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Case Report

Greither's disease: a case report

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ABSTRACT

Griether's disease, also known as, Transgrediens et Progrediens Palmoplantar Keratoderma, is an autosomal dominant disorder, clinically characterized by diffuse palmoplantar keratoderma associated with hyperhidrosis and progressive extension of keratoderma to the dorsum of hands and feet, knees, elbows with characteristic involvement of Tendo-Achilles. We report a case of a 9 year old male with typical features of Greither's disease.

Keywords: Greither's disease, Palmoplantar keratoderma

INTRODUCTION

Palmoplantar keratoderma are a heterogeneous group of disorders characterized by hyperkeratosis of the palms and soles.[1,2] In 1952, Greither[3] originally described keratosis extremitatum hereditaria progrediens, a new kind of palmoplantar keratoderma (PPK), inherited in an autosomal dominant pattern with variable expression. Mutations in the gene encoding keratin 1 are responsible.[4] Onset is in late childhood. Diffuse palmoplantar keratoderma extends onto the dorsal aspects of the hands and feet (transgrediens), knees, elbows, with characteristic involvement of Tendo-Achilles. Hyperhidrosis is common. Histopathology is usually nonspecific in these cases.

CASE REPORT

A 9 year old boy, presented to our department, with complaints of thickening and fissuring of the palms and soles since one year of age. Initially, the thickening was noted as small raised lesions over fingertips and plantar surface of the toes, which gradually progressed to diffuse thickening of the palms and soles with extension to the dorsum of hands and feet. Patient also had excessive sweating and fissuring of the palms and soles. He was born of non-consanguineous marriage and there was no similar illness in the family. Physical examination revealed diffuse keratoderma of the palms with erythematous border (Figure 1) extending to the dorsa of the hands (Figure 2). Diffuse plantar keratoderma was associated with deep fissures and hyperhidrosis with maceration of the skin, more prominent in area of weight bearing (Figure 3). The erythematous border was seen extending to the dorsum of the feet (Figure 4) and to the skin over the Achilles tendon (Figure 5). There was no hyperkeratosis over the knees or elbows. Hair, nails and teeth were normal. Routine investigations revealed mild anaemia (Hb-10 gm/dl). LFT, RFT and urine examination were within normal limits.

DISCUSSION

In 1952, Greither described transgrediens et progrediens PPK, and since then, few cases of this variant of PPK have appeared in the literature.[5-8] Greither's disease is characterized by diffuse keratoderma of the palms and soles, which may be slight, but by definition extends to the dorsal aspects (transgrediens), characteristically involving the skin over the Achilles tendon. Hyperkeratotic erythematous plaques may also be present over the knees and elbows.

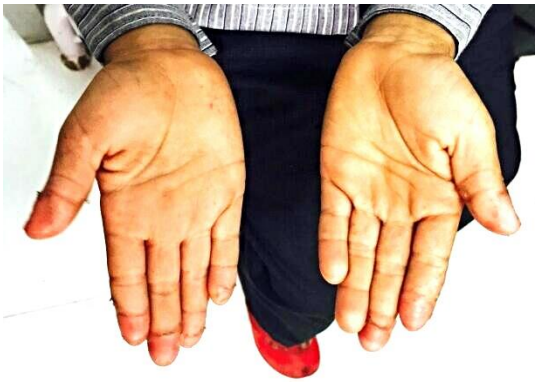

Figure 1: Keratoderma involving the palms.

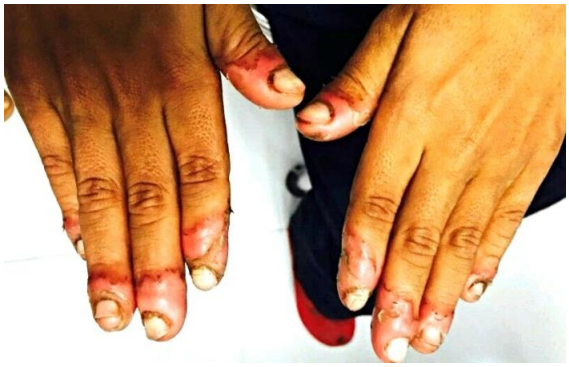

Figure 2: Lesions extended to the dorsa of the hands (transgadiens).

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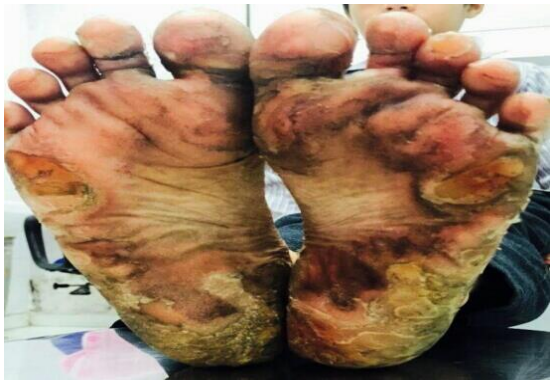

Figure 3: Diffuse plantar keratoderma.

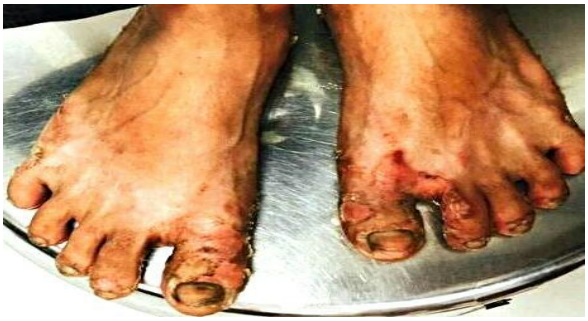

Figure 4: Hyperkeratosis extending to the dorsal aspect of the feet.

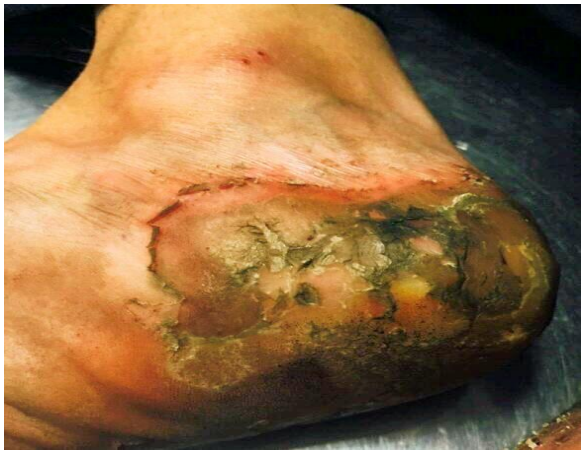

Figure 5: Hyperkeratosis extending to the Achilles tendon.

In classic cases, lesions of Greither's disease start after the second year of life and tend to involute after the sixth decade. In cases described in the literature, the histopathologic findings are nonspecific and consist of orthokeratotic hyperkeratosis. Flückiger and Itin studied a case of Greither's disease ultrastructurally and found cytoskeleton aberrations, filament aggregations, and cell-to-cell junctions with an imbricated pattern.[5] Some consider it as a distinct entity while others consider it as a variant of the Unna-Thost type of PPK.[9] 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.[10] Greither's disease has several clinical similarities with Mal de Meleda syndrome.[11] However, 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; the palmo-plantar keratoderma 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 features of Greither's disease. The histology described by Greither and others is not specific.[12] 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[13] showed aggregated monofilaments 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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