Libman-Sacks Endocarditis and Oral Anticoagulation

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Case Report

Libman-Sacks endocarditis was initially reported in 1924 as the presence of bacteriUmfree valvular vegetations; and later as a manifestation of systemic lupus erythematosus. Its incidence varies and may even reach 60% in postmortem studies. It is usually asymptomatic, but fragmentation of the vegetations may occur with systemic embolization and a predilection to infective endocarditis. The simultaneous presence of antiphospholipid antibodies has been reported in a small number of studies, and this association is still controversial. Corticosteroids and immunosuppressants are known not to have an effect on the valvular lesions of Libman-Sacks endocarditis. On the other hand, anticoagulation may be used in the treatment of antiphospholipid antibody syndrome, and some authors have suggested the use of this therapeutic modality when the association of antiphospholipid syndrome and Libman-Sacks endocarditis occurs.

We report the case of a patient with systemic lupus erythematosus, secondary antiphospholipid antibody syndrome, and Libman-Sacks endocarditis, whose vegetations disappeared after anticoagulation therapy.

Case report

The patient is a 34-year-old female with systemic lupus erythematosus and secondary antiphospholipid antibody syndrome, who evolved with convulsive crises, partially controlled with an anticonvulsant, and auscultation of a cardiac murmur, whose investigation showed the presence of a mitral valve vegetation.

Once the diagnosis of Libman-Sacks endocarditis was established, therapy with warfarin sodium was initiated, and, after 6 months of oral anticoagulation, the patient had total control of the convulsive crises and the valvular vegetation disappeared on echocardiography. This study discusses the occurrence of Libman-Sacks endocarditis in systemic lupus erythematosus, its association with antiphospholipid antibody syndrome, and the anticoagulant therapy. A literature review is also provided.

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Valvular heart disease is the most frequent and important cardiac manifestation of systemic lupus erythematosus. The...
The mitral valve is the most frequently affected, followed by the aortic valve. Impairment of the tricuspid and pulmonary valves is rarely reported. 

The pathogenesis of Libman-Sacks endocarditis has not yet been completely elucidated. The major mechanisms proposed are as follows: 1) formation of fibrin and platelet thrombi on the impaired valve, whose organization leads to fibrosis, distortion, and subsequent valvular dysfunction. The thrombotic phenomena may result from the following biological effects of anti phospholipid antibodies: an increase in platelet activity, a reduction in antithrombin III levels, inhibition of prostacyclin release by endothelial cells, inhibition of thrombomodulin in protein C-protein S system, and decreased activity of the clotting factor X activator released by endothelial cells; 2) immunologic injury as an initial insult to the valvular apparatus, triggering the sequence of pathogenetic events. Deposits of immunoglobulins and complement were shown in the subendothelial layer of the valves in patients with anti phospholipid antibodies. 

Some studies have suggested an association between valvular heart disease and the presence of anti phospholipid antibodies, although other studies have not confirmed this relation. These divergences partially result from the different methods used for detecting anti phospholipid antibodies, as well as from variations in the echocardiographic techniques used and in the interpretation of the results. A correlation between the type and the titer of anti cardiolipin antibodies and the probability of developing valvular heart disease seems to occur: patients with moderate to high IgG anti cardiolipin antibody titers have a higher incidence of valvular alterations when compared with patients whose IgG and IgM anti cardiolipin antibody titers are low. However, in some patients with valvular disease, the lupus anticoagulant may be the only anti phospholipid antibody detected. 

The ideal treatment for patients with anti phospholipid syndrome has not yet been defined, partly due to the scarcity of information on the natural history of the disease in untreated patients. Most authors recommend high-intensity (INR>3) anticoagulation as secondary prevention for thromboembolic phenomena. Due to the high risk of recurrence of thrombotic episodes, especially in the first 6 months after the interruption of anti coagulant therapy, indefinite anticoagulation is indicated in patients with persistently high titers of anti phospholipid antibodies. Primary prevention of thrombotic episodes in patients with moderate to high titers of anti phospholipid antibodies is controversial. These patients usually receive low doses of acetylsalicylic acid, although no evidence exists about the efficacy of this approach. Corticosteroids and immunosuppressants are not used in patients with anti phospholipid syndrome because they do not influence the hypercoagulable state. Our patient had anti phospholipid syndrome secondary to systemic lupus erythematosus, the use of the immunosuppressant being justified for controlling the clinical manifestations of the primary disease not related to the syndrome. 

Five cases have been reported about patients with primary anti phospholipid antibody syndrome manifested as stroke or acute myocardial infarction, who underwent oral anticoagulation. These patients had vegetations in the mitral valve and no evidence of infection, as in our case. After approximately 6 weeks to 4 months of treatment, all patients evolved with resolution of the vegetations.
valve and on the vascular face of the aortic valve. In addition to valve leaflet abnormalities, 2 patients had evidence of myocardial infarction and a defect in the atrial septum. All 11 patients used an oral anticoagulant or antiplatelet agent for 1 year, and 13 patients ended up undergoing a new transesophageal echocardiogram. The second examination showed unchanged lesions in 6 patients and new lesions in the other 7. The authors concluded that treatment with an oral anticoagulant or antiplatelet agent does not contribute to the disappearance of noninfectious valvular vegetations, despite the sporadic reports on the resolution of vegetations with the use of high-intensity oral anticoagulation for less than 1 year. Our patient evolved with disappearance of the vegetations with oral anticoagulation for 6 months, and, in addition, control of the convulsive crises, which was partial up to then.

One complication of systemic lupus erythematosus is the thromboembolic phenomenon, the brain being the most affected site. In most cases, the embolic episodes are known to be subclinical, but sometimes they may manifest as signs and symptoms of ischemia of the affected organ. Convulsive crises may be a sign of cerebral ischemia. In our patient, the control of convulsive crises coincided with the beginning of oral anticoagulation, and one may infer that the medication may have acted on one of the pathophysiological processes (hypercoagulable state) involved in the formation and release of thrombi from the vegetation. However, epilepsy is one of the most common neuropsychiatric manifestations of lupus and is associated with a high prevalence of anti-phospholipid antibodies, in whose pathogenesis the occlusion of small vessels of the cerebral circulation is implicated, as a result of the hypercoagulable state. Therefore, in our case, more than 1 etiology for the convulsive crises may have existed, all of which related to the presence of anti-phospholipid antibodies and to a hypercoagulable state.

Considering the ease of transesophageal Doppler echocardiography and the current difficulty of indicating long-term anticoagulation for patients with systemic lupus erythematosus, this case is worth reporting. In our patient, anticoagulation may have played an efficient therapeutic role, especially considering the presence of an antiphospholipid antibody and the manifestations of this disease. Our efforts should be directed to the study of the prevalent ence of the disease, its association with antiphospholipid antibodies and their manifestations, as well as to more adequate therapy and duration of treatment.

References

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