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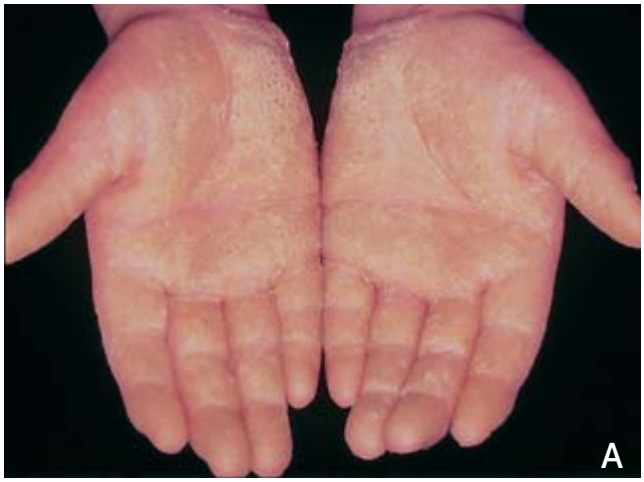
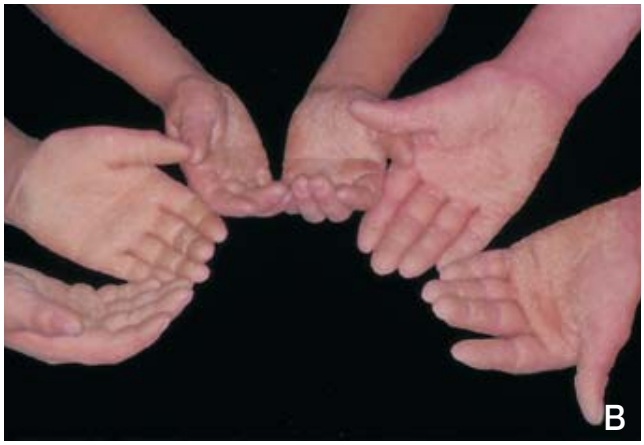


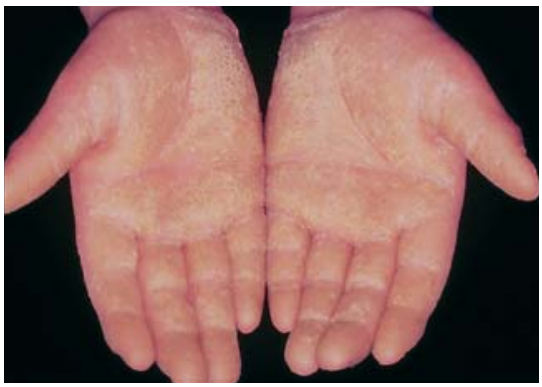


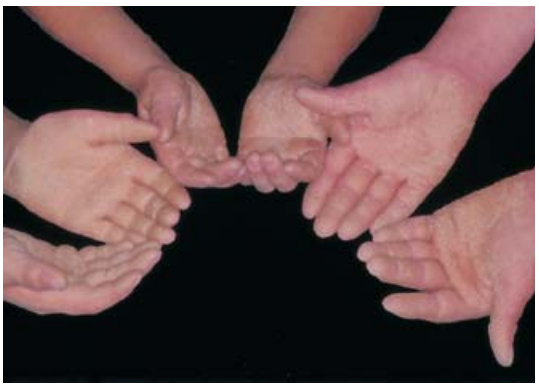
This patient (A) complains of thick palmar skin. His father and many other members of his family (B) have a similar condition.

What is your diagnosis?

PLEASE TURN TO PAGE 95 FOR DISCUSSION

The Diagnosis: Inherited Palmoplantar Keratoderma (Unna-Thost Disease)





Our patient presents with a classic example of the most common type of tylosis, hereditary diffuse palmoplantar keratoderma (PPK), also known as Unna-Thost disease. Because this condition is inherited in an autosomal dominant fashion, it is not surprising that the patient's father is similarly affected.[1] Presentation always occurs within the first 3 years of life, usually within the first few months.[1,2] Palms and soles show a bilaterally symmetrical and diffuse, smooth, white to yellow hyperkeratosis, often with indistinct erythematous margins, and there is no transgrediens (ie, no extension onto the dorsal surfaces).[1] Hyperhidrosis and dermatophytosis are common findings that lead to a foul odor. Desquamation and fissures can be painful, and treatment is supportive for this lifelong condition. Emollients are the essential mainstay of therapy. Preparations containing the alpha hydroxy acids can be of some benefit. Topical retinoids are generally ineffective, and although oral retinoids may help, long-term toxicity limits their use. Regular daily manual abrasion with pumice stones or sand paper is often helpful.[2]

When evaluating a patient with PPK, it is useful to categorize the disorder based on inheritance pattern, age of onset, clinical morphology, and histopathology. In 1901, Vörner[3] first described PPK with epidermolytic hyperkeratosis (granular degeneration in the malpighian layer), which typifies the cases now bearing his name. It has been suggested that Thost's original kindred manifested this histologic pattern and that the name should be changed to Unna-Vörner.[1] We now know that the underlying defect in Vörner's pattern of PPK is in keratin 9.[1] The majority of cases demonstrate the expected tonofilamentous cytoskeleton, but at least one has been described with tonotubular keratin.[4]

There are a large number of PPKs, and the entire scope cannot be adequately covered in this forum. For comparison with our cases, we include a discussion of the most similar entities.

Vohwinkel's syndrome, or keratoderma hereditarium mutilans, is inherited as an autosomal dominant trait, but it is quite rare.[1] Onset is also in infancy, and a loricrin defect is causative.[2] The hyperkeratosis is

clinically "honeycombed" rather than smooth, and "starfish" keratoses form on the dorsal hands, feet, elbows, and knees. Constricting fibrous bands later develop circumferentially around digits (pseudoainhum) resulting in autoamputation.[5] Scarring alopecia and high frequency hearing loss also occur. Oral retinoids can be helpful, but hand surgeons are often needed to save digits threatened by autoamputation.[1]

Howell-Evans' syndrome also is inherited in an autosomal dominant fashion, but onset is in the adolescent or early adult years. Clinically, the PPK in this syndrome is quite smooth, diffuse, symmetric, and non-transgrediens. There is a clear association with esophageal carcinoma after the third decade, and oral leukoplakia has been described in these patients. Fortunately, this type of PPK is rare.[1]

Punctate (spiny) keratoderma is an autosomal dominant type of PPK that usually presents after puberty (age 12–70 years at first onset). Lesions are described as resembling "music box spines," and some cases improve with topical application of 5-fluorouracil.[6]

Huriez syndrome can be a very mild PPK, associated with sclerodactyly (without Raynaud's phenomenon) and with atrophic skin of the dorsal hands, prone to squamous cell carcinoma. In this autosomal dominant condition, there is often hypohidrosis and nail changes such as clubbing, aplasia, and ridging.[7]

Mal de Meleda PPK is named for the Adriatic island where it was first described in an inbred population.[1] Inheritance is autosomal recessive, and there is a stocking and glove distribution (with transgrediens) of scaly red lichenification.[8]

Papillon-Lèfevre syndrome is an autosomal recessive PPK that shows transgrediens, but these patients also have periodontosis, as well as calcified dura at the tentorium and choroid attachments. Systemic retinoids have been reported to improve the dental disease and PPK.[1]

Richner-Hanhart syndrome also is known as tyrosinemia type II, a hepatic enzyme deficiency in which tyrosine accumulates in all tissues. An autosomal recessive PPK with painful, focal, or diffuse keratoses at weight-bearing sites (sometimes with erythema, bullae, and erosions), this syndrome includes corneal findings of pseudoherpetic (dendritic) keratitis caused by inflammation from the tyrosine crystals.[9]

Our patient's clinical appearance, age of onset, autosomal dominant inheritance and lack of associated abnormalities are typical of the Unna-Thost variant of PPK.

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