Constrictive Pericarditis Complicating Disseminated Lupus Erythematosus

By Peter M. Yurchak, M.D., Samuel A. Levine, M.D., and Richard Gorlin, M.D.

Disseminated lupus erythematosus, with its manifold complications, is being recognized with increasing frequency. Therefore, it is of some interest to describe a fully documented case with a hitherto unreported complication—constrictive pericarditis.

Case Report

H.B., age 58, was first admitted in 1958 because of exertional dyspnea and palpitations. He gave no clear-cut history of prior rheumatic fever, although an episode of pain beneath the left arm at age 18 was diagnosed as such. Physical activity was voluntarily restricted thereafter. A murmur was first heard when he was 19 years of age. He was asymptomatic until 3 months before his admission, when rapid palpitations first appeared. Apparently, these were related to documented episodes of atrial flutter and fibrillation, which reverted to sinus rhythm with digitalis and quinidine. Also, he had experienced brief spells of substernal oppression unrelated to exertion, which subsided spontaneously. There was no history of hemoptysis, or recurrent pulmonary infections. In 1930 he had contracted syphilis, which was treated with a series of injections. Subsequent blood and spinal fluid serologic tests for syphilis were allegedly negative.

On physical examination he appeared quite well. Venous pressure was normal. Heart rhythm was regular at a rate of 90 per minute. The first sound was increased; the second sound was described as normal. A grade-I systolic murmur was noted, along with a third sound in diastole. No definite presystolic rumble or other diastolic murmur was audible at any time. The lungs were clear, and there was no edema. The electrocardiogram showed first-degree atrioventricular block, with a P-R interval of 0.40 second. The P waves were broad and notched. The pattern of right ventricular hypertrophy and incomplete right bundle-branch block was present. The ST segments and T waves showed changes of digitalis effect. Chest roentgenograms (fig. 1) with fluoroscopy, showed enlargement of the transverse cardiac diameter to 25 per cent above predicted. Biventricular and slight left atrial enlargement were present. Pulmonary vasculature and hilar markings were prominent without hilar dance. Moderate pulmonary congestion was present. Calcification was seen in the region of the mitral valve. Blood serology was negative. The clinical impression was slight rheumatic mitral valvulitis without significant stenosis and myocardial disease. He was discharged on a medical regimen.

Subsequently he did well, having occasional palpitations as well as substernal distress precipitated by exertion, emotional upset and heavy meals. Seven months before re-entry (March 1962) he was seized by sudden, severe substernal pleuritic pain. A friction rub was heard.

Figure 1

Posteroanterior x-ray of heart showing generalized enlargement and pulmonary plethora.
and the diagnosis of acute viral pericarditis was made elsewhere. He was treated with prednisone and the pain abated. Persistent night sweats and intermittent low-grade fever appeared. Chest pain recurred as the dose of prednisone was reduced, but subsided with increase in dose. When steroid therapy was again terminated, he noted persistent lassitude, easy fatigue, anorexia, and slight weight loss. On close questioning subsequently, he recalled the appearance of an erythematous, scaly rash over the malar eminences and bridge of the nose.

On re-admission (October 1962) the history of rash described above drew attention to its faint residual. Jugular venous pressure and wave form were normal. Carotid pulsations were normal. The heart showed a normal left ventricular impulse. The first sound was increased. Aortic and pulmonic closure sounds were normal. Contrary to previous impression, the second sound was thought to show constant splitting, which widened only slightly with inspiration. A grade-II systolic murmur and grade-II late diastolic rumble were audible along the left sternal border. A to-and-fro friction rub was present in the third left interspace initially, but subsequently vanished. The liver edge was felt 6 cm. below the right costal margin, and the spleen was felt 4 cm. below the left margin. The remainder of the examination was unremarkable.

Preliminary laboratory studies showed a normal hematocrit value, white blood count, and differential. The corrected erythrocyte sedimentation rate (Wintrobe) was 20 mm./hr. The electrocardiogram and chest roentgenograms were unchanged. The amplitude of cardiac pulsations was thought to be normal.

The clinical impression on this admission was disseminated lupus erythematosus, possible Libman-Sacks endocarditis of the mitral valve, and possible constrictive pericarditis. The possibility of an atrial septal defect was also strongly considered for the first time because of the finding of fixed splitting of the second heart sound (previously undetected), and continued evidence on the electrocardiogram of incomplete right bundle-branch block.

Subsequent studies included a positive fluorescent antibody test for lupus erythematosus and two positive LE-cell preparations. The anti-streptolysin-O titer was less than 100 Todd units, and the C-reactive protein was 3+. The latex fixation test for rheumatoid factor was negative. Venous pressure was 15 cm. of saline, and circulation time (arm-to-tongue) was 22 seconds. The intermediate strength PPD skin test was positive.

At right heart catheterization, the catheter traversed a large, high-lying atrial defect into the left atrium and ventricle. Relevant data are given in table 1. There was no gradient across the mitral valve, but an 8-mm. gradient was measured across the aortic valve. Aortic valve area was calculated at 1.9 cm. Oxygen saturation values showed marked step-up at the atrial level, and oxygen saturation in the pulmonary artery approached that in the systemic artery. Calculation of pulmonary blood flow indicated an enormous shunt. A 4-mm. tricuspid gradient was measured, probably related to the torrential flow. Systemic output was 2.8 L./min./M.² (calculated from arterial dye-dilution curves following left ventricular injection). Because of difficulty in manipulating the catheter, it was not possible to appose the tip to the lateral wall of the right atrium in order to demonstrate thickening. No anomalous pulmonary veins were entered by the catheter.

Left ventricular volume was measured at rest and during intravenous infusion of isoproterenol, with thermodilution technics. The end-diastolic volume was 96 ml./M.² at rest (normal), and diminished to 81 ml./M.² during isoproterenol infusion. End-diastolic pressure, however, rose from 18 mm. to 23 at the same time.

After cardiac catheterization, a spiking fever to 102 F. appeared. Four blood cultures showed no growth. On the assumption that the stress of the procedure had activated the patient's collagen disease, Prednisone was begun in a dose of 10 mg. daily. Ten days later the patient underwent open-heart surgery by Dr. Robert Gross. The preoperative diagnosis was atrial septal defect, probable pericardial constriction, and rheumatic mitral valvular disease. At operation the heart beat was very weak and the pericardium quite thick and diffusely adherent to the heart. The portion overlying the right atrium was said to be "shoe-leathery" in character, be-

<table>
<thead>
<tr>
<th>Catheterization Data</th>
<th>Pressure (mm. Hg)</th>
<th>O2 saturation (%)</th>
</tr>
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<tbody>
<tr>
<td>Superior vena cava</td>
<td>—</td>
<td>52.0</td>
</tr>
<tr>
<td>Right atrium, mean</td>
<td>15</td>
<td>56.0 (high)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>91.0 (low)</td>
</tr>
<tr>
<td>Right ventricle, s/d</td>
<td>48/15</td>
<td>86.0</td>
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<tr>
<td>Right ventricle, diastolic mean</td>
<td>11</td>
<td></td>
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<tr>
<td>Pulmonary artery, s/d</td>
<td>41/15</td>
<td>87.0</td>
</tr>
<tr>
<td>Pulmonary artery, mean</td>
<td>25</td>
<td></td>
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<tr>
<td>Left atrium, mean</td>
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<tr>
<td>Left ventricle, s/d</td>
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<tr>
<td>Left ventricle, systolic mean</td>
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<tr>
<td>Left ventricle, diastolic mean</td>
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<tr>
<td>Brachial artery</td>
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<tr>
<td>Brachial artery, systolic mean</td>
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</table>
ing 6 to 8 mm. thick in some areas. The pericar-
dium was dissected free, resulting in good ven-
tricular contractions. A high secundum-type sepal defect was found on opening the right atrium, measuring 2.0 by 2.5 cm. The pulmonary veins from the right lung were found to enter the right atrium. The defect was closed and the anomalous venous drainage diverted into the left atrium. The mitral valve was explored digitally through the defect before closure. The surgeon described “some irregular calcifications in its annulus, and some nubby, hard material in part of the valve leaflet.” He added, “on the whole the leaflets moved with considerable mobility and softness.” The mitral orifice admitted approximately two fingers. Microscopic examination of the pericar-
dium removed (fig. 2), showed fibrous tissue with a high degree of hyalinization in some areas and, in others, coarse bundles of collagen. Foci of round-cell infiltration were seen.

Postoperatively the patient developed a vari-
ety of supraventricular arrhythmias, eventuating in atrial fibrillation refractory to all attempts at reversion. He was discharged taking daily digox-
in, chlorothiazide, potassium chloride and predni-
sone.

Discussion

The diagnosis of disseminated lupus ery-
thematosus in this patient appears well sub-
stantiated by clinical and serologic features recounted above. However, several features of this case are unusual. Most notable is the occurrence of pericardial constriction follow-
ing acute pericarditis due to disseminated lupus erythematosus. Careful search of the literature discloses no previously reported case. Acute pericarditis is said to occur in about one third of all cases of disseminated lupus erythematosus. When it is the initial sign of the disease, it is often misdiagnosed as the idiopathic variety. McCuiston and Moser have recently drawn attention to this pitfall and stress the importance of obtaining appropriate laboratory studies when confron-
ted with a case of acute pericarditis. Pericardial effusion, occasionally with tamponade, is known to occur in cases of clinically detectable lupus pericarditis, and pericardio-
centesis may be life-saving. Information available from the referring hospital indicated that effusion was probably not present in our patient. Fibrinous adhesions are usually found in the pericardial space at postmor-

![Figure 2](http://circ.ahajournals.org/)

**Figure 2**
High-power microscopic view from junction of pleura and pericardium, showing connective-tissue changes and inflammatory disease. Note the thick-walled, abnormal arteriole (center).

stem examination, but it has been said that constriction does not occur. Most cases of constriction follow infection, especially tuberculosis or trauma. Indeed, the occurrence of constriction following acute nonspecific pericarditis is rare. Azar found only 11 instances in his review of the literature, and added two cases of his own. Constriction de-
veloped within 6 months of the acute episode in eight of these 13 cases, as in ours. Subsequently, five additional cases have been mentioned elsewhere. It is notable that several cases of constrictive pericarditis complicating typical documented rheumatoid arthritis have been reported. Acute pericarditis may occur in rheumatoid arthritis, though less commonly than in disseminated lupus erythematosus, and may antedate manifestations of the
Constrictive pericarditis is said to occur uncommonly in patients with established rheumatic heart disease. The cardiac lesion traditionally associated with disseminated lupus erythematosus is so-called “verrucous endocarditis,” originally described by Libman and Sacks. It is said to occur in 30 to 50 per cent of cases coming to autopsy. These small ovoid vegetations are found most often on the mitral and tricuspid valves. No case of anatomic stenosis due to the lesion has yet been reported. Standard textbooks of pathology indicate that the lesions do not calcify. In our case, the “nubby, hard material in part of the valve leaflet” felt by the surgeon was not biopsied. However, the presence of calcification in the annulus and coexistence of mild aortic stenosis suggest that it represents old, healed rheumatic valvulitis.

Special Note on the Value of Ventricular Volume Measurement

The hemodynamic features of pericardial constriction have been well described previously. Basically, the diffuse pericardial lesion imposes the same filling pressure on both ventricles, resulting in a “plateau of pressures”—atrial and ventricular diastolic mean pressures being within a few millimeters of mercury of the same value. Furthermore, stroke output remains fixed and cardiac output changes only as a function of heart rate. This is well seen in our case in which cardiac output rose from 1.6 to 2.6 L./min./M. in response to isoproterenol, although stroke volume changed hardly at all (21 and 24 ml./beat/M., respectively).

The effect of an atrial defect on mean atrial pressures has also been discussed in detail. With an intact septum, left atrial pressure is a few millimeters higher than that of the right. A sizable defect results in equalization of pressures, usually at a normal level. The mean level is abnormally high when any obstruction to left ventricular filling coexists such as mitral stenosis, failure, or restricted compliance of the left ventricle. In our case, left ventricular end-diastolic pressure was 18 mm. Hg, and no mitral gradient was measured. The unusually high end-diastolic pressure could be ascribed to one or more possibilities: (1) true left ventricular failure; (2) constriction of the left ventricle by pericarditis; (3) encroachment on the left ventricle by displacement of the septum from an overfilled right ventricle. An unusually small left ventricle has been found at surgery in a few cases of atrial defect, supporting this latter possibility. It was in excluding one of these latter possibilities that measurement of left ventricular volume proved valuable. In left ventricular failure, both a rise in end-diastolic pressure and in end-diastolic volume (due to dilatation) would be expected. In both pericardial constriction and right ventricular encroachment, end-diastolic volume should be low in relation to end-diastolic pressure. Volume measurements do not differentiate between constriction and encroachment, however. Constriction was presumed to be present over both ventricles and the small difference in left ventricular and right ventricular diastolic pressures probably was due to non-simultaneity of pressure measurement. Despite the apparently equal distribution of restriction to ventricular filling (which should influence distensibility of both ventricles equally), enormous left-to-right shunting still occurred. Since filling pressure with sizable atrial defects is equal, shunting from left-to-right is usually explained by the greater compliance (or distensibility) of the right as compared to the left ventricle. The size of the shunt in our case would suggest that compliance of the two ventricles did differ in early diastole, and were only affected by pericardial rigidity late in diastole, after rapid filling and left-to-right shunting had already occurred. The paradoxical reduction in left ventricular end-diastolic volume with increase in end-diastolic pressure during isoproterenol infusion suggests some factor causing an additional increase in rigidity of the left ventricle, so that a lower volume of fill-
ing resulted in a higher pressure. Short of technical error, this could only occur with increased external compression. Conceivably, an increase in volume of the shunt with increased right ventricular end-diastolic volume within the fixed pericardial volume, could cause reduction in left ventricular volume. As both chambers fill at nearly equal pressures, the more distensible (right) chamber will fill earlier to a greater volume. Pericardial constriction complicating atrial septal defect has been reported previously.\(^4,^6\) As in our case, cardiac catheterization showed both a left-to-right shunt at the atrial level and the “plateau of pressures” of constriction.

**Summary**

A case of constrictive pericarditis complicating disseminated lupus erythematosus is reported in a 58-year-old man. Hemodynamic studies documented the diagnosis and also confirmed the clinical suspicion of coexistent atrial septal defect. Measurement of left ventricular volume by a thermodilution technic augmented information available from routine studies. Pericardiectomy and closure of the atrial septal defect relieved the mechanical lesion.

The case is also unique in its documentation of the coexistence of three different forms of heart disease—congenital atrial septal defect, pericardial constriction from lupus erythematosus, and rheumatic valvular disease.

**Acknowledgment**

We are indebted to Dr. Robert Vawter, Department of Pathology, Children’s Hospital Medical Center, for the photomicrograph of the surgical specimen.

**References**


As no two faces, so no two cases are alike in all respects, and unfortunately it is not only the disease itself which is so varied, but the subjects themselves have peculiarities which modify its action.—Sir William Osler. Aphorisms From His Bedside Teachings and Writings. Edited by William Bennett Bean, M.D. New York, Henry Schuman, 1950, p. 34.
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