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We hope that making available the relevant information on Pachyonychia Congenita will be a means of furthering research to find effective therapies and a cure for PC.
Diagnosis: Angiokeratosis naeviformis (see K. Dammert, Dermatologica 130: 17 [1965]).

14. J. D. Woerdeman-Evenhuis, Amsterdam

Angiokeratosis naeviformis

The patient is a young woman, 22 years old, who observed in February, 1964, a bright red, small papule, almost pea-sized, on the left anterior part of her waist. This small tumour was excised (Dr. Stijn) and shortly afterwards (±10) analogous elevated efflorescences developed in the same area. Some of the lesions are slightly desquamating.

The lesions caused no complaints and the patient is in good general health.

Biopsy: The lesion is raised above the surface of the epidermis and consists of many thin-walled capillaries and a non-specific infiltrate consisting of lymphocytes and a few eosinophils. There is a slight hyperkeratosis.

Treatment: No treatment was given. After 9 months most lesions became skin-coloured and slightly shrunken. Only two of the small tumours are still red.

The diagnosis: angiokeratosis naeviformis was made (see K. Dammert, Dermatologica 130: 17, Fig. 4 and 6 [1965]).

15. W. P. de Groot, Amsterdam

Pachyonychia Congenita with Sebocystomatosis (Sertoli)

Male, 55 years old, was demonstrated in 1929 by Mendes da Costa as a case of pachyonychia congenita. Changes of all nails were noticed at or shortly after birth and have always remained unchanged.

Already in 1929 the patient suffered from hyperhidrosis palmarum et plantarum. His chief complaint is now the appearance of blisters after walking especially during warm weather. Besides the pachyonychia there is a localized hyperkeratosis plantarum (and a coincidental tinea pedis).

Hair: hair growth is scanty in the axillae, on the breast and in the pubic region.

Teeth: prosthesis. According to the patient his teeth were normal. Trunk, scalp, some fingers: sebocystomatosis (as described by Sertoli in a similar case).

Family: patient is one of a triplet, a sister died at birth, a brother is healthy and has none of the patients symptoms. As far as the patient knows, he has no relatives with this affection.


Lymphadenosis Cutis Benigna Bäfverstedt?

Woman, 25 years old, suffering for 4 years from a slowly progressive, painless, non-itching affection of the nose, consisting of a dark red, slightly elevated plaque (Fig. 4),

Biopsy: Infiltration of the cutis with lymphocytes and centres of reticulum cells, partly of epidermoid type. Systemic examination: essentially normal; liver, spleen,