

# Cardiac involvement in polymyositis: a case report and review of the literature<sup>1</sup>

N. V. Ramana Raju, M.D.<sup>2</sup>

Neil Hart, M.D.

James Maloney, M.D.

Adnan Zaidi, M.D.

Kenneth Adams, M.D.

**Cardiac involvement in polymyositis has recently been found to be more prevalent than had been believed, and its clinical spectrum to be wide and variable. Recent studies suggest that cardiac abnormalities are seen in approximately 75% of patients with polymyositis. The usual manifestations are congestive heart failure, arrhythmias, heart blocks, and electrocardiographic abnormalities. This case report describes the diagnosis and clinical course of cardiac polymyositis in a 57-year-old woman.**

**Index terms:** Heart diseases • Myositis

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Myocardial involvement in polymyositis, although long recognized, has been thought to be rare. Recent studies suggest that cardiac abnormalities are seen in approximately 75% of patients with polymyositis.<sup>1,2</sup> We present an unusual case of polymyositis with severe cardiac problems.

## Case report

A 57-year-old white woman was first seen at The Cleveland Clinic Foundation in December 1980, with a history of heart-pounding and severe fatigue and weakness. Her cardiac irregularity dated from 1970 when she was told of having "skipped heartbeats." She had been symptomatic with a history of a pounding sensation in the heart and occasional fainting spells since 1972. Cardiac catheterization, performed in 1976, revealed slight cardiomegaly with mild left ventricular dysfunction. Left ventricular end-dia-

stolic pressure was 24 mm Hg, and she was diagnosed as having primary myocardial disease. A Holter monitor reading, obtained in 1976, revealed sinus rhythm with occasional ventricular ectopy and paroxysms of atrial fibrillation. She was treated with propranolol and digoxin with some benefit. However, she developed a rash while taking quinidine and could not tolerate procainamide. At the time of her visit to the Cleveland Clinic, she was on disopyramide, propranolol, and digoxin.

Physical examination in December 1980 revealed an anxious white woman with an irregular pulse of 80 beat/min. Blood pressure was 150/90 mm Hg. There was no jugular venous distention. The lungs were clear. Heart examination revealed the point of maximal intensity (PMI) in the fifth left intercostal space at the anterior axillary line. The first heart sound (S1) varied in intensity, and the second heart sound (S2) appeared normal. A third heart sound (S3) was heard at the apex. There was no fourth heart sound (S4). The abdomen was soft and nontender without masses or distention. A slight decrease in motor strength was noted in both upper and lower extremities.

Laboratory studies disclosed the following results: complete blood count (CBC) was normal. Chest radiograph revealed mild cardiomegaly. An electrocardiogram (ECG) revealed atrial fibrillation with variable ventricular rate. Sequential multiple analysis-18 (SMA-18) profile revealed an increased creatine phosphokinase (CPK) enzyme to 922 U/L (normal, 20-180 U/L), and the fractionation of CPK isoenzymes showed greater than 95% MM fraction, suggesting its origin from skeletal muscle. Nuclear ejection fraction and wall motion studies were suggestive of left ventricular dysfunction. Electromyography of the upper and lower extremities was suggestive of polymyositis with florid myopathic changes. Muscle biopsy of the right vastus medialis showed histological changes compatible with polymyositis. Extensive investigations for connective tissue disorders and occult malignancy were negative.

At this time she was started on prednisone, 10 mg/day. Disopyramide was discontinued. Digoxin and small doses of

<sup>1</sup> Department of Cardiology, The Cleveland Clinic Foundation. Submitted for publication Feb 1983; revision accepted Sept 1983.

<sup>2</sup> Present address: 2500 N. Rockton Avenue, Rockford, IL.

propranolol were continued. She had significant symptomatic improvement, and the CPK enzyme decreased to 297 U/L. She was admitted in August 1981, with recurrence of palpitations and fatigue. The CPK enzyme was over 1000 U/L, and serum aldolase was 17.7 mU/ml. Her cardiac status was essentially unchanged except that she was having ventricular ectopy, mainly in the form of couplets. A stress test at this time provoked ventricular tachycardia. Electrophysiologic studies revealed dissociated atrial rhythm, accelerated idioventricular rhythm with retrograde conduction, and an accessory atrioventricular (AV) nodal bypass tract. Amiodarone was prescribed, and there was initial improvement in cardiac rhythm. Prednisone was increased to 60 mg/day at this time. In October 1981, she was admitted for profound sinus bradycardia. A repeat electrophysiologic study confirmed the presence of sick sinus syndrome (SSN), and a permanent transvenous pacemaker was implanted. Despite clinical improvement, ventricular ectopy persisted. In April 1982, extreme fatigue and palpitations recurred. Muscle enzymes were markedly elevated (CPK, 2500 U/L), and methotrexate was started. Although the symptoms of fatigue and weakness are reasonably well controlled, the ventricular ectopy and paroxysms of atrial fibrillation have persisted. She is presently being followed in the Cardiology Department.

## Discussion

Polymyositis was first described by Wagner in 1886.<sup>3</sup> The clinical syndrome refers to an inflammatory myopathy of unknown cause, characterized by weakness of the proximal muscles of the pelvis and shoulder girdle. The incidence is 0.1–0.6/100,000 persons per year.<sup>4</sup> It is most common in those between 40 and 60 years of age and in females (female to male ratio, 2:1).<sup>4</sup> Underlying malignancy is noted in 10% to 15% of cases,<sup>4</sup> common malignancies being those of the stomach, colon, rectum, and ovaries. Steroids are the mainstay of therapy. Methotrexate and azathioprine are used with variable success.

Cardiac involvement was first described by Oppenheim in 1899.<sup>5</sup> Reported ECG abnormalities are first-, second-, and third-degree heart blocks, bundle branch blocks, left axis deviations, and atrial and ventricular dysrhythmias.<sup>1,6</sup> Myocardial involvement with congestive heart failure<sup>7</sup> and pericardial involvement with pericarditis<sup>8</sup> are common.

Age distribution and sex incidence follow the classic concept of polymyositis, which is predominately a disease of middle-aged women. Female to male ratio is 2:1, and the mean age ranges from 44 to 53 years.<sup>1,2</sup> Cardiovascular symptoms as presenting complaints are clinically apparent in only 10% to 15% of biopsy-proved polymyositis, even though cardiac involvement is seen in 70% of the cases.<sup>1</sup> This suggests symptomatic

cardiac involvement is infrequent. Presenting cardiovascular manifestations are palpitations, nonspecific chest pain, congestive heart failure, and systolic murmur of mitral regurgitation.<sup>1,2</sup> In a prospective study done on 21 patients at George Washington Hospital,<sup>1</sup> the frequency of cardiovascular involvement discovered with non-invasive techniques appeared to be high. Echocardiogram, phonocardiogram, and ECG were done on all patients studied, more than one half of whom were female. Fifty percent had ECG abnormalities, 33% had an ejection click and systolic murmur on phonocardiogram, and 50% had mitral valve prolapse on echocardiogram. All patients in that study had normal left ventricular end-diastolic dimensions. It is also reported that the mean velocity of circumferential fiber shortening and fractional shortening was greater than for the control group, which is suggestive of high cardiac output.<sup>1</sup> One of the patients underwent cardiac catheterization, and the hemodynamic data, which tend to support this, show a CI of 6.8 lit/m<sup>2</sup> (4 lit) and stroke volume (SV) index of 90.5 ml (40 ml). The cause of this high output remains unclear. Postulated mechanisms include: (1) increased metabolic demand, (2) increased AV shunting in the affected muscles, and (3) production of a vasoactive amine.<sup>9</sup> In a clinicopathologic study done at The Mayo Clinic on 20 patients,<sup>2</sup> 40% had clinical evidence of congestive heart failure, 45% had abnormal chest radiographs, 72% had ECG abnormalities, and the female to male ratio was 2:1. At autopsy the mean heart weight was 355 g compared to 405 g in controls. Histological evidence of myocarditis and small vessel disease was present in 30% of patients. Diffuse interstitial and perivascular mononuclear cell infiltrate was present. Muscle degeneration and fibrous replacement of sinoatrial (SA) node and bundle branches were noted. In a study done in Finland,<sup>10</sup> 69% of the patients showed cardiac involvement. An interesting finding in this study involves a group of 6 patients who also had coronary artery disease in addition to polymyositis. In 5, angina symptoms appeared after polymyositis was diagnosed. Three had myocardial infarctions, and 2 died. At autopsy, no scar of myocardial infarction or arteriosclerosis was found. Macroscopically the changes in the coronary arteries are suggestive of polymyositis involvement. None of these patients had coronary arteriography. The ECG changes could be secondary to myopathy.

All the classic manifestations of cardiac involvement in polymyositis, namely, abnormal ECG, atrial and ventricular arrhythmias, varying degrees of heart block, and left ventricular dysfunction were noted in this case. There was no evidence of ischemic or other etiologic factors to account for the cardiac abnormalities. Myocardial biopsy would have been helpful in establishing a definitive diagnosis, but it was refused by the patient. However, the combination of elevated muscle enzymes, abnormal electromyography suggestive of polymyositis, typical skeletal muscle biopsy findings, and myocardial disease strongly suggests that the cardiomyopathic process represents involvement secondary to polymyositis. It is extremely rare for polymyositis to present in this way, because the clinical picture was dominated by cardiac abnormalities and skeletal muscle involvement was of minor clinical importance.

Cardiac involvement in polymyositis is frequent, and the clinical spectrum is wide and variable. Available data are incomplete and inconclusive. Further studies are needed to evalu-

ate the natural history, detection, management, and prognosis of the disease and to prevent myocardial involvement. The role of antiarrhythmic drugs, steroids, and immunosuppressive drugs should be assessed.

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