Acrodermatitis chronica atrophicans

D NADAL, R GUNDELFINGER, U FLUELER, AND E BOLTSUHAUSER

Department of Pediatrics, University of Zurich, Switzerland

SUMMARY Two cases of acrodermatitis chronica atrophicans associated with Borrelia burgdorferi infection are reported; to our knowledge these are the first cases reported in children.

Lyme disease is a complex multisystem disorder which usually begins with erythema migrans, a creeping annular erythematous skin lesion. Weeks to months later some patients develop meningitis, cranial or peripheral neuropathies, myocarditis, or arthritis. The illness is caused by the tick borne spirochete Borrelia burgdorferi. Because acrodermatitis chronica atrophicans is also associated with B burgdorferi infection, it is now considered to be a late manifestation of Lyme disease. Acrodermatitis chronica atrophicans is usually seen in adults aged 30 to 60 years; we now report two cases in children.

Case reports

Case 1

A 10 year old boy was referred because of darkened...
Acrodermatitis chronica atrophicans skin over the right lower leg that had been present for a few weeks. No accompanying symptoms such as fever or pain were reported but the boy remembered a tick bite a year before. On examination the skin over the knee, lower leg, and dorsum of the foot was livid with some oedematous and hyperaemic areas. Over the ankle the skin was dry and dystrophic. The erythrocyte sedimentation rate was 22 mm in the first hour, and the white cell count and differential were normal. The B burgdorferi titre in the serum was 1/4096 (normal below 1/64). Histological examination of a skin biopsy specimen showed a thin epidermis with a strongly pigmented stratum basale and perivascular patchy infiltration by lymphocytes, plasma cells, and histiocytes. Culture of the skin for B burgdorferi was negative. Treatment was started with benzylpenicillin 12 million units/m²/day intravenously for four weeks, and within a week the hyperpigmentation decreased and the oedema and hyperaemia disappeared. Six months later the skin was almost normal. The B burgdorferi titre remained high at 1/4096.

CASE 2
A 12 year old boy was referred because of violet discoloration of the skin of his left leg. A year earlier the patient had noticed induration of the subcutaneous tissue over the left quadriceps muscle. Later the skin over this area darkened and the lesion spread to the lower leg. There was no pain, functional impairment, or fever, and no history of a tick bite. In addition to this hyperpigmentation and induration of the skin and subcutaneous tissue, a few vitiligo like patches were noted, some of which were atrophic (fig 1). Some areas felt warm. The circumferences of the left upper and lower leg were 2 cm less than the right. The erythrocyte sedimentation rate was 20 mm in the first hour; white cell count and differential were normal. The B burgdorferi titre in the serum was 1/4096. Histological examination of the affected skin showed pronounced oedema of the corium, and perivascular patchy round cell infiltrates consisting mainly of lymphocytes. Cultures of the skin for B burgdorferi were negative. Treatment was started with benzylpenicillin 12 million units/m²/day for four weeks, and within a week the dark skin areas lightened and the induration decreased. After six months the skin had reverted to normal (fig 2) but the differences in leg circumferences still persisted. The B burgdorferi titre in serum remained high at 1/2048.
Progressive disease or initial migrans, and to dermal burgdorferi because West Thames the in cytops. After periods varying from weeks to months, hyper- and hypopigmentation develop, and gradually the skin becomes frail like cigarette paper so that vessels and subcutaneous tissue become visible; on the other hand fibrosis and sclerosis also develop. In this late or atrophic stage biopsy specimens are pathognomonic showing the typical signs of epidermal atrophy, damaged and degenerated elastin and collagen, and dilated dermal vessels surrounded by plasma cell infiltrates.

In our patients the cutaneous lesions corresponded to the clinical picture of acrodermatitis chronica atrophicans, and the histological findings were compatible with the inflammatory stage of the disease. High antibody titres against B burgdorferi (as found in both our cases) are diagnostic, the negative skin biopsy culture is not surprising, because isolation of the organism is notoriously difficult. A history of tick bite (absent in case 2) is not mandatory in diagnosing diseases caused by B burgdorferi because the bite is often overlooked or forgotten, and tabanids and mosquitoes may also harbour the pathogen. Our patients had no other signs or symptoms of Lyme disease.

Benzyldenicillin has been reported to be effective in the inflammatory stage of acrodermatitis, but recommended treatment regimens differ. Because high doses of penicillin given parenterally have been successful in the later stages of Lyme disease, we prescribed intravenous benzylpenicillin 12 million units/m²/day for four weeks. Clinically this treatment was successful, although the skin changes had not completely disappeared at the time of writing. Raised antibody titres may persist for six to 12 months after treatment. As in late syphilis, it may be that the time taken to achieve seronegativity after acrodermatitis depends on the duration of the untreated infection.

Our observations show that acrodermatitis chronica atrophicans associated with B burgdorferi may occur in childhood. Paediatricians should therefore be aware of this disorder because early diagnosis and treatment are important.

References

Correspondence to Dr D Nadal, Department of Pediatrics, University of Zurich, Steinwiesstrasse 75, CH-8032 Zurich, Switzerland.

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Blood counts in extremely low birthweight infants

N McIntosh, C Kempson, and R M Tyler

South West Thames Regional Neonatal Unit, St George's Hospital, London

Summary White blood, neutrophil, and platelet counts were higher in 101 infants with appropriate weight for gestational age than in 42 infants who were small for gestational age. The recognised postnatal rise in counts was seen in the infants of appropriate weight, but in the infants who were small for gestational age the counts fell for the first three days.