Greither's disease

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ABSTRACT

A 14-year-old girl with diffuse palmoplantar keratoderma with hyperhidrosis and progressive extension of keratoderma to the dorsum of the hands and feet is reported. The inheritance pattern was autosomal dominant.

Key Words: Greither's disease, Palmoplantar keratoderma

INTRODUCTION

Hereditary palmoplantar keratodermas (PPK) are a heterogeneous group of diseases characterized by hyperkeratosis of the palms and soles with thickening of the stratum corneum, usually distinguishable by the mode of inheritance and by associated clinical findings.[1] We report a case of diffuse palmoplantar keratoderma associated with hyperhidrosis and progressive extension of the keratoderma to the dorsa of the hands and feet.

CASE REPORT

A 14-year-old girl presented to the dermatology OPD with the complaint of thickening of the palms and soles since the age of 6 years. Initially, thickening was noted in the centre of the palms and soles, and gradually spread to the periphery. Thickened skin was also noted over the knees and just below the lower lip. She was born of a non-consanguinous marriage. There was a history of her paternal grandfather and father suffering from a similar illness. The patient had three siblings of whom one sister had a similar ailment.

Cutaneous examination revealed diffuse palmoplantar keratoderma with hyperhidrosis (Figure 1). An erythematous border was found around the keratoderma (Figure 2) that continued along the Achilles tendon. The knee and the area just below the upper lip exhibited hyperkeratotic plaques. Routine hematological and urine examination findings were within normal limits. The patient refused a biopsy. A clinical diagnosis of Griether's disease was made.

DISCUSSION

Greither first described this disorder in 1952.[1] The most important signs and symptoms[2] are diffuse PPK with an erythematous border. The lesions tend to extend to the dorsum of the hands and feet (transgradiens), and hyperkeratotic plaques are present on the elbows and knees. Marked hyperhidrosis is said to be typical of this condition.

The nosological situation of this keratoderma is under scrutiny since the publication of Greither.[3] Some consider it as a distinct entity, while others consider it as a variant of the Unna-Thost type of PPK.[4] It is claimed to differ from the Unna-Thost variety by showing extension to the extensor surface of the hands, knees and elbows and by showing a tendency to improve in the fifth decade.[5] Greither's disease has several clinical similarities with Mal de Meleda syndrome.[6] However,

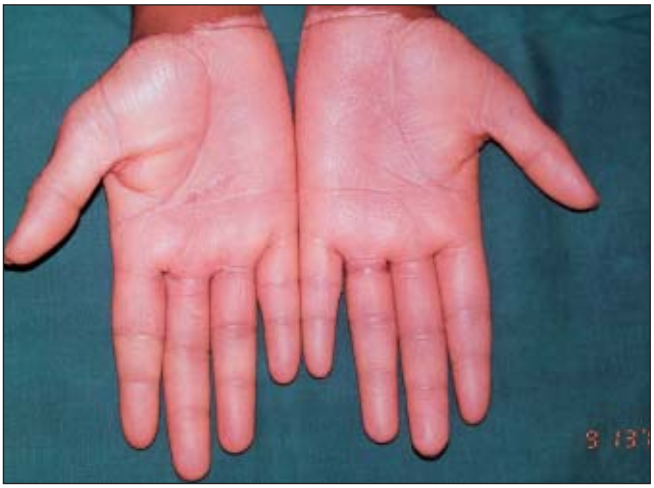

Figure 1: Diffuse palmar keratoderma

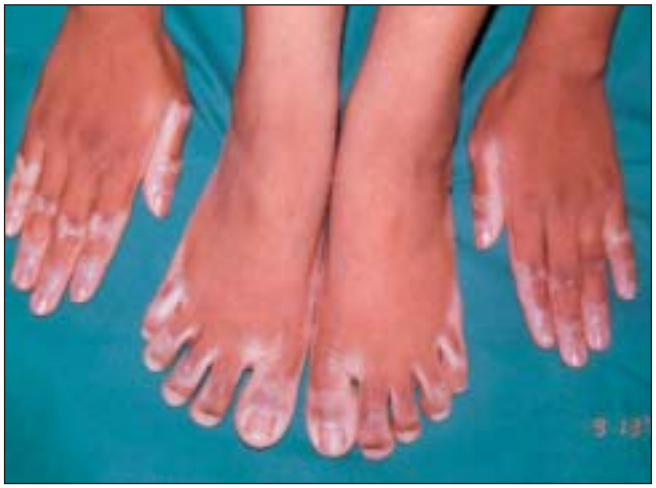

Figure 2: Transgradiens

in contrast to Mal de Meleda syndrome, the palms and soles may be spared in Greither's disease. Mal de Meleda syndrome is autosomal recessive; PPK appears early after birth and progressively involves other regions of the body without a tendency for spontaneous involution. It also has typical nail changes.

To conclude, we report a case of PPK consistent with the clinical and genetic definitions of Greither's disease. The histology described by Greither and others is not specific.[1] We think that the natural history and the unique clinical findings justify retaining the entity as Griether's disease. An electron microscopic study by Beylot-Barry et al[7] showed aggregated tonofilaments around the nucleus, without true clump formation. Desmosomes were numerous and cell-cell junctions showed an imbricated pattern. Molecular genetic investigation with localization of the responsible gene may clarify the nosologic situation of this type of keratoderma.

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