

Nephrotic Syndrome Associated with Libman-Sacks Endocarditis as a First Manifestation of Systemic Lupus Erythematosus: A Case Report and Review of the Literature

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ABSTRACT

Libman-Sacks endocarditis is a rare but well-recognized cardiac manifestation of systemic lupus erythematosus, characterized by sterile, verrucous vegetations on heart valves. We report an exceptional case of a 29 year old male patient who presented with profound nephrotic syndrome and mitral aortic endocarditis as the initial manifestation of previously undiagnosed systemic lupus erythematosus associated with antiphospholipid syndrome. The clinical presentation included inflammatory arthritis, constitutional symptoms, prolonged fever, and chronic inflammatory left subpalpebral cellulitis. Echocardiography revealed mitral and aortic valve thickening with fine vegetations on the aortic side of the posterior sigmoid cusp, along with significant mitral and aortic regurgitation. Immunological workup was strongly positive for antinuclear antibodies with a speckled pattern, IgG anti-cardiolipin antibodies, and IgG anti-beta-2-glycoprotein I antibodies. Renal biopsy was consistent with lupus glomerulonephritis. High dose corticosteroids combined with double antibiotic therapy led to significant clinical and biological improvement within 21 days. This observation is remarkable because cardiac involvement typically occurs years after systemic lupus erythematosus onset, whereas here it presented as an initial, aggressive manifestation in a male subject, associating renal involvement, endocarditis, and antiphospholipid syndrome. Treatment was based on a combination of antibiotics, corticosteroids, and synthetic antimalarials. This case highlights the importance of considering systemic lupus erythematosus in the differential diagnosis of culture negative endocarditis with nephrotic syndrome.

Keywords: *Libman-Sacks endocarditis, nephrotic syndrome, systemic lupus erythematosus, antiphospholipid syndrome, case report*

INTRODUCTION

Libman-Sacks endocarditis is a non-infectious, thrombotic endocarditis first described by Libman and Sacks in 1924. It is characterized by sterile, verrucous valvular lesions that predominantly affect the mitral valve, followed by the aortic valve, while involvement of the tricuspid and pulmonary valves is rare ^{1, 2}. Libman-

Sacks endocarditis is a well-recognized cardiac manifestation of systemic lupus erythematosus and is frequently associated with antiphospholipid syndrome³. The vegetations consist of immune complexes, mononuclear cells, fibrin, and platelet thrombi deposited on damaged endothelium⁴.

Although cardiac involvement is common during the course of systemic lupus erythematosus, its presentation

as the initial manifestation of the disease is rare. When it occurs, it is often clinically silent and may only be discovered incidentally on echocardiography⁵. However, Libman-Sacks endocarditis can lead to serious complications including valvular insufficiency, systemic thromboembolic events such as stroke, and secondary infective endocarditis^{1, 5}. The association of Libman-Sacks endocarditis with nephrotic syndrome as a first presentation of systemic lupus erythematosus is particularly exceptional, especially in male patients. We report an original case illustrating this complex clinical presentation and review the recent literature on diagnosis and management.

Case Presentation

A 29-year-old male patient, with no significant past medical history, presented to our nephrology department with a progressive and profound nephrotic syndrome. Clinical examination revealed significant bilateral lower limb edema extending to the legs, associated with exertional dyspnea. The patient also reported a history of inflammatory arthritis affecting the ankles, wrists, and knees, accompanied by constitutional symptoms

including asthenia and weight loss, all occurring in a context of prolonged fever. Additionally, the patient presented with a chronic inflammatory left subpalpebral cellulitis. On physical examination, the patient was in average general condition but asthenic. Cardiac auscultation revealed a significant systolic murmur at the aorto-mitral focus, suggestive of valvular involvement. No other remarkable findings were noted on systemic examination. Laboratory investigations confirmed a deep nephrotic syndrome with massive proteinuria exceeding 3 grams per 24 hours, hypoalbuminemia, and hypercholesterolemia. Microscopic hematuria was also present. Biological workup revealed a marked inflammatory syndrome with C-reactive protein at 80 milligrams per liter and a significantly elevated erythrocyte sedimentation rate. Complete blood count showed inflammatory anemia and lymphopenia.

The baseline laboratory and immunological findings are summarized in Table 1.

Table 1. Timeline of clinical events

Time	Clinical Event
Day 1	Admission with generalized edema and nephrotic syndrome
Day 2	Laboratory evaluation confirmed nephrotic syndrome and inflammatory syndrome
Day 3	Transthoracic echocardiography revealed mitral and aortic valve vegetations with regurgitation
Day 4	Immunological investigations confirmed systemic lupus erythematosus and antiphospholipid syndrome
Day 5	Renal biopsy confirmed lupus glomerulonephritis
Day 6	High-dose corticosteroids and empirical double antibiotic therapy initiated
Day 21	Significant clinical and biological improvement observed

Transthoracic echocardiography was performed and demonstrated mitral and aortic valve thickening, with the presence of fine vegetations on the aortic side of the posterior sigmoid cusp. The examination also revealed significant mitral and aortic regurgitation. Immunological workup was strongly positive, confirming the diagnosis of antiphospholipid syndrome. Testing revealed positive IgG anti-cardiolipin antibodies, positive IgG anti-beta-2-glycoprotein I antibodies, and positive antinuclear factor antibodies with a speckled appearance. A percutaneous renal biopsy was subsequently performed and was consistent with lupus glomerulonephritis, showing proliferative changes with immune complex deposition.

The patient was started on high-dose corticosteroids combined with double antibiotic therapy, after blood cultures were obtained to rule out infective endocarditis. Within 21 days of initiating corticosteroid therapy, the patient showed significant clinical see (figure 1) and biological see (figure 2) improvement. Notably, there was marked clinical improvement of the inflammatory cellulitis. Biological parameters also improved, with reduction in proteinuria and inflammatory markers. Synthetic antimalarials were added for long-term disease control.

The chronological sequence of clinical presentation, investigations, diagnosis, treatment, and follow-up is summarized in Table 2.

Table 2. Baseline laboratory findings

Parameter	Result
Proteinuria	>3 g/24 hours
Serum albumin	Decreased
Total cholesterol	Increased
Microscopic hematuria	Present
C-reactive protein	80 mg/L
Erythrocyte sedimentation rate	Elevated
Hemoglobin	Inflammatory anemia
Lymphocyte count	Lymphopenia
ANA	Positive (Speckled pattern)
IgG anticardiolipin antibodies	Positive
IgG anti- β 2 glycoprotein I antibodies	Positive
Renal biopsy	Lupus glomerulonephritis



Figure 1: Clinical improvement of inflammatory cellulitis of the left lower eyelid before and after corticosteroid therapy

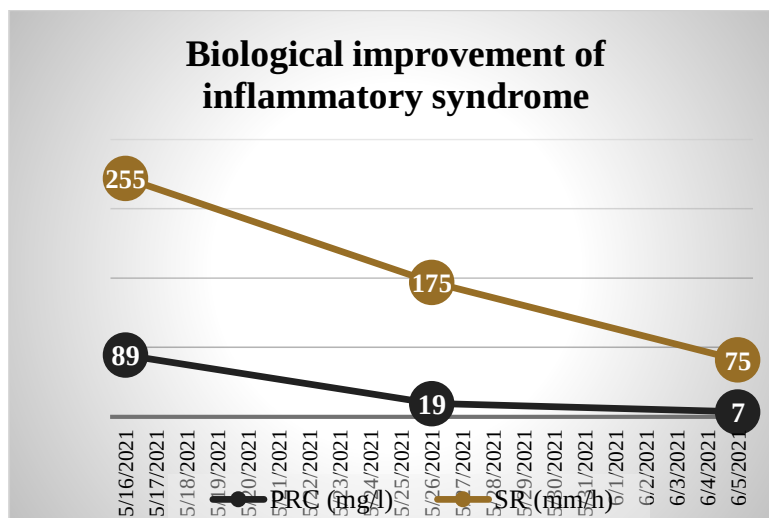


Figure 2 : biological improvement after 21 days of Corticoids therapy

DISCUSSION

This case presents several notable features that make it exceptional in the literature. The first remarkable aspect is the initial presentation of systemic lupus erythematosus. Classically, cardiac involvement in systemic lupus erythematosus, including Libman-Sacks endocarditis, occurs years after the initial diagnosis¹. In our patient, however, Libman-Sacks endocarditis and nephrotic syndrome were the inaugural manifestations of the disease. This presentation is rare and challenging, as the diagnosis may be delayed if systemic lupus erythematosus is not considered. Libman-Sacks endocarditis can present with a mask of infective endocarditis, making the distinction crucial². This case emphasizes the need to consider systemic lupus erythematosus in the differential diagnosis of non-infective endocarditis, especially in young patients with negative blood cultures and unexplained systemic symptoms.

The second notable feature is the male gender of the patient. Systemic lupus erythematosus predominantly affects women, with a female to male ratio of approximately 9 to 1. Severe forms with renal and cardiac involvement in men are less frequently reported, making this observation remarkable. Studies have shown that male patients with systemic lupus erythematosus may have a more aggressive disease course and higher rates of renal involvement⁶. Our case underscores the importance of vigilance regardless of gender.

The third important aspect is the association of systemic lupus erythematosus, antiphospholipid syndrome, and glomerulonephritis. The presence of antiphospholipid syndrome is a major risk factor for thromboembolism and valvulopathy³. Libman-Sacks endocarditis is now regarded as a manifestation of both systemic lupus erythematosus and antiphospholipid syndrome⁷. In our patient, the immunological workup was strongly positive for antiphospholipid antibodies, highlighting the close relationship between these conditions. The renal involvement, presenting as a proliferative glomerulonephritis with nephrotic syndrome, is a serious complication of systemic lupus erythematosus. Lupus nephritis with nephrotic syndrome can be the first manifestation of systemic lupus erythematosus and requires prompt diagnosis and treatment⁸.

The diagnostic challenges in this case deserve discussion. Excluding infective endocarditis is a critical first step in the diagnosis of Libman-Sacks endocarditis. This involves obtaining multiple blood cultures, which will be negative in Libman-Sacks endocarditis, and applying the modified Duke's criteria⁹. Echocardiography is the primary imaging modality for diagnosis. Transesophageal echocardiography is more sensitive than transthoracic echocardiography for detecting Libman-Sacks

vegetations and is recommended when suspicion remains high after initial imaging⁵.

Another important consideration is the risk of secondary infection. Patients with Libman-Sacks endocarditis are at risk of developing secondary infective endocarditis, which can occur after immunosuppressive therapy¹⁰. The combination of Libman-Sacks endocarditis with infective endocarditis has been described and should be suspected when patients with systemic lupus erythematosus develop new cardiac symptoms or embolic phenomena. Ramiandrisoa and colleagues reported a case of superinfected Libman-Sacks endocarditis, emphasizing the need for vigilance¹¹.

The management of Libman-Sacks endocarditis involves several therapeutic strategies. Immunosuppressive therapy with corticosteroids and other immunosuppressants is the cornerstone of treatment, as it reduces inflammation and disease activity. In our patient, high dose corticosteroids led to significant improvement within 21 days. Anticoagulation is an important therapeutic strategy, especially in patients with concurrent antiphospholipid syndrome, because Libman-Sacks lesions are thrombotic³. Warfarin has been shown to be effective, with cases reporting resolution of mitral valve vegetations after six months of therapy¹². Antibiotic therapy is not indicated for Libman-Sacks endocarditis itself given its sterile nature, but antibiotic prophylaxis for secondary infective endocarditis may be recommended during procedures precipitating bacteremia². Empirical antibiotics are justified when infection cannot be immediately ruled out, as in our case. Synthetic antimalarials such as hydroxychloroquine have anti-inflammatory and antithrombotic properties and have been shown to reduce the risk of thromboembolic complications in patients with systemic lupus erythematosus¹³. Valve surgery may be required for hemodynamically significant valvular dysfunction or recurrent embolic events¹⁴, but conservative management with anticoagulation and immunosuppression can be successful in many cases.

The prognosis of Libman-Sacks endocarditis is variable. Some patients respond well to medical therapy with resolution of vegetations¹², while others may have a poor outcome. Hussain and colleagues reported a case of multivalvular Libman-Sacks endocarditis in a 43 year old male who ultimately died from complications of acute on chronic kidney injury¹⁵. Vigilant monitoring is essential, as other serious conditions can rarely mimic Libman-Sacks endocarditis.

Study Limitations

This report describes a single clinical case, which limits the generalizability of the findings. Long-term echocardiographic follow-up after treatment was not

available. Nevertheless, the rarity of Libman-Sacks endocarditis presenting simultaneously with nephrotic syndrome as the first manifestation of systemic lupus erythematosus in a young male patient makes this observation clinically valuable.

CONCLUSION

Our observation is quite exceptional because cardiac involvement usually occurs years after the onset of systemic lupus erythematosus, but in this case the onset was aggressive, first in a male subject, associating renal involvement, endocarditis, and antiphospholipid syndrome. Libman-Sacks endocarditis should be considered in any patient presenting with culture negative endocarditis, particularly in the context of systemic symptoms and renal involvement. Early diagnosis and multidisciplinary management involving nephrologists, cardiologists, and internists are essential to improve prognosis. Treatment was based on a combination of antibiotics, corticosteroids, and synthetic antimalarials, which led to significant improvement. Further studies are needed to establish standardized treatment guidelines for this challenging condition.

Ethics approval and consent to participate

Ethical approval was waived by the institutional ethics committee because this study reports a single anonymized clinical case. Written informed consent was obtained from the patient prior to publication.

Consent for publication

Written informed consent for publication of this case report and any accompanying clinical images was obtained from the patient.

Availability of data and materials

All data generated or analyzed during this study are included in this published article. Additional information is available from the corresponding author upon reasonable request.

Competing interests

The authors declare that they have no competing interests.

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Authors' Contributions

GK: Conceptualization, patient management, data collection, literature review, manuscript drafting, and final approval.

AB: Clinical supervision, interpretation of findings, critical revision of the manuscript, and final approval.

LD: Data collection, literature review, manuscript editing, and final approval.

All authors read and approved the final manuscript.

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Abbreviation	Full Form
ANA	Antinuclear Antibody
APS	Antiphospholipid Syndrome
CRP	C-reactive Protein
ESR	Erythrocyte Sedimentation Rate
IgG	Immunoglobulin G
SLE	Systemic Lupus Erythematosus
TTE	Transthoracic Echocardiography
LSE	Libman-Sacks Endocarditis

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