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CASE REPORT

LIBMAN SACKS VEGETATION PRESENTING AS SYSTEMIC LUPUS ERYTHEMATOSUS

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ABSTRACT

A case of 14 year-old female patient presented to our hospital complaining of loss of weight and low grade fever for one year. Echocardiography showed the presence of sterile vegetation on the mitral valve known as Libman-Sacks Endocarditis (LSE). Patient was later diagnosed to have systemic lupus erythematosus (SLE).

KEY WORDS: Sterile vegetation; SLE; Libman-sacks.

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INTRODUCTION

Libman–Sacks endocarditis (LSE) is sterile fibrinous vegetation that preferentially develops 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]

CASE REPORT

The patient was a 14-year-old girl, student, single, has presented with low grade fever for 1 year with significant loss of weight of 10 kg in one year. She has also complained of dyspnoea on moderate exertion. Clinical examination showed her BP as 100/70 and heart rate 90 beats per minute. Peripheral pulse was well felt in all four limbs. Cardiac examination revealed pan systolic murmur at the apical area. Chest and abdominal examination was normal. No skin, oropharyngeal lesions, arthritis or any joint deformities were observed. The electrocardiogram was normal. Transthoracic echocardiography was done for the patient showing two vegetation. First one is attached to the ventricular surface of the posterior mitral valve leaflet measuring 12×11 mm, and causing mild anteriorly directed eccentric mitral regurgitation (MR). The other one is smaller and attached to the left ventricular (LV) side of the anterior mitral leaflet and measures 5×6 mm (Figure 1, 2).

The left ventricular ejection fraction was normal measuring 64%; the right sided chambers were not dilated. Other valves showed normal morphology and flow with no masses or vegetation attached. Three pairs of blood culture samples were collected from different sites and all had negative results. Haemoglobin =10.3 mg/dL, platelets =292,000 and leukocytes =7100, creatinine =0.7 mg/dL, normal urinary sediment. Anti-nuclear antibody (ANA), Anti-Ds-DNA antibody was positive and anticardiolipin IgG antibodies were negative, C-reactive protein (CRP) =3.95 mg/L, C3 was decreased and C4 was normal. Urine routine examination 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.

This is a case report of a patient with SLE who presented with fever and exertional dyspnoea. The patient had aseptic vegetation in the mitral valve with mild MR.

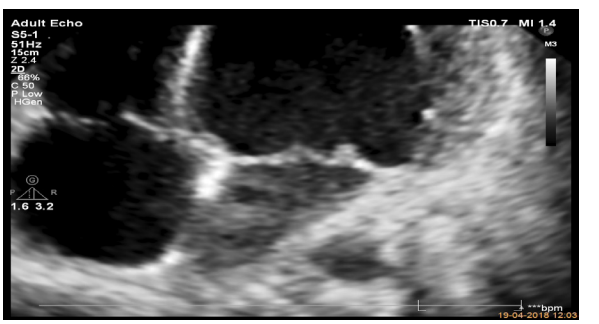

Figure 1: Libman sacks vegetation presenting as SLE on echocardiography.

Figure 2: Libman sacks vegetation presenting as SLE on echocardiography.

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Libman–Sacks endocarditis (LSE) (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 vegetation in 1924.[3] Libman-Sacks endocarditis most commonly involve mitral and aortic valves. However, all four cardiac valves and the endocardial surfaces can be involved[4] .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.[4]

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.[2]

DISCUSSION

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.[1] Thus, the diagnosis of LSE is important and the echocardiogram is currently the best imaging procedure for diagnosis.[5] 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[2]. 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.[6] 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 vary from 1% to 8% of cases.[7] 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.[2] Our patient has presented with long standing fever with weight loss. The only positive findings in cardiac examination are presence of vegetation in the ventricular aspect of mitral valve. Normally cardiac vegetation is of infective origin, but in this case all blood culture reports were negative.

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CONFLICT OF INTEREST

Authors declare no conflict of interest.

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None declared.

AUTHORS' CONTRIBUTION

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Conception or Design:	PKC
Acquisition, Analysis or Interpretation of Data:	PKC, BM, LA
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All the authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.