Clinical and Experimental Rheumatology 2014; 32: 121-122.

A boy with tight skin: borrelia-associated early-onset morphea

R.M. Verberkt[1], M. Janssen[2], J. Wesseling[1]

[1]Department of Paediatrics; [2]Department of Rheumatology, Rijnstate, Arnhem, the Netherlands.

Roos M. Verberkt, MD
Matthijs Janssen, MD
Judit Wesseling, MD, PhD

Please address correspondence to:
Judit Wesseling, MD, PhD,
Department of Paediatrics,
Rijnstate,
Wagnerlaan 55,
6815 AD Arnhem, The Netherlands.
E-mail: jwesseling@rijnstate.nl

Received on January 26, 2013; accepted in revised form on April 30, 2013.

Key words: morphea, localised scleroderma, Borrelia burgdorferi, rheumatology

ABSTRACT

We present a case of a 16-year-old boy with morphea caused by Borrelia burgdorferi. We re-emphasise an immunohistochemical method, focus floating microscopy (FFM), to detect Borrelia burgdorferi spirochetes in tissue sections. Focus floating microscopy (FFM) proved to be more sensitive than polymerase chain reaction (PCR) and nearly equally specific.

Introduction

Morphea, or localised scleroderma, is an inflammatory connective tissue disease that affects the skin and leads to a pronounced sclerosis with increased collagen deposition and atrophy of the skin. It displays a wide clinical spectrum ranging from single localised patches to disseminated devastating forms. Lesions usually present as ivory sclerotic patches surrounded by an inflamed lilac ring.

Involvement of subcutaneous fat and underlying muscles may result in profound atrophy. In children this may lead to growth retardation of the affected limb or anatomic region. The possible involvement of *Borrelia burgdorferi* as a causative agent in morphea shows conflicting results, obtained by different studies using serological, immunohistochemical, culture and polymerase chain reaction (PCR) approaches (1). We present a patient with morphea caused by *Borrelia burgdorferi*.

Case report

Herein, we report the case of a 16-year-old, previously healthy, boy presented to the department of paediatric rheumatology in November 2009, with severe progressive limitation of flexion and extension in shoulders, knees, ankles, elbows, wrists and fingers over a period of six months. He also noticed a brownish discoloration of the skin around the large joints. There were no signs of arthritis and the movements were not painful. The patient was extremely tired. Our patient did not recall a tick bite or erythema migrans over the previous months. There was no Raynaud's phenomenon. Inflammatory autoimmune diseases were unknown to his family.

Physical examination was normal. A severe limitation of movement in the shoulders, knees, ankles, elbows, wrists and finger joints was seen with circumscribed ivory sclerotic plaques around these joints, surrounded by areas of hyperpigmentation. We suspected morphea. The skin around his feet, knees and elbows was tight (Fig. 1), but the facial skin was normal. The capillaries in the cuticles were normal and there was no sclerodactyly. There were no digital pitting scars or ulcera. His blood pressure was normal.

Laboratory studies showed no infectious parameters besides a slightly elevated erythrocyte sedimentation rate (16 mm/hour). There was a mild elevation of alkalic phosphatase (157 U/L). X-rays of the joints were all normal. However, testing for anti-Borrelia burgdorferi IgG was extremely positive (>400 U/ml). Immunoblot tested positive for p39, p41, p58, p100, OspC and VlsE. IgM was negative. A treponemal screening test was negative. Antinuclear antibodies (ANA) tested negative. A skin biopsy showed extensive lymphocytoplasmic infiltration in the (epi)dermis and subcutis, no spirochetes were found. A polymerase chain reaction (PCR) for *Borrelia burgdorferi* was negative in the skin biopsy material. The positive Borrelia serology suggested a causative role in the presence of morphea and oral Doxycyclin (200 mg for 30 days) was started. Three months later, a deep skin biopsy (*i.e.* skin, muscle and fascia) showed a lymphoplasmocytic infiltration with increased fibrosis in the dermis, subcutis, fascia as well as in striated muscle tissue. He was started on treatment with methotrexate and steroids.

Eisendle *et al.* used focus-floating microscopy (FFM) for detection of *Borrelia*. This is a modified immunohistochemical technique that combines several strategies to detect minuscule organisms in tissue sections. Spirochetes can be shown by staining with polyclonal rabbit antibodies to *Borrelia burgdorferi* (2, 3). FFM proved to be more sensitive than PCR (96.0% *vs.* 45.2%) and nearly equally specific (99.4% *vs.* 100%). Furthermore, FFM rejected the diagnosis Borrelia infec-

Competing interests: none declared.

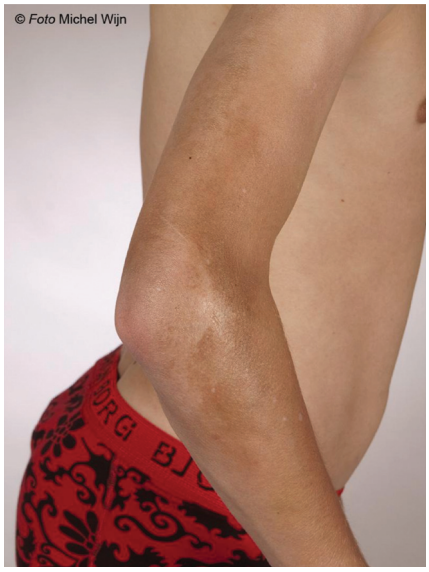

Fig. 1. Morphea on the patient's right elbow.

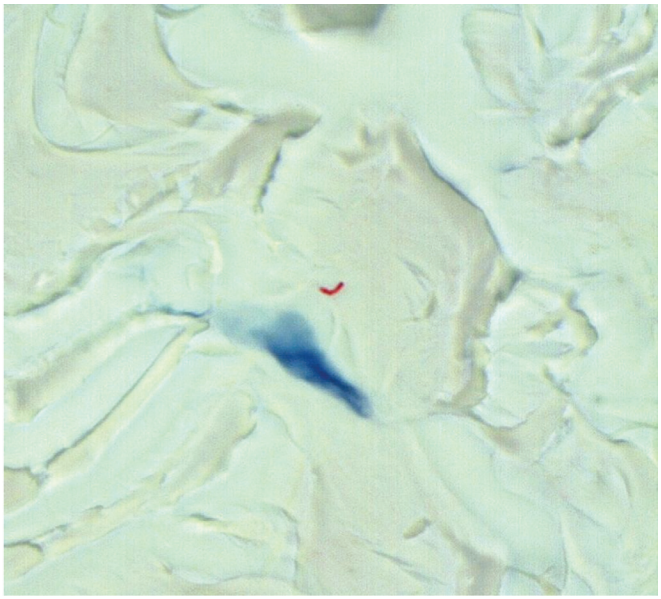

Fig. 2. Focus floating microscopy detected spirochetes in the skin biopsy material.

tion with more consistency: negative predictive value is 97.1% *vs*. 66% (3). Focus floating microscopy in our patient detected spirochetes in the skin biopsy material (Fig. 2) ultimately proving Lyme disease. The second biopsy material, after treatment for Borrelia infection tested negative for spirochetes by FFM indicative for an effective antibiotic treatment.

Lyme borreliosis is the most common vector-borne bacterial infection in temperate areas of the Northern hemisphere (4). Erythema migrans and acrodermatitis chronica atrophicans are well known skin manifestations of Lyme disease. This case history shows that morphea should also be considered as a skin manifestation of Lyme disease in young patients. It had to be added to the differential diagnosis of scleroderma-like diseases presenting to paediatric rheumatology practices.

References

1. PRINZ J, KUTASI Z, WEISENSEEL P *et al*.: "Borrelia-associated early-onset morphea": A particular type of scleroderma in childhood and adolescence with high titer antinuclear antibodies? *J Am Acad Dermatol* 2009; 60: 248-55.
2. EISENDLE K, GRABNER T, ZELGER B: Morphoea: a manifestation of infection with Borrelia species? *Br J Dermatol* 2007; 157: 1189-98.
3. EISENDLE K, GRABNER T, ZELGER B: Focus floating microscopy: "gold standard" for cutaneous borreliosis? *Am J Clin Pathol* 2007; 127: 213-22.
4. O'CONNELL S: Lyme borreliosis: current issues in diagnosis and management. *Curr Opin Infect Dis* 2010; 23: 231-5.