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Case Report

Unna Thost syndrome: a case report

Geetha M.[1]*, Rajeswari Bhat[2], Kiran B.[2], Madhusmitha Jena[3], Libni D. Angel[1]

[1]Department of Paediatrics, [2]Department of Dermatology, [3]Department of Pathology, East Point College of Medical Sciences and Research Centre, Bangalore, Karnataka, India

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***Correspondence:**
Dr. Geetha M.,
E-mail: gee_festoon@yahoo.co.in

ABSTRACT

Unna thost syndrome is Palmo Plantar Keratoderma (PPK) of diffuse non epidermolytic type inherited in autosomal dominant fashion. Authors had a seven-year-old boy born to second degree consanguineous parents had palmoplantar keratoderma with hyperhidrosis with positive family history. He was promptly diagnosed and treated with acitretin and showed improvement.

Keywords: Hyperhidrosis, Epidermolysis, Palmo plantar keratoderma, Unna Thost Syndrome, Vorner

INTRODUCTION

Skin lesions are common presenting complaints of children with causes ranging from the common to bizarre, innoxious to lethal, with visual identification many times being diagnostic. Authors present one such example. Palmoplantar keratodermas (PPKs) is a heterogeneous entity where some forms occur with specific clinical, pathological and genetic features.[1] Unna Thost is diffuse non-epidermolytic type inherited in an autosomally dominant manner, without associated organ involvement. The disease develops with hyperkeratosis of palms and soles in infancy, is evident by early childhood and persists throughout life.[2]

CASE REPORT

A seven-year-old boy born to second degree consanguineous parents presented with dry scaly lesions in the palms and soles noted since early infancy. Hyperhidrosis had been noted for one year. There was a family history of hyperkeratotic palms in the elder sibling. Other family members were normal. On examination, there was diffuse non-trans-gradient thickening thick yellow keratoderma with sharp cut off at wrist along with hyperhidrosis (Figure 1). A provisional diagnosis of Palmoplantar Keratoderma with likely Unna Thost Syndrome was made. The diagnosis was confirmed by skin biopsy. Histopathological examination revealed extensive non-epidermolytic hyperkeratosis, hyper-granulosis with focal vacuolar degeneration of the spinous layer (Figure 2). He was managed with oral acitretin along with topical emollients after relevant blood investigations. He showed rapid improvement in two months (Figure 3).

There is reduction of hyperkeratosis, healing of cracks and no hyperhidrosis. There is smoothening of the skin and no fissures. There is reduction of ichthyosis and no signs of inflammation (Figure 3).

DISCUSSION

PPK though with the spectacular and often alarming presentation is a heterogeneous benign indolent entity.

Unna first described a family with this peculiar syndrome in 1880.[3] Today it is known that it follows autosomal dominant inheritance with a mutation in the keratin 1 and 9 genes, with almost no difference in the non-epidermolytic Unna Thost and epidermolytic type of keratoderma of Vorner.[4]

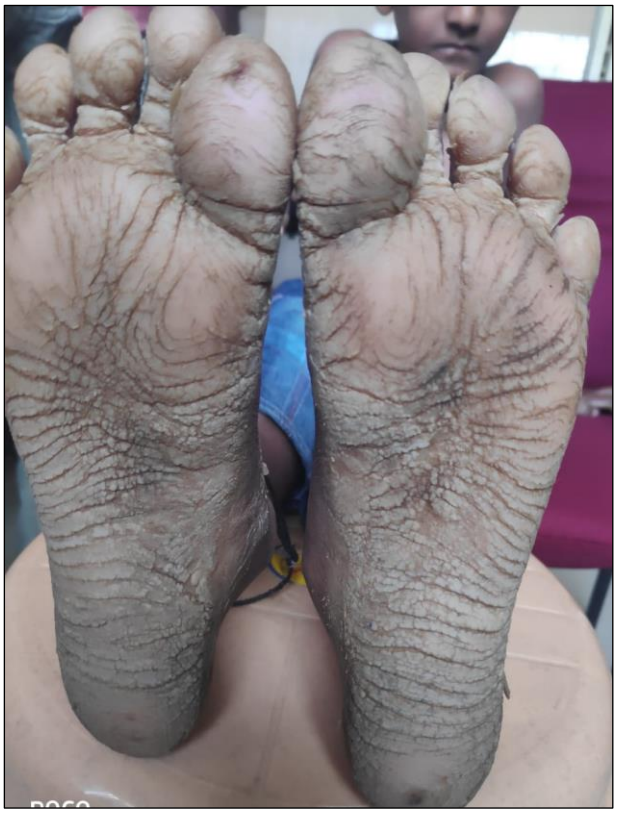

Figure 1: Clinical photograph of the boy showing symmetric hyperkeratosis involving both palms and soles.

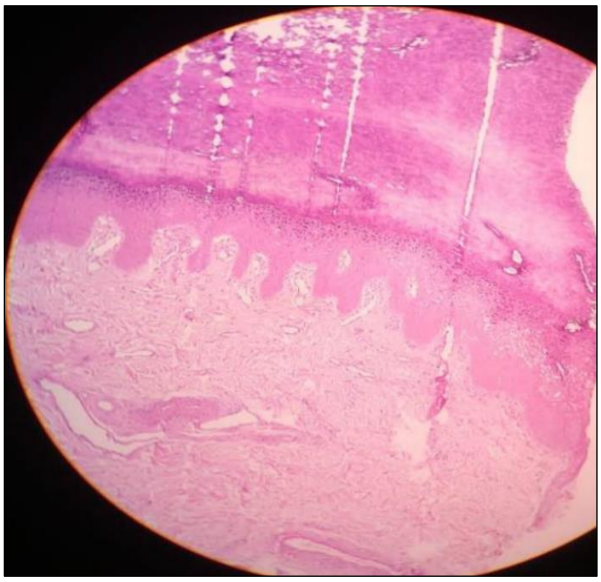

Figure 2: Histopathological picture showing hyperkeratosis, hypergranulosis with rete pegs.

Unna-Thost PPK is a hereditary form of palmoplantar keratoderma. Osvaldo Costa in 1962, considered it to be the main representative of all types of palmoplantar keratosis. Thost in 1880 described it as a form of palmoplantar ichthyosis. In 1883, Unna classified it as a variant of keratoderma.[5] Isolated cases of the precocious or late-onset emergence of the disease, which may be related to genetic factors, such as spontaneous mutation, reduced expression, and incomplete penetrance were described by Croatia.[6,7] Researchers observed alterations present in the filaments of keratin type II gene (codified as 12q11-13) after family screening.

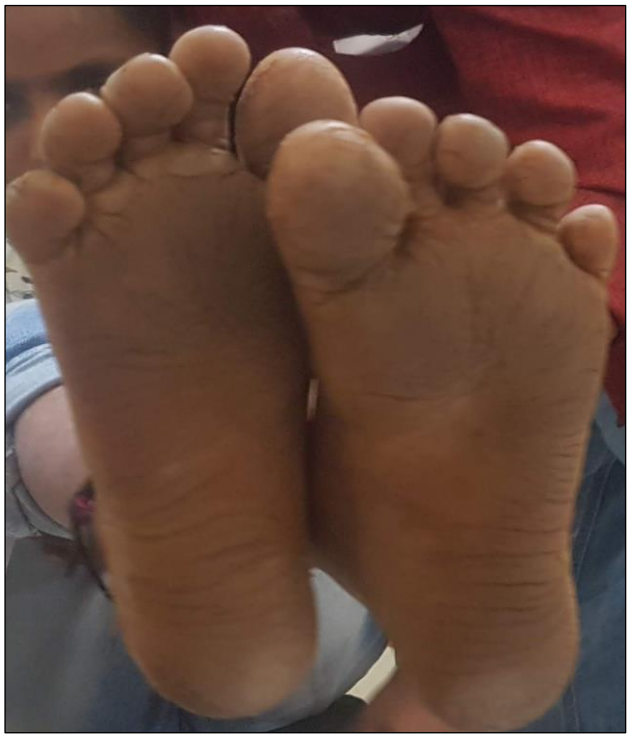

Figure 3: Follow up photograph of the same boy after treatment with acitretin.

Clinically, Unna-Thost PPK is characterized by symmetrical keratosis circumscribed to the palms and soles.[8] Absence of pruritus can be attested, and palmoplantar hyperhidrosis is a frequent symptom. When there is no such symptom, the disease leads to hardening and drying of the corneal layer, which ends up producing grooves and deep fissures.[9,10]

When intense, the keratosis may reach the lateral edges of the feet and hands. In some cases, at the shifting point between the diseased skin and healthy skin, a red-bluish colored band can be verified as beginning at the outer edge of the palms and quickly reaching the center and inner sides of the fingers and toes. Still, these are not obligatory signs.[11,12] This disease typically presents in infancy and is conspicuous by years of age, usually associated with hyperhidrosis.[13] It has distinct points on histological examination with the feature of non-epidermolytic hyperkeratosis believed to be the bedrock

of diagnosis from other forms of palmoplantar keratoderma, though this has now been disproven.[14,15]

The onset of the disease virtually always takes place during the first year of life. It may also appear in adolescence and even in adulthood. There is no predilection for race, though incidence is slightly higher in men.[16] The disease is related to the influence of climatic factors. Worsening of the clinical condition or increasing relapses occurs during the cold periods of the year.[17] The histopathologic alterations in Unna-Thost PPK are non-specific. They typically lack epidermolysis, which is an important factor of differentiation from Vorner's epidermolytic palmoplantar keratoderma. Despite its hereditary pattern and the similar clinical alterations to Unna-Thost PPK, the latter keratoderma shows typical histopathologic alterations, such as vacuolar alterations of the granular layer.[18]

Numerous therapies have been tried with less than satisfactory results with the disease being incurable and lasting lifelong. The mainstay of treatment of palmoplantar keratodermas is aromatic retinoids.[19] Previously topical keratolytic agents were used with no satisfactory results on the long term.

Systemic retinoids, such as etretinate and recently, acitretinate, have been used on severe and diffuse forms of palmoplantar keratodermas. In various cases, it has proved to have a good therapeutic response in spite of it presenting some adverse effects, such as increased cutaneous fragility and sensitivity, which has contributed to discontinuing treatment in many cases. At this point in time, genic therapy remains only a theoretical perspective due to the extreme difficulty of repairing the transformed allele.[20]

CONCLUSION

Unna Thost Palmo Plantar Keratoderma is a rare disorder of early childhood with distinct clinicopathological features caused due to mutations in the keratin gene. Thus, skin manifestations may hold the key to disease diagnosis and may unlock the path to prevention and gene therapy.

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