Keratosis Palmaris ET Plantaris

Three major autosomal dominant forms and two autosomal recessive forms of keratosis palmaris et plantaris have been discovered. The three dominantly inherited forms include the following:
Palmoplantar keratoderm = اﻠاﺨﻤﺼﻲ اﻠراﺤﻲ اﻠﺠﻠدﻲ اﻠﺘﻘرﻦ

1. Keratosis palmaris et plantaris of Unna-Thost showing either diffuse or localized, occasionally linear hyperkeratosis of the palms and soles {Fig. 6-6}. A division of the condition into two types, based on the sites involved, one affecting the hands and wrists and the other the feet, the ankles and wrists, and the elbows and knees-is not tenable, because both types may occur in the same family.
Palmoplantar keratoderma = اﻠاﺨﻤﺼﻲ اﻠراﺤﻲ اﻠﺠﻠدﻲ اﻠﺘﻘرﻦ

2. Epidermolytic keratosis palmaris et plantaris, although clinically indistinguishable from the Unna-Thost type, histologically shows epidermolytic hyperkeratosis. This type is caused by a mutation in keratin type 9 localized within the keratin gene cluster on chromosome 17q12-q21 and keratin type 1 on chromosome 12q13.

3. Keratosis palmoplantaris punctata (or papulosa) has multiple keratotic plugs.

The two recessively inherited forms include the following:
1. *Keratosis palmaris et plantaris* of the Meleda type, showing diffuse involvement of the palms and soles and a marked tendency toward progression to the dorsa of the hands and feet, the ankles and wrists, and the elbows and knees [67].

2. The *Papillon-Lefèvre syndrome* shows the clinical characteristics of the Meleda type in association with periodontosis resulting in the loss first of teeth and subsequently of bone. Mutations of the *ERCP1* gene on chromosome 11q14.1 have been found as well as keratins 6b, 9, 16, 17 on 12q13 and 17q12-q21 and connexin 30 on 13q12.

In addition, keratosis palmaris et plantaris occurs in three syndromes: {a} pachyonychia congenita, {b} hidrotic ectodermal dysplasia, and {c} the Richner-Hanhart syndrome associated with tyrosinemia (tyrosine tyrosinase on 12q24).
16q22, 1-q22.3) (69).

*Histopathology*

In keratosis palmaris et plantaris of the Unna-Thost type and the Meleda type, a
In epidermolytic keratosis palmaris et plantaris, the histologic picture is identical with that seen in epidermolytic hyperkeratosis. There is massive hyperkeratosis with rupture of the superficial epidermal cells, and scattered cavities are present as a result of ruptured cell walls. Keratohyaline granules are numerous and large.

In keratosis palmoplantaris punctata, there is massive hyperkeratosis over a sharply limited area, with depressed irregular keratotic papules with central keratotic plugs.
Palmoplantar keratoderma = اﻠاﺨﻤﺼﻲ اﻠراﺤﻲ اﻠﺠﻠدﻲ اﻠﺘﻘرﻦ