SHORT REPORTS

Acrodermatitis chronica atrophicans in Association with Lichen sclerosus et atrophicans: Tubulo-Interstitial Nephritis and Urinary Excretion of Spirochete-like Organisms

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We report about a 38-year-old male patient with coexisting acrodermatitis chronica atrophicans, lichen sclerosus et atrophicans and recurrent diabetic metabolic disorders since 9 years. Serologically IgG antibodies against *Borrelia burgdorferi* could be detected. Moveless winded structures, morphologically resembling borreliae could be demonstrated in the urine sediment by dark field microscopy. Additionally a tubulo-interstitial nephritis was diagnosed by the presence of a dysmorphic hematuria, a pathological polyacrylamid gel electrophoresis and raised alpha 1- and beta 2-microglobulin in the urine. We suggest that the excreted spirochete-like structures are borreliae. They may be the putative infectious agent for the development of lichen sclerosus et atrophicans in the genital area. (Received May 5, 1986.)

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Acrodermatitis chronica atrophicans (ACA) is regarded as a manifestation of Lyme disease, a systemic borreliosis (1). High antibody titers to *B. burgdorferi* could be detected in the serum of 100% of ACA patients by indirect immunofluorescence or enzyme-linked immunosorbent assay (ELISA) (1).

Besides coexistence of ACA with scleroderma, which has been claimed to be also a spirochetal disease (2), coexisting lichen sclerosus et atrophicans (LSA) or LSA similar dermatoses have been described in 5 of 41 ACA patients by Åsbrink (3). Atrophic genital manifestations in ACA have been reported by Gans (4) and Sweitzer (5).

In this paper we want to present a 38-year-old male patient with a 9-year history of ACA and coexisting LSA.

CASE REPORT

The patient remembered an insect bite on the back of his right hand in 1975. In September 1976 a blue reddish swelling appeared on the dorsal aspect of his right hand and lower arm, which then was diagnosed as acute ACA. At the same time he developed a balanoposthitis with thickening of the preputium. Penicillin-V 1 mill was administered orally three times a day through 24 days. Under this treatment the skin lesions improved. However, the balanitis recurred leading to xerosis of the preputium which made circumcision necessary. A diabetic metabolic disorder was diagnosed at this occasion. Subsequent evaluations of serum glucose, however, were within normal range till 1986.

In July 1985 the patient again came for advice because of a blue reddish swelling of the lateral aspect of his right upper arm (Fig. 1). He reported pains in his right elbow joint since half a year. At this time IgG antibodies against *B. burgdorferi* have been investigated for the first time by ELISA and were found to be highly positive.

Biopsy specimens were taken from the righ upper arm. The histological picture revealed an atrophic epidermis and a dense patchy lympho-plasma-histiocytic infiltrate in the dermis. The elastic tissue was absent in the area of infiltrate. Additionally a homogenization of collagen fibres could be observed. All these findings were consistent with the diagnosis of ACA with pseudosclerodermatous changes. Under treatment with penicillin V 1.5 mill. three times a day for 10 days his skin lesions

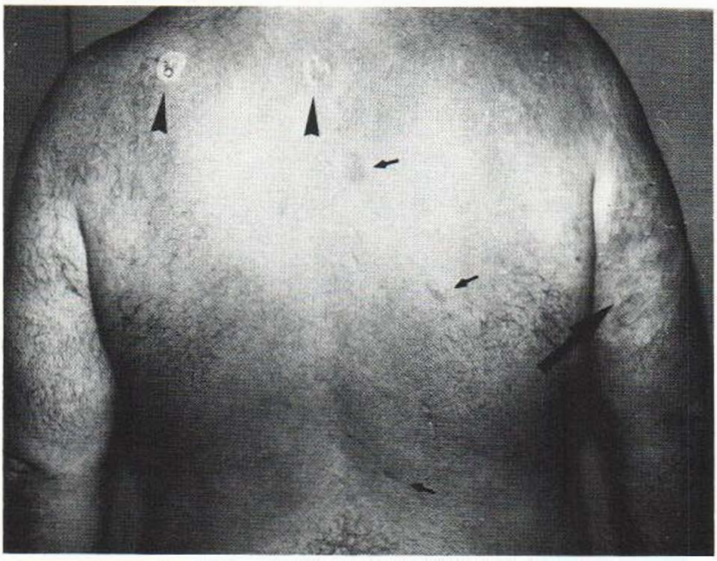

Fig. 1. ACA: Patchy discoloration of the right upper arm, loss of hair (↟). LSA: 2 ivory coloured patches on the back (▲); 3 lipatrophic lesions on the back (↟).

improved. A month later the same therapy was repeated and in September, 1985, a treatment with 500 mg Oxytetracyclin twice a day for 10 days was added. One month later serology showed a decreased IgG-titer. The swelling has disappeared leaving a blue reddish discoloration, but increased again in January, 1986. At the same time 2 ivory coloured patches, 3 cm in diameter, developed on the upper back. They were diagnosed as LSA clinically and histologically (Fig. 1). The glans penis also revealed the picture of LSA. Additionally 2 lipatrophic oval lesions, about 2 : 1.5 cm in diameter, appeared on the back (Fig. 1).

Laboratory findings

VDRL, TPHA negative; Erythrocytes 6.5 mill., Hb: 19.0 g; WBC, platelet count and ESR: normal; blood sugar: 304 mg%; triglycerides 406 U/l, GOT 20 U/l, GPT 53 U/l; ANA negative; Immunoelectrophoresis: normal.

Cultivation of B. burgdorferi

Skin samples taken from ACA and LSA lesions were cultured in modified Kelly's medium at 33°C. After incubation for 2 months no *B. burgdorferi* could be detected (performed at the Institute of Hygiene, Vienna).

Urinalysis

1% glucose, albumen negative; urine sediment: some hyaline cylindres, epithelial cells, leucocytes, erythrocytes.

In interference contrast microscopy dysmosphic erythrocytes and tubular epithelial cells could be demonstrated (Fig. 3). The polyamidgel electrophoresis showed a low molecular proteinuria, which can be found in several inflammatory tubular kidney diseases such as typhus (Fig. 2). High molecular proteinuria would be present in glomerular kidney diseases. Beta 2-microglobulin with 4 900 µg/l (normal: 0-400 µg/l) and alpha 1-microglobulin with 1.95 mg/100 ml (normal: 0-0.30 mg/100 ml) were elevated in the urine. These findings indicated a defective function of the proximal tubules and were in combination with dysmorphic hematuria and the low molecular proteinuria a sign for a tubulointerstitial nephritis.

Darkfield microscopy of the urine sediment revealed many motionless winded structures possibly representing spirochete-like bacteriae (Fig. 4).

DISCUSSION

In this case report we want to demonstrate three additional symptoms, associated with ACA, which can be interpreted as further manifestations of Lyme disease:

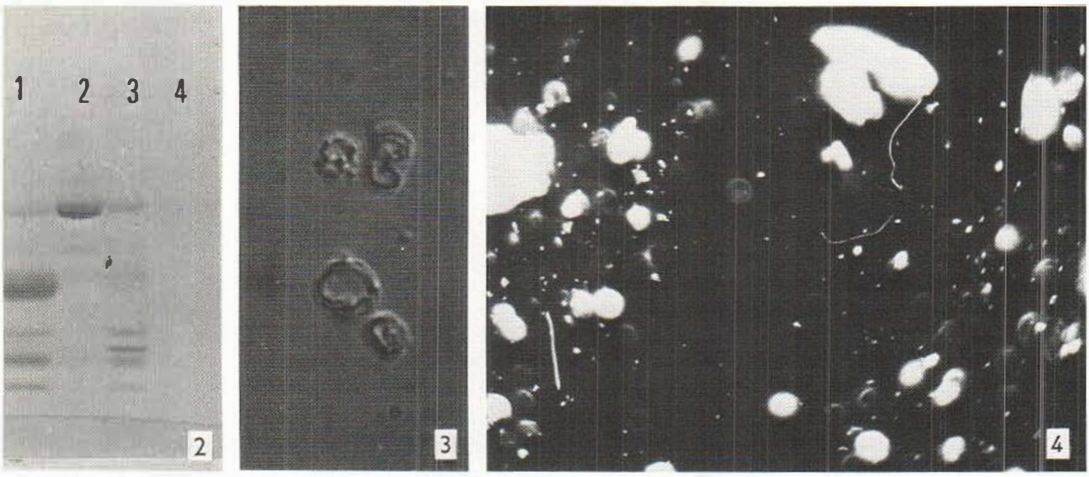

Fig. 2. 8% polyacrylamid gel electrophoresis of urine: 1) patient, 2) glomerulonephritis (high molecular proteinuria), 3) typhus (tubulo-interstitial nephritis, low molecular proteinuria), 4) healthy control person.

Fig. 3. Dysmorphic erythrocytes in the urine sediment. Interference contrast. ×320.

Fig. 4. Darkfield microscopy of urine. ×320.

1. Coexisting LSA has already been reported by Åsbrink (3). In our patient the genital LSA appeared at the same time with ACA and improved after penicillin therapy but recurred almost simultaneously with ACA.

2. The excreted spirochete-like bacteria in the urine might represent borreliae, since we could detect the same structure in 4 other patients (2 with ACA, 2 with LSA). In one case of ACA these organisms could be stained by an indirect avidin-biotin-immunoperoxidase method, using a highly positive serum of a patient with ACA as antibody. With the same technique borreliae could be detected on histological sections from 5 out of 9 patients suffering from genital LSA (E. Aberer: Spirochetes: The causative agent of morphea and LSA? presented at the 7th International Dermatopathological Colloquium, Graz, May 1986).

These additional findings let us suggest that the winded structures in the urine are borreliae, which are excreted through the kidney after hematogenic spreading. Spirocheturia had already been reported in feral reservoir animals by Bosler (6). It could be demonstrated that borreliae are transmitted to other laboratory animals by urine causing subsequent spirochetemia (6).

3. The dysmorphic hematuria, the pathologic polyacrylamid electrophoresis and the elevated alpha 1- and beta 2-microglobulin in the urine indicate an inflammatory parenchymatous process in the proximal tubulus of the kidney (7, 8), whereas in diabetes, glomerular defects could be expected. *B. burgdorferi* was isolated from kidneys in certain mice and hamsters (9, 10) and also from kidneys in fetal borreliosis in humans (11).

The raised serum glucose levels during exacerbation of disease suggest an additional pancreatic involvement, since the diabetic metabolic changes were concomitant with recurrences of the skin lesions. The development of lipatrophic lesions on the back might also be of borrelial origin.

In our patient ACA appeared at the site of an insect bite after a latency of one year. Spirochetemia, kidney involvement, spirocheturia and LSA developed. The occurrence of LSA in the genital area might suggest a borrelia infection by urine and different infectious

or local immune mechanisms might play a role in its development. Despite a clinical improvement after each treatment, recurrences of ACA lesions, new LSA patches and pains in the elbow joint occurred.

These new findings enlarge the spectrum of Lyme disease. LSA should also be looked upon as a possible borrelia infection. The exact infectious mechanism has to be elucidated. For preventing further spreading of the disease a sufficient antibiotic therapy remains to be established.

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Abnormal Vitamin D Metabolism in Patients with Psoriasis

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Staberg B, Oxholm A, Klemp P, Christiansen C. Abnormal vitamin D metabolism in patients with psoriasis. Acta Derm Venereol (Stockh) 1987; 67: 65–68.

To elucidate if psoriatic skin involvement induces changes in vitamin D metabolism, the serum concentrations of the major vitamin D metabolites (25-hydroxy-vitamin $D_{(2+3)}$ (25OHD), 1,25-dihydroxyvitamin $D_{(2+3)}$ ($1,25(OH)_2D$), and 24,25-dihydroxyvitamin $D_{(2+3)}$ ($24,25(OH)_2D$)) were studied in a group of patients with psoriasis, who had not been exposed to ultraviolet radiation at least three months before the investigation. Serum concentrations of $1.25(OH)_2D$ were significantly reduced in 17 patients with disseminated psoriasis compared to healthy age and sex matched controls (22.3 pg/ml versus 35.0 pg/ml ($p<0.001$)) and compared to 15 patients with moderate extended psoriasis (22.3 pg/ml versus 38.3 pg/ml ($p<0.005$)). Serum concentrations of the two other metabolites were not