

PALMOPLANTAR KERATODERMA OF PUNCTATE TYPE: ACROKERATOELASTOIDOSIS COSTA

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Abstract. A special type of punctate palmo-plantar keratoderma occurring in 10 patients from six Finnish families is described clinically, histologically and ultra-structurally. Eight of the patients were women. The patients had symptomless, slightly elevated, transparent, round or oval, hyperkeratotic papules, 2 to 5 mm in diameter, located at the edges of the palms and fingers, the entire palms and wrists, and at the edges of the soles. The clinical picture resembled acrokeratoelastoidosis Costa. Six of the patients also had knuckle pad-like lesions on the interphalangeal joints of the fingers and toes. Three of the patients had recalcitrant warts and no wart virus antibodies were found in their sera. The pedigrees of three families are presented and an autosomal dominant inheritance pattern is suggested. The histology of the lesions revealed undulating hyperkeratosis with slight depressions on the epidermis, which was otherwise normal. The dermis was of normal thickness and both the elastic and the collagen fibres seemed to be microscopically normal. Ultrastructurally, however, the elastic fibres in the deep dermis showed pathological alterations in some cases. In conclusion, we consider the condition to be acrokeratoelastoidosis Costa, a variant of hereditary palmo-plantar keratodermas.

Key words: Palmo-plantar keratoderma; Acrokeratoelastoidosis Costa; Viral warts; Ultrastructural findings

In 1953 Costa described a new variant of hereditary palmo-plantar keratoderma, which he called acrokeratoelastoidosis (3). The patient, an 18-year-old Brazilian woman, had hyperkeratotic papules on the skin of the palms and soles and small smooth patches over the interphalangeal joints of the fingers and toes. Epidermal hyperkeratosis, thickening of the dermis and fragmentation of dermal elastic fibres were demonstrated histologically. Subsequently, Costa presented similar skin symptoms in another Brazilian woman (4) and in three siblings (5). Most of the patients reported hitherto have been from South America. Only a few cases from Europe have been reported (2, 8, 11) and as far as we know none from Scandinavia. In 1973 Jung

presented a German family with 21 affected members in three generations (8). The pedigree of this family suggests an autosomal dominant mode of inheritance in this disease. Acrokeratoelastoidosis was classified with the group of hereditary palmo-plantar keratodermas by Costa (6) and later also by Greither (7) and Jung and his co-workers (9).

In the present paper we describe a clinical, histological and ultrastructural study of 10 patients with palmo-plantar keratoderma clinically similar to acrokeratoelastoidosis Costa.

MATERIALS AND METHODS

The patients were examined at the Department of Dermatology, University Central Hospital, Helsinki, Finland, during 1977-78. For light microscopy, the skin biopsies were cut and stained with hematoxylin and eosin, PAS, and Weigert resorcin-fuchsin for elastic tissue. For electron microscopy, additional specimens were fixed in 2.5% glutaraldehyde, postfixed in 1% osmium tetroxide, embedded in epoxy resin and cut with an LKB ultramicrotome. The sections were stained with lead citrate (Reynolds 1:10 NaOH) and 5% uranyl acetate and examined with a Jeol 100 S electron microscope.

At the time of the examination 3 of the patients had also viral warts; thus the sera of all patients were studied for wart virus antibodies, twice with a 1-2 month interval. The antibodies were measured with immunodiffusion tests carried out by a micromodification of gel double diffusion and complement fixation tests performed with microplates and 2 full units of complement as previously described for wart virus serology (12). In one relative with recalcitrant plantar warts, material was taken from the plantar lesion for studying the presence of virus under the electron microscope with the negative staining technique.

Clinical evaluation

The clinical details of the 10 patients are presented in Table 1. In all 10 patients the eruption was localized on the thenar and antithenar area of the palms and in 4 patients also on the entire palm and the inner surfaces of the fingers (Fig. 1). Three of the patients had lesions on the feet as well, mostly on the lateral and medial side of the feet. The eruption was composed of round or oval, slightly elevated, hyperkeratotic, transparent papules, 2-5 mm in



Fig. 1. Punctate, transparent, hyperkeratotic and coalescent lesions on the palm.

diameter, some of which had coalesced into plaques separated by furrows. In one patient the papules formed a diffuse hyperkeratotic plaque over the entire palm. Six of the patients had also knuckle pad-like smooth patches on the skin over the interphalangeal joints of the fingers and toes (Fig. 2). The lesions were symptomless, except in one case of itchy dermatophyte superinfection.

The patients did not have palmo-plantar hyperhidrosis or congenital abnormalities. They had no history of medication with arsenic. The wart virus antibodies were negative in all the patients, including the 3 cases of clinically typical viral warts.

Family study

The disorder occurred in three families. The mode of inheritance seems to be autosomal dominant with incomplete penetrance and variable expressivity in our series, as reported in most palmo-plantar keratodermas (7). The pedigrees are presented in Fig. 3. The patients are designated by the same initials as in the Table.

In family A, 2 members were affected, the mother (IA) and her son (PA). Both had similar punctate lesions on the hands, and also clinically typical viral warts. In 1976 the brother of IA (OS) had been treated at our clinic for prominent plantar warts, which had persisted for 30 years and failed to respond to any treatment. In electron microscopical examination, no virus particles were seen.

In family L, the mother (EL) and her daughter (LV) had numerous punctate lesions over the entire palms, the mother having them also on the soles. They both had knuckle pad-like lesions on the interphalangeal joints of the toes. Another daughter (UK) had only a single lesion.

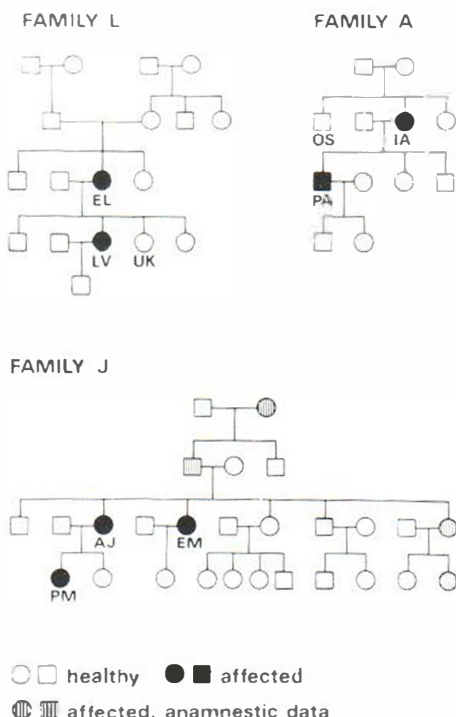


Fig. 3. Pedigrees of the three families with affected members in 2 to 4 generations.



Fig. 2. Small, hyperkeratotic papules and knuckle pad-like patches on the interphalangeal joints of the fingers.

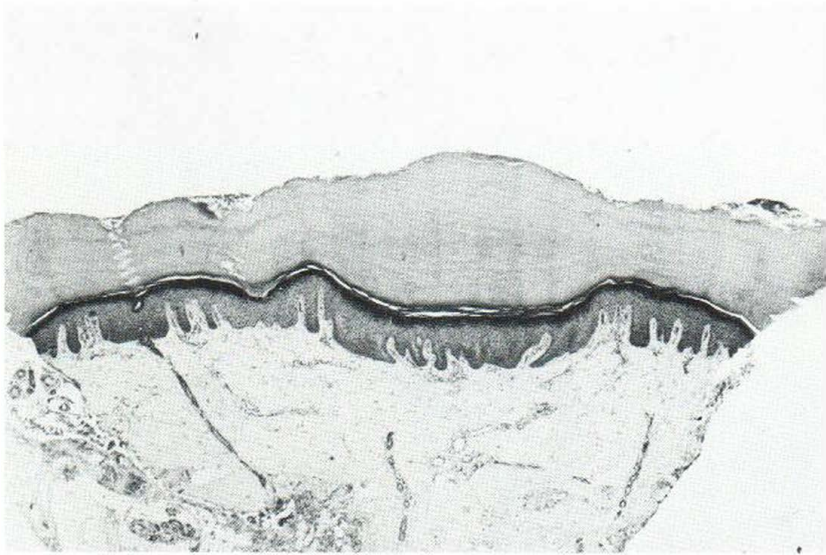


Fig. 4. Beaded band of hyperkeratosis (HE 12.5×10).

on her right thenar, the skin of the palms being otherwise normal.

In family J, 8 persons were affected. We were able to examine two sisters (AJ and EM) as well as a daughter (PJ) of one of them. Similar hyperkeratotic lesions had been reported also on the palms of the deceased father and his deceased mother. The younger of the two affected sisters (EM) had numerous confluent papules affecting the entire palm, the clinical picture greatly resembling the diffuse palmar keratoderma. Only on the thenar and antithenar area were separated papules seen. We were not able to examine the other affected siblings, who live abroad.

Histological evaluation

The skin biopsies were taken from a skin papule on the antithenar area of the palms. In light microscopy all the sections exhibited a marked hyperkeratosis embedded as a shallow pit in the epidermis (Fig. 4). There was no parakeratosis. The granular layer below the hyperkeratosis was prominent, with 4–6 cell layers. Otherwise

the epidermis was not altered. The basement membrane area stained normally with PAS and resorcin-fuchsin. The thickness of the dermis was normal and the collagen and elastic fibres were not altered when compared with the normal palmar skin.

Electron microscopical examination was performed in 7 cases. The epidermal layers did not show any abnormal morphological alterations. The superficial dermal collagen was composed of normal fibres mixed with well preserved, thin anchoring filaments abutting on the basement membrane. In some cases the elastic fibres in the reticular dermis were disaggregated and the microfibrils were fragmented (Fig. 5). However, these alterations were not found in every case.

DISCUSSION

The clinical findings in our patients greatly resemble those in acrokeratoelastoidosis Costa. Most of our patients were women, as also reported in other

Table 1. *Clinical findings of the ten patients*

Patient	Sex	Age	Age at onset	Localization of lesions	Knuckle pad lesions	Warts
1. IA	F	60	50	Entire palms and soles	—	On the hands
2. PA	M	32	22	Thenar and antithenar area	—	On the hands
3. EL	F	55	10	Entire palms and soles	On the toes	—
4. LV	F	32	10	Whole palms	On the toes	—
5. AJ	F	54	—	On the thenar and antithenar area	—	—
6. PJ	F	15	10	On the thenar and antithenar area	On the fingers	—
7. EM	F	49	10	Hyperkeratotic plaques on whole palms	On the fingers	—
8. OM	M	36	10	Thenar and antithenar area	—	On the soles
9. RAH	F	33	25	Thenar and antithenar area	On the toes	—
10. KR	F	64	64	Thenar and antithenar area	On the left hand	—

Cases 1 to 7 were from three families and 8 to 10 non-related.

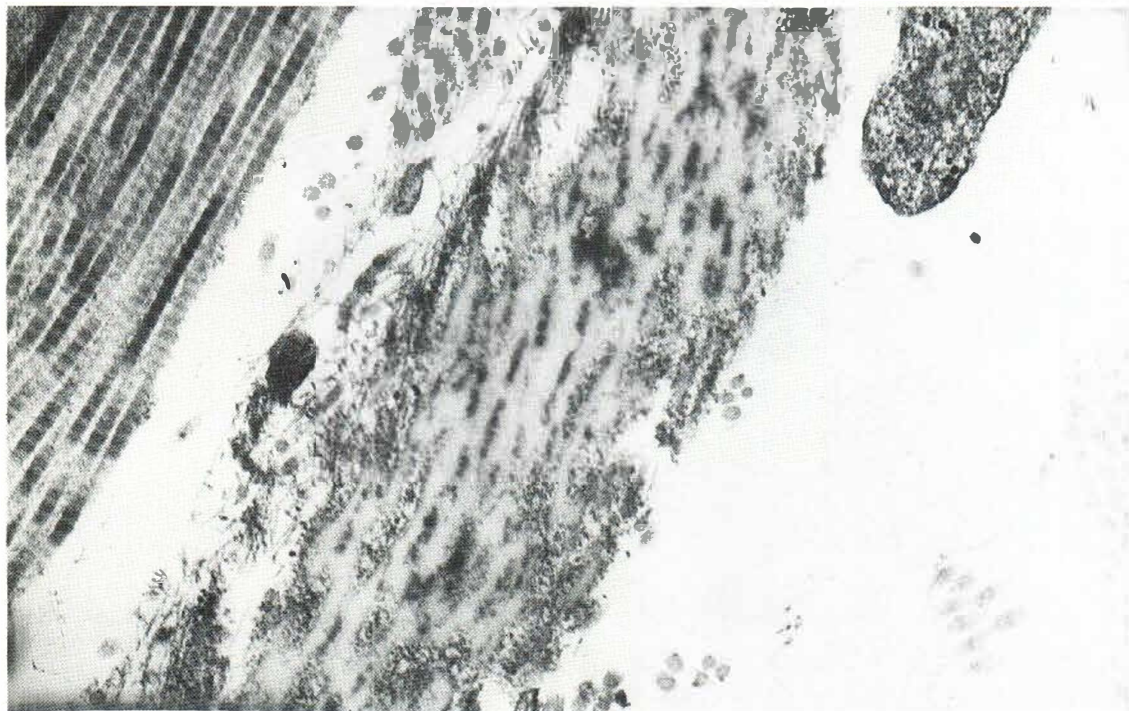


Fig. 5. In the reticular dermis the elastic fibre is disaggregated and contains granular electron-dense fragments.

The microfibrils at the periphery of the fibre are disarranged. Original magnification 20 000.

series (6, 8). The lesions had usually appeared already in childhood.

The histological diagnosis of acrokeratoelastoidosis Costa has been based on both epidermal and dermal alterations. In the epidermis, hyperkeratosis and thickening of the granular layer occur (3). The diagnostic features, however, appear in the dermis. According to Costa, the whole dermis is thick, the collagen fibres are homogenized, and the elastic fibres fragmented and sparse (3, 4, 5). Ultrastructural alterations have been reported in the elastic fibres and in the fibroblasts (9, 11), but not in the collagen fibres. The elastic microfibrils were considered to be scanty and fragmented and the fibroblasts were shown to contain dense granules in their cytoplasm, without extracellular elastic fibres (11). The authors considered these findings to be suggestive of a failure in the secretion of the elastic material.

The epidermal changes in our patients are similar to the findings in acrokeratoelastoidosis Costa. In light microscopy we were not able to find any alterations in the thickness of the dermis or in the morphology of the elastic and collagen fibres in our

patients. However, ultrastructural investigation revealed alterations in the elastic fibres in some (but not in all) cases. These alterations are similar to those reported earlier by Jung and his co-workers (9).

Confusingly, some authors have used the name acrokeratoelastoidosis to designate degenerative collagenous papules of the hands (14). This disease, mentioned in the literature also by the name of keratoelastoidosis marginalis (1, 10, 13) occurs mainly in elderly men at the juncture of the dorsal and palmar skin of the hands and represents localized solar elastosis of the dermis.

In family A both affected members suffered from recalcitrant viral warts, and a relative had been treated for prominent plantar warts or hyperkeratotic plaques, which clinically resembled warts. Wart virus particles, however, could not be found in the plantar material. The connection between viral warts and palmoplantar hyperkeratosis is interesting: either the hyperkeratotic epidermis is susceptible to the growth of wart virus, or the virus, importing its genome to the epidermal cells, may lead to changes with abnormal keratinization and growth of

the epidermis. The fact that the sera of the patients did not contain antibodies against wart virus, is in agreement with this hypothesis.

According to both the clinical and histopathological findings we regard the disease in our patients as acrokeratoelastoidosis Costa, an example of the very complex group of hereditary palmoplantar keratodermas. In this variant the hyperkeratosis appears mostly as punctate or papulous lesions, sometimes diffusely.

REFERENCES

1. Burks, J. W., Wise, L. J. & Clark, W. H.: Degenerative collagenous plaques of the hands. *Arch Dermatol* 82:362, 1960.
2. Civatte, J., Hinchy, M., Barranger, Ch., Vivier, O. & Degos, R.: Acrokérato-Élastoïdose. *Ann Dermatol Vénéréol* (Paris) 104:877, 1977.
3. Costa, O. G.: Akrokerato-Elastoidosis. *Dermatologica* 107: 164, 1953.
4. Costa, O. G.: Acrokeratoelastoidosis. *Arch Dermatol* 70: 228, 1954.
5. Costa, O. G.: Acrokératoélastoïdose. *Ann Dermatol* 83:146, 1956.
6. Costa, O. G.: Acroceratoses (ceratodermias palmo-plantares). Impr Univ Minas Gerais, Belo Horizonte, Brazil, 1962.

7. Greither, A.: Erbliche Palmoplantarkeratosen. *Hautarzt* 28: 395, 1977.
8. Jung, E. G.: Acrokeratoelastoidosis. *Humangenetik* 17: 357, 1973.
9. Jung, E. G., Beil, F. U., Anton-Lamprecht, I., Grenten, H. & Nemetschek, Th.: Akrokeratoelastoidosis. *Hautarzt* 25: 127, 1974.
10. Kocsard, E.: Keratoelastoidosis marginalis of the hands. *Dermatologica* 131: 169, 1965.
11. Massé, R., Quillard, A., Héry, B., Toudic, L. & Le Her, G.: Acrokérato-Élastoïdose de Costa. *Ann Dermatol Vénéréol* (Paris) 104: 441, 1977.
12. Pyrhönen, S. & Penttinen, K.: Wart-virus antibodies and the prognosis of wart disease. *Lancet* ii: 1330, 1972.
13. Ritchie, E. B. & Williams, H. M.: Degenerative collagenous plaques of the hands. *Arch Dermatol* 93: 202, 1966.
14. Rook, A.: Disorders of connective tissue. In *Textbook of Dermatology* (ed. by A. J. Rook, D. S. Wilkinson and F. S. Ebling), 2nd edn, vol. 2, p. 1491, Blackwell Scientific Publications, Oxford, 1972.

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