

Libman-Sacks Endocarditis and Primary Antiphospholipid Syndrome

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Cardiac involvement is a not uncommon complication in patients with antiphospholipid syndrome (APS). Herein, the case is reported of cardiac failure in a female patient with Libman-Sacks endocarditis and with primary APS diagnosed eight years previously. Aggressive anticoagulation therapy and medical treatment for the cardiac failure over a 12-month period resulted in a partial regression of the severe

mitral regurgitation. Close clinical and echocardiographic surveillance during the follow up of patients with APS and heart valve disease is mandatory. Optimal treatment, including adequate aggressive anticoagulation therapy and specific treatment for heart failure, may play a pivotal role in reducing the severity of valve dysfunction in these patients.

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Antiphospholipid syndrome (APS) has been defined as venous or arterial thrombosis, recurrent fetal loss, or thrombocytopenia accompanied by an increased level of antiphospholipid antibodies (1). The syndrome can be either primary or secondary to an underlying condition, most commonly systemic lupus erythematosus (1). Clinical manifestations, including heart involvement, neurological manifestations, as well as skin, kidney and hematologic abnormalities, have been described in patients with APS. Cardiac manifestations include valvular heart disease (defined as Libman-Sacks non-bacterial endocarditis), cardiomyopathy, intracardiac thrombus and coronary bypass graft and angioplasty occlusions (1-5). In both primary and secondary APS, the probability of developing valvular heart disease seems to be increased with higher levels of circulating anti-phospholipid antibodies (6,7).

Case report

A 49-year-old woman diagnosed with primary APS eight years previously attended the authors' institution for cardiac evaluation. She had presented two episodes of pulmonary embolism during the year

before admission. On admission, her medical treatment consisted of oral anticoagulant treatment with acenocumarol and low doses of prednisone (5 mg/day). During the last month before admission, the patient complained of dyspnea (NYHA functional class III). Physical examination disclosed a 3/6 systolic cardiac murmur on the mitral area and right basilar pulmonary rales. Routine biochemistry laboratory parameters and a full blood cell count were normal, and the International Normalized Ratio (INR) was 2.4. A chest radiograph disclosed left cardiac failure, but the 12-lead electrocardiogram was normal. Transthoracic and transesophageal echocardiography showed severe mitral regurgitation caused by two anterior and posterior leaflet mitral valve masses on the auricular side, measuring 6×5 mm and 5×8 mm, respectively (Fig. 1A and B). The left ventricular size and ejection fraction were normal, and the systolic pulmonary artery pressure was 40 mmHg.

Although there was no clinical suspicion of infection, six blood cultures were taken, and all yielded negative results. Additional laboratory evaluations performed showed negative antinuclear and anti-double-stranded DNA antibodies, and normal serum levels of C3 and C4 complement. However, IgG (measured by ELISA) anticardiolipin antibodies were positive (100 GPL units; normal value <20).

A diagnosis of mitral valve Libman-Sacks endocarditis was made. Aggressive anticoagulation with

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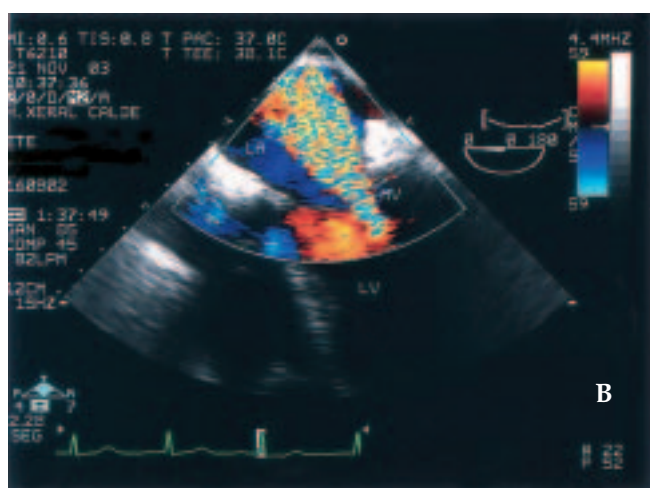


Figure 1: A) Diastole frame horizontal transesophageal echocardiographic view of the mitral valve, showing two leaflet vegetations (arrows). B) Horizontal transesophageal echocardiographic view showing a mitral regurgitation color jet almost reaching the posterior wall of the left atrium. The mitral regurgitant volume (64 ml) was calculated using the PISA method. LA: Left atrium; LV: Left ventricle; MV: Mitral valve.

acenocumarol was aimed to achieve an INR between 3.0 and 4.0. In addition, medical treatment for the cardiac failure, including diuretics and angiotensin-converting enzyme inhibitors, was commenced. During the next four weeks a progressive improvement of the patient's condition was observed. At 12 months after diagnosis of Libman-Sacks endocarditis, the severe mitral valve regurgitation had partially regressed and surgery was no longer considered. In this regard, further transthoracic echocardiography, performed one year after the initial investigation, showed a reduction in size of the vegetations on the mitral valve. A Doppler study demonstrated a significant decrease in mitral regurgitation and normalization of the systolic pulmonary artery pressure (Fig. 2).

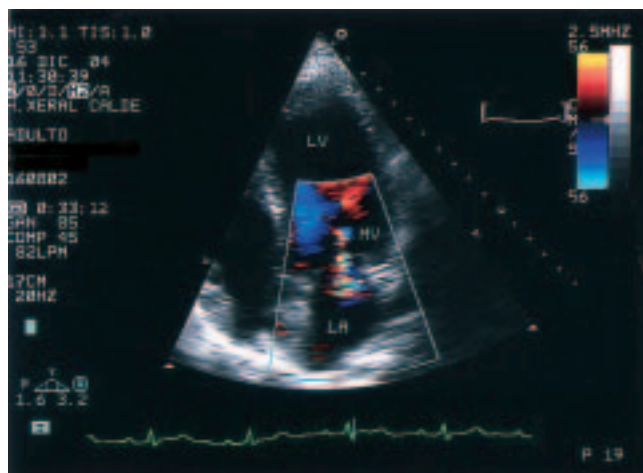


Figure 2: Transthoracic echocardiographic four-chamber view during the follow up period showing mild-moderate mitral regurgitation. Abbreviations as Figure 1.

Discussion

Cardiac involvement in primary APS is a not uncommon complication. Echocardiographic studies have disclosed heart valve abnormalities in about one-third of patients with primary APS, with the mitral valve most often involved (1,2,4,5).

In patients with APS associated with heart valvular disease who have experienced a thromboembolic event, antithrombotic therapy is indicated as a secondary prevention. High-intensity oral anticoagulant therapy (to achieve an INR >3.0) has proved to be more effective than low-intensity anticoagulation (INR <3.0) in preventing further venous and arterial thrombotic events associated with APS in these patients (8).

The presence of severe mitral regurgitation in cases of APS is relatively uncommon, and the course of the valvular lesions in these patients is generally stable or slowly progressive (1). However, the present case showed severe mitral valve regurgitation and partial regression of this abnormality during the follow up. Regression of mitral valve vegetations has also been reported in a patient with systemic lupus erythematosus and secondary APS (9).

Emphasis should be placed on the potential role of optimal anticoagulation treatment in reduction of the thrombotic component in sterile fibrofibrinous vegetations, improving the systolic coaptation of mitral leaflets, followed by - as in the present case - a reduction in the grade of mitral valve regurgitation.

Close clinical and echocardiographic surveillance during the follow up of patients with APS and heart valve disease is mandatory. Optimal treatment, which includes adequate aggressive anticoagulation therapy and specific treatment for heart failure, may play a piv-

otal role in reducing the severity of valve dysfunction and, consequently, the need for cardiac surgery in these patients.

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