Case report

CONCURRENT ACRODERMATITIS CHRONICA ATROPHICANS AND CIRCUMSCRIBED SCLERODERMA IN A FEMALE FARM HELPER

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SUMMARY

A 53-year old female farm helper presented with concurrent acrodermatitis chronica atrophicans and circumscribed scleroderma, both histopathologically proven. The serum IgG antibody titre against *Borrelia burgdorferi* was highly positive. The patient was treated with intravenous ceftriaxone (2x2 g/d) for 21 days. At the end of treatment, skin lesions of acrodermatitis chronica atrophicans had diminished, whereas the circumscribed scleroderma plaques remained unchanged. Acrodermatitis chronica atrophicans is a late chronic manifestation of Lyme borreliosis of proven borrelial etiology.

Circumscribed scleroderma, however, is only suspected to be a cutaneous manifestation of Lyme borreliosis. The simultaneous appearance of acrodermatitis chronica atrophicans and circumscribed scleroderma in the same patient has only been reported in a few cases, and may be taken as evidence for the spirochetal origin of circumscribed scleroderma.

KEY WORDS

Lyme borreliosis, acrodermatitis chronica atrophicans, circumscribed scleroderma

INTRODUCTION

Lyme borreliosis (LB) is a spirochetosis that can involve nearly all organs. Skin manifestations are seen most frequently, and occur during all stages of LB. The typical cutaneous manifestations of LB are erithema migrans (EM), borrelial lymphocytoma (BL), and acrodermatitis chronica atrophicans (ACA). In addition there are several skin diseases in which *Borrelia burgdorferi (Bb)* is being discussed as the etiologic agent, most importantly circumscribed scleroderma (CS). However, definite proof of *Bb* infection has only been possible in few cases of CS (1,2,3).

We report on a female farm helper who presented with concurrent ACA and CS lesions, and who had a positive *Bb* serum IgG antibody titre.

CASE REPORT

A 53-year-old female farm helper, who works in an area endemic for LB was seen at the Department of Dermatology in Graz, Austria in the summer of 1992. On admission she presented with extensive, poorly demarcated, painless livid erythemas (partly with signs of skin atrophy) distributed over the entire trunk and the extremities (Fig. 1, 2). This picture corresponded with the clinical diagnosis of ACA. Moreover, some whitish oval indurated plaques on both upper arms and the thorax were observed, consistent with the diagnosis of CS (Fig. 3). The onset of these skin changes could not be ascertained, and the patient could not recall any tick or insect bite. A punch biopsy was obtained from an erythema on the right thigh, as well as from a plaque on the right upper arm. The histopathologic examination of the biopsy from the right thigh showed a moderately dense perivascular and interstitial inflammatory infiltrate within the dermis (Fig. 4). Cytomorphologically, a predominance of lymphoplasmocytoid elements and plasma cells was noted. In the biopsy from the right upper arm, distinctly thickened and sclerotic collagen fibres were observed, in addition to a superficial and deep lymphohistiocytic perivascular infiltrate (Fig. 5).

In conjunction with a positive ELISA IgG serum antibody titre against *Bb*, the diagnosis of ACA in association with CS was established.

Blood chemistry was within normal limits. The BSR was 30/59, and the serum electrophoresis showed an elevation of IgG (1820 mg/dl) and IgM (500 mg/dl) antibodies. ECG, echogram of the heart, X-ray of the chest, a neurologic and an ophthalmologic examination were all without pathologic findings.

Intravenous treatment with ceftriaxone (2x2 g/d) was initiated. At the end of therapy (3 weeks), the ACA lesions were distinctly diminished, whereas the CS plaques remained unchanged.

DISCUSSION

ACA is a late skin manifestation of LB with a chronic and usually progressive course. It begins months to years after *Bb* infection with an acute inflammatory stage. The predilection sites are the acral parts of the body, and it predominates in women in the later decades of life. The skin lesions gradually develop into a chronic atrophic stage. Symmetric manifestation in either the upper or lower extremities may develop; involvement of all four extremities is rare (4). ACA lesions on the trunk are infrequent (5). The diagnosis of ACA is primarily based on clinical criteria. The histopathologic picture helps to confirm the diagnosis. Elevated IgG antibody titres against *Bb* are found in nearly all patients with ACA (6). On the other hand, seroprevalence in Europe lies between 3% and over 40% (7,8). Regarding non-specific laboratory parameters, a high BSR, as well as elevated IgG, IgA, and IgM antibodies in serum electrophoresis have been described (9).

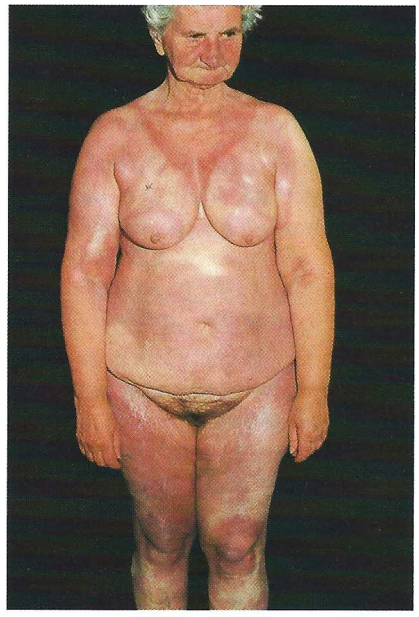
Of the three main types of localized scleroderma (plaque type, linear type, profound type), the plaque type, observed in our patient, is the most common (70 %) (10). The diagnosis of CS is based on clinical and histopathologic features. *Bb* antibody titres may he either positive or negative.

The borrelial etiology of ACA is proven. *Bb* has been isolated from ACA lesions (11), spirochetes have been demonstrated in histological sections of ACA (12), and Bb-specific DNA has been amplified by polymerase chain reaction (PCR) from an ACA lesion (13).

EM may develop into CS (14). ACA can occur together with EM (15), BL (16), lichen sclerosus et atrophicus (17), Shulman's syndrome (18) and CS (19,20,21).

The skin lesions in our patient are of interest because ACA was manifest on all four extremities as well as on the trunk, but particularly owing to the concurrence of ACA and CS. Besides its coexistence with, and development from typical dermatological LB manifestations (e.g. EM), a number of arguments exist in favor of the spirochetal origin of CS. (i) Various studies from Central Europe have demonstrated distinctly elevated antibody titres against *Bb* in a significantly higher percentage of CS cases than in controls (22, 23). Indeed, Aberer et al. have demonstrated Western Blot results with a

Fig. 1: 53-year-old woman with acrodermatitis chronica atrophicans lesions distributed over the entire trunk and the extremities.
Fig. 2: Typical acrodermatitis chronica atrophicans lesions on both thighs.
Fig. 3: Circumscribed scleroderma plaque on the right upper arm
Fig. 4: Histopathology of acrodermatitis chronica atrophicans. H&E, scanning magnification.
Fig. 5: The histopathological features from a biopsy specimen of the right upper arm are consistent with circumscribed scleroderma. H&E, scanning magnification.



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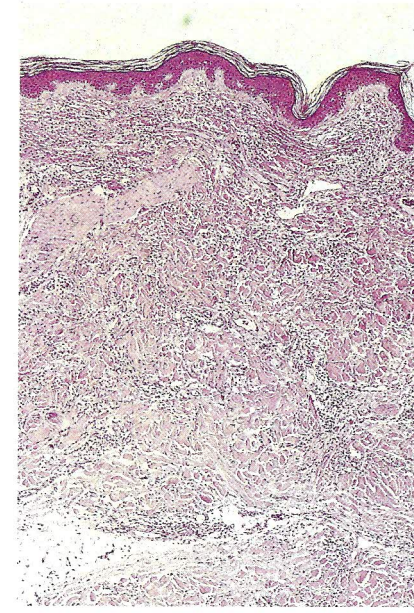

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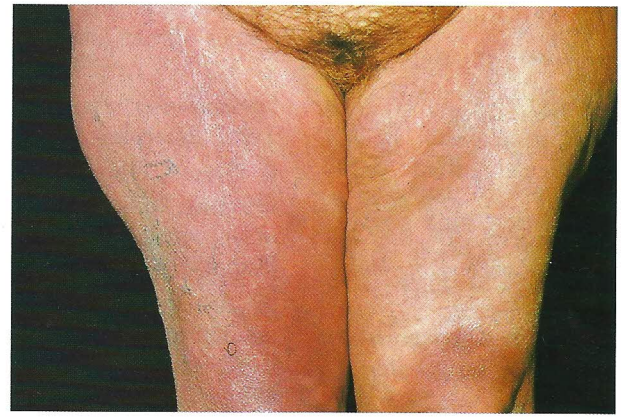

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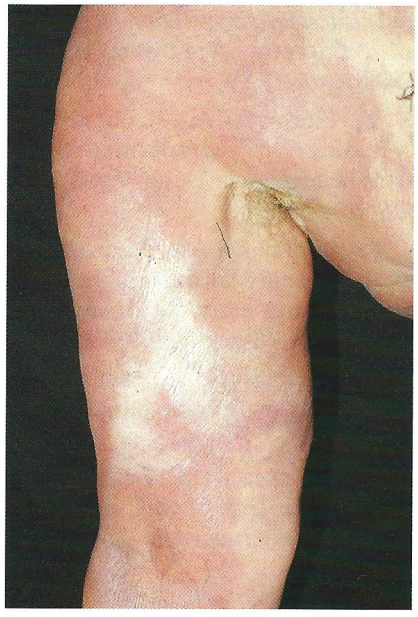

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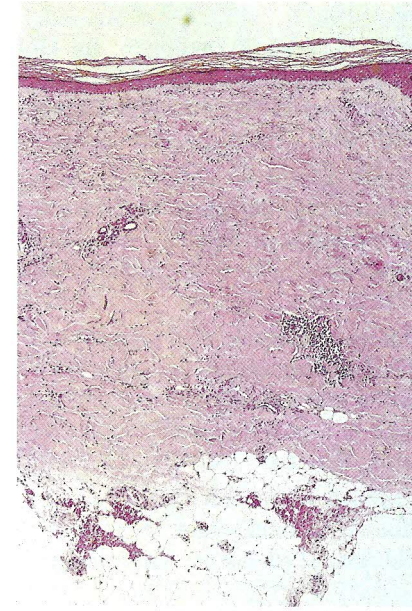

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pattern typical for LB in 40% of CS-patients (24). (ii) Borrelia-like organisms have been demonstrated in histopathologic sections of CS by silver staining (1), as well as by the avidin-immunoperoxidase method (25). (iii) The cultivation of spirochetes from CS lesions has been described (2). (iv) Using nested PCR, Schempp et al. were able to demonstrate Bb specific DNA in a CS lesion of a young woman (3). (v) Response to penicillin therapy in CS patients is also an intimation of its spirochetal origin. (24, 26). Considering all these arguments, it may be concluded that CS represents yet another manifestation within the widening spectrum of LB.

Recent work on LB indicates the possibility that only certain borrelia subtypes may induce CS (27). Therefore, a borrelial etiology for CS is not necessarily uniform, and may vary in accordance with the geographical distribution of the different subtypes.

Studies from other parts of Europe, which were not able to demonstrate increased antibody titres against *Bb* in CS, lend credence to this explanation (28, 29). PCR with its ability to differentiate borrelia subtypes (30), should help to clarify these open questions and thus promote understanding of the pathogenesis of CS.

In conclusion, our patient represents another example of the concurrent manifestation of ACA and CS which may be interpreted to imply a borrelial etiology for at least a subset of CS cases.

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