Heart transplant for dilated cardiomyopathy associated with polymyositis

A Afzal, R S D Higgins, E F Philbin

Abstract
Cardiac involvement is one of the most significant factors in the poor clinical outcome of polymyositis. The case of a 39 year old African American woman with polymyositis, cardiomyopathy, and severe heart failure who had orthotopic heart transplantation is described. Review of the literature reveals that cardiac manifestations of polymyositis are frequent and include conduction system abnormalities, myocarditis, cardiomyopathy, coronary artery atherosclerosis, valvar disease, and pericardial abnormalities.

Keywords: polymyositis; cardiomyopathy; heart failure; heart transplantation

A 39 year old African American woman developed proximal muscle pain and weakness in 1990. Clinical evaluation, including muscle biopsy, confirmed polymyositis and corticosteroids were given. Shortly afterwards she had evidence of congestive heart failure (dyspnoea and fatigue). An echocardiogram revealed modest impairment of left ventricular contractile function with an ejection fraction of 45%, and mitral and tricuspid valve regurgitation. An ECG showed first degree atrioventricular block, left bundle branch block, and non-specific abnormalities of the ST segments and T waves. Chest radiography showed cardiomegaly and bilateral pleural effusions.
and echocardiography revealed four chamber cardiac enlargement with an end diastolic dimension of the right ventricle of 5.5 cm and a left ventricular ejection fraction of 20%. Cardiac catheterisation showed moderate to severe elevation of right atrial and left atrial pressures, low cardiac output, severe left ventricular systolic dysfunction, and normal coronary angiograms.

Ten days after her first visit to the clinic, the patient was admitted to hospital for evaluation and management of worsening heart failure, and symptomatic sustained monomorphic ventricular tachycardia, which required resuscitation with direct current cardioversion. Due to severe haemodynamic compromise and recurrent cardiac arrhythmias, an automatic cardioverter defibrillator pacemaker device was implanted and she needed a prolonged stay in hospital. Continuous intravenous infusions of dobutamine hydrochloride, dopamine hydrochloride, and milrinone lactate were used to maintain haemodynamic stability. Renal insufficiency due to low cardiac output was treated with peritoneal dialysis. After 118 days in hospital, she had successful and uncomplicated orthotopic heart transplantation. Histopathological examination of her explanted heart (fig 3) showed areas of diffuse fibrosis and mononuclear inflammatory infiltrate in both the right and left ventricles; the right ventricle myocardium had largely been replaced by adipose tissue. These findings were consistent with myocardial involvement of polymyositis.

The onset of heart failure secondary to cardiomyopathy or myocarditis usually occurs in patients with active peripheral muscle involvement. It may, occasionally, be the presenting symptom of polymyositis or develop in a patient with improving peripheral muscle involvement. The most consistently observed histological findings include focal myonecrosis, interstitial oedema, round cell infiltration, and patchy fibrosis of both the myocardium and conducting tissues. The coexistence of fibrosis and myocarditis in the same patient suggests that cardiac involvement may be occurring in a relapsing fashion, resulting in a combination of acute inflammatory infiltrates and chronic replacement fibrosis. The onset of heart failure secondary to cardiomyopathy or myocarditis usually occurs in patients with active peripheral muscle involvement. The most consistently observed histological findings include focal myonecrosis, interstitial oedema, round cell infiltration, and patchy fibrosis of both the myocardium and conducting tissues. The coexistence of fibrosis and myocarditis in the same patient suggests that cardiac involvement may be occurring in a relapsing fashion, resulting in a combination of acute inflammatory infiltrates and chronic replacement fibrosis. The most consistently observed histological findings include focal myonecrosis, interstitial oedema, round cell infiltration, and patchy fibrosis of both the myocardium and conducting tissues. The coexistence of fibrosis and myocarditis in the same patient suggests that cardiac involvement may be occurring in a relapsing fashion, resulting in a combination of acute inflammatory infiltrates and chronic replacement fibrosis. The most consistently observed histological findings include focal myonecrosis, interstitial oedema, round cell infiltration, and patchy fibrosis of both the myocardium and conducting tissues. The coexistence of fibrosis and myocarditis in the same patient suggests that cardiac involvement may be occurring in a relapsing fashion, resulting in a combination of acute inflammatory infiltrates and chronic replacement fibrosis.
with acute myocardial infarction secondary to obstructive coronary artery disease, two of whom had a myocardial infarction shortly after the diagnosis of polymyositis. Disease of the small myocardial vessels with encroachment of the lumen by medial smooth muscle hyperplasia and little or no intimal proliferation has also been reported.\(^3\)

Valvar abnormalities are often limited to the mitral valve—for example, mitral insufficiency and mitral valve prolapse.\(^6\) Of 17 patients studied by Gottdiener,\(^4\) 65% had echocardiographic evidence of prolapse; an audible late systolic click was present in seven. Pericarditis or pericardial effusion diagnosed by echocardiography may be seen in up to 25% of patients;\(^5\) constrictive pericarditis and pericardial tamponade are seen rarely.\(^7\)

Close clinical follow up, routine screening (including electrocardiography and echocardiography), and prompt treatment may alter the natural course of the cardiac involvement of polymyositis and may reduce morbidity and mortality from this disorder.\(^7\)

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