Case report

Libman-Sacks Endocarditis as the first manifestation of Systemic Lupus Erythematosus :a case report .

Abstract

Libman-Sacks endocarditis (LSE) is a rare cardiovascular manifestation of systemic lupus erythematosus/antiphospholipid syndrome and has been described as a sterile, verrucous, non-bacterial vegetative. These lesions can cause progressive damage to heart valves and lead to valve surgery. The most commonly affected valves are the aortic and mitral valves. The majority of LSE patients are usually asymptomatic. Case overview , we describe a 29-year-old women patient who presented with dyspnea , increased pulmonary congestion and severe mitral regurgitation with vegetation revealed by transthoracic echocardiography and was diagnosed as Libman-Sacks endocarditis after laboratary test including blood cultures, autoimmune profile, and testing for hypercoagulability..

Keywords : Libman-Sacks endocarditis , Systemic lupus erythematous

Introduction

Libman-Sacks endocarditis (LES) was first described by Libman and Sacks in 1924 as a cardiac manifestation of Systematic Lupus Erythematous (SLE) and antiphospholipid antibody syndrome (APS) (1). LSE is an uncommon disorder that might be confused with infective endocarditis . It is characterized by thrombus deposition primarily in the aortic and mitral valves. Involvement of other heart valves is less common (2).

The majority of patients with LSE are usually asymptomatic, but when patients are symptomatic, this is usually due to embolic infarction, either due to cerebrovascular or systemic thromboembolism. Patients with SLE and APS may show signs and symptoms of underlying conditions, such as cheek rashes and recurrent miscarriages (3,4)

We report the case of an adult patient who presented with LSE as the first symptom of SLE.

Case report

A 29-year-old women , with no medical history or cardiac disease , presented to the emergency with a two-month history of exertional dyspnea classified as class III of the New York Heart Association (NYHA) with cough ,palpitations and migratory multiple small and large joints pain. The patient reported five undefined syncope attacks during the past year. She reported also 3-month history of lethargy, anorexia, and weight loss. The patient denied fever, or substance abuse. Family history was unremarkable for malignancy or heart disease.

Physical examination revealed that her temperature was 37.3°C, her blood pressure was 118/75 mmHg, her heart rate was 115 beats/min, her oxygen saturation in room air was 97%, and her respiratory rate was 23 beats/min. Cardiovascular examination revealed a Levine grade II/VI systolic murmur at the apex . Chest examination reveals decreased right lower zone breath sounds, bilateral

basal crackles. Upon arrival, an electrocardiogram (ECG) was performed. and showed sinus tachycardia and normal voltage (Figure 1) . Subsequent transthoracic echocardiography (TTE) showed thickening of the anterior mitral leaflet with severe mitral regurgitation , left atrium dilatation , and no left ventricular (LV) dilatation and normal LV systolic function (Figure 2). Regurgitation and vegetation was evident and blood cultures was négative . Laboratory tests showed that the white blood cell count and C-reactive protein levels were normal andrevealed hypochromic microcytic anemia with thrombocytopenia and acute kidney injury (Table 1).. To explore the renal dysfunction , a Renal biopsy was made and showed: Localized proliferative lupus nephritis class III and membranous lupus nephritis class V (ISN/RBS).A biological investigation in the context of SLE were performed and showed low C3 and C4 complement and positive anti-dsDNA and negative antinuclear body .

Based on clinical and physical findings , SLE with nephritis and Libman-Sachs endocarditis was definitively diagnosed . She was hospitalized for 20 days, during which her SLE flare-ups were adequately managed with intravenous methylprednisolone and cyclophosphamide. After that , she took 6-month oral prednisolonewithesomeprazole, calcium, and vitamin D and monthly intravenous cyclophosphamide. Her heart failure symptoms were reduced with furosemide, spironolactone, and captopril.

Echocardiography made 4 months later revealed mild mitral regurgitation and reduced thickness of the anterior mitral leaflet. For this reason; no surgical intervention was performed She clinically returned to normal life and was slowly weaned off medication for heart failure.SLE were under control and she is still being followed up by a team of rheumatologists, cardiologists and nephrologists.

Discussion

According to the literature, the prevalence of cardiovascular disease in patients with SLE is estimated to exceed 50% (5,6). SLE manifests as pericarditis, arrhythmias, conduction abnormalities, myocarditis ,increased pulmonary pressure and coronary artery disease (7). In SLE the left heart valve was most frequently affected especially the mitral valve, followed by the aortic valve,(6,8). Mitral valve involvement in SLE patients has been classified as cusp thickening, vegetations , regurgitation, and valvular stenosis(8).

Because some SLE or APS patients are asymptomatic, the approach to diagnosing LSE may not be straightforward and can be difficult in the absence of cardiac symptoms, routine echocardiographic screening is recommended for SLE patients, even in the absence of cardiac symptoms (9). TTE is the best initial modality for diagnosing LSE, but transesophageal echocardiography is more sensitive (9,10). TTE detects 18% to 50% of valve disease (1, 10), and transesophageal echocardiography detects up to 74% (8, 9). TheLSE imaging is described as an irregular and non-uniform echo density without independent motion of the verrucous vegetation on the heart valve and endocardium [8, 9]. The development of valvular lesions in SLE is closely associated with the presence of antiphospholipid antibodies (9), but not in other SLE and LSE patients (8) or non-bacterial thrombotic endocarditis without underlying disease. Even negative test results have been reported in the literature.

The diagnosis of LSE is difficult to confirm with laboratory tests. However, patients with suspected LSE must undergo a complete blood count, comprehensive metabolic assessment, blood cultures, autoimmune profile, and testing for hypercoagulability(8).

Treatment of valvular symptoms of SLE depends on the type and severity of the lesion. Treatment options remain challenging due to the lack of large systematic studies.

There is controversy about the place of steroids in this context. Some investigators have suggested that the introduction of corticosteroids as a basis for SLE treatment may reduce symptom frequency and disease activity (3,11). Steroids do not prevent LES , but they promote healing of the lesion over time

by reducing inflammation and valve damage. However, they can promote fibrosis, scarring , and hyperplasia, leading to additional damage and valve dysfunction (3,5,12).There was a case report of the rapid onset of severe mitral regurgitation in this SLE patient receiving high-dose corticosteroid therapy for recurrence of acute disease (11).

Surgical valve replacement is recommended in symptomatic and severe cases of LSE. Mechanical valve replacement in women of childbearing potential is discouraged because it requires anticoagulant therapy with increased fetal and maternal side effects. However, it is still recommended as many authors believe that patients with SLE/APS will eventually require anticoagulants for disease-related thromboembolism (12 , 13).

Conclusion

LSE is rare as the first manifestation of SLE/APS. We presented a 29-year-old women with LSE as first-onset SLE. She was successfully diagnosed with echocardiography and treated with medical therapy . This case highlights SLE as a differential diagnosis when a healthy person presents with new-onset valvular disease.

References

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List of abbreviations:

LSE :Libman-Sacks endocarditis

SLE : Systematic Lupus Erythematous

ECG : electrocardiogram

APS :and antiphospholipid antibody syndrome

TTE :transthoracic echocardiography

LV: left ventricular

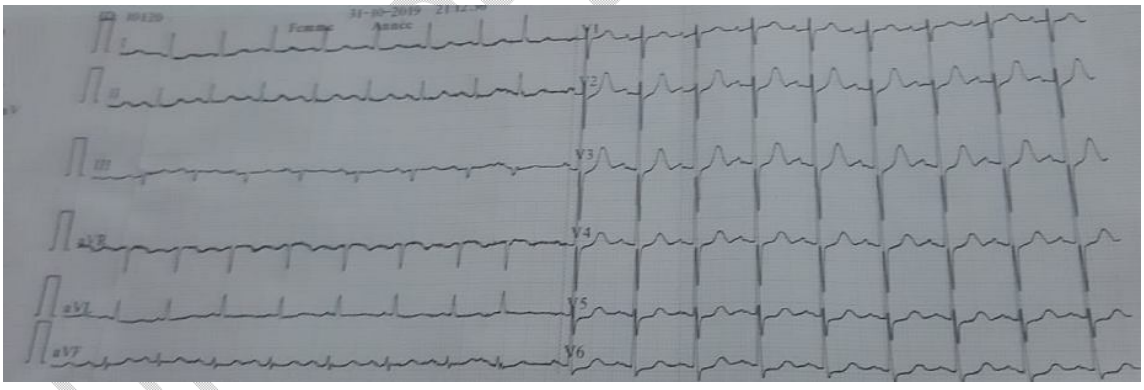

Figure 1: ECG with sinus tachycardia and normal voltage

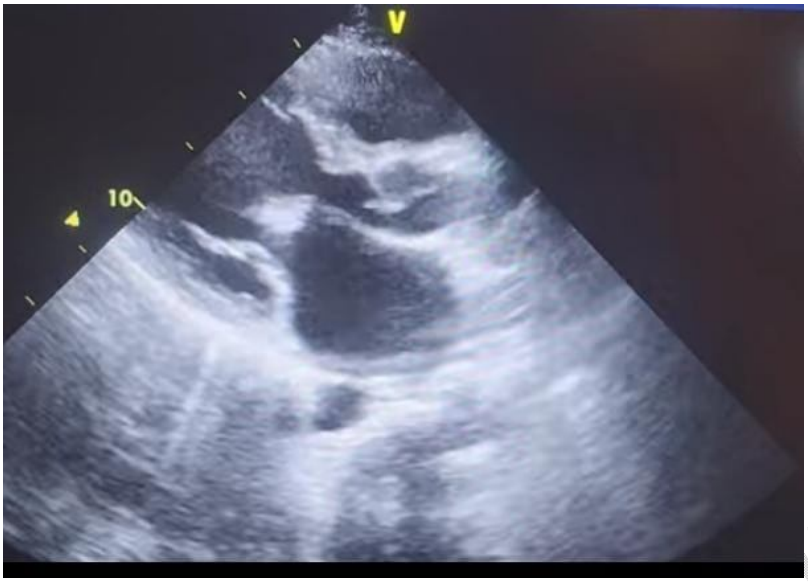

Figure 2: TTE showedvegetation on mitral valve .

Labs	**Results**	**Reference range**
Hemoglobin	7.8g/dl	11.5-16.5g/dl
Platet	131*109/L	150-450 *109/L
White cell count	10*109/L	4.5-13.5*109/L
Creatinin	150 μmol/L	45-102μmol/L
Brain natriuretic peptide	980 pg/ml	<100 pg/mL
Troponin T	92 ng/ml	<14ng/ml
C-reactiveprotein	65 mg/l	<5 ng/l
Antinuclearantibody	Positive	Negative
Anti-double Stranded DNA	>500UI/ml	0-10UI/ml
C3 complement	0.64 g/dl	0.9–1.9 g/dL
C4 complement	0.04g/dl	0.1-0.4g/dl

Table 1 : laboratory data for the patient .