Libman-Sacks Endocarditis Involving a Bioprosthesis in the Aortic Valve Position in Systemic Lupus Erythematosus

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Described herein is a 39-year-old man with systemic lupus erythematosus not receiving corticosteroid therapy who developed Libman-Sacks endocarditis causing stenosis of a bioprosthesis in the aortic valve position. **(Am J Cardiol 2019;124:316−318)**

Libman and Sacks described what later became known as Libman-Sacks (L-S) endocarditis in 1924,[1] and its relation to systemic lupus erythematosus (SLE) was described by Gross in 1940.[2] Lipman and Sacks initially described 4 patients at necropsy and noted that the endocarditis could involve any of the 4 cardiac valves. The "endocarditis" was described as deposits of fibrin not containing microorganisms or leukocytes on either side of a valvular leaflet but more commonly on the atrial side (atrioventricular valve), or aortic side (semilunar valve). Once patients with SLE were treated with corticosteroids the L-S fibrin lesions were uncommonly seen, the medication presumably converting the fibrin deposits into fibrous thickenings.[3,4] We recently encountered a man with known SLE not treated with corticosteroids who developed aortic valve regurgitation, underwent replacement of that valve with a bioprosthesis, which became stenotic because of development of L-S endocarditis on both sides of the bioprosthetic cusps within 8 months of its implantation. A description of this unusual patient is the purpose of this report.

Case description

A 39-year-old Hispanic man, who was born in April 1979, had been well until age 26 when he noted tender nodules and a rash on his legs, and pain in some joints. A diagnosis of SLE was made. He was advised to take prednisone but during the next 13 years he failed to do so. At age 39, he developed signs of heart failure, was found to have aortic regurgitation, and in May 2018 underwent replacement of his aortic valve with a bioprosthesis and insertion of a bypass conduit in his narrowed left anterior descending coronary artery. About 6 months later, symptoms of heart failure reoccurred and it rapidly progressed. On admission to Baylor University Medical Center in December 2018, echocardiogram showed the velocity across his bioprosthesis to be 3.9 m/s; mean transbioprosthetic gradient 35 mm Hg; left ventricular ejection fraction 30%; left ventricular end-diastolic diameter 6.7 cm, and systolic diameter, 6.3 cm. Mild mitral regurgitation was also found. The left internal mammary arterial conduit to the left anterior descending coronary artery was wide open. The left circumflex ostium was narrowed about 50%. The right coronary artery was wide open. His blood pressure was 115/70 mm Hg. His body mass index was 37 kg/m^2. The bioprosthesis was replaced with a mechanical prosthesis (#25 On-X). Multiple laboratory values just before replacement of the bioprosthesis are listed in Table 1.

The operatively-excised bioprosthesis is shown in Figure 1. Thrombi are present on both the aortic and ventricular aspects of the cusps. Histologic study of the thrombi revealed that they consisted of fibrin within which were scattered monocytes, mainly plasma cells (Figure 2). The surgeon incised the bioprosthesis to facilitate its removal.

Discussion

Described herein is a patient who at age 26 years was diagnosed with SLE. Although advised to do so, he never took a corticosteroid drug. At age 39, he underwent replacement of his native aortic valve because it was purely regurgitant. It is unclear if the operatively-excised native aortic valve contained superimposed fibrin deposits. Several months after replacement of the native aortic valve he developed heart failure and the bioprosthesis was found to be stenotic. Accordingly, just 8 months after implanting the bioprosthesis in the aortic valve position it was excised and replaced with a bileaflet mechanical prosthesis. The cusps of the operatively-excised bioprosthesis contained fibrin thrombi typical of L-S endocarditis.

Although it has been reported on native aortic and mitral valves on many occasions and leading to valve replacement,[5−15] L-S endocarditis involving a bioprosthesis in the aortic or mitral valve position is rare.[16−20] Niaz and Butany[17] described a 32-year-old woman who developed L-S endocarditis causing stenosis on a bioprosthesis in the aortic valve position in place for 8 years. The patient had not been on corticosteroid therapy. Both Moriski et al[19] and Sladak and Ausla[20] described young (ages 45 and 36 years)

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See page 318 for disclosure information.

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Table 1
Laboratory valves in the 39-year-old patient just before replacement of the bioprosthesis in the aortic valve position

Variable	Case
Hemoglobin (g/dl)	9
Hematocrit (%)	28
Platelets per mcL (μl)	109,000
Erythrocyte sedimentation rate (mm/hour)	79
C-reative protein (mg/dl)	2.1
Creatinine (mg/dl)	2.1
Total cholesterol (mg/dl)	154
Low-density lipoprotein cholesterol (mg/dl)	116
Urine protein/creatinine ratio	1:6
Antinuclear antibodies (ANA)	Negative
Antineutrophil cytoplasmic antibodies (ANCA)	Negative
Anti-DNA antibody titer	01:40
Anti-SSA	Negative
Anti-SSB	Negative
Anti-RNP	Negative
Anti-SM	Negative
Anti-Jol	Negative
Anti-Scl70	Negative
HIV	Negative
Anti-cardiolipin IgA (APL 'U/ml)	14
Anti-cardiolipin IgG (GPL 'U/ml)	72
Anti-cardiolipin IgM (MPL 'U/ml)	<10
B2-glycoprotein Ab IgA (SAU)	>150
B2-glycoprotein Ab IgG (SGU)	143
B2-glycoprotein Ab IgM (SMU)	<10
Phosphatidylserine Ab IgA (APS 'U/ml)	7
Phosphatidylserine Ab IgG (GPL 'U/ml)	>100
Phosphatidylserine Ab IgM (MPL 'U/ml)	6
Complement C3 (mg/dl)	108
Complement C4 (mg/dl)	18
Lupus anticoagulant	Positive
Prothrombin time (s)	11
Activated partial thromboplastin time (s)	39
Thrombin time (s)	16
Dilute Russell viper venom time	139

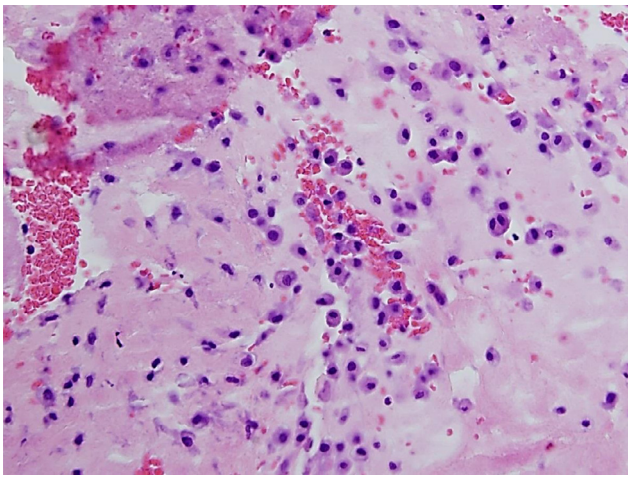

Figure 2. Photomicrograph of the Libman-Sacks lesion from the patient described. The lesion consists of fibrin within which are some mononuclear cells, mainly plasmacytes. Hematoxylin/eosin stain; × 400.

patients with L-S endocarditis involving a bioprosthesis in the mitral valve position and in each patient the bioprosthesis was made stenotic. L-S endocarditis occurs not only in patients with well-documented SLE but also in patients with the antiphospholipid syndrome without SLE.[21–23]

It is important to emphasize the apparent effect on corticosteroid drug therapy in patients with either SLE or the antiphospholipid syndrome. These medications appear to convert the fibrin thrombi of L-S endocarditis to fibrous lesions leading to thickening of the underlying valvular leaflet. The present patient never received corticosteroid therapy and presumably as a consequence developed full-blown L-S lesions without much underlying fibrous thickening of the cusps.

As a final note, coronary artery disease, as occurred in the present patient, is far more common in SLE patients than in subjects of similar age and sex without SLE.[24]

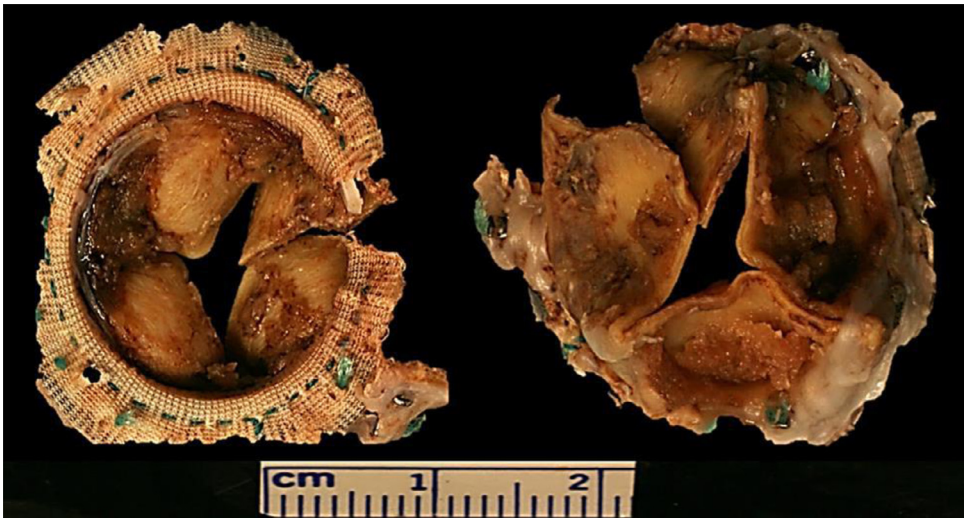

Figure 1. Bioprosthesis viewed from both the ventricular (left) and aortic (right) aspects in the patient described. Typical Libman-Sacks lesions are present on both sides of the bioprosthetic cusps.

Disclosures

All authors have participated in the work and have reviewed and agree with the content of the article and have no conflict of interests to disclose.

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