International Journal of Advances in Medicine
Dhillon KS et al. Int J Adv Med. 2015 Aug;2(3):288-290
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: http://dx.doi.org/10.18203/2349-3933.ijam20150564

Case Report

Unna-Thost type of palmoplantar keratoderma: a case report

K. S. Dhillon*, Vidhi Agarwal, Tarunveer Singh, Deepak Sharma, Areeba Khan, Ritika Srivastava, Sweksha Srivastava, Soni Yadav

Department of Dermatology, Era's Lucknow Medical College, Lucknow, UP, India

Received: 16 May 2015
Accepted: 19 June 2015

***Correspondence:**
Dr. K. S. Dhillon,
E-mail: kanwarjit29@yahoo.co.in

ABSTRACT

Non-epidermolytic variety of hereditary palmoplantar keratoderma with autosomal dominant inheritance characterised by uniform, thick, yellow hyperkeratosis over palms and soles was first described separately by Thost and Unna and named as Unna-Thost Keratoderma. We present a case of Unna-Thost keratoderma in a 9 year old male child who presented with waxy, thick, yellow palms and soles with fissures and contractures in hands.

Keywords: Unna-Thost, Palmoplantar keratoderma

INTRODUCTION

Unna-Thost keratoderma is an autosomal dominant inherited disorder of keratinization, with mutation in Keratin 1 gene.[1] The eponym refers specifically to non-epidermolytic keratoderma, although the original Thost family is now known to have had epidermolytic keratoderma.[2] The condition usually presents in the first few months of life and is usually obvious by the age of 4 years. It rarely appears in the third decade. An even, thick, yellow hyperkeratosis occurs over the whole of the foot, starting on the heel and anterior arch, spreading later to the palms with a sharp demarcation at the wrist, sparing the dorsal surfaces of hand and feet (non-transgredient). The margins show a reddish border with a waxy consistency. Hyperhidrosis is common and dermatophytic infections and pitted keratolysis are frequent but there is no systemic involvement.[3,4] Diagnosis is mainly clinical and prognosis is poor as condition persists for a lifetime. Various treatment modalities have been tried for the condition with unsatisfactory results and it is paramount to monitor the results of treatment by liver function test, kidney function test and lipid profile in addition to clinical examination.[5]

CASE REPORT

A 9 year old boy presented to our department with one year history of uniform thickening of both palms (Figure 1) and soles (Figure 2) with a reddish appearance and waxy consistency. This was associated with diffuse fissures on the palms and contractures in both hands since 3 months. As stated by the patient, he experienced uniform thickening of both soles since one year with gradual reddening and fissuring spread over next two months. Subsequently, after a period of around one month off involvement of the feet, both his palms were also involved with similar features. Later patient experienced difficulty in movement of digits and contractures in hands (Figure 3) developing over a period of about 7 months. There was no history of excessive sweating or any significant complaint suggestive of organ involvement. No similar complaints in the family. Personal history was unremarkable.

On examination, well demarcated, non transgredient hyperkeratosis of both palms and soles (Figure 1 and 2) was seen with waxy consistency, presence of scales and multiple fissures. It had sharp erythematous cut off borders limited till ankle and wrist.

There was conspicuous sparing of the dorsal aspect of hands and feet. Flexion at the distal interphalangeal joints of all the fingers was present (Figure 3). There was absence of infective foci on the lesions, pitted keratolysis or hyperhydrosis. The oral mucosa, nails and teeth were normal. All routine investigations were within normal limit. He is being treated with keratolytics including 6% salicylic acid with urea and oral isotretinoin 10 mg/day.

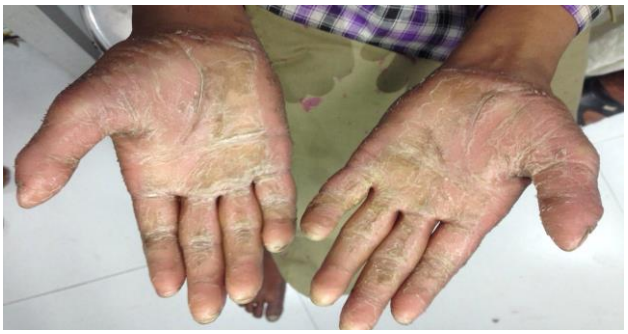

Figure 1: Diffuse, waxy keratoderma of palms with fissures.

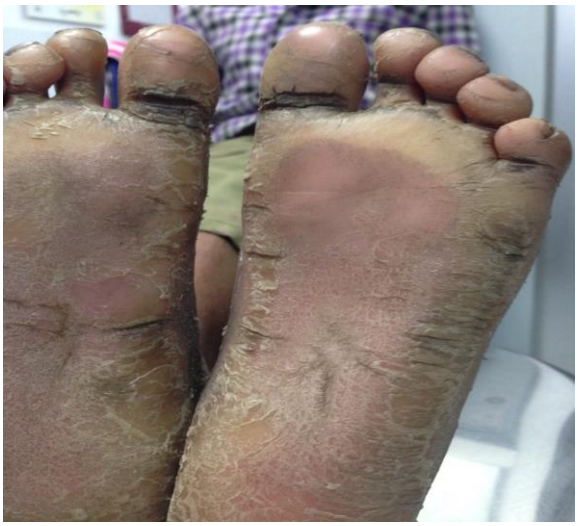

Figure 2: Waxy plantar keratoderma with scaling and fissures.

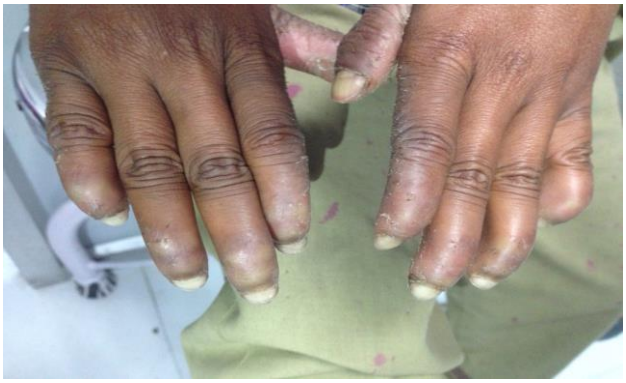

Figure 3: Flexion at the distal interphalangeal joints of all fingers with scaling.

DISCUSSION

As it is already known, the Unna–Thost PPK follows autosomal dominant inheritance with a mutation in the keratin 1 and 9 genes.[6] Recently, it has been shown that epidermolytic PPK due to keratin 9 mutation can lead to digital mutilation. The Unna-Thost form of PPK is clinically identical to localized epidermolytic hyperkeratosis of the palms and soles as described by Vorner.[7] Although they were earlier thought to be histopathologically distinguishable, reappraisal of the kindred originally analyzed by Thost[8] and study of a descendant supported separate identities for the two conditions.[9] By reinvestigation of the family originally seen by Thost[8] in 1880, the features of epidermolytic hyperkeratosis were found histologically, confirming the diagnosis PPK by Vorner.[9] This proves the identity of PPK type Thost with PPK of Vorner. Also, it should be noted that both epidermolytic and nonepidermolytic forms of PPK have been observed with various mutations in the keratin 1 gene.[10] Indeed, mutations of the KRT1 and KRT9 genes that are associated with the epidermolytic form of PPK affect the central regions of the protein that are important for filament assembly and stability, and for that reason lead to cellular degeneration or disruption.[10,11] On the other hand, the mutation of the KRT1 gene that was found in association with nonepidermolytic PPK involved the amino-terminal variable end region, which may be involved in supramolecular interactions of keratin filaments rather than stability.[12] A recent molecular analysis gives further evidence that PPK of Vorner represents the same entity as PPK of Thost.[10]

A recent re-evaluation of Thost's original family revealed that the condition was caused by a similar mutation, R162W, in the same segment of KRT9 as in epidermolytic PPK. Studies[13,14] demonstrated substitution of tryptophan for arginine-162 as the result of a mutation in exon 1 of the KRT9 gene in large German epidermolytic PPK kindred and in five other apparently unrelated families. The R162W mutation was seen in one of the five families originally described by Thost.[9] This is convincing evidence that the disorder previously listed as a separate entity in these catalogs is in fact the same as epidermolytic hyperkeratosis.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Kimyai-Asadi, A'Kotcher LB, Jih MH. The molecular basis of hereditary palmoplantar keratodermas. J Am Azad Dermatol.2002;47;327-43.
2. Kuster W, Becker A. Indication for the identity of palmoplantar keratoderma Unna Thost with type Vörner. Acta Derm Venereol (Stockh) 1992;72:120–2.

3. Kansky A, Durinovic-Belló I, de Jongh BM et al. HLA antigens in Yugoslav patients with palmoplantar keratoderma, type Unna–Thost: a family study. Acta Derm Venereol (Stockh). 1982;62:313–6.
4. Gamborg-Nielsen P. Two different clinical and genetic forms of hereditary palmoplantar keratoderma in the northernmost county of Sweden. Clin Genet. 1985;28:361–6.
5. Kanitakis J, Tsoitis G, Kanitakis C. Hereditary epidermolytic palmoplantar keratoderma (Vorner type). Report of a familial case and review of the literature. J Am Acad Dermatol. 1987;17:414-22.
6. Umegaki N, Nakano O, Tamai K, et al. Vörner type palmoplantar keratoderma: novel KRT9 mutation associated with knuckle pad-like lesions and recurrent mutation causing digital mutilation. Br J Dermatol. 2011;165:199–201.
7. Vorner H. Zur Kenntniss des Keratoma hereditarium palmare et plantare. Arch Derm Syph. 1901;56:3-31.
8. Thost A. Ueber Erbliche Ichthyosis Palmaris et Plantaris Cornea. Dissertation. Heidelberg, Germany: University of Heidelberg; 1880.
9. Kuster W, Becker A. Indication for the identity of palmoplantar keratoderma type Unna-Thost with type Vorner. Thost's family revisited 110 years later. Acta Derm Venereol. 1992;72:120-2.
10. Kanitakis J, Tsoitis G, Kanitakis C. Hereditary epidermolytic palmoplantar keratoderma (Vorner type). Report of a familial case and review of the literature. J Am Acad Dermatol. 1987;17:414-22.
11. Irvine AD, Paller AS. Molecular genetics of the inherited disorders of cornification: an update. Curr Probl Dermatol.2002;14:84-6.
12. Lucker GP, van de Kerkhof PC, Steijlen PM. Topical calcipotriol in the treatment of epidermolytic palmoplantar keratoderma of Vorner. Br J Dermatol. 1994;130:543-5.
13. Hennies HC, Kuster W, Langbein L, et al. Keratin 9 gene mutations in epidermolytic palmoplantar keratoderma (EPPK). Am J Hum Genet. 1993;53:A211.
14. Reis A, Hennies HC, Langbein L, et al. Keratin 9 gene mutations in epidermolytic palmoplantar keratoderma (EPPK). Nat Genet. 1994;6:174-9.

Cite this article as: Dhillon KS, Agarwal V, Singh T, Sharma D, Khan A, Srivastava R, Srivastava S, Yadav S. Unna-Thost type of palmoplantar keratoderma: a case report. Int J Adv Med 2015;2:288-90.