Polymyositis is a progressive inflammatory myopathy which can affect the heart.14 Cardiac involvement during the course of polymyositis has been increasingly recognized in the last decade. Gottdiener et al., in a noninvasive study, found that 76 percent of their patients with polymyositis had evidence of cardiac involvement. Patients with conduction abnormalities are prone to develop complete heart block because of the widespread progressive destruction of specialized conducting tissue and replacement by fibrotic tissue.15

CASE REPORT

A 59-year-old man was hospitalized in March 1980 because of chest pain and dyspnea. We noted cardiomegaly, normal heart sounds and no murmurs. Neurologic examination results were normal. Chest x-ray films showed moderate cardiomegaly with mild pulmonary congestion. The ECG revealed bifascicular block (right bundle branch block and left anterior hemiblock) and first degree A-V block. Results of laboratory studies, including erythrocyte sedimentation rate, serum glutamic-oxaloacetic transaminase (SGOT), creatinine phosphokinase (CPK) and lactate dehydrogenase (LDH) were all normal. An echocardiogram showed a dilated, poorly contracting left ventricle. Cardiac catheterization revealed an enlarged, poorly contracting left ventricle, mild mitral regurgitation and 40 percent obstruction of the left circumflex artery. The patient was discharged with a presumptive diagnosis of idiopathic dilated cardiomyopathy.

In February, 1982 he was readmitted because of palpitations and vertigo. The ECG showed the same conduction defects and, in addition, periods of second degree A-V block (Wenckebach periodicity). An electrophysiologic study demonstrated an A-V nodal level of block. No treatment was given. Twenty months later (October, 1983), he was readmitted because of syncope and pulmonary edema. The ECG showed intermittent second and third degree A-V block. Physical examination revealed hepatosplenomegaly and symmetric proximal muscle wasting and weakness in both upper and lower limbs, as well as wasting of the paravertebral muscles. Laboratory studies showed marked elevation of serum muscle enzymes: CPK (of muscle origin, MM band) 2300 μ/l (normal under 145 μ/l), aldolase—16 μ/l (normal under 7.6 μ/l), SGOT—127 μ/l (normal under 40 μ/l), LDH—290 μ/l (normal under 225 μ/l). The erythrocyte sedimentation rate, hemogram, 

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creatinine, calcium, phosphorus, alkaline phosphatase, protein electrophoresis and thyroid gland function tests all gave normal results. The latex test, antinuclear factor and LE cells all provided negative results. Electromyography demonstrated a characteristic myopathic pattern. Right deltoid muscle biopsy confirmed the clinical diagnosis of polymyositis. Right ventricular endomyocardial biopsy was attempted, but for technical reasons did not succeed. A ventricular demand pacemaker was implanted and prednisone 60 mg a day was started and reduced gradually to 20 mg daily. A complete work-up revealed no evidence of underlying malignancy. Over the following nine months, steroid therapy suppressed his symptoms, but persistent raised levels of serum muscle enzymes indicated active, progressive skeletal muscle myositis. Noninvasive cardiac evaluation revealed no change in ventricular function or A-V conduction.

**DISCUSSION**

Polymyositis is an inflammatory myopathy of unknown etiology, characterized by acquired symmetric proximal muscle weakness, muscle pain and atrophy. Cardiac manifestations include mitral valve prolapse; congestive heart failure; hyperkinetic state; pericarditis; pulmonary hypertension; dissecting aneurysm of ascending aorta extending into the coronary arteries; atrial and ventricular arrhythmias; the sick sinus syndrome; nonspecific ST-T wave changes and conduction abnormalities, among them A-V block, bundle branch block and intraventricular conduction defects.

There is an apparent correlation between the duration and severity of skeletal muscle involvement to the cardiac manifestations and the extent of conduction system disease. However, the exact relationship between them is not well established. Characteristically, cardiac involvement during the course of polymyositis has been recognized once the typical features of skeletal muscle myositis are apparent. The present patient is very unusual in that he manifested typical cardiac disturbances associated with polymyositis three years before demonstrating any clinical or laboratory evidence of polymyositis. In the absence of endomyocardial biopsy, we cannot confirm the presumptive diagnosis of polymyositis heart disease. However, the gradual progression over a three-year period of conduction system disease that is typical for polymyositis cardiomyopathy raises the possibility of the existence of polymyositis heart disease in the absence of clinical or laboratory evidence of active skeletal muscle myositis.

The association between cardiac and skeletal muscle myositis has been observed in a variety of clinical entities. In cardiomyopathies, neuromuscular disorders and collagen vascular diseases, there is an undetermined relation between cardiac and skeletal muscle myopathy. On the basis of electromyographic and histologic studies, hypertrophic cardiomyopathy can be considered a generalized myopathy with early cardiac manifestations. Dunnigan et al. performed skeletal muscle evaluation in young patients with cardiomyopathy with no clinical evidence of skeletal muscle involvement and found abnormal electromyograph and histologic findings. They supported the concept that cardiomyopathy is not an isolated disease, but rather a partial clinical feature of a generalized myopathy with early cardiac manifestations. Cardiac involvement is also an integral part of neuromuscular disorders such as progressive muscular dystrophy of Duchenne and myotonic muscular dystrophy, where progressive conduction system disease or atrial arrhythmias are occasionally recognized before the systemic neuromuscular disorder is diagnosed.

From the accumulation of reported cases, including one recently reported patient who had complete heart block requiring a pacemaker installation several years before the development of polymyositis, it is obvious that striated muscle (both skeletal and cardiac) may be affected by diverse myopathic and neuropathic processes. This growing awareness of concomitant cardiac and skeletal muscle abnormalities suggests that more comprehensive evaluation (eg, electromyographic examination and skeletal muscle biopsy) should be taken in patients with primary cardiomyopathy in order to detect asymptomatic skeletal muscle abnormalities.

Cardiac involvement is encountered with a high incidence in advanced cases of polymyositis. However, some patients may have an early predisposition for cardiac manifestations rather than skeletal muscle symptoms. They present unexplained asymptomatic cardiomegaly, nonspecific ECG changes or conduction abnormalities while the clinical picture of polymyositis is nonapparent and undetectable by laboratory examinations. Our observation suggests that patients with unexplained cardiac manifestations, as mentioned above, should be followed for skeletal muscle symptoms and transvenous endomyocardial biopsy should be considered in order to evaluate the possible association of polymyositis.

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