Case report

Constrictive pericarditis complicating dermatomyositis

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SUMMARY Chronic constrictive pericarditis may complicate both rheumatoid arthritis and systemic lupus erythematosus. To our knowledge, however, this is the first time it has been described in a patient with dermatomyositis. This association should be kept in mind as constrictive pericarditis should be considered in the differential diagnosis of liver cirrhosis and Budd-Chiari syndrome in rheumatic diseases.

Key words: autoimmune disorders, serositis, cardiac disease, ascites.

Dermatomyositis is an inflammatory disease of the muscle and skin. Although cardiac involvement may occur in the course of the disease, constrictive pericarditis has not been found before in large series of patients with dermatomyositis and polymyositis.^{1–4} Herein is a case where constrictive pericarditis developed in a patient with longstanding dermatomyositis.

Case report

A 54 year old white man presented in July 1985 with progressive weakness, anasarca, and moderate anaemia. During the several months preceding his admission he had had a noticeable weight gain, abdominal distension, and bilateral ankle swelling.

PAST HISTORY

The patient was first admitted in March 1973 for intermittent fever, arthralgia, symmetric, proximal muscle weakness, and a skin rash which he had had for three months. On admission his temperature was 38°C. Pertinent physical findings were an oedematous, lilac, scaly rash over his eyelids, nose, forehead, elbows, knees, and knuckles; the fingers were

oedematous; and there was generalised periarticular tenderness. The liver edge was palpable 4 cm below the costal margin; the spleen was not palpable.

Laboratory tests showed the following serum concentrations: bilirubin 29.2 µmol/l (direct 20.5), transaminase 132 IU (normal range (NR) <40), lactic acid dehydrogenase 273 IU (NR 50–200), creatinine phosphokinase 90 IU (NR 10–60), aldolase 12.2 IU (NR 2–9.6), immunoglobulin G 29 g/l (NR 8.5–12.5), immunoglobulin A 10 g/l (NR 1.9–3.5), immunoglobulin M 1.4 g/l (NR 0.7–1.5). The Rose-Waaler test was positive at a titre of 1/80 and the latex fixation titre was 1/160. Repeated immunofluorescent antinuclear antibody tests were negative and the complements C3, C4, and CH₅₀ were normal. Bromosulphophthalein retention was 27% after 45 minutes. An electrocardiogram disclosed multiple ventricular premature beats. Electromyographic examination of the deltoid, biceps, quadriceps, and thenar muscles was consistent with active myositis. A deltoid muscle biopsy specimen examined by light microscopy was normal, and liver biopsy showed macrovesicular fatty changes. The patient was diagnosed as having dermatomyositis. Because of progressive deterioration in muscle strength and severe arthralgia treatment was started with prednisone 60 mg/day, and rapid improvement followed. After three months the dose was gradually tapered to 15 mg/day, which was the minimal dose required to keep him symptom free.

Accepted for publication 25 March 1988.

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He was admitted again in 1976 for muscle weakness in the extremities and stiffness of the biceps and deltoid muscles. He had diffuse subcutaneous indurations that had gradually developed over the previous 12 months. There was a diffuse musculoskeletal tenderness and limitation of movement. The liver was palpated 3 cm below the costal margin.

An x ray examination disclosed patchy and diffuse calcification of soft tissues over the chest, neck, shoulders, arms, pelvis, and thighs. Treatment with intravenous corticotrophin (ACTH) and oral azathioprine was started, and prompt relief of the muscle and skin stiffness ensued shortly. After three months prednisone was substituted for ACTH, and gradual absorption of the calcium tissue deposits continued until their almost complete resolution.

Physical examination in July 1985 disclosed congested jugular veins. The lungs were clear on auscultation with dullness over the right lung base on percussion, and the abdomen was distended by ascitic fluid. The liver was palpated 7 cm below the costal margin and had a span of 17 cm. The spleen was palpated 2 cm below the costal margin. Bilateral ankle oedema was present. Firm nodules were noted under the skin of his face, chest, arms, and thighs.

Haematological investigation showed normochromic and normocytic anaemia with haemoglobin concentration of 109 g/l and normal white blood cell and platelet counts. The serum bilirubin was 37.6 $\mu\text{mol/l}$ and urea 8.6 mmol/l. Alkaline phosphatase, transaminase, and lactic acid dehydrogenase were within the normal ranges. The Rose-Waaler test was positive (1/640), also the latex agglutination test (1/128). Roentgenograms of the chest showed a large right pleural effusion and few nodular calcifications in soft tissues. The cardiac silhouette was slightly enlarged. Abdominal ultrasound showed a large amount of ascitic fluid and an enlarged liver and spleen. The portal and hepatic veins were normal. Isotope scanning with technetium-99m phytate demonstrated a slightly enlarged liver and markedly enlarged spleen. The isotope was also concentrated in the bone marrow. An upper gastrointestinal series was normal and no oesophageal varices were found. Ascitic fluid was clear, yellowish, and contained 45 g/l protein. Cytological and bacteriological studies were normal and echocardiography disclosed normal ventricular function.

A diagnosis of liver cirrhosis was made, and the patient was discharged with salt restriction and diuretic treatment. No improvement ensued and he was referred again to our hospital in September 1985. Cardiac catheterisation was performed and disclosed a markedly increased pulmonary wedge pressure of 30 mmHg, comparable with the right

ventricular and left ventricular end diastolic pressures. Right ventricular diastolic pressure was half the systolic pressure and there was no atrial kick. The pressure curve of the right ventricle showed an early dip with plateau and the pressure curve in the right atrium demonstrated equal A and V waves with marked Y descent. Constrictive pericarditis was diagnosed, and pericardiectomy was performed. The pericardium was markedly thickened (1 cm) and diffuse adhesions were present between the pericardium and the myocardium with no loculated fluid. No calcium deposits were present. Histological examination showed marked fibrosis with diffuse lymphocyte and plasma cell infiltration.

After the operation normal cardiac contraction was restored. The patient had a marked diuresis, lost 9 kg in weight, and the ascites and oedema disappeared. He regained his strength rapidly, and the haemoglobin concentration increased to 152 g/l within three months.

Discussion

Our patient fulfilled the criteria for dermatomyositis according to Bohan *et al.*¹ He did not have any additional manifestations of an associated connective tissue disorder and had no previous history of acute pericarditis. Symptoms of constrictive pericarditis had developed 12 years after the onset of dermatomyositis.

In reviews analysing large numbers of patients with clinical manifestations of dermatomyositis and polymyositis, cardiac involvement is not uncommon.¹⁻³ Myocardial disease, heart block, and acute pericarditis have been described.⁵ None of the reported patients had constrictive pericarditis, however. Also, necropsies of 20 patients with cardiac involvement in polymyositis⁴ did not reveal constrictive pericarditis.

Constrictive pericarditis is a well known feature of rheumatoid arthritis and was reported in 0.64% of male and 0.06% of female patients.⁶ In all these patients rheumatoid factor was positive and the mean duration of the rheumatoid disease before the diagnosis of constrictive pericarditis was 9.4 years; the longest duration was 20 years. Rheumatoid arthritis was the cause in four out of 32 patients with constrictive pericarditis.⁷

The presence of rheumatoid factor is not unusual in dermatomyositis. It was found in 5% of patients with pure dermatomyositis¹ and in other classes of dermatomyositis/polymyositis.^{1,3} In our patient antinuclear antibodies were not present, and we therefore suggest that the constrictive pericarditis is secondary to dermatomyositis, a finding which, to our knowledge, has not previously been described.

The pathogenesis of constrictive pericarditis in autoimmune disorders is unknown. It may represent a late complication of acute pericarditis which passed unnoticed. As constrictive pericarditis has been described previously in autoimmune diseases it is reasonable to assume an association between the two diseases in our patients. We cannot, however, exclude entirely a coincidence of two unrelated disorders.

Constrictive pericarditis is known to mimic the signs of liver cirrhosis⁷ and the Budd-Chiari syndrome.⁸ In our case the diagnosis of cirrhosis was suggested by the presence of hepatosplenomegaly, ascites, abnormal liver function tests, and a suggestive isotope scan. The absence of oesophageal varices and the presence of jugular venous congestion, however, led to the suspicion of a cardiac origin for the findings.

In summary, chronic constrictive pericarditis may complicate various rheumatic diseases as well as dermatomyositis. In the presence of liver damage there should be a high degree of awareness of the existence of this complication as it is completely reversible if surgery is performed in time.

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