Case Report

Eosinophilic Fasciitis Presenting with a Reactive Hepatitis

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Summary

We report a patient with eosinophilic fasciitis, reactive hepatitis and splenomegaly. Administration of prednisone resulted in normalization of liver enzymes and spleen-size and an improvement of his clinical condition. These uncommon manifestations are only part of the broadening clinical spectrum, which is discussed briefly.

Key words: Eosinophilic Fasciitis, Hepatitis, Splenomegaly.

INTRODUCTION

Eosinophilic fasciitis (EF) was first described as a syndrome, characterized by scleroderma-like skin changes, peripheral eosinophilia and hypergammaglobulinemia in the presence of a rapid onset of diffuse fasciitis (1). EF has a characteristic histological picture of inflammation and sclerosis, primarily affecting the deep fascia and subcutis. Initially, it was distinguished from systemic sclerosis by the absence of Raynaud's phenomenon, visceral involvement and a favourable response to corticosteroid therapy. Since the original publication by Shulman, more than 200 cases have been reported and the full extent of the clinical spectrum is becoming clear (2,3). Systemic involvement is more common than previously thought. There have been isolated cases of oesophageal, pulmonary or cardiac involvement, but in contrast to systemic sclerosis, the viscera are rarely affected (4). Here we report a patient with eosinophilic fasciitis who presented with jaundice, due to a reactive hepatitis.

CASE REPORT

After a common cold, a 21-year old student noted slowly progressive muscle pains and a gradual onset of muscle stiffness in his upper and lower extremities. After a few weeks he developed malaise, night-sweating, heat-intolerance and pretibial pitting oedema. Four months after the onset of his complaints the serum level of liver enzymes was elevated. Serologic studies for causative microorganisms were negative; there was no history of hepatotoxic medication. Microscopic examination of a liver biopsy specimen showed non-specific, reactive hepatitis (Fig. 1). One month later he developed jaundice with discoloration of his stools and dark brown coloration of his urine. At this time the values of his liver enzymes had further risen to ASAT 453 IU/l (normal: <30 IU/l), ALT 514 IU/l (normal: <30 IU/l), Alkaline Phosphatase 423 IU/l (normal: 35-95 IU/l), Gamma GT 58 IU/l (normal: 5-44 IU/l), bilirubine 147 µmol/l (normal: <17 µmol/l). Because of a marked progressive functional impairment due to limitation of the movements of his arms and legs and worsening of his general condition, he was referred to our department. Until then he had lost 15 kg bodyweight and he needed help with most activities in daily life. On physical examination we saw a slightly icteric man with striking contractures in virtually every joint (Fig. 2). His skin was firm and showed hyperpigmentation.

On elevation of the arms there was retraction of the subcutaneous tissue along the veins — referred to as “sunken veins” — and between muscle groups -referred to as “the groove sign” — (5). All over his body the muscles were firm, preventing deep palpation of abdominal organs and limiting his thoracic excursions. Some pretibial pitting oedema was noted. Further examination revealed no abnormalities, except for hepatosplenomegaly on percussion.

On admission to our unit, laboratory values showed an ESR of 40 mm after 1 hour, a haemoglobin level of 7.7 mmol/l, a white blood cell count of 4.100 with 19%
Eosinophilic fasciitis presenting with a reactive hepatitis

Fig. 1: Section of the liver with marked reactive changes. The sinusoids are prominent as a result of clusters of Kupfer cells and diffusely located inflammatory cells (arrow). (original magnification 240 x ).

eosinophils, 1% basophils, 52% neutrophils, 22% lymphocytes, 6% monocytes, an ASAT of 283 U/l, an ALAT of 300 U/l, an AF of 699 U/l, a GGT of 130 U/l, a bilirubin of 49 µmol/l and a protein of 72 gr/l. Routine laboratory values, including CK were normal. Agargel electrophoresis disclosed a normal pattern with an increase of gamma globulin. Quantitation of the immunoglobulins showed a marked elevation of IgG to 35.6 gr/l (upper limit 11.2 gr/l); IgA and IgM were normal. Serologic testing for hepatitis A, B, C, EBV, CMV, HIV, toxoplasma, Borrelia Burgdorferi, Yersinia and Brucella was negative, as were tests for rheumatoid factor, anti-nuclear antibodies, antibodies to dsDNA and extractable nuclear antigen.

Upper, abdominal ultrasound examination confirmed hepatosplenomegaly. The chest X-ray was normal, but spirometry disclosed a restrictive lung function. Oesophageal manometry and a swallow cinegram revealed no abnormalities. A muscle-fascia biopsy specimen from the right quadriceps showed lymphoplasmocytic infiltration of a thickened fascia with perivascular infiltration, without destruction of vessel walls; no eosinophilic cells were seen in the infiltrate (Fig. 3). A diagnosis of diffuse fasciitis with peripheral oesinophilia was made and treatment with 60 mg/day prednisone was initiated. After six weeks of treatment his condition improved considerably. The limitations of his extremities had decreased and the serum values for ASAT, ALAT, GGT, AF, bilirubin, gammaglobulin and the ESR returned to normal. Control ultrasound examination showed a normalization of the size of the spleen but no change in liver size. After one year of treatment the prednisone medication has been tapered off to 10 mg daily at the present time. His motor function, although improving, is still hampered by residual contractures.

DISCUSSION

The diagnosis eosinophilic fasciitis is made by a combination of clinical features, laboratory results and a full thickness biopsy specimen of the muscle and fascia. Classically, there is a rapid onset of disease after a prodromal stage of fatigability, low grade fever and muscle-aches and cramps following extreme physical exertion. Subsequently, sclerodermoid skin changes may develop. These appear to evolve through three stages starting with pitting oedema of arms and legs, followed by a “peau d’orange” appearance of the skin and ultimately thickening of subcutaneous tissues leading to induration of the skin and limitation of joint motion, often resulting in contractures. Virtually any part of the body may be