Mitral and Aortic Valve Replacement in Valvular Rheumatoid Heart Disease*

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Specific endocarditis involving the aortic and mitral valves in a patient with peripheral rheumatoid arthritis is reported. The patient underwent prosthetic replacement of both valves. Typical rheumatoid nodules were detected histologically in the valvular tissues.

Peripheral rheumatoid arthritis may involve the heart either specifically, with nodules located anywhere in the heart and similar to those found in the subcutaneous tissue, or nonspecifically, with pericarditis, myocarditis, endocarditis and coronary arteritis, without typical granulomas. The aspecific involvement may be considered a healed variety of the specific form.1-4 However, specific valvular involvement has been reported rarely.1-7

We describe a patient with peripheral rheumatoid arthritis in whom mitral and aortic valve involvement was so severe that surgical replacement of both valves was necessary.

CASE REPORT

Our patient, a 38-year-old-woman, had been receiving corticosteroid therapy since age 22 for serosopitive peripheral rheumatoid arthritis. Early in 1976 she began complaining of exertional dyspnea, cough and frequent hemoptysis; signs of left cardiac failure were detected and a mitro-aortic disease was suspected.

In December, 1976, as her condition markedly deteriorated with appearance of episodes of paroxysmal nocturnal dyspnea, she was admitted to our service for hemodynamic evaluation and possible surgical correction.

Physical examination showed orthopnea and mild ankle edema; blood pressure was 140/40 mm Hg. There was a fusiform swelling of the proximal interphalangeal joints and ulnar deviation of the 4th and 5th fingers of both hands, due to subluxation of the metacarpophalangeal joints with hypotrophy of the tenar and hypotenar muscles. The ankles were tender and swollen; passive and active movements of the hands, shoulders and knees were partially limited. Pressure on the carpal, tarsal and metatarsal bones was painful.

A grade 3/6 holosystolic murmur at the apex, radiating to the left axillary region, and a grade 3/6 decrescendo diastolic murmur on the Erb point were noted. Bilateral basal rales were also present.

Pertinent laboratory findings were: no bacterial growth on numerous blood cultures; positive latex test for rheumatoid

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chest, 3+, an increased erythrocyte sedimentation rate. The antistreptolysin 0 titer was 186, a lupus erythematosus cell preparation was slightly positive, and antinuclear antibodies were detected.

An anteroposterior chest x-ray film showed an enlarged cardiac shadow; no radiologic evidence of ankylosing spondylitis was observed, while gross deformities of the carpal and metacarpal bones were noted. ECG disclosed sinus tachycardia, and signs of left ventricular hypertrophy.

Right and left cardiac catheterization was then performed showing an aortic pressure of 140/50 mm Hg, a left ventricular pressure of 150/0-40 mm Hg, a pulmonary wedge pressure of 25 mm Hg average and a pulmonary pressure of 45-15-30 mm Hg. An injection of contrast material in the aortic root was done revealing a severe reflux in the left ventricular cavity. The angiographic study of the left ventricle, which appeared dilated and hypokinetic, showed a moderate reflux of contrast material into the left atrium through the mitral orifice. The diagnosis of severe aortic incompetence, moderate mitral regurgitation and impairment of left ventricular function was reached and surgical replacement of mitral and aortic valves was indicated.

On December 11, 1976, the patient underwent open heart surgery. The mitral valve, which presented retracted and thickened leaflets and partial fusion of the chordae tendineae, was replaced with a Hancock 29 mm xenograft; the aortic valve, whose cups were rigid and thickened, was severely incompetent and was replaced with a Lillehei-Kaster 23.5 mm prosthesis. No evidence of calcification, commissural fusion, and perforation was found (Fig 1).

Histologic examination of the leaflets disclosed a moderate fibrous thickening of the cusps and the presence of typical rheumatoid nodules at their basal attachment in both valves. The nodules were formed by central necrotic debris surrounded by histiocytes, not arranged in the characteristic palisade-like fashion, and occasional lymphocytes, polymorphs and multinucleated giant cells of the foreign body type (Fig 2).

Culture of the valvular tissue did not lead to any bacterial growth, and a final diagnosis of granulomatous rheumatoid endocarditis of the aortic and mitral valves was advanced. The postoperative course was uneventful and at this writing, three months following surgery, the patient is doing well.

**FIGURE 1.** Gross anatomy of the excised aortic valve.

Specific valvular endocarditis seldom complicates the natural history of peripheral rheumatoid arthritis. To our knowledge, 24 cases with specific rheumatoid granulomas involving the cardiac valves have been described in the English literature.4-7 Involvement was univalvular in 15 patients, bivalvular in three, trivalvular in three, and quadrivalvular in the remaining three, with a definite preference for the mitral, followed by the aortic, tricuspid and pulmonary. The nature of the cardiac lesions was recognized at autopsy, and the exact location of the nodules was within the substance of the leaflets, surrounded by a border of fibrous tissue.6,4 Perforation of the valvular leaflets has never been described in peripheral rheumatoid valvulitis. Our histologic findings are in agreement with these observations.

Most patients with valvular rheumatoid heart disease have only mild aortic and mitral involvement and they do not warrant valvular replacement. Our case is the first one of its kind to undergo surgery for valvular dysfunction due to rheumatoid arthritis of the peripheral type. A second patient, who presented with the same type of rheumatoid arthritis, underwent aortic valve replacement at our center because of acute regurgitation due to perforation of two of the cusps. Unfortunately, the specific nature of the valvular lesion was not clearly demonstrated histologically, and therefore, this case could not be considered in this report.

The prognosis of our patient, whose conditions at present are markedly improved, depends upon the natural course of the rheumatoid cardiac involvement, as well as upon the function of the valvular prostheses. In this regard, we wish to stress that all reported autopsy cases with specific rheumatoid valvulitis showed extensive cardiac involvement by rheumatoid granulomas.

**FIGURE 2.** Histologic section of the anterior leaflet of the mitral valve. The cusp is thickened and fibrotic; on its basal portion a typical rheumatoid nodule with central zone of necrosis is present (Hematoxylin and eosin, original magnification x 60).

**DISCUSSION**

Specific valvular endocarditis seldom complicates the natural history of peripheral rheumatoid arthritis. To our knowledge, 24 cases with specific rheumatoid granulomas involving the cardiac valves have been described in the English literature.4-7 Involvement was univalvular in 15 patients, bivalvular in three, trivalvular in three, and quadrivalvular in the remaining three, with a definite preference for the mitral, followed by the aortic, tricuspid and pulmonary. The nature of the cardiac lesions was recognized at autopsy, and the exact location of the nodules was within the substance of the leaflets, surrounded by a border of fibrous tissue.6,4 Perforation of the valvular leaflets has never been described in peripheral rheumatoid valvulitis. Our histologic findings are in agreement with these observations.

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Thus, we cannot exclude the presence of myocardial involvement in our patient even though no clinical signs of myocarditis, such as rhythm disturbances, have been detected. We also ignore whether there was any pulmonary and tricuspid valve specific involvement. However, the clinical findings seemed to be related to a dysfunction of the mitral and aortic valves only.

We suggest that prosthetic replacement may be regarded as an effective treatment also in this variety of valvular disease.

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Dissimilar Atrial Rhythms Diagnosed by Echocardiography

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A 27-year-old woman with ectopic atrial rhythm and apparent atrioventricular dissociation was found to have A waves on the mitral echocardiogram that bore no relation to the P wave on the surface electrocardiogram. The ECG did not reflect the presence of dissimilar atrial rhythms, and their presence became apparent only during the echocardiographic studies. The mechanism of this phenomenon and its clinical implications are discussed.

Dissimilar atrial rhythms were first described by Hering.1 Recent reports by Wu et al2 by Zipes and DeJoseph3 and by Friedman et al4 have stressed the limitations of surface electrocardiographic studies in the diagnosis of dissimilar atrial rhythms and have described invasive techniques used to detect such rhythms. In this report, we describe a patient with an ectopic atrial rhythm and apparent atrioventricular dissociation who echocardiographically had mitral A waves which were dissociated from the electrocardiographic P waves. Left atrial mechanical systole was not related to the electrocardiographic inscription of atrial activity and therefore represents an example of dissimilar atrial rhythms. To the best of our knowledge, this is the first report of dissimilar atrial rhythms diagnosed by echocardiographic studies.

CASE REPORT

The patient, a 27-year-old woman, was admitted to the William Lifikoff Cardiovascular Institute of Hahnesmann Hospital, Philadelphia, for evaluation of a cardiac murmur and fatigue. Following a syncopal episode at the age of 13 years, the patient had undergone cardiac catheterization, which demonstrated mild mitral insufficiency. She was well until five months prior to admission, when she complained of fatigue and also admitted to occasional episodes of palpitations.

On physical examination, the heart rate was 76 beats per minute and regular, and the blood pressure was 110/70 mm Hg. The point of maximal impulse was in the fifth intercostal space, 1 cm to the left of the midsclavicular line. The first and second heart sounds were normal, and a soft intermittent third sound was audible. A midystolic murmur was best heard at the apex and radiated to the left sternal border. No clicks were audible.

An electrocardiogram (Fig 1) showed a normal sinus rhythm at a rate of 73 beats per minute. The P-R interval was 0.18 second, and the axis was +50°. Voltage criteria for left ventricular hypertrophy and diffuse minor nonspecific ST-segment and T-wave abnormalities were present. Chest x-ray film was normal. Phonocardiogram confirmed the presence of the mid-to-late systolic murmur, and an echocardiogram was consistent with the possibility of prolapse of the mitral valve (Fig 2). Electrocardiographic monitoring (lead 2) during the echocardiographic study showed several episodes of an intermittent ectopic atrial rhythm at a rate of 94 beats per minute and atrioventricular dissociation, with the ventricles controlled by a different pacemaker at a rate of 66 beats per minute. There was no relationship between P waves and mitral A waves or QRS complexes, but the A waves on the echocardiogram had a constant relation to the ventricular complexes (Fig 2).

FIGURE 1. Standard ECG shows normal durations of P-R interval and QRS complex, minimal nonspecific changes in ST segment and T wave, and voltage criteria for left ventricular hypertrophy.

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