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# Heart Valve Involvement (Mitral Valve Regurgitation) Caused by Libman–Sacks Endocarditis in Systemic Lupus Erythematosus : A Case Report and Review of the Literature

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**ABSTRACT**

Libman–Sacks (LS) endocarditis is one of the most common cardiac manifestations of both systemic lupus erythematosus (SLE) and the antiphospholipid syndrome (APS).
We report a case of mitral valve replacement for mitral regurgitation (MR) caused by Libman-Sacks endocarditis in young patient with systemic lupus erythematosus (SLE).
In conclusion, Although typically mild and asymptomatic, the disease can lead to serious complications, including superimposed bacterial endocarditis, thromboembolic events, and severe valvular regurgitation and/or stenosis requiring surgery.

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**KEYWORDS:** Libman–Sacks endocarditis, mitral regurgitation, vegetation, lupus carditis.

## INTRODUCTION

Valvular disease is one of the most prevalent and important forms of cardiac involvement in patients with systemic lupus erythematosus, representing a relatively common cause of morbidity (1).

In 1924, Libman and Sacks first described four cases of non-bacterial verrucous vegetative endocarditis (2), and that was the first introduction of Libman-Sacks endocarditis (LSE). The sterile verrucous lesions of Libman-Sacks (LS) endocarditis shows a clear predisposition for the mitral and aortic valves, tricuspid lesions were very rare, chordae tendinae, and endocardium surface can also be affected . These lesions are nowadays seen as both a cardiac manifestation of autoimmune diseases such as SLE and antiphospholipid syndrome (APS) (3) (4). SLE is an autoimmune disorder resulting in multiorgan inflammatory damage.

The pathologic changes of LSE involve the formation of fibrin-platelet thrombi on the altered valve, the organization of which leads to valve fibrosis, edema, diffuse thickening, mild inflammatory changes, valve distortion, scarring, and subsequent valvular dysfunction such like stenosis or regurgitation (3)(5).

Over the last decades with prolonged survival and improvement in diagnostic techniques, particularly in echocardiography, cardiac disease associated with SLE has become more apparent (6)(7). A recent echocardiographic study in patients with SLE revealed that LS vegetations can be found in approximately 11% of patients with SLE (8). In 63% of these patients with vegetations the mitral valve was involved (8). Earlier echocardiographic studies reported a higher prevalence of LS vegetations in patients with SLE, ranging from 53% to 74% (9)(10).

Antiphospholipid syndrome (APS) has been defined as venous or arterial thrombosis, recurrent fetal loss, or thrombocytopenia accompanied by increased levels of antiphospholipid antibodies (aPLs) (anticardiolipin antibodies and the lupus anticoagulant) (11)(12). This syndrome can be either primary or secondary to an underlying condition (most commonly SLE) (11)(12). An echocardiographic study in patients with primary APS showed that approximately one third of these patients have LS valvular lesions (14). SLE is frequently accompanied by the presence of aPLs, which is associated with a higher prevalence of valvular abnormalities in SLE patients (5)(13).

Although typically mild, asymptomatic and hemodynamically insignificant, LS endocarditis can lead to serious and potentially fatal complications, including superimposed bacterial endocarditis, thromboembolic events including stroke and transient ischaemic attacks, and severe valvular regurgitation and/or stenosis which may require emergency surgical management.

The literature on mitral valve surgery for mitral regurgitation (MR) caused by LS endocarditis is comparatively sparse .

In this study we report a case of mitral regurgitation caused by LS endocarditis in patient with SLE history, and we provide a systematic review of the literature .

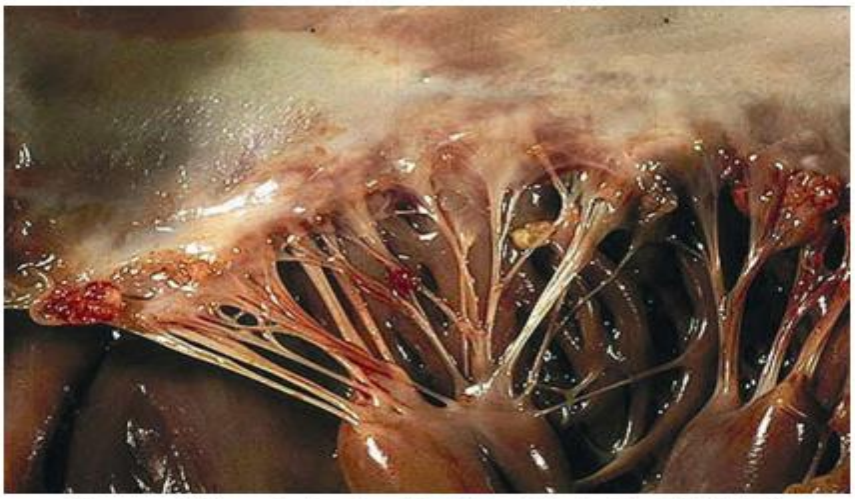


**Figure 1 Verrucous vegetations seen in Libman-Sacks endocarditis of the mitral valve.** The sterile fibrofibrinous vegetations seen in LS endocarditis of the mitral valve may vary in size and typically have a wart-like morphology. They can be found near the edge of the leaflets along the line of closure; both on the atrial and ventricular sides of the leaflets. They can even be found on the chordae and the endocardium. In this case several microthrombi are present on the free edge of the leaflet and on the chordae. *Reproduced with permission from Dr. S. Gonzalez. Copyright 2009, department of Pathology, Pontifical Catholic University of Chile, Santiago, Chile.*

**CASE REPORT**

A 45-year-old woman presented at our institution with SLE that had been diagnosed originally in 1998 and treated with long-term plaquenil and prednisone. The patient was admitted with progressive exertional dyspnoea, and a mitral systolic murmur in a jet of steam at the apex radiating to the left axilla.

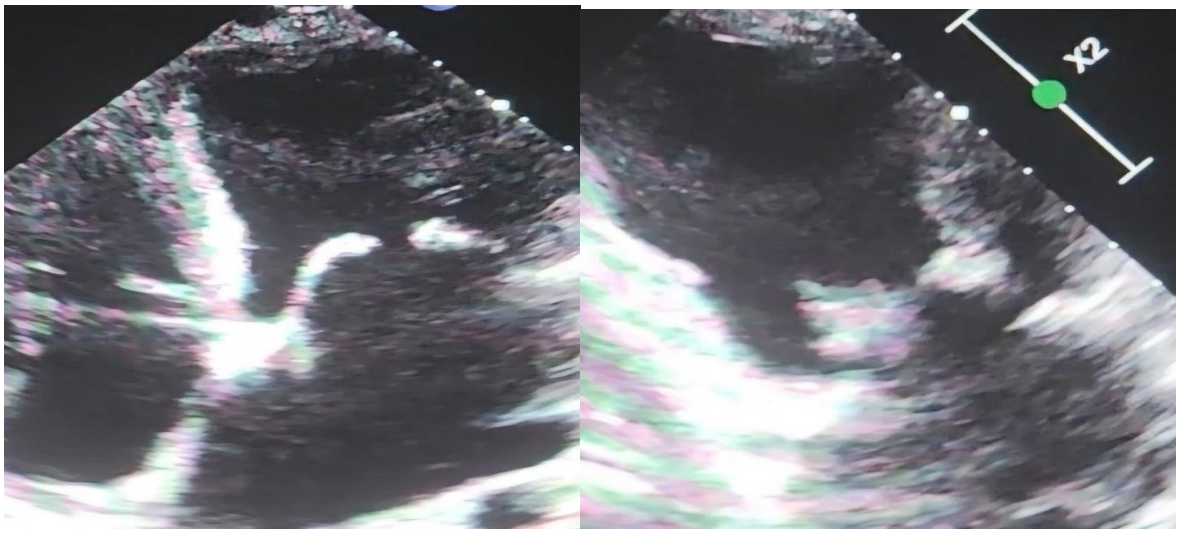
Transthoracic (TTE) and transesophageal echocardiography (TEE) revealed mitral valve leaflet thickening with small vegetations on the edges of both mitral valve leaflets and severe mitral regurgitation with backflow into the pulmonary veins, dilated left atrium and a normal left ventricular (LV) function.

Repeated blood cultures were negative and there was no other evidence of infective endocarditis.

Previous echocardiographic has revealed a rapid increase in left ventricular diameters and mitral regurgitation. The patient was referred to cardiovascular surgery department for further management.

The patient underwent mitral valve replacement with bioprosthesis. The excised mitral valve was thickened, fibrotic with focal vegetations and microscopic pathologic examination has confirmed LS endocarditis of the mitral valve.

The patient's recovery from surgery was uneventful, and she was discharged on the seventh postoperative day.

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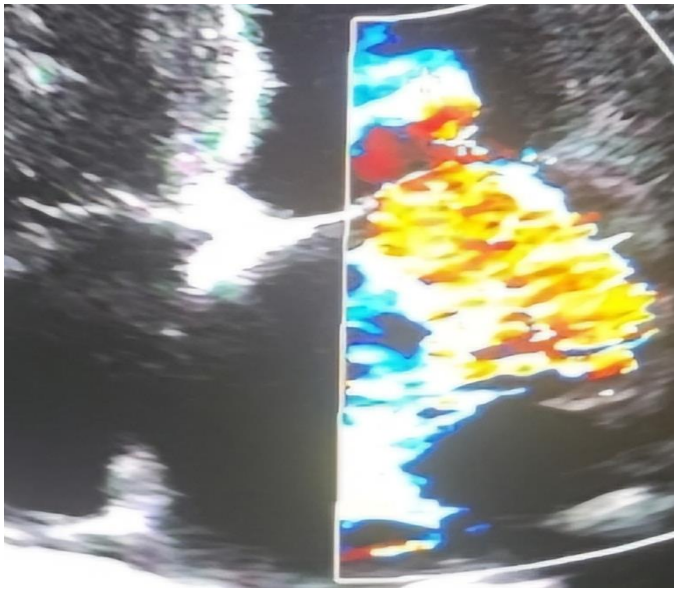

*C*

**Figure 2 : Two-dimensional TTE examination (A) Apical four-chamber view. (B) Apical two-chamber view. Morphologic examination of the mitral valve leaflets in both views revealed leaflet thickening and vegetations on the edges of both leaflets mitral valve. (C) Severe mitral regurgitation with left atrial area (29 cm²)**

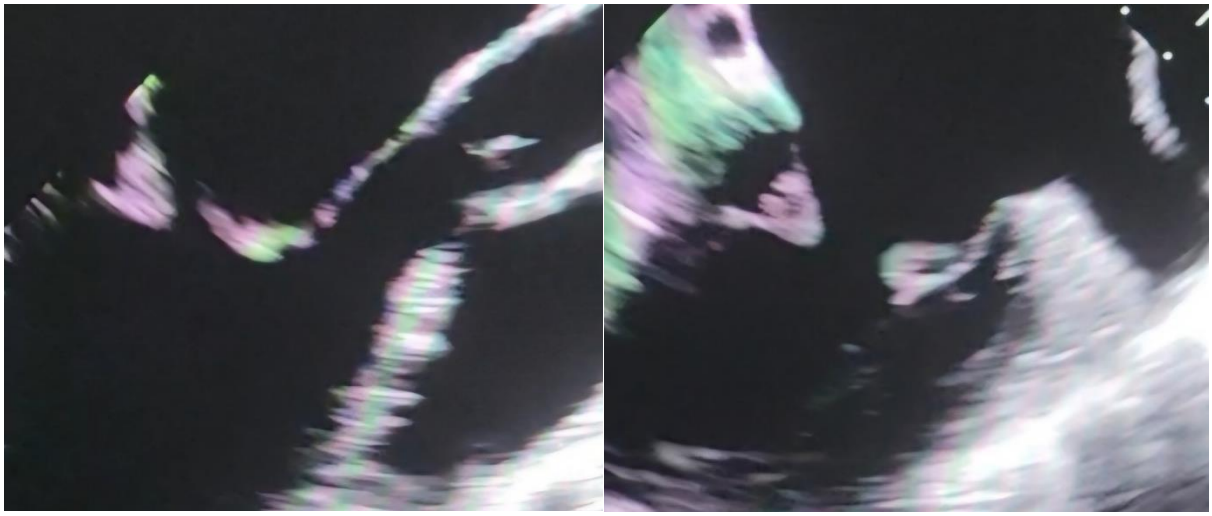

**Figure 3 : Transesophageal echocardiography (TEE) examination**

## DISCUSSION

Systemic lupus erythematosus is a chronic autoimmune disorder characterized by auto antibody-initiated, complement mediated multi-organ tissue injury. Cardiac involvement in lupus is estimated to occur in greater than 50 % of cases and to involve all three layers of the heart : the conduction system, coronary arteries, and valves (15)(16).

SLE is a complex and protean disease, and factors such as the presence of other antibodies and immunologic disturbances, duration of active disease, and immunomodulatory and antithrombotic therapy may all influence the expression of endocardial lesions. Recently, a significantly higher prevalence of valvular involvement was observed in patients with APS secondary to SLE than in primary APS patients.

Libman-Sacks endocarditis was originally described in 1924 in four patients with atypical sterile verrucose lesions of the valvular and mural endocardium. The lesions, pathologically distinct from endocarditis of other etiologies, were believed to be characteristic of SLE. Libman-Sacks vegetations were found in 35% to 65% of lupus patients in early autopsy studies but routinely were clinically silent and of minor hemodynamic importance. Subsequent postmortem series showed smaller incidence and size of vegetations (17).

Valvular heart disease in SLE occurs along a spectrum from leaflet thickening to large vegetation, as in Libman–Sacks endocarditis (18) (19) .LS endocarditis has been reported to be associated with higher disease activity, longer disease duration, and positive anticardiolipin and antiphospholipid antibodies (20).

At this point the exact pathogenesis of LS endocarditis is still unclear. However, LS endocarditis has been assumed to involve the formation of fibrin-platelet thrombi on the altered valve, the organization of which leads to valve fibrosis, edema, diffuse thickening, mild inflammatory changes, valve distortion, scarring, and subsequent valvular dysfunction . Both valve thickening and formation of vegetations represent different stages of the same pathological proces . Rather than playing a more direct pathogenetic role, aPLs are thought to promote thrombus formation on the endothelium of valves already compromised by immune complex deposition, leading to further valvular damage and inflammation.

Microscopy valvular LS lesions are microscopically characterized by fibrin deposits at various stages of fibroblastic organization, neovascularization, occasional haematoxylin bodies, and by a variable extent of inflammation with mononuclear cell infiltration.

Two morphological echocardiographic patterns can be discerned : valve masses (vegetations) and valvular thickening. These two morphological alterations can be combined and both can be associated with valve dysfunction. The predominant functional abnormality is regurgitation, whereas stenosis is rarely seen. The mitral valve is mainly affected, followed by the aortic valve. Involvement of the tricuspid or pulmonary valve was seldom identified. (17)

Moyssakis reported 38 LSE in 342 SLE patient which were diagnosed by echocardiography, among which there were 24 mitral, 13 aortic involvement and only one tricuspid involvement. Doppler echocardiography can be considered as the diagnostic technique of choice. But sometimes it is very difficult to identify LSE and true infectious endocarditis (IE), for the former may also have fever due to the original immunology diseases and the latter may also have vegetation. The role of transesophageal echocardiography (TEE) had been emphasized in assessing vegetation size in a patient with LSE. And in our case the diagnostic role of TEE had been also highlighted, which might be more sensitive to the very special verrucous vegetations (21).

Due to its asymptomatic nature establishing a diagnosis of LS endocarditis can be rather difficult. This is further complicated by the fact that the condition can mimick intracardiac tumors and bacterial endocarditis ("pseudoinfective" endocarditis) or may coexist with (superimposed) bacterial endocarditis (also known as "double-decker" endocarditis).

The modified Duke criteria can be useful in helping differentiate between true infective endocarditis and LS endocarditis. Helpful laboratory markers in distinguishing infective endocarditis from LS endocarditis are the white blood cell count (elevated in infective endocarditis and often decreased in LS endocarditis), C-reactive protein levels (elevated in infective endocarditis and relatively low in LS endocarditis), aPL levels (normal in infective endocarditis and moderate to high in LS endocarditis), and (repeated) blood cultures (positive in infective endocarditis and negative in LS endocarditis). Echocardiographically, LS vegetations appear as valve masses of varying size and shape with irregular borders and echodensity, they are firmly attached to the valve surface and exhibit no independent motion. Contrary to the vegetations of infective endocarditis, which typically exhibit independent motion.

Although LS vegetations are usually typically sessile, wartlike, and small, varying from pinhead size to 3-4 mm, they can become rather large making them difficult to distinguish (echocardiographically) from a typical mitral valve tumor such as papillary fibroelastoma. Recently, a prospective randomized controlled study showed that TEE was superior to TTE in diagnosing LS endocarditis. Nevertheless, establishing the diagnosis remains challenging. The prevalence of Libman– Sacks vegetations is <10% by transthoracic echocardiogram and up to 30% by TEE (22).

Several studies have reported an association between Libman-Sacks vegetations and systemic lupus erythematosus, current studies showed immunoglobulins and complement deposition in the valvular structure that subsequently led to Libman-Sacks vegetations and thickening (23).

The literature regarding the treatment of LS endocarditis is scant. Because the disease is often associated with active SLE, treatment focuses on the management of active lupus using immunosuppressive agents, while steroids are thought to reduce inflammation, they may lead to scarring and valvular dysfunction. It is imperative to note that immunosuppressive therapy often used in this setting may lead to infection and poor wound healing and tissue integrity for those who need surgical valve replacement or repair. In patients with LS endocarditis associated with a positive aPL, antithrombotic therapy is recommended for the prevention of thromboembolic events. Supportive therapy for associated heart failure and arrhythmias may also be necessary.

Evidently, no definite consensus has been reached at this point as to whether or not these valves should be replaced or repaired and whether a mechanical prosthesis is more advantageous than a bioprosthesis (24).

## CONCLUSION

LS endocarditis should be strongly suspected when significant valve dysfunction, such as MR, developed during the course of SLE and/or APS. Differentiation from infective endocarditis and intracardiac tumors can be difficult, but is important and has different therapeutic implications. After establishing the diagnosis, periodic echocardiographic follow-up is recommended to detect detoriation of valvular function. Consequently, an increased

awareness of this entity may lead to careful cardiovascular examination, early diagnosis, and proper intervention.

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