Case Reports

Isolated Tricuspid Valve Libman-Sacks Endocarditis

in Systemic Lupus Erythematosus with Secondary Antiphospholipid Syndrome

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Libman-Sacks endocarditis, one of the most prevalent cardiac presentations of systemic lupus erythematosus, typically affects the aortic or mitral valve; tricuspid valve involvement is highly unusual. Secondary antiphospholipid syndrome increases the frequency and severity of cardiac valvular disease in systemic lupus erythematosus. We present the case of a 47-year-old woman with lupus and antiphospholipid syndrome whose massive tricuspid regurgitation was caused by Libman-Sacks endocarditis isolated to the tricuspid valve. In addition, we discuss this rare case in the context of the relevant medical literature.
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Systemic lupus erythematosus (SLE) is an inflammatory disease with multiorgan involvement. In patients with SLE, Libman-Sacks endocarditis most frequently affects the aortic and mitral valves. The echocardiographic presentation of Libman-Sacks endocarditis includes leaflet thickening, and masses or verrucous vegetations that cause regurgitation.[1] Tricuspid valve (TV) involvement, particularly in isolation, is rarely seen in SLE.[1,2] Secondary antiphospholipid syndrome (APLS) increases the severity of valvular disease in SLE.[3] Here, we discuss the case of a patient who had SLE, APLS, and Libman-Sacks endocarditis isolated to the TV.

Key words: *Antiphospholipid syndrome/complications; disease progression; endocarditis/etiology/pathology/surgery; heart valve prosthesis; lupus erythematosus, systemic/complications; treatment outcome; tricuspid valve insufficiency/etiology/surgery*

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Case Report

In July 2012, a 47-year-old woman was admitted to our cardiology department with exertional dyspnea, peripheral edema, and chest discomfort. Fifteen months earlier, she had been diagnosed with SLE and APLS; her therapy included prednisolone and chloroquine.

The patient's medical history was notable for multiple miscarriages and for unexplained thrombocytopenia, which had been treated with corticosteroids. Rheumatologic evaluation yielded elevated antinuclear antibody levels, double-stranded DNA, anticardiolipin antibody, low levels of complement components 3 and 4, and a positive lupus anticoagulant test, all significant for SLE with secondary APLS. Her physical examination revealed moderate jugular venous distention, a pulsatile liver, an S_3 gallop, and a parasternal pansystolic murmur increasing with inspiration. Transthoracic echocardiograms showed right atrial enlargement (45 mm), right ventricular enlargement (48 mm), suspected vegetations on all 3 of the thickened TV leaflets, and a massive central jet of tricuspid regurgitation without significant pulmonary hypertension. The aortic, pulmonary, and mitral valve leaflets appeared to be normal. Negative blood cultures suggested noninfective endocarditis. Left ventricular function and size were normal. Coronary angiograms showed no coronary artery stenosis. The central venous pressure was 20 mmHg with cv waveforms. Right-sided heart catheterization was not performed. The patient was scheduled for surgery.

We instituted cardiopulmonary bypass and performed surgery on the normothermic beating heart. Intraoperatively, we found thickened and retracted TV leaflets with nodules and vegetations on the atrial side and an annular dilation of 43 mm (Fig. 1). We replaced the TV with a size 29 Mosaic® bioprosthesis (Medtronic, Inc.; Minneapolis, Minn), which the patient preferred over a mechanical valve with audible clicks.

The patient recovered uneventfully. Microbiological cultures of the leaflet tissue remained sterile. Histopathologic analysis confirmed Libman-Sacks endocarditis with vegetations consisting primarily of fibrin peripherally and platelets centrally, fibrinoid and hyaline deposits atop the vegetations, and mononuclear inflammatory infiltration at the base of the vegetations (Fig. 2). As of March 2017, the patient had no cardiac symptoms, and annual echocardiograms had consistently shown a normally functioning bioprosthetic TV.

Discussion

Isolated TV Libman-Sacks endocarditis necessitating valve replacement is exceedingly rare.[4] Multivalvular involvement prompts surgical treatment more frequently,[5] but conservative treatment has also been reported.[2,3,6,7]

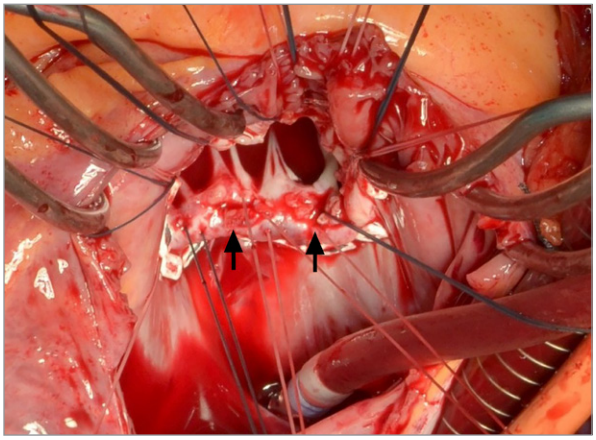

Fig. 1 *Intraoperative photograph shows verrucous vegetations on the tricuspid leaflets (arrows).*

The pathogenesis of valve lesions in Libman-Sacks endocarditis involves the formation of fibrin-platelet thrombi on the altered valve. Endothelial damage on the valvular surface is the presumed initial insult that leads to the fibrin-platelet formation. Further organization of such lesions, as well as the deposition of immunoglobulins—including anticardiolipin antibody and complement components—leads to cusp fibrosis, thickening, and scarring, and ultimately to distortion and valvular dysfunction. Blood-flow turbulence and jet effects are more pronounced and prevalent in the left-sided heart valves. Because thrombotic mechanisms are impaired in APLS, valvular pathologic conditions are more severe in patients with both SLE and APLS.[8]

The isolated TV involvement in our patient might be explained by existing but clinically negligible changes in TV morphology and function, which, after the endothelial damage occurred, provided an environment for typical vegetations to form.

Valvular thickening and vegetations are often found in patients with SLE, but they usually cause mild, clinically irrelevant valvular regurgitation,[1] which can be successfully treated with antithrombotic and anticoagulation therapy.[2] However, in some patients, abnormalities can cause ischemic strokes, peripheral embolization, and severe valvular regurgitation—all of which are indications for surgery like that for infective endocarditis.[6]

In a prospective clinical and echocardiographic study,[7] approximately 10% of SLE patients developed severe valvular regurgitation associated with high levels of anticardiolipin antibody; symptoms of heart failure and progressive ventricular dysfunction indicated surgery. Vianna and colleagues[3] reported that valve lesions in Libman-Sacks endocarditis are more severe in patients with secondary APLS than in patients with APLS alone. Patients with secondary APLS needed surgery more

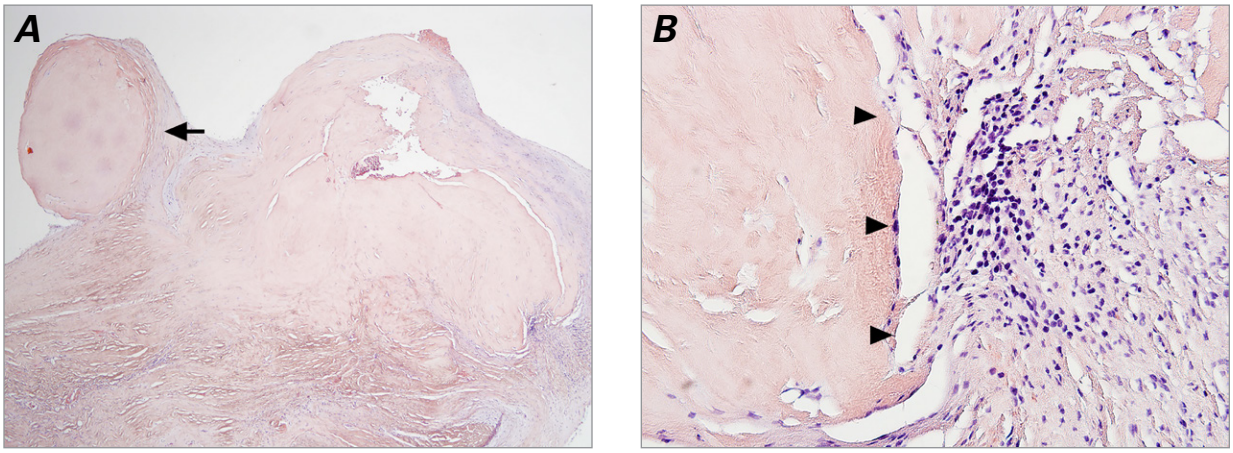

Fig. 2 *Photomicrographs of cross-sections of resected tricuspid valve leaflet tissue show* ***A****) a vegetation (arrow) on the atrial side of the tricuspid leaflet (H & E, orig. ×40) and* ***B****) mononuclear inflammatory infiltration (arrowheads) at the base of the vegetation, confirming Libman-Sacks endocarditis (H & E, orig. ×400).*

often, and they had a higher perioperative risk. They also had a higher incidence of autoimmune hemolytic anemia, neutropenia, low complement component 4 levels, thrombocytopenia, and antiphospholipid manifestations,[3] all of which conditions our patient also had.

Close clinical and echocardiographic monitoring are recommended in such patients, as is aggressive anticoagulant or antiplatelet therapy to prevent further valvular degeneration and thromboembolic events. Upon clinical deterioration and disease progression, surgery is indicated.

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