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KYRLE - FLEGEL DISEASE IN SIBLINGS WITH CHILDHOOD ONSET AND KOBNER PHENOMENON

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This is a report of Kyrle - Flegel disease occurring in 3 siblings. Two of them were girls (13 and 9 years) and the third patient was a boy (7 years). They had developed hyperkeratotic papules since the ages of 9, 6, and 5 years respectively. Lesions were present mainly on the extensor surfaces of limbs. Some lesions were linear, they had appeared at the sites of trauma. Histology showed laminated hyperkeratosis, follicular plugging, and flattening of the dermoepidermal junction. Patients partially responded to oral vitamin A in high doses.

Key Word : Kyrle's disease

Introduction

The exact relationship between some disorders with idiopathic warty papules is unclear. Kyrle's and Flegel's diseases are perhaps the same condition. The small papule pattern (Flegel) is much more common than the larger lesions (Kyrle).[1] We report Kyrle - Flegel disease in 3 siblings with certain peculiar features.

Case Reports

Three siblings (2 sisters aged 13 and 9 years, and 1 brother of 7 years) presented with asymptomatic hyperkeratotic papules and nodules mainly on the extensor surfaces of limbs and buttocks. The lesions started appearing at the ages of 9, 6, and 5 years respectively. Individual lesion was a horny papule with a central protruding plug. Removing the central plug left a crateriform depression. There was no discharge from the lesions. Some lesions were linear (Fig. 1) and had appeared at the sites of trauma or scratch. Face, palms, and soles were spared and oral cavity, hair, and nails were normal in all cases. All other family members (including 2 other siblings 4 1/2 years - old -brother and 2 - year - old sister) were normal. Glucose tolerance and liver function tests were normal in all patients.

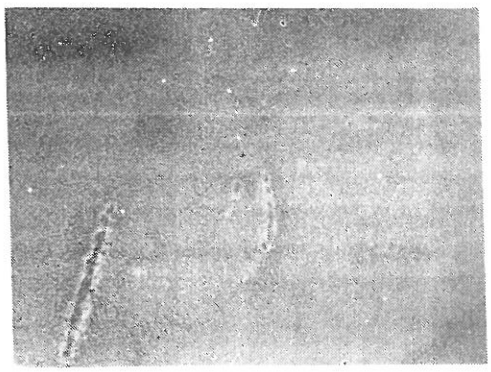

Fig. 1. Linear lesions

Biopsy of a lesion on the lower limb of

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the eldest patient was taken. Histology showed marked laminated hyperkeratosis, follicular plugging, and flattening of dermoepidermal junction. There was no downgrowth of epidermis and dermis was normal. The diagnosis of Kyrle - Flegel disease was made. All patients were given oral vitamin A 50,000 IU bid for 15 days and the number of lesions decreased almost by half in each case.

Comments

Till 1969 Kyrle's disease had been reported only 5 times in siblings.[2] A few reports of such cases have appeared after that.[3,4] Onset of Kyrel's disease is usually between the ages of 35 and 60 years.[5] In our cases the onset was much earlier. All our patients exhibited Köbner phenomenon, which has not been described with this disease.[1] The reasons for reporting the cases are (1) occurrence in siblings, (2) early age of onset, (3) presence of Köbner phenomenon, and (4) response to oral vitamin A.

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