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Systemic Lupus Erythematosus revealed by Libman-Sacks Endocarditis: A Case Report

Asmaa Elamraoui [a++*], Adam Fadoul [a++], Yousra Hamine [a++], Ghali Bennouna [a#], Leila Azzouzi [a#], Abdenasser Drighil [a#] and Rachida Habbal [a†]

[a] *Cardiology Department, CHU Ibn Rochd, N°1 quartier des Hopitaux-24200, Casablanca, Morocco.*

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Libman-Sacks endocarditis (LSE) is a rare cardiovascular manifestation of systemic lupus erythematosus and has been described as an aseptic verrucous non-bacterial autonomic disease. Theselesions can cause progressive damage to heart valves and may need surgery. The aortic and mitral valves are the most commonly affected 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.

[++] *Resident Doctor;*
[#] *Associate Professor;*
[†] *Head of Service;*
**Corresponding author: Email: asmaa.elamraoui12@gmail.com;*

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ABBREVIATIONS

LSE : Libman-Sacks endocarditis
SLE : Systematic Lupus Erythematous
ECG : Electrocardiogram
APS : And antiphospholipid antibody syndrome
TTE : Transthoracic echocardiography
LV : Left ventricular

1. INTRODUCTION

In 1924, Libman and Sacks, first desribe LSE as a cardiac manifestation of systemic lupus erythematosus (SLE) and antiphospholipidanti-body syndrome (APS) [1]. LSE is a rare condition that can be confused with infective endocarditis. It is characterized essentially by thrombus deposition in the aortic and mitral valves. Involvement of other heart valves is not 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.

2. 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. She also 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 (Fig. 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 (Fig. 2). Regurgitation and vegetation was evident and blood cultures was négative. Laboratory tests showed normal white bloodcellcounts and C-reactiveproteinlevels, consistent with hypochromic micro cyticanemia with thrombocytopenia and acute kidney injury (Table 1). To investigate renaldys function, a renal biopsy was performed and showed f ocalproliferative lupus nephritis class III and membranous lupus nephritis class V (ISN/RBS). Bioassays associated with SLE were performed and showed low C3 and C4 complement, positive anti-dsDNA, and negative antinuclear 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 prednisolone with esomeprazole, calcium, and vitamin D and monthly intravenous cyclophosphamide. Her heart failure symptoms were reduced with furosemide, spironolactone, and captopril.

Echocardiography performed 4 months later revealed mild mitral regurgitation and reduce danterior mitral leaflet thickness. For this; no surgical intervention was performed. Shereturne dclinically to a normal life and was slowly weaned off medication for heart failure.SLE is under control and is still being followed up by a team of cardiologist, rheumatologists and nephrologists.

3. DISCUSSION

The literature estimates that the prevalence of cardio vascular disease in SLE patients exceeds 50% [5,6]. SLE manifests as mypcarditis, pericarditis, increased pulmonary pressure, arrhythmias, conduction disturbances, and coronary artery disease [7]. In SLE, the

leftheart valve, especially the mitral valve, was most commonly affected, followed by the aortic valve [6,8]. Mitral valve involvement in SLE patients has been classified as apical thickening, warts, regurgitation, and valvular stenosis [8].

The approach to LSE diagnosisis not straightforward and can bedifficult in the absence of cardiacsymptoms. Routine echocardiographic screening is recommended for all SLE patients, even cardiacsymptoms are not present [9]. The best initial modality for diagnosing LSE is TTE, but transesophageal echocardiography is more sensitive [9,10]. TTE detects 18% to 50% of valve disease [1,10] and transesophageal echo cardio graphydetects up to 74% [8,9]. LSE imaging has been described as irregular and non-uniformechodensities without the inherent motion of warty vegetation on heart valves and endocardium [8,9]. The development of valvular lesions in SLE is closely associated with the presence of anti-phospholipid antibodies [9], whereas other her SLE and LSE patients [8] or non-bacterial thrombotic endocardium without under lyingdisease not so with flames. In the literature, evennegative test results have been reported.

It is diffucult to confirm the diagnosis of with laboratory tests. However, when LSE issus pected, patients should undergo a complete blood count, bloodcultre, comprehensive metaboli cassessment, autoimmune profile and hyper coagulability testing [8]. Treatment of SLE heart valve symptoms depends on the type and severity of the lesion. Treatment options remain challenging due to the lack of large-scale systematic studies.

The role of steroids in this context is controversial. Some investigators have suggested that the introduction of corticosteroids as a basis for SLE treatment may reduce symptom frequency and disease activity [3,11]. The steroids do not prevent her LES, but they do acceleratehealing of the lesions 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 one case report of the rapidonset of severe mitral regurgitation in this SLE patient who was receiving high-dose corticosteroid therapy for acute diseasere currence [11].

In symptomatic and severe cases of LSE, surgical valve replacement is recommended. Mechanical valve replacement in women of child bearing potentialis not recommended because it requires anticoagulant therapy with in creasedfetal and maternalside effects. However, many authors believe that patients with SLE/APS will eventually require anticoagulants for disease-related thromboembolism and are still recommended [12,13].

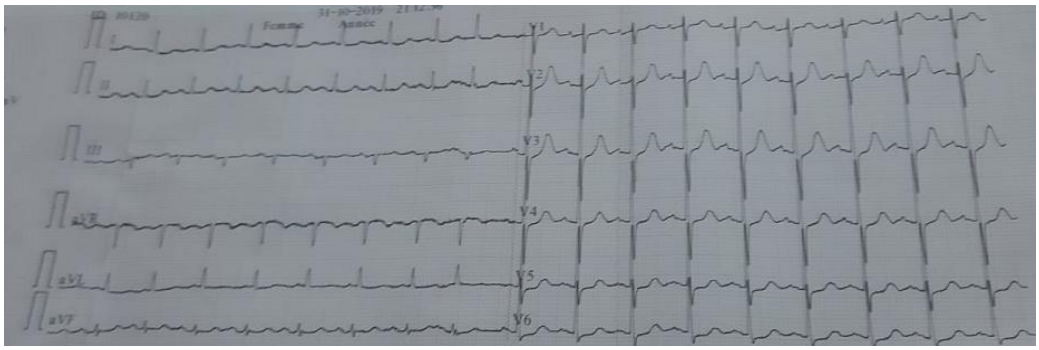

Fig. 1. ECG with sinus tachycardia and normal voltage

Table 1. Laboratory data for the patient

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 *µ*mol/L	45-102*µ*mol/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

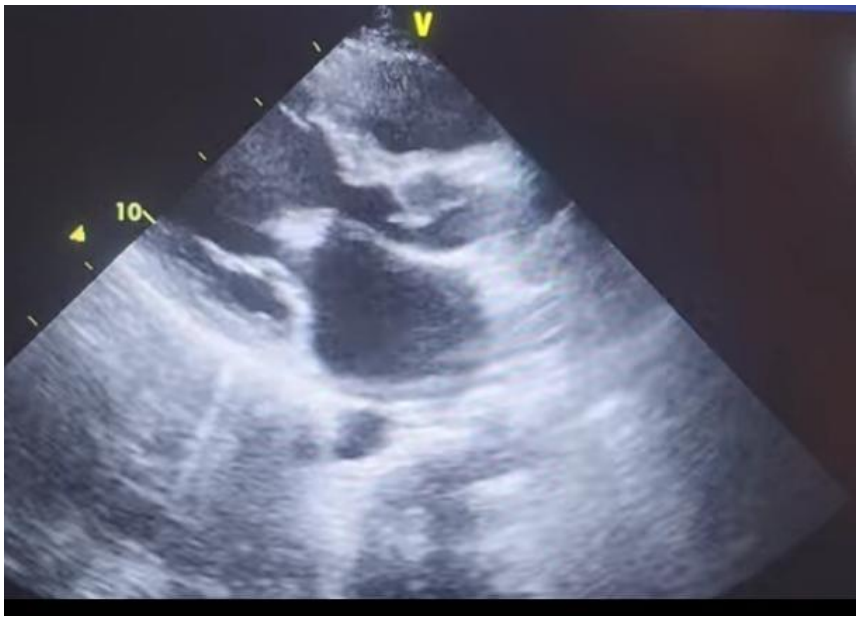

Fig. 2. TTE showed vegetation on mitral valve

4. CONCLUSION

LSE is rare as the first manifestation of SLE/APS. We introduced a 29-year-old woman with LSE as first-onset SLE. She was diagnosed normally on herechocardiogram and treated with medication. This case highlights her SLE as a differential diagnosis when a healthy person presents with new-onset valvular heart disease.

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).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Bourré-Tessier J, Huynh T, Clarke AE, Bernatsky S, Joseph L, Belisle P, Pineau CA. Features associated with cardiac abnormalities in systemic lupus erythematosus. *Lupus*. 2011;20(14):1518–1525.
DOI: 10.1177/0961203311420318
2. Ginanjar E, Yulianto Y. Autoimmune disease with cardiac valves involvement: Libman-sacks endocarditis. Acta Med Indones. 2017;49(2):148-150.
3. Ferreira E, Bettencourt PM, Moura LM. Valvular lesions in patients with systemic lupus erythematosus and antiphospholipid syndrome: An old disease but a persistent challenge. *Revista* Portuguesa de Cardiologia. 2012;31(4):295–299.
DOI:10.1016/j.repc.2012.02.005
4. Yamashita G, Kanemitsu N, Nakashima Y, Matsuo T, Nakane T, Honda M, Okabayashi H. Hypertrophic obstructive cardiomyopathy and mitral regurgitation in Libman-Sacks endocarditis. Gen Thorac Cardiovasc Surg. 2020;68(2):181-184.
5. Akhlaq A, Ali TA, Fatimi SH. Mitral valve replacement in systemic lupus erythematosus associated Libman- Sacks endocarditis. Journal of the Saudi Heart Association. 2016;28(2):124–126.
DOI:10.1016/j.jsha.2015.09.003
6. Ibrahim AM, Siddique MS. Stat Pearls. Treasure Island FL: Stat Pearls Publishing. Libman *Sacks endocarditis*; 2020.
7. Ferreira E, Bettencourt PM, Moura LM. Valvular lesions in patients with systemic lupus erythematosus and antiphospholipid syndrome: An old disease but a persistent

challenge. Revista Portuguesa de Cardiologia. 2012;31(4):295–299.
DOI: 10.1016/j.repc.2012.02.005.

8. Roldan CA, Tolstrup K, Macias L, Qualls CR, Maynard D, Charlton G, Sibbitt WL Jr .Libman-Sacks endocarditis: detection, characterization, and clinical correlates by three-dimensional transesophageal echo-cardiography.. J Am Soc Echocardiogr. 2015;28:770–779
9. Yoo BW, Lee SW, Song JJ, Park YB, Jung SM. Clinicalcharacteristics and long-term out comes of Libman-Sacks endocarditis in patients with systemic lupus erythematosus. Lupus. 2020;29(9):1115-1120.
10. Chen S, Smith B, Ilina M, Bowen C, Martin N. PReS-FINAL-2307: Libman-Sacks endocarditis as a presentation for systemic lupus erythematous in an adolescent with isolated mitrial regurgitation and Noonan syndrome. Pediatric Rheumatology. 2013;11(Supplement2) DOI: 10.1186/1546-0096-11-s2-p297
11. Gorki H, Malinovski V, Stanbridge RDL. The antiphospholipid syndrome and heart valve surgery. European Journal of Cardio-Thoracic Surgery. 2008;33(2): 168–181.
DOI: 10.1016/j.ejcts.2007.11.004
12. Akhlaq A, Ali TA, Fatimi SH. Mitral valve replacement in systemic lupus erythematosus associated Libman- Sacks endocarditis. Journal of the Saudi Heart Association. 2016;28(2):124–126.
DOI: 10.1016/j.jsha.2015.09.003
13. Vassileva CM, Swong MN, Boley TM, Markwell SJ, Hazelrigg SR. Influence of systemic lupus erythematosus on procedure selection and outcomes of patients undergoing isolated mitral valve surgery. Journal of Cardiac Surgery. 2012; 27(1):29–33.
DOI: 10.1111/j.1540-8191.2011.01393.x

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