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CASE REPORT

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Noninfective Endocarditis in a Young Patient with Systemic Lupus Erythematosus

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Abstract

Noninfective endocarditis, particularly in association with systemic autoimmune diseases like systemic lupus erythematosus (SLE), presents a diagnostic challenge due to its nonspecific symptoms and the potential for severe cardiac complications. We describe a case of a 16-year-old female diagnosed with SLE who presented with fever, malaise, and a new heart murmur. Subsequent investigations revealed vegetations on the mitral valve consistent with noninfective endocarditis without any evidence of infectious etiology. This case underscores the importance of considering noninfective endocarditis in the differential diagnosis for patients with systemic autoimmune disorders presenting with fever and cardiac symptoms. Early recognition and management are crucial to prevent serious outcomes.**(International Journal of Biomedicine. 2024;14(2):338-340.)**

Keywords: systemic lupus erythematosus • noninfective endocarditis • Libman-Sacks endocarditis

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Abbreviations

LSE, Libman-Sacks endocarditis; **LVH**, left ventricular hypertrophy; **SLE**, systemic lupus erythematosus; **CRP**, C-reactive protein.

Introduction

Libman-Sacks endocarditis (LSE), also known as verrucous, marantic, or nonbacterial thrombotic endocarditis, was first described by Emanuel Libman and Benjamin Sacks in 1924.[(1)] Heart valve abnormalities can be found in 1 of every 3 patients with systemic lupus erythematosus (SLE), while valvular vegetations, such as LSE, are present in 1 of every 10 SLE patients.[(2)] Libman-Sacks endocarditis is rare in children and adolescents, more so as a first manifestation of SLE. They are clinically silent in most cases. The presence of antiphospholipid antibodies in SLE is associated with a three times higher prevalence of mitral valve nodules and significant mitral regurgitation.[(3)] The diagnosis of LSE becomes challenging, especially in differentiating it from infective endocarditis, as both diseases may present similarly.

Case Presentation

A 16-year-old female was admitted with fatigue, fever, and shortness of breath. During the physical examination, she presented tachypnea, tachycardia (150 bpm), temperature of 38.8°C, blood pressure of 150/90 mmHg, increased central venous pressure, and a grade V/VI pan systolic mitral murmur. An electrocardiogram demonstrated sinus tachycardia without other associated anomalies. Three pairs of blood culture samples

were collected from different sites, and all had negative results. A cardiac ultrasound revealed a large pericardial effusion, and the decision to pericardiocentesis was made. The patient tolerated the procedure well, and 220 cc of serous fluid was aspirated and sent for laboratory studies. Blood test: WBC - $10.9\times10^3/\mu L$, RBC - $4.93\times10^6/\mu L$, Hb - 7.3 g/dL, platelets - $140\times10^3/\mu L$, lymphocytes -12%, urea – 123 mg/dL, creatinine - 3.1 mg/dL, CRP - 33.8 mg/L, AST - 25 U/L, ALT - 10 U/L, GGT - 72 U/L, LDH - 410 U/L, total protein - 7.3 g/dL. A urine analysis revealed proteinuria and hematuria. Workup showed positive antinuclear antibody, anti-dsDNA, anti-ENA screen, and slightly depressed serum complement levels. Chest X-ray revealed bilateral pleural effusion. During the hospital stay, the patient developed an episode of seizure. According to the diagnostic criteria, the diagnosis of SLE was made, and the patient started treatment with methylprednisolone and cyclophosphamide. The patient was sent for transthoracic echocardiography (2 days after pericardiocentesis), which revealed a diffuse infiltration of anterior mitral leaflet and a nodular thickening on it, sized 20×10 mm (Figures 1 and 2), with severe mitral regurgitation (Figure 3). Also, a nodular thickening was seen on the pulmonary valve, and there was mild regurgitation (Figure 4). A concentric left ventricular hypertrophy (LVH) was noticed with a mild dilatation of the left atrium; both ventricles' size and systolic function were preserved (Figure 5). A small amount of pericardial effusion was detected. Considering possible infective endocarditis and elevated CRP and WBC levels, the patient was placed on a regimen of ampicillin and gentamycin for 2 weeks. As the urea and creatinine levels rose and the patient became oliguric, she was transferred to the nephrology ward, where hemodialysis was started. Under the treatment with hemodialysis, methylprednisolone, cyclophosphamide, lercanidipine, carvedilol, ampicillin, gentamycin and levetiracetam, her symptoms improved. The patient remained afebrile, and her creatinine level, after reaching 7 mg/dl, went down to 3 mg/dl. Another transthoracic echocardiography was performed after 2 weeks, with the same results as the previous one, but without pericardial effusion. Echocardiographic studies have yet to be repeated as of writing this article.

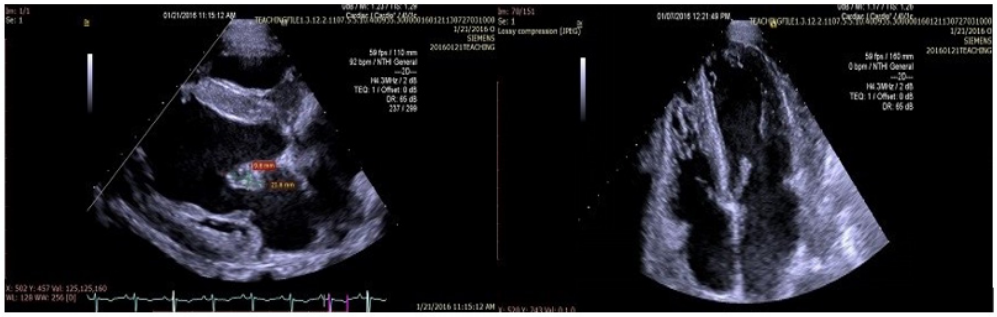

Fig. 1 and 2. *Diffuse infiltration of the anterior leaflet of the mitral valve with nodular thickening in the parasternal and apical 4-chamber view.*

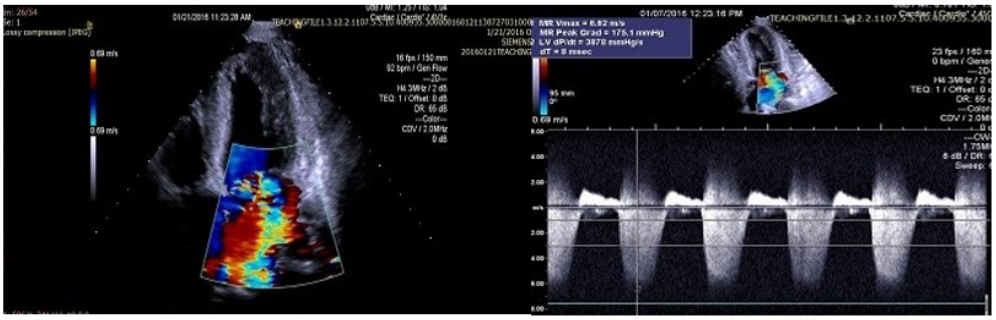

Fig. 3. *Severe mitral regurgitation.*

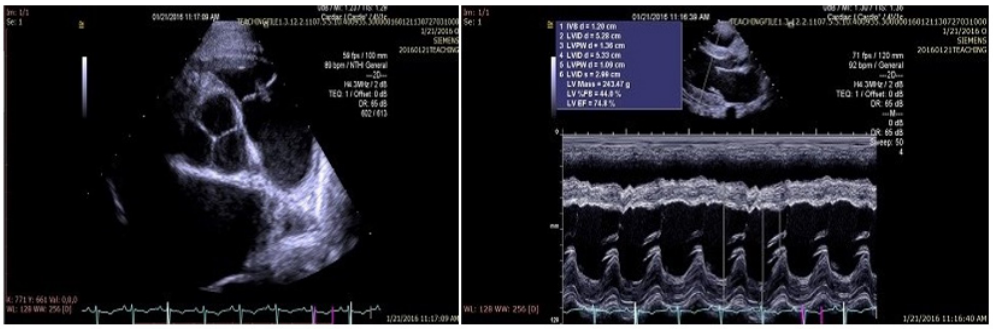

Fig. 4. *Nodular thickened on the pulmonary valve.* ***Fig. 5.*** *Concentric hypertrophy of LV with normal diameters and systolic function.*

Discussion

This case of a young patient with SLE presented with progressive dyspnea (which may have been caused by pericardial effusion) and fever was also characterized by the presence of aseptic vegetation on the mitral valve with severe mitral regurgitation. The lesions primarily consist of accumulations of immune complexes and mononuclear cells. These subendothelial deposits may eventually lead to deformed valves. The most involved valve is the mitral valve, followed by the aortic valve.[3] Characteristic valvular pathology can also distinguish infective endocarditis vegetations from Libman-Sacks endocarditis, but this may not always hold as vegetative lesions may evolve throughout the disease. Infective endocarditis is characterized by large, irregular masses on the valve cusps, which can extend onto the cords. Libman-Sacks endocarditis has small or medium-sized vegetation on either or both sides of the valve leaflets. The vegetation seen on the patient's echocardiogram was on the anterior mitral valve leaflet. Still, the exact size and extent of involvement in this leaflet could not be distinguished because of the infiltration of the entire leaflet.

Laboratory parameters such as WBC count, CRP, and blood cultures can also be useful in distinguishing infective endocarditis from Libman-Sacks endocarditis.[4] Leukocytes tend to decrease during lupus activity, and the opposite occurs in infectious endocarditis. Very high CRP levels suggest an infectious cause; blood cultures are more important for a definitive differential diagnosis.[5]

In our case, a diagnosis of Libman-Sacks endocarditis was more probable, as the leukocyte count was slightly elevated, the CRP was not highly elevated, and blood culture samples had negative results. SLE patients have an increased prevalence of left ventricular hypertrophy that is not exclusively a result of concomitant coronary artery or valvular heart disease, renal involvement, or other traditional stimuli, including hypertension. Our patient has an acute onset of renal failure and hypertension that doesn't explain the existing left ventricular hypertrophy. Results of studies suggest that inflammation-mediated arterial stiffening, a strong determinant of left ventricular hypertrophy, is likely the underlying mechanism for Libman-Sacks endocarditis in systemic lupus erythematosus.[6,7] Libman-Sacks endocarditis is known to lead to an increased risk of stroke, coronary artery disease, and sudden cardiac death in varied populations and, therefore, is likely to be a prognostic indicator of cardiac morbidity and mortality in SLE patients.[8,9] Our findings suggest that more aggressive and targeted therapy may be needed to control the inflammation-mediated effects on vascular stiffening that lead to left ventricular hypertrophy.

In conclusion, SLE patients will be more likely to develop cardiac manifestations, such as valvular regurgitation and possible Libman-Sacks endocarditis.

Competing Interests

The authors declare that they have no competing interests.

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