Double-valve Replacement for Mitral and Aortic Regurgitation in a Patient with Libman-Sacks Endocarditis

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Abstract

A 53-year-old woman with systemic lupus erythematosus and antiphospholipid syndrome presented with central nervous system (CNS) lupus and vegetation of the mitral and aortic valves. Her CNS lupus was relieved with methylprednisolone pulse therapy; however, her mitral regurgitation worsened, and she developed acute decompensated heart failure. The mitral and aortic valves were replaced with mechanical heart valves. Microscopic examination of the excised valves showed no bacterial invasion, and Libman-Sacks (LS) endocarditis of both valves was confirmed. This was a case of LS endocarditis with clear vegetation that spread over the mitral and aortic valves.

Key words: antiphospholipid syndrome, double-valve replacement, Libman-Sacks endocarditis, regurgitation, systemic lupus erythematosus

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Introduction

Libman-Sacks (LS) endocarditis is a well-known cardiac manifestation of systemic lupus erythematosus (SLE) in which sterile valvular vegetation or masses are observed. Although typically mild and asymptomatic, LS endocarditis can lead to serious complications, including superimposed bacterial endocarditis, thromboembolic events such as a stroke and transient ischemic attacks, and severe valvular regurgitation and/or stenosis. Reports on valvular surgery for LS endocarditis are fairly rare, and simultaneous double valve surgery is even rarer. We herein describe a case of rapidly degenerated mitral valve due to LS endocarditis and/or methylprednisolone pulse therapy for central nervous system (CNS) lupus. The patient underwent surgery to replace the mitral and aortic valves.

Case Report

A 53-year-old woman first presented at our hospital with SLE and antiphospholipid syndrome in 1989. Manifestations of her disease included malar rash, photosensitivity, and arthritis. She also had a history of recurrent pregnancy loss. Before beginning corticosteroid therapy, her anti-ds-DNA antibody level was 43.7 IU/mL (normal ≤12 IU/mL), and her anti-CL-β²GPI antibody level was 63 U/mL (normal <3.5 U/mL). Her prothrombin time was 13.6 sec, and her activated partial thromboplastin time was 85.3 sec. The activated partial thromboplastin time was remarkably lengthened, but the partial thromboplastin time was in the normal range. She was diagnosed with antiphospholipid antibody syndrome. The patient had been treated with long-term prednisolone and tacrolimus hydrate for SLE. An echocardiographic examination documented moderate mitral regurgitation (MR) (Fig. 1A) and aortic regurgitation (AR). The intensity of AR and MR murmur was Levine II/IV. The quantitative data of AR and MR severity were as follows: the pressure half time (PHT) of the AR jet was 389 ms (Fig. 2A). Using the method to measure the proximal isovelocity surface area (PISA) of a regurgitant color flow jet, the effective regurgitant orifice area (EROA) of MR was 0.30 cm², and the regurgitant volume was 53.2 mL (Fig. 1C). The left atrium and left ventricle were not dilated, and the left
Figure 1. Color Doppler echocardiographic images of the parasternal long-axis view obtained before admission (A) and in the ICU (B). Mitral regurgitation was observed, but its grade was not severe before admission. The mitral regurgitation worsened to a severe grade in the ICU. Quantitative analysis of mitral regurgitation severity observed before admission (C) and in the ICU (D). Using the method to measure PISA of a regurgitant color flow jet, the effective regurgitant orifice area of mitral regurgitation was 0.30 cm², and the regurgitant volume was 53.2 mL before admission. These values were simply calculated as the sum of the values derived from two PISAs. In the ICU, the effective regurgitant orifice area of mitral regurgitation was 0.42 cm², and the regurgitant volume was 60.7 mL. Both of the ICU values were apparently worse than those before admission.

Figure 2. Quantitative analysis of aortic regurgitation grade using the method that measures the PHT of aortic regurgitation jets before admission (A) and in the ICU (B). The PHT was shortened from 389 ms to 320 ms.

Ventricular ejection fraction was preserved. In early 2013, she was hospitalized with symptoms of nausea and vomiting. On the seventh day in the hospital, she experienced recurrent seizures. She was diagnosed with CNS lupus and was thereafter treated with methylprednisolone pulse therapy, followed by an increased dose of oral prednisolone. On
the 30th day in the hospital, she suddenly suffered from orthopnea and was transferred to the intensive care unit (ICU) with a diagnosis of acute decompensated heart failure. In the ICU, her blood pressure was 128/96 mmHg, her pulse rate was 100/min, and her percutaneous oxygen saturation concentration (SpO2) was 90% when inhaling 7 L/min O2 using a face mask. Rales were heard in the bilateral lung fields. The intensity of AR murmur remained at Levine II/IV; however, that of MR murmur increased to Levine III/IV. Her chest X-ray film showed pulmonary edema, and her electrocardiography revealed sinus tachycardia. Laboratory tests showed anemia, thrombopenia, elevated serum creatinine, and elevated N-terminal prohormone of brain natriuretic peptide (NT-proBNP). The patient's laboratory findings are shown in Table. In the ICU, she did not adequately respond to the initial therapy of noninvasive positive pressure ventilation and carperitide administration for acute decompensated heart failure. The patient was intubated and put on mechanical ventilation due to her progressively worsening condition. Transthoracic echocardiography revealed thickened valve leaflets and vegetation on the anterior leaflet of the mitral valve (Fig. 3); however, no specific finding was observed on the aortic valve. Color Doppler echocardiography demonstrated severe grade MR (Fig. 1B). The quantitative data with MR grade were apparently worse than before (EROA: 0.42 cm², regurgitant volume: 60.7 ml) (Fig. 1D). The mitral valve area calculated using the PHT (142 ms) of the early diastolic transmitral flow velocity was 1.55 cm², which meant that the patient also had mitral stenosis; however, it was not considered hemodynamically significant. The PHT of the AR jet was slightly shortened to 320 ms (Fig. 2B), indicating that the patient’s AR was marginally worsened. Repeated blood cultures were continuously negative, and there was no other evidence to indicate infective endocarditis. At that time, we thought that valve replacement surgery was mandatory for her heart; however, her platelet count decreased to <40,000/mm³. She was diagnosed with thrombotic microangiopathy because her activity of disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13 (ADAMTS-13) was decreased to 44.8% (normal range: 86.4-140.6% in women). We initiated daily plasma exchange, and her platelet count recovered to 100,000/mm³ after three sessions. On the 49th day, she underwent mitral valve replacement with a 25-mm On-X mechanical heart valve and aortic valve replacement with a 19-mm On-X mechanical heart valve using minimally invasive cardiac surgery (MICS). The MICS procedure was chosen to prevent complications due to sternotomy after long-term predisolone therapy. Intraoperative inspection showed that the anterior mitral valve leaflet was calcified, and the posterior mitral valve leaflet was shortened. Vegetation was observed over the mitral anterior and posterior leaflets and the noncoronary cusp of the aortic valve (Fig. 4A, B, 5A). Microscopic examination of the excised mitral valve segment revealed hyaline degeneration, calcification, and infiltration of a few mononuclear cells (Fig. 4C). The excised aortic valve segment showed hyaline degeneration and an edematous region (Fig. 5B). No bacterial invasion was identified in either valve, and the cultures of excised valves were negative for infection. LS endocarditis of the mitral and aortic valves was confirmed. The patient recovered from valve replacement surgery uneventfully.

Discussion

In 1924, Libman and Sacks first described four cases of nonbacterial verrucous vegetative endocarditis (1). The sterile verrucous lesions of LS endocarditis were mainly observed in the mitral and aortic valves. A significant association was previously found between LS endocarditis and SLE duration and activity, thromboses, stroke, thrombocytopenia, anticardiolipin antibodies, and antiphospholipid syndrome (2). An echocardiographic study in patients with SLE
revealed that LS endocarditis occurs in approximately 11% of SLE patients (2). The diagnosis of LS endocarditis is quite difficult because the clinical syndrome and images often mimic those of bacterial endocarditis. Infective endocarditis and LS endocarditis have been reported to coexist (3, 4). In the present case, we diagnosed LS endocarditis on the basis of repeated negative blood cultures and negative cultures from the excised valves.

Corticosteroid therapy is necessary in the treatment of SLE; however, there is no evidence that corticosteroid usage can prevent valvular damage. Hojinik et al. (5) reported that corticosteroid therapy could increase the extent of fibrosis and ultimately worsen valve deformity. A case was reported in which the rapid development of severe MR occurred after a few weeks of high-dose corticosteroid therapy (6). In the present case, methylprednisolone pulse therapy and an in-

Figure 4. The mitral valve at the time of operation. Vegetation was observed on the anterior mitral leaflet (A). The resected mitral valve (B). Vegetation was clearly seen over the anterior and posterior mitral leaflets. Microscopic examination of the excised mitral valve (C) showed hyaline degeneration, calcification, and infiltration of a few mononuclear cells. Hematoxylin and Eosin stain, original magnification ×200.

Figure 5. The patient’s aortic valve at the time of operation. Vegetation was obvious on the non-coronary cusp (A). Microscopic examination of the excised aortic valve (B) showed hyaline degeneration and an edematous region. Hematoxylin and Eosin stain, original magnification ×200.
creased dose of oral prednisolone for CNS lupus may have led to worsened MR. In contemporary cardiac surgery, mitral valve repair has become the mainstay of treatment for most cases of MR. However, some studies suggest that results from mitral valve replacement are usually superior to mitral valve repair for LS endocarditis (7, 8). Accordingly, severe calcification and fibrosis of the mitral valve may lead to rapid recurrence of MR after repair with subsequent reoperation for the mitral valve replacement. In the present case, we performed a double-valve replacement for the mitral and aortic regurgitation because the mitral valve was severely deformed and a concomitant aortic valve replacement was needed.

The authors state that they have no Conflict of Interest (COI).

References