Heart transplant for dilated cardiomyopathy associated with polymyositis

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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 an intraventricular conduction delay of the left bundle branch block type. The patient was evaluated by a cardiologist who prescribed diuretics, digitalis, and angiotensin converting enzyme inhibitors. Over the following years, the signs of heart failure became progressively worse and in September 1997 the patient was referred to our heart failure and transplantation clinic for management of severe dyspnoea, fatigue, chest pain, and palpitations.

The patient’s medical history was notable for large, symptomatic, uterine fibroid tumours as well as cardiac disease and polymyositis. She had two healthy children, aged 10 and 19 years, who were born by uncomplicated, spontaneous, vaginal delivery. Her medication included digoxin, metolazone, frusemide (furosemide), benazepril, prednisone, amiodarone, and potassium chloride. She did not smoke cigarettes or drink alcohol.

Her blood pressure was 92/60 mm Hg, pulse was 84 beats/min and irregular, and she was in mild respiratory distress. Her symptoms suggested decompensated biventricular heart failure, and included pulmonary rales, pedal oedema, distended neck veins, an enlarged and pulsatile liver, ascites, a third heart sound, and murmurs of mitral and tricuspid regurgitation. Liver transaminases were very abnormal, indicating congestive hepatopathy, and serum creatinine was 100 g/l. An ECG (fig 1) showed normal sinus rhythm, first degree atrioventricular block, left bundle branch block, and non-specific abnormalities of the ST segments and T waves. Chest radiography (fig 2) showed cardiomegaly and bilateral pleural effusions.

Figure 1  Admission 12 lead ECG.

Figure 2  Admission chest radiograph, anteroposterior view.
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 patient was discharged 38 days after the transplant in stable medical condition; she was medically well more than 14 months after the operation in New York Heart Association functional class II. She has experienced no episodes of severe cardiac allograft rejection and her left ventricular ejection fraction is 60%. Her liver function tests are normal, serum creatinine is 150 g/l, and she does not need dialysis.

**Discussion**

Polymyositis is a rare disorder, with an estimated annual incidence in the USA of five to 10 new patients per million population. The clinical features include proximal muscle pain and weakness. Typical laboratory findings include raised serum creatine kinase and elevation of other muscle enzymes including serum aldolase, which is reflective of the skeletal muscle injury caused by the inflammatory process. Biopsy of a proximal muscle such as the deltoid usually confirms the presence of inflammation. Cardiac involvement is now being described with increasing frequency and is one of the most significant factors linked to the poor clinical outcome of this disorder. Heart failure has been reported with variable frequency; Bohan et al noted an incidence of 3%, while Denbow et al and Oka and Rassakka noted a higher incidence of 45% and 25%, respectively. We are unaware of any previous report of a case of cardiomyopathy associated polymyositis which culminated in successful heart transplantation.

Cardiac involvement in polymyositis was first recognised in 1899. It was considered uncommon until recently, but as survival of patients with this disorder has improved, myocardial involvement has become increasingly important and is now recognised as the third leading cause of death from this condition after sepsis and malignancy. Studied prospectively, as many as 75% of patients with polymyositis have been reported to have definable cardiac abnormalities, including myocarditis, cardiomyopathy, conduction system abnormalities, coronary artery disease, valvar disease, and pericardial abnormalities.

Involvement of the myocardium, which is pathologically similar to skeletal muscle, can result in heart failure but is only rarely a cause of serious morbidity. 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. It may, occasionally, be the presenting symptom of polymyositis or develop in a patient with improving peripheral myositis.

Electrocardiographic abnormalities are seen in up to 70% of patients with polymyositis and are most commonly non-specific abnormalities of the ST segments and T waves (23% of patients). Other abnormalities include abnormal Q waves, atrial and ventricular arrhythmias, fascicular block, bundle branch block, first, second, or third degree atrioventricular block, and sick sinus syndrome.

Studies of coronary arteries have shown inflammatory changes (arteritis), and arteritis obliterans. Conversely, the patient may have angiographically normal coronary arteries, even in the presence of ECG changes which have been attributed to myocardial ischaemia. Oka and colleagues reported three patients...
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; 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