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Fatal myocarditis with acute polymyositis in a young adult

A. Bhan[1], S.I. Baithun[1,2], P. Kopelman[1,2] and M. Swash[2]

Departments of Histopathology and Medicine at [1]Newham General Hospital, London E13 and [2]The London Hospital, London E1, UK.

Summary: A 21 year old woman presented with acute polymyositis associated with fatal myocarditis. The significance of cardiac involvement in polymyositis is discussed in relation to this unusually fulminant case.

Introduction

Cardiac involvement may occur with polymyositis but is usually in elderly patients and after several months illness. In many instances, this chronic myocarditis proves fatal. In contrast, polymyositis presenting with acute, fatal myocarditis is exceptional; we report such a case.

Case report

A 21 year old caucasoid woman, who had lived all her life in East London, presented with a 4-day history of myalgia and muscular weakness starting in the legs and spreading to affect all skeletal muscles. There was no past or family history of muscular disorder, or drug abuse and no history of any recent illness or contact with an infectious disease. The patient had taken no medication. Examination showed severe generalized weakness in limb and axial muscles with marked muscular tenderness. There was no sensory abnormality and no associated skin rash. The patient was apyrexial, the blood pressure was 130/80 mmHg and there was sinus tachycardia (100 beats/min). The remainder of the examination was normal, in particular there was no clinical evidence of pericarditis, no audible cardiac murmurs and the patient did not have a gallop rhythm. An electrocardiogram (ECG) and chest X-ray were both normal. A presumptive diagnosis of a viral illness was made and the patient admitted for observation. The possibility of polymyositis was raised because of the myalgia and appropriate investigations requested. Urinary myoglobin was not detected.

Initial laboratory investigations showed a normal haemoglobin (12.8 g/dl), and a leucocytosis (12×10^9 WBC/l). The plasma urea was moderately raised (12.4 mmol/l) but plasma electrolytes were normal. An intravenous infusion of isotonic saline was commenced and the patient was given paracetamol (1 g/6 h) for her pain. By the following morning (16 hours after admission) her condition had improved and she no longer required analgesia. However, the laboratory telephoned the results of the admission creatine kinase which was markedly elevated (452,000 IU, normal range 5–200 IU) indicating the probable diagnosis to be acute polymyositis. The patient was immediately given a bolus intravenous injection of 200 mg hydrocortisone and an oral dose of prednisolone (80 mg); a muscle biopsy was also performed. Despite this treatment, the urine output began to diminish and by 36 hours after admission the patient had become oliguric. The plasma urea was 26.5 mmol/l, the plasma creatinine 799 μmol/l and potassium 6.2 mmol/l. Once again, urinary myoglobin was not detected. She was treated with intravenous glucose and insulin, calcium resonium by mouth, and fluid restriction and arrangements made for her immediate transfer to a renal unit for dialysis. Unfortunately before this could be accomplished the patient suffered a cardiac arrest from which she could not be resuscitated. At the time of death, the plasma potassium was normal. Subsequent autoantibody and viral antibody titres showed no significant elevations.

Pathology

Autopsy revealed gross pulmonary oedema with an enlarged heart, and normal coronary arteries. There was no evidence of infarction, atheroma or thrombus. Histological examination of muscle showed features of acute polymyositis, with widespread foci of muscle fibre necrosis associated with infiltration of small mononuclear cells. The cardiac

Correspondence: S.I. Baithun, M.R.C.Path., Histopathology Department, St Andrews Hospital, Devons Road, London E3 3NT, UK.

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muscle showed widespread foci of necrosis (Figure 1), with a prominent macrophage response and marked oedematous swelling of the myocardium. Microscopy of the lungs showed changes of adult respiratory distress syndrome. The kidneys showed acute tubular necrosis. Some tubules were filled by hyaline casts formed of rounded brown granules which showed a positive reaction for myoglobin.

Discussion

Polymyositis may be acute or chronic. Our patient suffered an acute illness of a few days duration, associated with a fatal myocarditis. Such a rapidly progressive illness in polymyositis is unusual.[1-4] In reported cases of myocarditis associated with polymyositis, post-mortem studies and endomyocardial biopsies show widespread myocardial necrosis with focal areas of relative sparing and evidence of regeneration.[5-8] The heart of our patient resembled this description, with widespread areas of myocardial necrosis, but there was no evidence of regeneration. Similar histopathological features were noted in the skeletal muscle biopsy taken prior to death. Absence of focal lymphocytic infiltration or microthrombi on histological examination makes it unlikely that these findings were the result of either viral infection or arteritis.

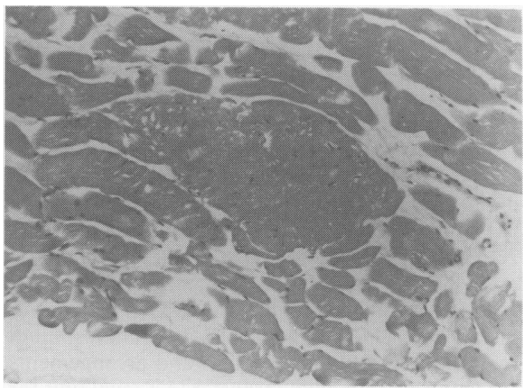

Figure 1 Section of heart showing muscle necrosis. Note absence of inflammatory cell infiltrate (H&E × 50).

It seems likely that the recognized, but rare, complication of rhabdomyolysis-induced acute renal failure in polymyositis[9] contributed to the terminal event although urinary myoglobin was not detectable during the course of the illness. Gabow and colleagues[10] have reviewed the aetiology, presenting features and biochemical abnormalities associated with acute rhabdomyolysis. They found alcoholism to be the most common aetiological factor followed by abdominal trauma and drug abuse. No case of polymyositis was included in the 87 patients reviewed. Nevertheless, their finding that myoglobinuria was absent in 50% of cases of rhabdomyolysis at the time of presentation is relevant to the present case report.

It is important for the clinician to be aware of the severity of acute myocarditis in association with acute necrotizing polymyositis in adults. In contrast to previous reports of fatalities (mean age 46 years), in this condition[11-13] our patient was much younger. Tachyarrhythmias and conduction abnormalities develop in more than 50% of acute and subacute cases of polymyositis, but these changes are almost invariably subclinical.[14]

Methods for monitoring the progress of myocarditis in polymyositis have been reported,[15] but continuous ECG assessment is possibly more important in acute cases than has been realized. Although the duration of polymyositis is not correlated with myocardial damage, the plasma creatine kinase level (isoenzyme MB) rises with progressive myocardial damage and may be a useful indicator. The rapid rise in muscle enzymes in our patient with a concomitant deterioration of renal function, probably due to rhabdomyolysis with myoglobinaemia, was indicative of severe acute polymyositis, and also emphasizes the vital importance of monitoring urine output and biochemical renal function in all suspected cases.

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