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. 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,
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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.
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.\(^7\)