Cardiac involvement in polymyositis: Report of case and review of literature

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The involvement of cardiac muscle in polymyositis is now being recognized more frequently. Routine diagnostic studies such as electrocardiography, echocardiography, and Holter monitoring have revealed evidence of cardiac abnormalities in as many as 76 percent of patients. Utilization of noninvasive scanning techniques (thallium-201 and gated blood pool studies), when indicated, may increase the diagnostic yield to as high as 100 percent. The case reported here illustrates characteristic polymyositic cardiac involvement incidentally associated with hypertrophic cardiomyopathy. Guidelines for the diagnosis and management of the various cardiac manifestations of polymyositis are also discussed.

Polymyositis and dermatomyositis may be described as inflammatory disorders of skeletal musculature that result in symmetric weakness and/or atrophy of the proximal hip and shoulder girdles.^{1,2} These diseases are fairly well defined by criteria³ adapted from Bohan and Peter (1975), which include proximal muscle weakness, serum muscle enzyme elevation (particularly creatine kinase CK), typical electromyographic changes, characteristic muscle biopsy pathology, and classic dermatologic findings. Many extramuscular manifestations, including articular symptoms,⁴ pulmonary interstitial disease,⁵ decreased gastrointestinal motility,⁶ and Raynaud's phenomenon,⁷ have been described.

Dermatomyositis has also been associated with an underlying malignancy in as many as 10-30 percent of adult patients, especially patients over

40 years of age. It is often mentioned that males may predominate, but recent studies have shown that females may be affected in the majority of cases.⁸⁻¹⁰

Cardiac involvement in polymyositis, although first described by Oppenheim¹¹ as early as 1899, was thought^{12,13} to be a rare manifestation of this disorder until recent studies¹⁴⁻¹⁷ emphasized the frequency of atrial and ventricular dysrhythmias,¹⁵ cardiac conduction defects,¹⁸ hyperkinetic heart disease,¹⁹ congestive heart failure,¹⁶ mitral valve prolapse,¹⁵ and pericarditis²⁰ (Table 1). Postmortem studies^{16,21} have noted the prevalence of inflammatory and fibrotic changes in the myocardium and conduction system, as well as the rare finding of coronary artery vasculitis.²¹ These histopathologic changes seem to correlate well with the various clinical presentations^{16,17,21} and are analogous to the inflammatory changes noted in proximal skeletal muscles.

The following report describes an adult with primary idiopathic polymyositis. Not only did electrocardiographic changes related to his underlying muscular disease develop, but he was incidentally noted to have hypertrophic cardiomyopathy.

Report of case

A 53-year-old black man presented with a 4-week history of symmetric polyarthritides and mild myositis, affecting the shoulders, wrists, hands, elbows, and ankles, as well as a mild muscle pain. He complained of morning stiffness lasting approximately 1 hour, and positive physical findings included a grade 2-3/6 aortic systolic murmur, bibasilar rales in the lungs, and mild synovitis with thickening of wrists, MCP and PIP joints bilaterally. Small knee effusions were present as well. No skin rash, alopecia, or mouth lesions were noted.

The creatine kinase (CK) value was 1,725 I.U. (normal, 80-180), the serum glutamic-oxaloacetic transaminase level was 91 I.U. (normal, 10-40), and the serum glutamic-pyruvic transaminase value was 105 I.U. (normal, 5-45). Lactic dehydrogenase (LDH) was 440 I.U. (normal, 110-220) and the erythrocyte sedimentation rate (ESR)

TABLE 1. CARDIAC ABNORMALITIES IN POLYMYOSITIS.

Type of finding	Abnormality
Electrocardiographic	Dysrhythmia
	Conduction defect
	Left atrial abnormality
	Nonspecific ST-T wave
	Old myocardial infarction
Echocardiographic	Axis deviation
	Mitral valve prolapse
	Hyperkinetic cardiac state
	Pericarditis (rare)
Clinical	Systolic murmur (mitral)
	Congestive heart failure
Pathologic	Myocarditis
	Fibrosis
	Coronary vasculitis (rare)

was 124 mm./hour. Bibasilar pulmonary fibrosis was noted on routine chest x-ray examination. The EKG revealed no sinus rhythm at 90 beats per minute with nonspecific ST-T wave changes.

The patient was a moderate to heavy drinker (approximately 1,500 ml. of rum per week). He was treated with Clinoril (200 mg. twice a day) for tentatively diagnosed rheumatoid arthritis with mild myositis. He was instructed to refrain from alcohol.

He returned to the hospital 8 weeks later because of progression of symptomatology, stating that he had completely abstained from alcohol. At this time, a rheumatology consultant also found symmetric polyarthritis, as well as severe proximal myopathy (Grade II/V). Laboratory data included a CK value of > 15,000 I.U. and a serum aldolase level of 135 units (normal 0-6 units). A unilateral electromyogram yielded myopathic changes characteristic of polymyositis; biopsy of the right deltoid muscle showed inflammatory cellular infiltrate, patchy necrosis and fibrosis of muscle fibers, and variation of fiber size. A thyroid profile, including determinations of triiodothyronine, thyroxine, free thyroxine index, and thyroid-stimulating hormone, was normal, and *Toxoplasma* antibody titers (IgG and IgM) were negative. Other diagnostic studies, including complete gastrointestinal series and intravenous pyelography (to exclude occult cancer), were unremarkable.

There was no prior history of atypical chest pain, angina pectoris, hypertension, diabetes, or myocardial infarction. An electrocardiogram (EKG) that had been obtained 5 years prior to admission was normal. The present EKG again revealed nonspecific ST-T wave changes, a left atrial abnormality (P-wave mitrale), and the suggestion of an old myocardial infarction (QS complexes in precordial leads). Physical examination also revealed an aortic, systolic murmur, varying in intensity. Echocardiography disclosed systolic anterior motion of the anterior mitral valve leaflet with concentric left ventricular hypertrophy, which is in keeping with a diagnosis of hypertrophic cardiomyopathy.^{22,23} Therapy was initiated with 60 mg. of prednisone (drug of choice) per day in divided doses with significant improvement in symptomatology noted at 2 weeks.

Discussion

Evidence for cardiac involvement in polymyositis has been greatly enhanced by the utilization of current, noninvasive techniques. The routine EKG has disclosed abnormalities from 33 to 70 percent of patients,^{16,24} with nonspecific ST-T wave changes, left atrial abnormalities, axis deviation, and varying degrees of atrioventricular block being recognized most frequently.¹⁶ Complete heart block necessitating pacemaker insertion has even been reported.¹² Other significant findings have included atrial and ventricular dysrhythmias as well as EKG evidence of old myocardial infarction^{16,25} (in 1 case²⁵ despite normal coronary arteries at angiography). One investigator²⁶ has shown that continuous 24-hour EKG monitoring (Holter) may even increase the diagnostic yield of potentially serious dysrhythmias in polymyositis, thereby recommending periodic Holter monitoring of this group of patients.

Gottdiener and coworkers¹⁵ have employed the echocardiogram and phonocardiogram in addition to the EKG in their prospective evaluation of cardiac function in polymyositis. Analysis of their findings unveiled evidence of cardiac abnormalities in 76 percent of patients (16/21), with an overall incidence of mitral valve prolapse in 65 percent (11 of 17 studied). Further evidence for the hyperkinetic cardiac state characterized by enhanced left ventricular systolic function in the etiology of congestive heart failure also was discussed.

Askari²⁶ prospectively evaluated 8 patients with polymyositis who had elevation of the CK-MB isoenzyme fraction to >3 percent of the total CK. Not only were EKG, Holter monitor, and echocardiogram recorded, but thallium-201 scintiscan and gated blood pool studies were performed. The inclusion of these noninvasive scanning techniques led to the detection of rather subtle myocardial abnormalities, which in turn led to the diagnosis of cardiac involvement in 100 percent of patients. Either patient selection (elevated CK-MB fraction) or improved diagnostic techniques may well have been responsible for this diagnostic frequency. Askari also suggested that elevation of the CK-MB band in this subgroup of patients served as an indicator of poor prognosis.

The patient in the case reported here certainly exemplifies some of the common cardiac abnormalities of polymyositis by the development of various EKG abnormalities in concert with the onset of inflammatory muscular disease. The electrical evidence of nonspecific ST-T wave changes, left atrial abnormality, and old myocardial infarction could theoretically be attributed to his hypertrophic cardiomyopathy, except that these findings were ab-

sent prior to the development of polymyositis. One could also reason that alcoholic myopathy, which has been well described²⁷⁻²⁹ and is often difficult to distinguish from polymyositis, was the cause of his muscular disorder. The patient, however, exhibited bibasilar pulmonary fibrosis, symmetric polyarthritides, and disease progression (CK elevation from 1,700 to >15,000 I.U.) despite abstinence from alcohol. These features, as well as the cardiac findings, are more characteristic of a connective tissue disorder than of acute or chronic alcoholic myopathy. Also, there was no echocardiographic evidence of alcoholic cardiomyopathy.

The histologic features of hypertrophic cardiomyopathy²² differ significantly from those of polymyositis, so a cause-effect relationship is difficult to postulate as the sole explanation for the cardiac picture in this case. Recent evidence,^{30,31} however, has also noted the association between cardiac and skeletal muscle myositis, supporting the theory that cardiomyopathy in some cases may be a partial clinical feature of a generalized myopathy. Nonetheless, this would appear to be one of the first reported associations of hypertrophic cardiomyopathy with polymyositis.

Comment

Cardiac involvement in polymyositis is remarkably more common than previously presumed. This extramuscular manifestation is now recognized in the majority of patients. The use of noninvasive cardiac studies, such as EKG, chest x-ray, CK-isoenzymes, Holter monitor, and echocardiogram, may better define polymyositis cardiac involvement and, therefore, guide its therapy. The specific management of the patient with an elevated CK-MB band may be enhanced further by use of noninvasive scanning (thallium-201 and gated blood-pool studies).

Treatment of a cardiac manifestation of polymyositis usually consists of aggressive control of the underlying disease process by utilization of corticosteroids and/or immunosuppressive agents, when indicated. Further therapy is individualized and dependent upon the specific clinical setting—for example, antiarrhythmics for dysrhythmias, pacemaker for significant heart block, and diuretics for congestive heart failure. Through an increased awareness of cardiac involvement and the appropriate use of diagnostic and therapeutic modalities as described, it is anticipated that decreased morbidity and mortality from polymyositis heart disease might result.

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