Case Report

Polymyositis Associated with Primary Biliary Cirrhosis

K.A. BOKI, S.P. DOURAKIS*

Summary The coexistence of polymyositis (PM) and primary biliary cirrhosis (PBC) is rare; only nine cases have been described in English literature. We report a case of a 46-year-old woman presenting with these two autoimmune diseases. The diagnosis of PM was based on the symmetrical, proximal limb muscle weakness, elevated muscle enzymes and was confirmed with the electromyography and muscle biopsy. The diagnosis of PBC was based on the increased serum levels of alkaline phosphatase, gamma glutamyltransferase, IgM immunoglobulin, the presence of antimitochondrial antibodies and diagnostic liver biopsy.

Key words Polymyositis, Primary Biliary Cirrhosis, Autoimmune Diseases

INTRODUCTION

Polymyositis (PM) is an inflammatory myopathy in the pathogenesis of which immunologic mechanisms seems to play a role (1). Polymyositis is frequently associated with other connective tissue diseases but has rarely been reported to occur simultaneously with primary biliary cirrhosis (PBC) (2-9). An association of PM and chronic active hepatitis with autoantibodies to mitochondrial proteins was previously described (10). PBC is a chronic, autoimmune liver disease, characterized by progressive, nonsuppurative, destructive cholangitis. Nonhepatic disorders, particularly thyroid and connective tissue disorders, are found in 69% of patients with PBC (11). The widespread use of automated biochemical screening tests has resulted in diagnosing an increasing number of asymptomatic patients. We report the rare coexistence of these two autoimmune diseases.

CASE REPORT

A 46-year-old woman was admitted to our hospital for assessment of a 6-month history of weakness primarily in the legs, expressed as difficulty in climbing stairs and rising from a sitting position. Recently she had noted difficulty in lifting objects from high places and dressing up. She did not have pruritus. Examination revealed proximal weakness, the pelvic muscles being more affected than the shoulders. The neurological examination was normal. There was no rash. The laboratory findings were: haemoglobin 12.9 mg/l, white blood count 5100/mm³, platelets 318000/mm³, and erythrocyte sedimentation rate 52mm/hr. Electrolytes were normal, urea 40mg/dl, creatinine 0.8mg/dl, glucose 74mg/dl, and cholesterol 290mg/dl. The creatinine phosphokinase was 890 Units/l (normal value 10-235), lactate dehydrogenase 353 Units/l (normal value 109-200), aldolase 19 Units/l (normal value 1-7.6), aspartate transaminase 128 Units/l (normal value 5-40), alanine transaminase 128 Units/l (normal value 5-40), gamma glutamyl transferase 164 Units/l (normal value 8-35), alkaline phosphatase 286 Units/l (normal value 12-100). Total bilirubin was normal. Hepatitis B surface antigen and antibodies were negative. Urinalysis was normal. Electrophoresis showed a diffuse increase in the gamma region and no paraproteinaemia. Quantitative determination of immunoglobulins showed: IgM 540mg/dl (normal value 80-200), while IgG and IgA were normal. Thyroid function tests were within normal limits. Antithyroid antibodies were negative. Antinuclear antibodies were positive at 1/640 with a diffuse and speckled pattern, while anti-double stranded DNA antibodies, antibodies to extractable nuclear antigens and rheumatoid factor were negative. Antimitochondrial antibodies were positive at 1/640 and antismooth muscle antibodies also positive at 1/160. Complement levels

From the Department of Rheumatology "KAT" Hospital, and the Department of Internal Medicine, "Ippokration" Hospital, Athens, GREECE.
(C3,C4) were normal. The HLA typing showed: A1, A24(9), B5(51), B40, DR5, DR7.

A muscle biopsy of the quadriceps showed necrotic and atrophic muscle fibers, variation in fiber size and centralized nuclei in some fibers, with a small number of focal chronic inflammatory cells infiltrating the necrotic areas and perivascular regions. Muscle cells showed more features of degeneration than of regeneration. The electromyography showed a complex of short duration, low amplitude, polyphasic, motor unit potentials, fibril-