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CASE REPORT

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A case of a childhood linear scleroderma with limb asymmetry

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Abstract We report the case of a child with unilateral linear scleroderma disturbing the right lower limb. It took three-quarters of a year to be diagnosed as linear scleroderma and another 8 months to be treated with oral steroids. Although functional disabilities of the right knee and foot were improved with the steroid therapy, the limb asymmetry has remained. We believe that early treatment with steroids is essential for childhood linear scleroderma with lesions over any joints.

Key words Anti-single-stranded DNA · Limb asymmetry · Linear scleroderma · Oral steroid therapy · Thermography

Introduction

Localized scleroderma always affects the skin and subcutaneous fat tissue, and occasionally the fascia, skeletal muscle, and bone, but rarely involves internal organs, which are usually involved in systemic sclerosis. Localized scleroderma is classified into three main subtypes, i.e., morphea, generalized morphea, and linear scleroderma. Linear scleroderma is considered a pediatric disease that usually occurs in the first decade of life. A band or bands of skin become thick and tight on the trunk or extremities. In some cases it may extend deep into the tissue and down to the bone, causing an associated loss of deep tissue or bone structure and interfering with normal growth. We report the case of a girl with linear scleroderma on the right lower limb, which has remained severely shortened.

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Case report

A 3-year-old girl presented with a brown macula that developed on her right proximal femur in July 2000, when she was 1 year and 11 months old. The same lesion was warm to the touch a few months later. She fell and broke her right thigh in March, 2001. The fracture was cured by traction with a plaster cast. When the plaster was removed, skin sclerosis was found. A pediatrician diagnosed her condition as localized scleroderma, and treated her with oral tranilast and diglorasone diacetate ointment. After visiting several clinics, she finally came to our clinic in October, 2001. We recognized at the first visit that her lower limbs were of unequal length, and that there was sclerosis from the dorsal right knee to the foot, and contraction of the right knee joint and foot joint (Fig. 1). The ranges of movement were 0°–90° of right knee flexion and 0°–130° of left knee flexion (Fig. 2). Radiography performed at the first visit showed cystic changes and sclerosis within the proximal and distal metaphysis of the right tibia, and cystic changes within the distal metaphysis of the right femur. The lengths of the right and left tibia were 22.3 cm and 22.9 cm, respectively, and those of the right and left fibula were 17.1 cm and 18.7 cm, respectively. The results of routine blood tests were normal, including the eosinophil count, and levels of transaminase and creatinine kinase. IgG and IgM levels were also normal. The antinuclear antibody (ANA) titers were 1:1280 (homogenous pattern), and there was an elevated level of anti-single-stranded (ss) DNA antibody, 800 < arbitrary units (AU) ml (normal range 0–25 AU/ml). Antibodies to *Borrelia burgdorferi* were negative, 0.94 (normal range <1). We started treating the patient with 0.2 mg/kg body weight of oral prednisolon per day, and tapered it as the joint contraction and immunological disorder improved. The ANA titers and level of anti-ss DNA reduced to 1:160 and around 100 AU/ml, respectively, in 3 months (Fig. 2). Inflammation on the reticular dermis or the lipid layer of the right knee was monitored by thermography (Fig. 3a,b). After the administration of prednisolon for 1 year, the stiffness in the right knee and right ankle was reduced, but the difference in

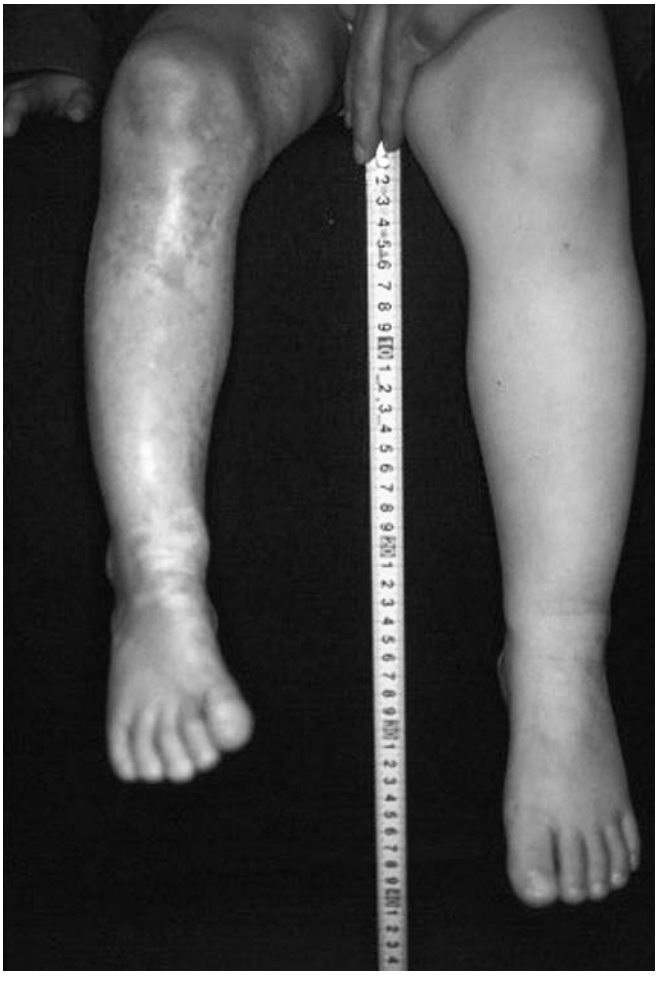

Fig. 1. Limb asymmetry and sclerotic skin from the dorsal right knee to the foot was observed at the first visit to our clinic

limb length had not improved. No systemic growing failure induced by steroids has been observed up to now.

Discussion

Localized scleroderma is a rare disease of unknown etiology, and its incidence was reported by Peterson et al.[1] to be 27 patients per million. It is divided into three subtypes: morphea, generalized morphea, and linear scleroderma, which may be more common in childhood.[2] Many cases are preceded by a history of trauma.[3] The lesion may develop gradually over several years, or rapidly over a few weeks or

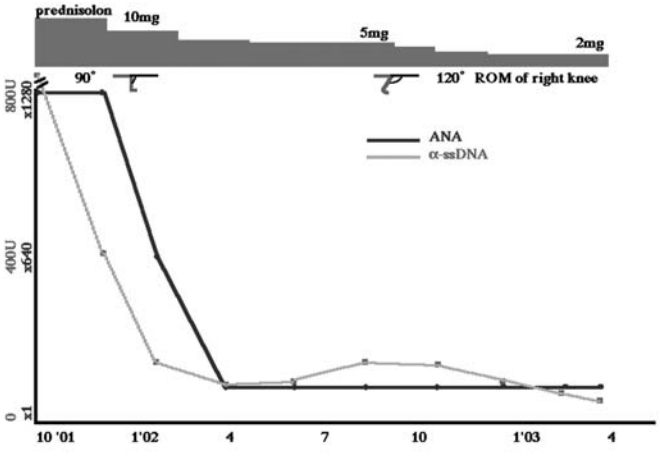

Fig. 2. Clinical course. *ROM*, range of movement; *ANA*, antinuclear antibody; *α-ssDNA*, anti-ss DNA

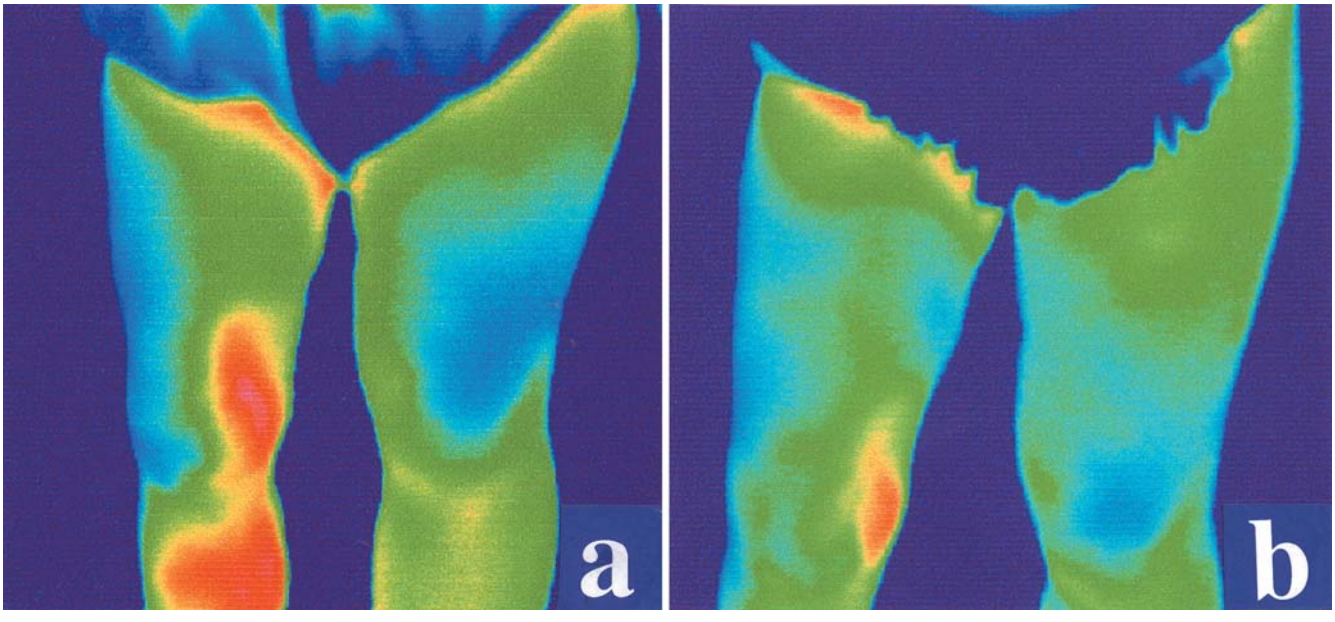

Fig. 3. Thermography. A reddish color indicates higher temperature. A reduction in the higher temperature area around the right knee was observed by comparing the pre- (**a**) and post- (**b**) steroid medication

months, sometimes having devastating effects on growth and development, such as limb asymmetry, flexion contracture, and psychological distress. The reason for the unilateral distribution is unclear, as in other skin diseases. However, Hauser et al.[4] suggested that scleroderma en coup de sabre may follow the line of Blashko.

In localized scleroderma, the frequency of positive ANA varies from 23% to 73%.[5–8] ANA are most commonly found in patients with generalized morphea, followed by linear scleroderma and morphea alone. More patients (73%) with generalized morphea had anti-ss DNA antibody than did patients with linear scleroderma (32%).[2] The presence of anti-ss DNA antibody was associated with joint contracture, disability, or more extensive, active and prolonged disease.[9]

When the lesion rapidly enlarges or involves functionally important areas, systemic therapy may be indicated. In such cases, oral corticosteroids are the first choice of treatment. Takehara et al.[10] described the indications for oral steroid therapy for localized scleroderma as: (1) clinically strong inflammatory findings with a rapidly enlarging lesion, (2) functional involvement or expectation of functional involvement, (3) possibility of growth failure, (4) a muscle lesion and high titer of anti-ss DNA antibodies. Other drugs such as penicillin, amoxicillin, salazopyrine, etretinate, phenytoin, cyclosporin A, colchicines, methotrexate, and PUVA therapy have been reported to be effective in isolated cases or in a small numbers of patients.[2,11,12] Topical corticosteroid therapy is often used for mildly involved skin. The monitoring of lesion activity in linear scleroderma is often conducted by thermography.[13] In our case, oral corticosteroid therapy for 1 year reduced the stiffness in the right knee and in the right ankle, although the limb asymmetry remained. The patient should have been treated with steroids before the appearance of the growth failure. We believe that early treatment with steroids is essential for childhood linear scleroderma with lesions on any joints.

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