CASE REPORT

Watcher's type of palmoplantar keratoderma

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Abstract

A 51-year-old man reported with progressive thickening of the skin of the hands and feet since the age of 6yrs. It was largely asymptomatic; however, brisk walking caused excessive sweating, pain, and widening of the fissures on the soles of the feet. He also had scaly raised lesions on legs and knees. His mother, maternal aunt, cousin and nephew had similar lesions. Examination of the soles of feet revealed pronounced thickening of the skin at the bony prominences which was well demarcated. In addition, marked fissuring was obvious. Both palms have similar lesions which were minimal. Nails showed subungual hyperkeratosis and onychogyrophosis. Histopathology from sole showed marked hyperkeratosis, parakeratosis, hypergranulosis, irregular elongation of rete rigdes and dense lymphocytic infilterate in upper dermis & papillary dermis. Patient was started on oral acitretin 25mg twice daily and topical 12% salicylic acid with minimal improvement. A rare and unusual case of palmoplantar keratoderma is presented.

Key words: *Palmoplantar keratoderma, focal*

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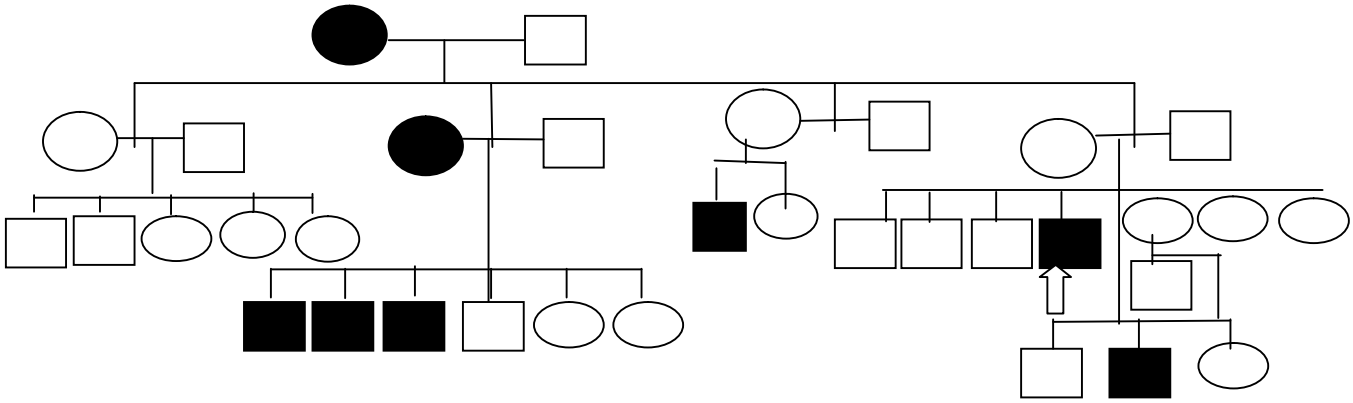
Introduction

Palmoplantar keratodermas form a rare but highly diverse group of hereditary skin disorders characterized by abnormal thickening of palms and soles that occur either isolated or in association with other cutaneous and extracutaneous manifestations (1). Palmoplantar keratodermas can be separated in the following functional subgroups: disturbed gene fuctions in structural proteins (keratins), cornified envelope (loricrin, transglutaminase), cohesion (plakophilin, desmoplakin, desmoglein1), cell-to-cell communication (connexins), and transmembrane signal transduction (cathepsin C) (2). Here we present an unusual case of watcher's type of palmoplantar keratoderma.

Case Report

A 51-year-old man reported with progressive thickening of the skin of the hands and feet since he was 6yrs. It was largely asymptomatic; however, brisk walking caused excessive sweating, pain, and widening of the fissures on the soles of the feet. He was unable to walk barefooted. He also had scaly raised lesions on legs and knees. His mother, maternal aunt, cousin and nephew had similar lesions (table 1 pedigree chart). Examination of the soles of feet revealed pronounced hyperkeratosis of the skin at the bony prominences which was well demarcated. The color of the skin was yellow and waxy (Fig. 1). In addition, marked fissuring was obvious. Both palms have similar lesions which was minimal (Fig. 2). Nails showed subungual hyperkeratosis and onychogyrophosis. Histopathology from sole showed marked hyperkeratosis, parakeratosis, hypergranulosis, irregular elongation of rete rigdes and dense lymphocytic infilterate in upper dermis & papillary dermis (Fig 3). Thus on history, positive family history, clinical examination and histopathology, a diagnosis of palmoplantar keratoderma of Watcher's type was made. Patient was started on oral acitretin 25mg twice daily and topical 12% salicylic acid with minimal improvement.

Table 1: Pedigree chart (arrow: patient, black: affected)



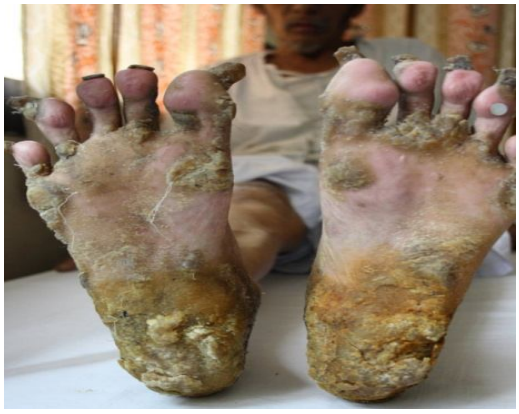


Figure 1: hyperkeratotic plaques of the skin at the bony prominences

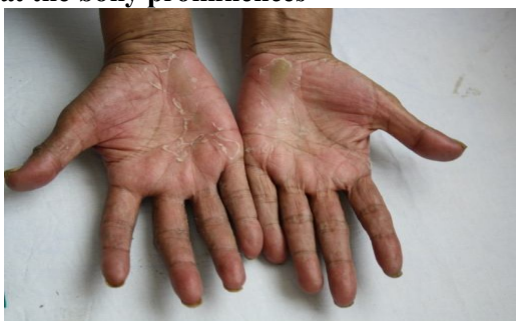


Figure 2: hyperkeratosis and scaling of palms

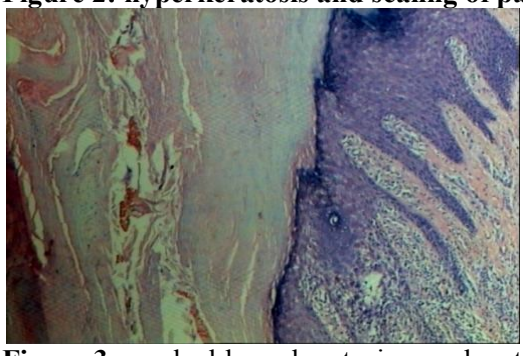


Figure 3: marked hyperkeratosis, parakeratosis, hypergranulosis, irregular elongation of rete rigdes and dense lymphocytic infilterate in upper dermis & papillary dermis

Discussion

Watcher's palmoplantar keratoderma also called as focal keratoderma or Palmoplantar keratoderma varians is an autosomal dominant keratoderma. It principally involves soles but hands may show callous formation. Sometimes knees and elbows are involved (3). Nails and hairs are invariably involved. It presents as compact masses of keratin developing at the site of friction on feet although also on palms and other sites. Mutations in the third K6 gene have been implicated as causal in focal palmoplantar keratoderma (4).

The pathogenesis of PPK remains unknown. The treatment is purely symptomatic; there is no definitive treatment or cure. Treatment modalities include topical and systemic therapy as well as surgical excision. Topical therapy has topical retinoids, corticosteroids, calcipotriol, or topical keratolytics such as 5% to 10% salicylic acid ointment, 30% propylene glycol, 20% to 30% lactic acid, and 10% to 12% urea ointment. Keratolytic agents may be useful in reducing the thickness of the keratoderma, but the lesions recur when treatment is stopped. Overall, the outcomes of treatment of PPK have been rather disappointing. Systemic treatment with oral retinoids, specifically isotretinoin, has been reported in some cases of PPK. However, there are significant risks and toxicities associated with long-term oral retinoid therapy; and like keratolytic agents, discontinuance of therapy causes the lesions to recur to their initial severity (5).

Conclusion

A rare and unusual case is presented where family history is important and also other system are to be examined to rule out other related syndrome.

Conflict of Interest: *None declared*
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