Libman-Sacks Endocarditis in a Congenital Valve Defect: A Case Report

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Case Report

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Abstract

**Background:** Libman-Sacks endocarditis (LSE) is a rare complication of systemic lupus erythematosus (SLE) characterized by non-infectious vegetations on normal heart valves. Here, we present a case of SLE with a congenital valve malformation.

**Case presentation:** The patient, a 20-year-old, had been undergoing treatment for SLE for one year with a good response. One year later, she presented with stage IV dyspnea and chest pain, accompanied by signs of right heart failure on clinical examination. Investigations revealed active lupus and vegetations on the mitral valve, along with agenesis of the posterior mitral leaflet and severe mitral insufficiency on echocardiography. The patient received corticosteroids, antibiotics, anticoagulants, and symptomatic treatment for heart failure. Despite initial clinical improvement, the patient’s condition worsened and did not respond to resuscitation measures.

**Conclusion:** While LSE typically responds favorably to treatment, severe valvulopathy, as seen in our patient with congenital valve malformation, can lead to fatal outcomes.

**Introduction**

Libman-Sacks endocarditis (or non-bacterial thrombotic endocarditis) is a rare cardiac complication of systemic lupus erythematosus (SLE) in which there is non-infectious vegetation on normal heart valves, particularly the mitral valve (1). This condition is generally asymptomatic, and diagnosis is based on echocardiography and/or histopathology. Treatment typically involves anticoagulation, corticosteroids, and sometimes surgery (2). Here, we report a case of Libman-Sacks endocarditis in the setting of a congenital heart malformation in a patient with lupus.

**Case Presentation**

A 20-year-old female, deaf-mute since birth, was followed in our department for one year for systemic lupus erythematous (SLE), presenting with joint involvement including polyarthritis affecting large, medium, and small joints, hemolytic anemia, and immunological involvement (Table I). She was on corticosteroids 20mg per day and synthetic antimalarials with a good response. One year later, she presented with stage IV dyspnea associated with chest pain. Clinical examination revealed signs of right heart failure. Investigations showed elevated inflammatory markers and abnormal liver function tests (Table I).

**Table I: Paraclinical Data of the Patient**
<table>
<thead>
<tr>
<th></th>
<th>Data at lupus diagnosis</th>
<th>Data at Decompensation</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Hemoglobin (g/dl)</strong></td>
<td>12.6</td>
<td>9.7</td>
</tr>
<tr>
<td><strong>MCV</strong></td>
<td>87</td>
<td>82</td>
</tr>
<tr>
<td><strong>MCHC</strong></td>
<td>32</td>
<td>32</td>
</tr>
<tr>
<td><strong>White Blood Cells</strong></td>
<td>3300</td>
<td>7430</td>
</tr>
<tr>
<td><strong>Lymphocytes (per mm3)</strong></td>
<td>780</td>
<td>743</td>
</tr>
<tr>
<td><strong>Platelets</strong></td>
<td>319000</td>
<td>442000</td>
</tr>
<tr>
<td><strong>ANA (UI/ml)</strong></td>
<td>&gt;160</td>
<td>NF</td>
</tr>
<tr>
<td>(Negative Typing)</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Anti-nDNA antibodies</strong></td>
<td>Positive</td>
<td>NF</td>
</tr>
<tr>
<td><strong>C3 (g/l)</strong></td>
<td>1.30</td>
<td>0.55 (0.90-2.10)*</td>
</tr>
<tr>
<td><strong>C4 (g/l)</strong></td>
<td>0.23</td>
<td>0.05 (0.10-0.40)*</td>
</tr>
<tr>
<td><strong>C1q (mg/l)</strong></td>
<td>NF</td>
<td>72 (118-244)*</td>
</tr>
<tr>
<td><strong>CRP (mg/l)</strong></td>
<td>11</td>
<td>22</td>
</tr>
<tr>
<td><strong>ASAT (UI/ml)</strong></td>
<td>23</td>
<td>74</td>
</tr>
<tr>
<td><strong>ALAT (UI/ml)</strong></td>
<td>16</td>
<td>171</td>
</tr>
<tr>
<td><strong>D-dimers (mg/l)</strong></td>
<td>NF</td>
<td>8.05 (&lt;0.28)*</td>
</tr>
<tr>
<td><strong>PASP (mmHg)</strong></td>
<td>18</td>
<td>80</td>
</tr>
</tbody>
</table>

MCHC: Mean Corpuscular Hemoglobin Concentration; MCV: Mean Corpuscular Volume; PASP: Pulmonary Artery Systolic Pressure; "NF" indicates that the data was not available; *Normal Range

Lupus workup revealed consumption of complement C3, C4, and C1q. Transthoracic echocardiography showed vegetation on the mitral valve associated with agenesis of the posterior mitral leaflet and severe mitral insufficiency with severe pulmonary hypertension. Thoraco-abdominal and cerebral CT angiography did not show signs of thrombosis. Screening for antiphospholipid syndrome and infectious workup were negative. The patient was started on corticosteroids at 1mg per kg per day, antibiotics (third-generation cephalosporin combined with gentamicin), anticoagulation, and symptomatic treatment for heart failure, with an indication for valve replacement. The patient initially showed clinical improvement but later experienced worsening heart failure complicated by cardiogenic shock resistant to resuscitation measures.

**Discussion**
Libman-Sacks endocarditis (LSE) is a rare but severe manifestation in patients with systemic lupus erythematosus (SLE) and other autoimmune diseases. It is primarily found at autopsy, with a prevalence of 0.9 to 1.6% (3). LSE typically affects lupus patients aged 40 to 80 years, with a predilection for the mitral valve, followed by the aortic and tricuspid valves, and exceptionally simultaneous involvement of two or three valves (4,5). Patients often present with asymptomatic valvular abnormalities, necessitating echocardiography for accurate diagnosis (6,7). Among the 135 reported cases of LSE, no author has reported an underlying cardiac malformation, highlighting the uniqueness of our case. The etiology of LSE is complex, involving immune mechanisms leading to a state of hypercoagulability and valve damage (1). Diagnosis requires clinical acumen, with echocardiography being the main evaluation method (8). Treatment focuses on managing the underlying disease, whether it be SLE in our case. Corticosteroids are used to reduce inflammation but may lead to tissue scarring and fibrosis, predisposing to further valve damage (9,10). Anticoagulation is considered for secondary prevention, and surgical valve replacement is recommended for severe cases. Regular follow-up is crucial to monitor disease progression (2). Surgical valve replacement is recommended for symptomatic and severe cases of LSE (11). Regular follow-up is crucial to monitor disease progression.

**Conclusion**

Libman-Sacks endocarditis typically responds favorably to treatment except in cases of severe valvulopathy, which can be fatal, as in our patient who presented with decompensated heart failure due to Libman-Sacks endocarditis on the background of a congenital heart malformation. Hence, strict clinical and echocardiographic monitoring of lupus patients is warranted.

**Declarations**

**Funding.**

None.

**Conflicts of interest.**

The authors declare that they have no competing interest.

**Ethics approval and consent to participate.**

The case report was approved by the Ethics Committee of the University Hospital of Ibn Rochd.

**Consent for publication.**

written informed consent to publish was obtained from the mother of the patient.

**Availability of data and material.**

Data concerning the patient’s record are available from the corresponding author on reasonable request.
Authors' contributions.

SZ: conception, data curation, interpretation, and writing of the manuscript.

KN, AA, SJ: conception, data curation, and supervision of the draft.

References


