic and parakeratotic plug with basophilic debris penetrating into the dermis where an inflammatory reaction consisting of lymphocytes and a few polymorphs was seen. (Fig. 4).

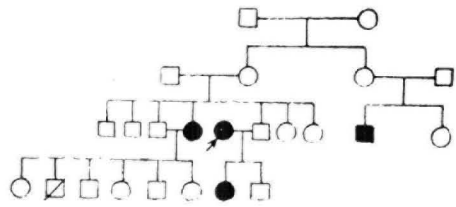

Fig. 2 Pedigree pattern of case 1

Discussion

The diagnosis of this rare disease is to be confirmed by biopsy. The microscopic changes are characteristic and the disease is aptly called 'hyperkeratosis follicularis et parafollicularis in cutem penetrans'.

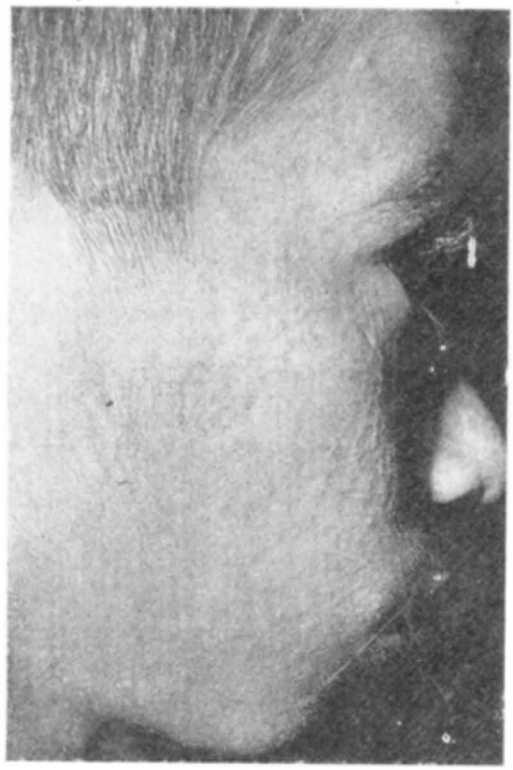

Fig. 3
Pitted atrophic scars over the face looking like pock marks.

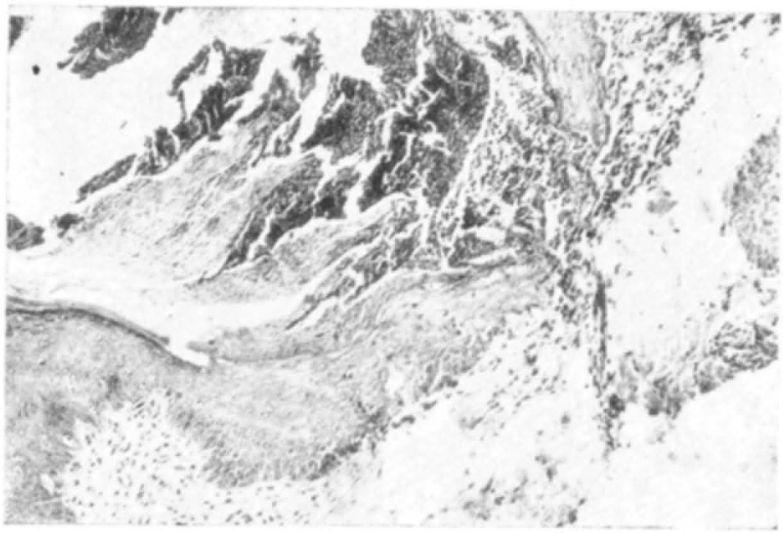

Fig. 4 Shows typical histopathologic features. Refer text.

Carter and Constantine in 1968 forwarded certain criteria for the diagnosis of Kyrle's disease[3,4] based on clinical and histological features. All our patients satisfied these criteria.

A few unusual features noted in our patients need to be highlighted. Out of the 3 cases 2 were males and one female. This sex ratio is in contrast to the usually reported female predominance The 2nd patient was reported to have started the disease at the age of 5 years, an unusually early start of the disease. The hereditary nature of the disease if any has been in dispute. Most cases have occurred as isolated cases[5,6]. Kyrle's disease when reported in families have occurred in siblings. Autosomal dominant inheritance has also been suggested[7]. The occurrence of the disease in 2 generations as in the first case reported here is extremely rare. The pedigree pattern in this instance was however not suggestive of any particular mode of inheritance.

Yet another unusual phenomenon was the koebering seen in our cases. Koebner-like phenomenon unrelated to trauma has been reported in some cases. All 3 cases showed lesions in linear arrangement but in cases 2 and 3 this was seen on sites not particularly prone to injury. Kyrle's disease appears to be associated with diabetes mellitus[3,8,9]. Such association is seen in case No. 3 though the skin lesions appeared in the 7th decade many years after the onset of diabetes. As has been the experience of many the response to Vitamin A therapy and topical keratolytics in our cases has not been encouraging.

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