Case report

Systemic Lupus Erythematosus revealed by Libman-Sacks Endocarditis: a case report .

Abstract

Libman-Sacks endocarditis (LSE) is a rare cardiovascular manifestation of systemic lupus erythematosus and has been described as an asepticverrucous non-bacterialautonomicdisease. Theselesions can cause progressive damage to heart valves and mayneedsurgery . The aortic and mitral valves are the mostcommonlyaffected valves . The majortiy of LSE patients are usuallyasymptomatic . 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

In1924 ,Libman and Sacks , first desribe LSE as a cardiac manifestation of systemic lupus erythematosus (SLE) and antiphospholipidantibody syndrome (APS) (1). LSE is a rare condition that can beconfusedwithinfectiveendocarditis. It ischaracterized essentially by thrombus deposition in the aortic and mitral valves. Involvement of otherheart valves isnot common (2).

The majority of patients with LSE are usually asymptomatic, but when they 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 joints pain. The patient reported five undefined syncope attacks during last year. Shealso reported 3-month history of anorexia , weight loss 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, blood pressure was 118/75 mmHg,, heart rate was 115 beats/min , oxygen saturation in room air was 97%, and 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 a severe miral regurgitation with thickening of the anterior mitral leaflet , 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 normal white bloodcellcounts and C-reactiveproteinlevels, consistent withhypochromicmicrocyticanemiawiththrombocytopenia and acute kidneyinjury (Table 1). To investigaterenaldysfunction, a renalbiopsywasperformed and showed f ocalproliferative lupus nephritis class III and membranous lupus nephritis class V (ISN/RBS). Bioassaysassociatedwith SLE wereperformed and showedlow C3 and C4 complement, positive anti-dsDNA, and negativeantinuclear bodies.

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

Echocardiographyperformed 4 monthslaterrevealedmild mitral regurgitation and reducedanterior mitral leafletthickness. For this; no surgical intervention wasperformed. Shereturnedclinically to a normal life and wasslowlyweaned off medication for heart failure.SLE is under control and isstillbeingfollowed up by a team of cardiologist ,rheumatologistsand nephrologists.

Discussion

The literatureestimatesthat the prevalence of cardiovasculardisease in SLE patients exceeds 50% (5,6). SLE manifests as mypcarditis , pericarditis, increasedpulmonary pressure , arrhythmias, conduction disturbances , and coronaryarterydisease (7). In SLE, the leftheart valve, especially the mitral valve, wasmostcommonlyaffected, followed by the aortic valve (6,8). Mitral valve involvement in SLE patients has been classified as apical thickening, warts, regurgitation, and valvularstenosis (8).

The approach to LSE diagnosisis not straightforward and can bedifficult in the absence of cardiacsymptoms. Routine echocardiographic screening isrecommendedfor all SLE patients, even cardiacsymptoms are not present (9). The best initial modality for diagnosing LSE isTTE , but transesophagealechocardiographyis more sensitive (9,10). TTE detects 18% to 50% of valve disease (1, 10) and transesophagealechocardiographydetects up to 74% (8, 9). LSE imaging has been described as irregular and non-uniformechodensitieswithout the inherent motion of wartyvegetation on heart valves and endocardium [8, 9]. The development of valvularlesions in SLE iscloselyassociatedwith the presence of antiphospholipidantibodies (9), whereasotherher SLE and LSE patients (8) or non-bacterialthromboticendocardiumwithoutunderlyingdisease Not sowith flames. In the literature, evennegative test results have been reported.

It isdiffucult to confirm the diagnosisof withlaboratory tests. However, when LSE issuspected , patients shouldundergo a completeblood count, bloodcultre , comprehensivemetabolicassessment, , autoimmune profile, and hypercoagulabilitytesting (8).
Treatment of SLE heart valve symptomsdepends on the type and severity of the lesion. Treatment options remainchallenging due to the lack of large-scalesystematicstudies.

The role of steroids in thiscontextiscontroversial. Someinvestigators have suggestedthat the introduction of corticosteroids as a basis for SLE treatmentmayreducesymptomfrequency and diseaseactivity (3,11). The steroids do not preventher LES, but they do acceleratehealing of the lesions over time by reducing inflammation and valve damage. However, they can promotefibrosis, scarring and hyperplasia, leading to additional damage and valve dysfunction (3,5,12). There was one case report of the rapidonset of severe mitral regurgitation in this SLE patient whowasreceiving high-dose corticosteroidtherapy for acute diseaserecurrence (11).

In symptomatic and severe cases of LSE ,surgical valve replacement isrecommended. Mechanical valve replacement in women of childbearingpotentialis not recommendedbecauseitrequires anticoagulant therapywithincreasedfetal and maternalsideeffects. However, manyauthorsbelievethat patients with SLE/APS willeventuallyrequire anticoagulants for disease-relatedthromboembolism and are stillrecommended (12, 13).

Conclusion

LSE is rare as the first manifestation of SLE/APS. Weintroduced a 29-year-old womanwith LSE as first-onset SLE. Shewasdiagnosednormally on herechocardiogram and treatedwithmedication. This case highlights her SLE as a differentialdiagnosiswhen a healthypersonpresentswith new-onsetvalvularheartdisease.

Consent

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

Ethical Approval:

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

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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

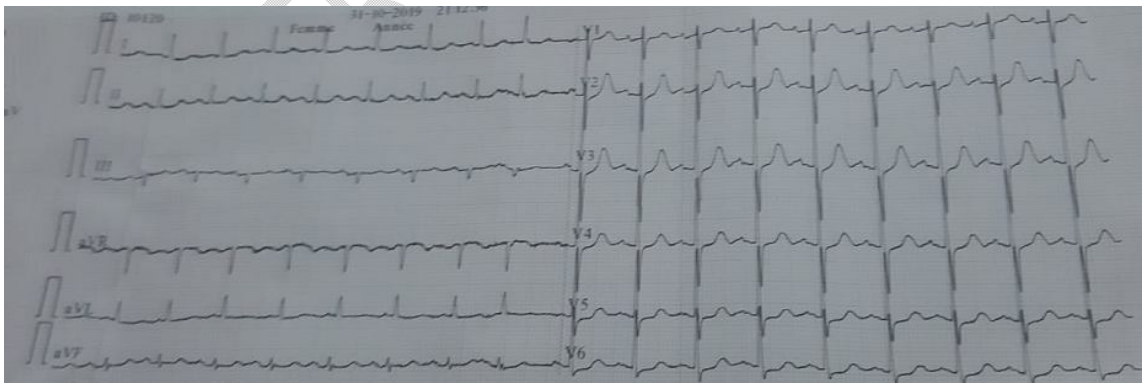

Figure 1: ECG with sinus tachycardia and normal voltage

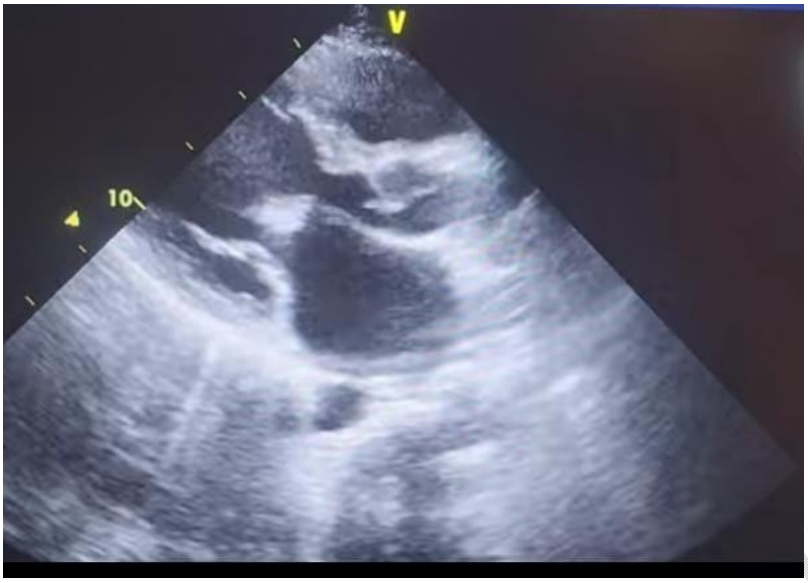

Figure 2: TTE showedvegetation on mitral valve .

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 .