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WIDESPREAD ACRODERMATITIS CHRONICA ATROPHICANS IN TWO SISTERS

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Abstract. Two elderly sisters with very extensive lesions of acrodermatitis atrophicans are described. The coincidental onset of the disease took place in about 1930. One of the sisters was treated 1950 with penicillin. Subsequently, peculiar permanent subcutaneous fat deposits developed around her knees. In the other, the disease spread during 40 years all over the extremities and also the skin of trunk and face became partly affected. IgM was increased in the immunoelectrophoresis of both patients.

The primary lesion of acrodermatitis chronica atrophicans (ACA) is usually localized to one of the lower extremities (11, 13, 26). In the course of years and decades the skin change spreads, finally covering both extremities rather symmetrically. The upper extremities may also be affected. The skin of the trunk is not often involved (Hauser: 8 cases in 234 patients; Koskimies: 1 case in 57 patients; Montgomery & Sullivan: 1 case in 45 patients) (9, 18, 24). Facial skin lesions in 5 patients was recorded by Hauser (11). A very rare case in which ACA had spread to the skin of extremities, trunk, scalp and forehead was described by Marshall (23). In connection with characteristics of ACA the following symptoms have also been described: bone atrophy (26), arthrotic changes in the joints (14, 16) and neurological symptoms (15).

More than two-thirds of the patients with ACA have been women (4). In some cases two or more members of the same family were involved: Grandmother and granddaughter (22); mother and daughter (27); mother, daughter and son (5, 25); two sisters (3, 9, 21); three sisters (18). The most remarkable report is that of Rasch (30) with 6 affected members: father, mother, three children, and father's sister. However, familial occurrence is on the whole rather uncommon. Thus, in a material of 234 cases of ACA reported by Hauser (9), only two sisters were mentioned.

CASE REPORTS

Case 1

H.E., an 80-year-old woman, born 1890 in Vestanfjärd, in the archipelago of Southwestern Finland. The patient spent her childhood and later on often the summer months there with her sister (case 2). During these summer months wood ticks often adhered to her skin.

The insidious onset of the disease took place in the beginning of 1930 with erythema on the lower extremities. Swelling in the legs and pain in the knees occurred periodically. In 1939 the affected areas were treated with UV-light which resulted in blister formation in the skin and a slowly healing ulcer about the left knee.

The disease spread to the thighs as well as to the lower abdomen and to the buttocks. The skin change on the lower abdomen was first noticed by a masseuse who treated the patient in 1947. The patient was admitted for examination to Salus Hospital (Wihuri Research Institute, Helsinki) by Dr Koskimies in 1950. She was administered 8 000 000 I.U. penicillin after which erythema disappeared and slight, gradual thickening of the skin took place. Rather soon after the penicillin treatment large, soft, occasionally tender, subcutaneous fat deposits began to develop around both knees (Fig. 1). The last few nodules formed about 2 years ago.

Status dermatologicus 12.11.1970: The skin of both lower extremities is light and so thin that the blood vessel network is clearly seen. The skin change is rather sharply restricted up to the groins and down to the ankles including the proximal half of the metatarsal skin. The skin of the lower third of the buttocks and lower abdomen is also thin and wrinkled.

Both knees are surrounded by band-like, large, subcutaneous clusters of doughy fat deposits (Fig. 1). In the lateral edge of both kneecaps the skin is attached to its base forming a funnel-shaped depression surrounded by tissue folds. The circumference of the right knee is 49 cm and that of the left 48 cm. On movement crepitation is noted in the knee joints.

In general examination the chest X-ray was normal.

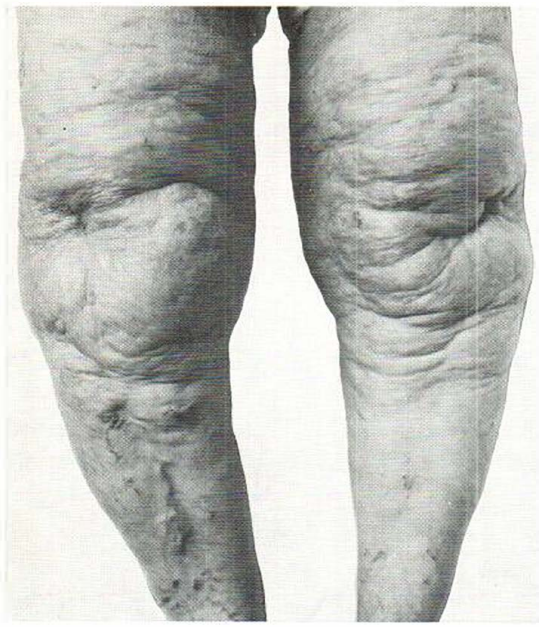

Fig. 1. Acrodermatitis chronica atrophicans. Large subcutaneous fat deposits around the knees (case 1).

No pathological changes were recorded by ECG. Blood pressure was 155/90. No palpable lymph nodes were found. X-ray examination of the knees showed on both sides strong arthrotic changes, narrowing of the interarticular space and numerous periarticular spur formations. Lumbar X-ray revealed marked spondylitic changes.

Laboratory tests showed the following: peripheric blood cell counts were normal; Sitolipin test negative; sedimentation rate 19 mm; urine normal; liver function tests normal. Serum proteins 7.3 g%. In the electrophoresis, albumin 52.6, alfa$_1$ 3.5, alfa$_2$ 7.9, beta$_1$ 10.5, beta$_2$ 6.1, gamma 19.2. In the immunoelectrophoresis, IgM was increased being quantitatively 195 mg/100 ml. IgG and IgA were normal.

Histological examination: The skin biopsy from the lateral side of the left knee revealed a ribbon-like, straightened, thin epidermis without any ridges. Pronounced atrophy of the corium was also noted and small sweat glands were seen immediately under the epidermis. No inflammatory reaction was observed. Absence of elastic tissue was recorded in the upper and central parts of the corium. Deformed, fragmented elastic fibres were seen in the very lowest parts of the corium. Loose, fat tissue was noted under the dermal tissue.

Case 2

D.W., a 76-year-old woman, born 1894 and lives in Vestanfjärd. The onset of the disease took place in about 1930 with purple patches on the thighs. Within 10 years the disease spread to the entire skin of the lower extremities. Subsequently the buttocks, the hips and the abdomen became involved. Extension to the upper extremities took place in 1967 when the disease spread to the skin of hands and upper arms. The skin changes spread over the chest and in August 1969 over the cheeks.

Status dermatologicus 5.2.1970: Both lower extremities exclusive of digital and plantar skin are involved. The skin of the legs and the thighs is purple, atrophied, papery and the subcutaneous blood vessels are clearly visible through the skin. Infiltrated scarlet patches, about one inch in diameter, are found about both knees.

On the back of the trunk the disease has spread over the buttocks and the lumbar area up to the waist. The abdominal skin is so affected that on the lower abdomen a triangular area confining to the groins is not involved. The upper surfaces of the breasts are affected.

The skin of upper arms, forearms and fingers is also affected. The cutaneous change on the right upper extremity is clearly stronger than on the left. The skin change is in the atrophic stage with the exception of two red, infiltrated patches in the extensor surface of the right upper arm. The skin of the cheeks is thin and purple, the colour of the left cheek is clearly more intensive.

In *the general health* of the patient no marked changes were recorded: X-ray examination of the lungs revealed a normal finding. Blood pressure was 160/90. No palpable lymph nodes were found. Descensus uteri was found on gynecological examination.

Laboratory tests showed the following: the peripheric blood cell counts were normal; Sitolipin test negative; sedimentation rate 9. Urine normal. Serum proteins 7.5 g%. In the electrophoresis, albumin 54.8, alfa$_1$ 1.9, alfa$_2$ 7.7, beta$_1$ 7.7., beta$_2$ 3.8, gamma 24.0. In the immunoelectrophoresis, IgM was markedly increased, quantitatively 380 mg/100 ml. IgG and IgA were about normal.

Histological examination 1: In the biopsy specimen taken from the infiltrated area above the left knee the epidermis is slightly hyperkeratotic and the rete ridges are completely flattened. Under the stratum basale there is a narrow layer of collagenic connective tissue and under it a dense, ribbon-like infiltration of inflammatory cells which consists of lymphocytes and histiocytes. The perivascular, inflammatory infiltrate also penetrates deeper into the corium. Elastic tissue is completely lacking in the upper parts of the corium. In its central and lower parts some fragmented, elastic fibres have accumulated into irregular groups. Atrophic hair follicles as well as sweat glands are found sporadically rather near the epidermis.

2. A slightly hyperkeratotic epidermis which is rather thin and produces only few rete pegs is seen in the biopsy taken from the healthy-looking skin of the upper back. Slight perivascular inflammatory infiltrate is found in the upper parts of the corium and around the small blood vessels in the subcutaneous fat tissue. The corium is not thinned. Both the collagen and the elastic tissue look normal.

Decursus morbi: The patient was administered 800 000 I.U. procain pencillin/day for 14 days in February 1970. Erythema began to disappear and the general health of the patient improved even during treatment. The patient was admitted again in September 1970. There were still some infiltrated, red patches above the knees. Tetracycline 500 mg × 3/day was given for 14 days. The skin became

lighter, but no reduction of atrophy was recorded. The results of the laboratory tests were the same after treatment as before, with the exception of IgM which had decreased slightly, being quantitatively 200 mg/100 ml. The histological examination showed in the affected skin areas the same as previously.

DISCUSSION

The histological picture of ACA with its special characteristics is very clear, differing slightly, however, according to the variations in the activity of the disease. A characteristic of the inflammatory stage is inflammatory infiltration, consisting mainly of lymphocytes and histiocytes which are localized in the upper part of the corium. It is separated from the atrophied, ridgeless epidermis by a thin band of collagen (20, 26). According to Montgomery & Sullivan (24) the collagen and elastic tissues around the inflammatory cellular band begin to degenerate. The behaviour of the elastic tissue in various atrophic dermatoses has been studied recently by Korting et al. (17). These authors described local reduction and sometimes even complete lack of elastic tissue in connection with ACA.

No histological findings of the healthy-looking skin of a patient with ACA are reported in the literature. In the healthy-looking skin of our patient (case 2), however, slight epidermal hyperkeratosis, narrowing of cellular layers and partial atrophy of rete pegs were found. Additionally, perivascular cellular infiltrate was recorded both in the corium and subcutaneously. Considering the advanced age of the patient, the atrophic changes could be caused by senility. The inflammatory changes, however, could not be explained thus. Corresponding mild tissue changes were recorded by Götz (6, 7) in the originally healthy persons in whom he succeeded in implanting ACA. The clinical symptoms of these patients represented the abortive forms of ACA.

The only abnormal laboratory finding in our patients was a marked increase in IgM in immunoelectrophoresis. In case 1 an increased IgM was observed despite the fact that the disease became arrested and 20 years had passed since the penicillin treatment. In case 2 a slight decrease in IgM was observed after the treatment. Previously an increase of IgM was recorded by Herrmann (12) and Brehm (1). A higher sedimentation rate and an increase in the alfa- and gamma-globulin fractions in the electrophoresis were often observed by Koskimies (18) and Kraus et al. (19). Inflammatory changes in lymph nodes as well as an increase in plasma cells and eosinophils in bone marrow were recorded by Hauser (10).

The etiology of ACA is still unknown. The possible heredity of the disease was considered by some of the authors who described ACA in two or more members of one family (3, 29). This, however, is not true, but must be considered a coincidence.

At present the infectious character of ACA is commonly approved, but the causative agent has not been isolated (6, 7, 8, 33). The causative agent may have been transmitted by ticks, *Ixodes ricinus*, which, in summertime, often adhered to the skin of the patients described here. The global distribution of this wood tick is somewhat larger than the endemic occurrence of ACA (2, 15).

Spread by direct contact from one person to another seems unlikely, since in that case the incidence of the disease within one family would presumably be greater. Coincidental involvement of several members of a family and especially coincidental onset of the disease, as in the case of the two sisters described in this study, is probably explained by the fact that in some years the vectors carry exceptionally large quantites of causative agents.

The effectiveness of antibiotic treatment also emphasizes the infectious etiology of the disease (28, 31, 32). In addition to penicillin, aureomycin and achromycin have been found effective (11). In a case which did not respond satisfactorily to penicillin the treatment was successfully continued with tetracycline in our Department of Dermatology (30). In case 2 we also administered tetracycline some 6 months after the penicillin treatment. In this case, however, the improvement through tetracycline was less impressive.

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