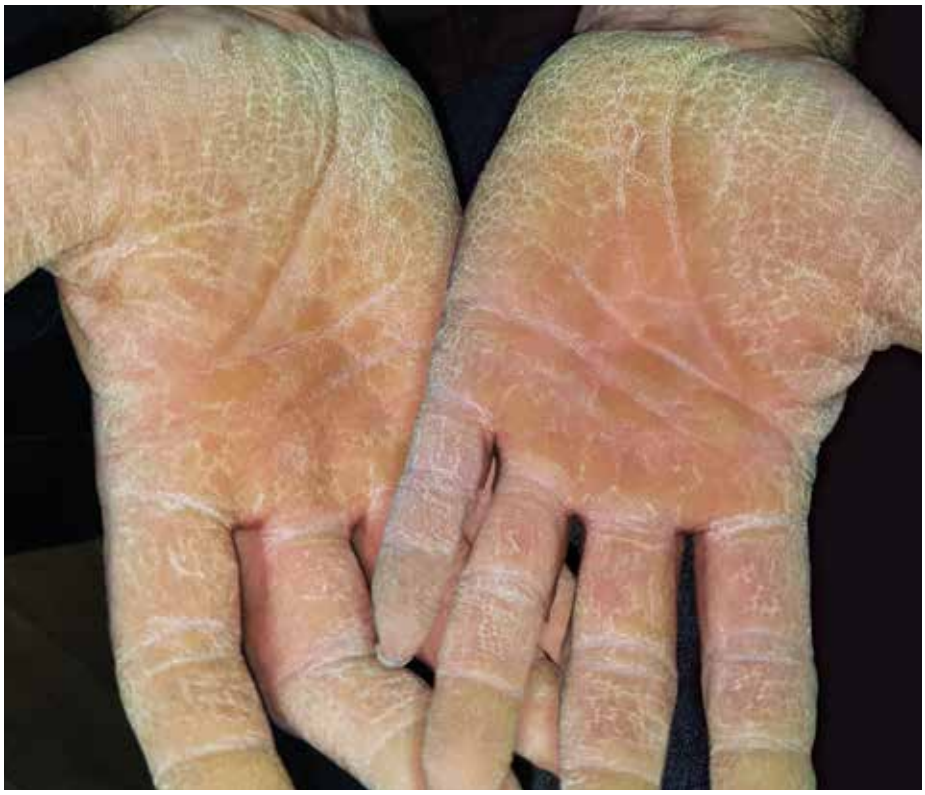
Man Is in Thick of Skin Problem

Since birth, a now 60-year-old man has been affected by the same dermatologic condition as several members of his mother's family. As he has aged, the problem has become more intrusive: His hands and feet are rough and painful, and he experiences loss of sensation, particularly on his soles.



Over the years, he has been evaluated in multiple venues, both private practices and medical schools, by a variety of providers, including pediatricians and dermatologists. Many treatments have been tried; few have produced any good effect. Still seeking an explanation for his condition, he self-refers to dermatology.

Evaluation reveals that the surfaces of both feet and hands are uniformly covered by rough and, in some focal areas, waxy hyperkeratotic skin. The nails, hair, teeth, and dorsal surfaces of the extremities are unaffected. The patient's affect and level of intelligence appear well within normal limits.

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Of the following, what is the term generally used to refer to this type of problem?

a) Palmoplantar psoriasis
b) Xerosis
c) Palmoplantar hyperkeratosis
d) Perineoplastic hyperkeratosis

ANSWER

The correct answer is palmoplantar hyperkeratosis (PPK; choice "c"). As a category, it includes many conditions that are characterized by a heterogeneous thickening of the palms and soles, although each has distinct features.

Psoriasis (choice "a") can present in somewhat similar fashion. However, it rarely appears so early in life and almost never involves such uniform hyperkeratosis.

Xerosis (choice "b") merely means dry skin—a common enough problem, but one that does not involve such uniform and deep hyperkeratosis. It is almost never congenital.

Occult cancers (eg, GI, breast) can occasionally trigger a hyperkeratotic "perineoplastic" reaction (choice "d") in the palms, soles, and other areas. However, it is acquired relatively late in life.

DISCUSSION

PPK is a relatively common problem, especially in those of northern European ancestry. The incidence in Northern Ireland, for example, is 4.4/100,000. In its more severe forms, such as this case, it can be debilitating.

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Categorization of this extraordinarily diverse group of disorders is a challenge, to say the least. For purposes of this discussion, let's start with this patient's condition and then clarify its place in the panoply of hyperkeratotic disorders.

Our patient has one of the more common forms of diffuse PPK, generically called *nonepidermolytic PPK*, which presents with a waxy, uniform yellowed hyperkeratosis confined to the palms and soles. The old eponymic term for it was *Unna-Thost disorder*, named for the Viennese and Norwegian dermatologists who first described it late in the 19th century. A more modern term is *tylosis*, but many variations exist. This particular form is inherited in autosomal dominant fashion, but other forms of PPK can be transmitted by autosomal recessive or even X-linked genes.

Other types include focal (thick calluses over points of friction, especially on the feet) versions that can present in linear configuration and punctate forms with deep calluses on palms and soles that can be discrete or widespread, with lesions that range in size from pinpoint ("speculated") to pea-sized.

New-onset PPK should prompt a search for an occult malignancy. In terms of establishing a firm diagnosis, punch biopsy can be performed for clarification when necessary.

As might be expected, treatment is difficult at best. Urea or salicylic acid–based topical preparations can help to thin it out. This patient was started on acitretin, an oral retinoid (artificial form of vitamin A) that may help; it was one of the few treatments he hadn't already tried. A one-month course of terbinafine had already been tried, without success, just in case there was any secondary fungal involvement—a fairly common complication of PPK. **CR**