SHORT COMMUNICATION

Libman–Sacks endocarditis, and other echocardiographic findings in systemic lupus erythematosus: Case report

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KEYWORDS
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Abstract
Case report of a 19 year-old female patient with systemic lupus erythematosus (SLE) who was presented to Ain Shams University Hospital complaining of dyspnea on moderate exertion. Echocardiography showed the presence of sterile vegetation on the mitral valve, Libman–Sacks endocarditis (LSE).

1. Introduction
Libman–Sacks valvular lesions are sterile fibrinous vegetations that preferentially develop at the left-sided heart valves. Echocardiography is a useful tool for documenting the valvular involvement and dysfunction as well as disease progression of the patients with systemic lupus erythematosus (SLE).1 It is usually asymptomatic; however, cases that needed valve replacement have been described.2

The present case report describes a patient with SLE, LSE, and other echocardiographic findings; like pericardial effusion, decreased left ventricular function and pulmonary hypertension.

2. Case report
The patient was a 19-year-old Egyptian lady, student, single, with the diagnosis of SLE, since 2008 when she presented with skin rashes all over the extremities and butterfly rash on her face, photosensitivity and arthritis in her left knee joint. At the time of the SLE diagnosis, she presented a normal blood count, normal urea/creatinine levels, increased ESR, decreased C3 fraction of complement, C4 at the lower normal range. The antinuclear antibodies (ANAs) and antinative DNA were positive.

Echo-Doppler study was done at that time revealing normal LV dimensions and function with EF = 60%, and a mild circular pericardial effusion. Treatment was initiated with prednisone 1 mg/kg/day, with the improvement of her symptoms.

However, the patient interrupted the treatment for one year. At 24/04/11, she started to complain of dyspnea on moderate exertion. She was presented to the Ain-Shams University Hospital to ask for medical advice. Clinical examination showed her BP = 100/70 and HR 90 bpm. Peripheral pulse was well felt. Cardiac examination revealed accentuated S2, and S4 on the apex. Normal chest/abdominal examination. No skin or oropharyngeal lesions, as well as no arthritis or any joint deformities were observed.

The electrocardiogram was normal. Transthoracic echocardiography was done for the patient showing two vegetations, first one is attached to the ventricular surface of the anterior mitral valve leaflet measuring 16 × 13 mm, and causing mild mitral regurgitation. While the other one is smaller and...
attached to the LV side of the interventricular surface adjacent to the first one and measures 0.7 × 0.6 mm.

The LVEF was mildly decreased measuring 48%, the right sided chambers were dilated with elevated RVSP measuring 60 mm Hg. There was also a mild circular pericardial effusion with no evidence of tamponade. Other valves showed normal morphology and flow with no masses or vegetations attached.

Three pairs of blood culture samples were collected from different sites and all had negative results. Hemoglobin = 11.3 mg/dL, platelets = 252,000 and leukocytes = 6400, creatinine = 0.6 mg/dL, normal urinary sediment. Antinuclear-DNA antibody was positive and anticardiolipin IgG antibodies (13.6 GPL) were undetermined, whereas IgM was negative, C-reactive protein (CRP) = 3.95 mg/L, C3 was decreased and C4 was normal.

The patient received prednisone (40 mg/day) and enalapril (10 mg/day) and was discharged after the symptomatic improvement with a plan of follow up for clinical and echo.

3. Discussion

This is a case report of a patient with SLE (established according to the criteria of the American College of Rheumatology), who presented with exertional dyspnea, which might have been caused by the decreased LV function or pulmonary hypertension. Also, the patient had aseptic vegetation in the mitral valve with mild MR.

Libman–Sacks endocarditis (otherwise known as verrucous, or nonbacterial endocarditis) is the characteristic cardiac manifestation of the autoimmune disease systemic lupus erythematosus (SLE). Libman and Sacks first published a description of these atypical, sterile, verrucous vegetations in 1924. Libman–Sacks endocarditis most commonly involves mitral and aortic valves. However, all 4 cardiac valves and the endocardial surfaces can be involved.

Valvular abnormalities are often clinically silent, without significant valvular dysfunction. Valvular regurgitation is more common than stenosis, which is rare. Valvular dysfunction can result in cardiac failure. Embolic phenomena and secondary infective endocarditis are uncommon but can result in neurological and systemic complications.

In a study with 342 SLE patients, LSE was detected by Doppler echocardiogram in 11% of the cases, more often in the mitral valve. After 4 years of follow-up, valve failure and/or stenosis were frequent, and two patients who were candidates for heart surgery died. Thus, the diagnosis of LSE is important and the echocardiogram is currently the best imaging procedure for diagnosis.

One must also recall that the infectious endocarditis is not unusual in SLE patients with LSE and a differential diagnosis is mandatory. In this aspect, three laboratory data are important: leukocyte count, CRP levels, and blood cultures. The leukocytes tend to decrease during lupus activity and the opposite occurs in infectious endocarditis. Very high CRP levels suggest an infectious cause, as lupus patients are less capable of presenting an exuberant response of this protein; however, for a definitive differential diagnosis, the blood cultures are more important.

In the present case, a diagnosis of LSE was attained, as the leukocyte count was normal, the CRP was not very elevated and blood culture samples had negative results.

The combined rate of heart failure, valvular replacement, thromboembolism, and secondary infective endocarditis has been reported to be as high as 22% in lupus patients with valvular disease, compared with 8% of patients without valvular disease. Most patients do not have clinically significant valvular dysfunction. Regurgitation is noted on echocardiography images in 25–61% of lupus patients. The reported patients who need valve replacement varies from 1% to 8% of cases.

The occurrence of clinically significant embolic phenomena is thought to be low. Although stroke rates are higher in patients with lupus and antiphospholipid syndrome, multifactorial etiologies for neurological events are often present, making the specific contribution of valvular abnormalities difficult to determine. The likely prevalence of secondary infective endocarditis is low, but it has not been widely reported. Potential contributing factors to infective endocarditis are systemic lupus erythematosus, medications prescribed for lupus, and underlying valvular abnormalities.

There is scarce information in the literature regarding the treatment of LSE. It is known that the use of corticoids and immunosuppressive drugs seems to have no effect on valve lesions; however, anticoagulation therapy must be used for the treatment of patients with thromboembolic events.

References