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:

Keratosis Palmaris ET Plantaris
Palmoplantar keratoderma = اﻠاﺨﻤﺼﻲ اﻠراﺤﻲ اﻠﺠﻠدﻲ اﻠﺘﻘرﻦ

1. *Keratosi 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 this disease into two types—palmar and plantar—is not tenable, because both types may occur in the same family.
2. *Epidermolytic keratosis palmaris et plantaris*, although clinically indistinguishable from the Unna-Thost type, histologically shows epidermolytic hyperkeratosis. This condition affects keratin type 9 localized within the keratin gene cluster on chromosome 17q12-q21 and keratin type 1 on chromosome 12q13.

3. *Keratosis palmoplantaris punctata (papulosa)* has multiple keratotic plugs. The two recessively inherited forms include the following:

The two recessively inherited forms include the following:
1. **Keratosis palmaris et plantaris** (the Meleda type)

   Keratosis palmaris et plantaris of the Meleda type shows 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.

2. The **Papillon-Lefèvre syndrome** shows...
Histopathology

In keratosis palmaris et plantaris of the Unna-Thost type and the Meleda type, a sparse inflammatory infiltrate of lymphocytes in the upper dermis.
In epidermolytic keratosis palmaris et plantaris, the histologic picture is identical with that seen in epidermolytic keratosis elsewhere. Keratinocytes are absent in the superficial epidermis, 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 depression of the surface and formation of punctate keratoses. The histologic picture is similar to that of epidermolytic keratosis.
Palmoplantar keratoderma