

Libman-Sacks endocarditis in a male patient with systemic lupus erythematosus

Endocardite de Libman-Sacks em Paciente Masculino com Lúpus Eritematoso Sistêmico

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Introduction

Libman-Sacks endocarditis (LSE), also known as marantic or verrucous endocarditis, is a form of non-bacterial thrombotic endocarditis (NBTE) that involves the presence of sterile vegetations in the heart valves. These vegetations may be associated with malignancy, systemic lupus erythematosus (SLE), or antiphospholipid syndrome (APS), and although they are more commonly found in the mitral and aortic valves, other valves may be involved. Diagnosis requires high clinical suspicion when evaluating specific populations.¹

Clinical case

A young 21-year-old illicit drug user diagnosed with SLE was admitted to the emergency unit with continuous non-traumatic epistaxis over the previous six hours. The patient reported

episodes of melena and the onset of macular, erythematous, diffuse lesions on the soles of the feet over the previous two weeks (Figure 1). The patient denied fever or chills associated with the condition. Physical examination identified tachycardia (heart rate, 115 beats per minute) and cardiac auscultation with a regular rhythm, normal heart sounds, and a 3+/6+ systolic murmur in the mitral focus radiating to the axilla. Admission tests suggested active SLE and showed reduced complements C3 and C4; thrombocytopenia (2,000 U/ μ L); anti-double stranded DNA, 1:640; and hemoglobin, 8.8 mg/dL. Exams collected 10 months earlier showed negative lupus anticoagulant and positive anticardiolipin antibodies at low titers. Pulse therapy with methylprednisolone and intravenous human immunoglobulin was indicated and started by the rheumatology team. Transthoracic echocardiography was also requested to assess the cardiac involvement.



Figure 1 – Macular, erythematous, and painless lesions with a two-week progression compatible with an embolic phenomenon. Description similar to Janeway lesions in infective endocarditis.

Keywords

Endocarditis; Echocardiogram; Lupus Erythematosus, Systemic.

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Case Report

Transthoracic echocardiography showed an enlarged left ventricle with eccentric hypertrophy and a preserved ejection fraction. The mitral valve with diffusely thickened leaflets and amorphous isoechoic vegetation with irregular surface adhered to the ventricular face of the posterior leaflet (Video 1) measured 1.0×1.2 cm (Figure 2), restricting its mobility and causing important eccentric reflux (Video 2) directed to the posterior atrial wall. In addition, an enlarged left atrium was noted with a volume indexed by the body surface of 67 mL/m^2 and normal right chambers.

Due to the vegetation identified in the mitral valve, associated with the clinical context of the patient, the following were listed as possible differential diagnoses: 1) NBTE; and 2) infective endocarditis (IE). For better elucidation of the case, transesophageal echocardiography was requested and three blood cultures were collected. Empirical antibiotic therapy was not started due to a lack of evidence of infection.

Transesophageal echocardiography complemented the transthoracic examination by better delimiting the mitral valve lesions. Thickened leaflets were observed (Videos 3 and 4, Figure 3), with an echogenic sessile well-delimited image in

the posterior portion of the valve annulus and ventricular face of the posterior leaflet (maximum thickness, 1.56 cm; Figure 4) encompassing all of their segments and part of the subvalvular apparatus, reducing the mobility of the posterior leaflet and causing moderate to severe reflux.

Blood cultures were negative after three days. Three other samples were collected that also showed negative bacterial growth results.

Considering complementary test findings, the patient's characteristics, and the absence of infectious evidence, the diagnosis of NBTE remained. The patient completed pulse therapy with methylprednisolone, underwent a course of intravenous immunoglobulin, and started therapy with cyclophosphamide.

Discussion

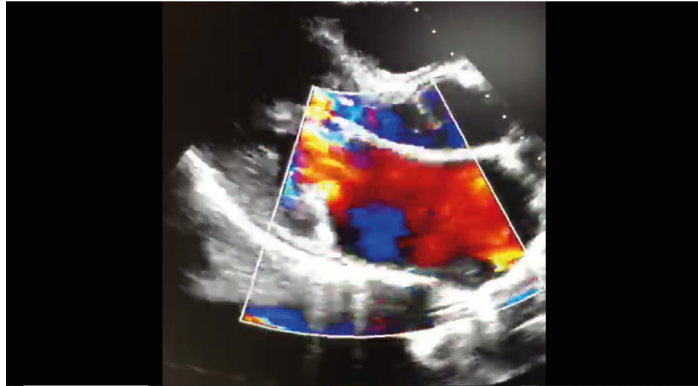
LSE is a rare condition, mostly found postmortem, with a prevalence of 0.9–1.6%. It commonly affects patients aged 40–80 years, but it can occur in any age group. NBTE is often associated with hypercoagulable states such as malignant



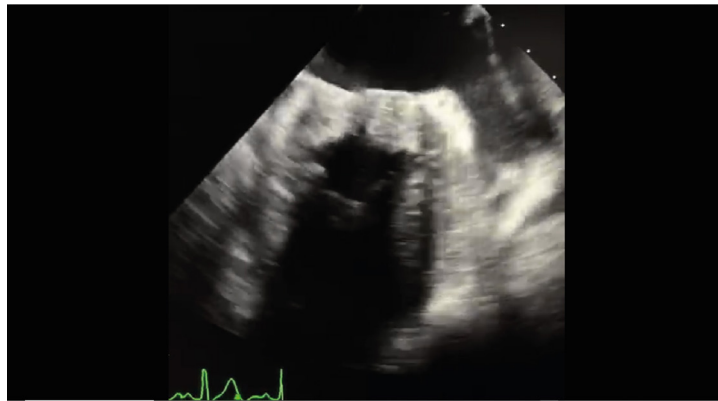
Video 1 – Long-axis transthoracic echocardiography of the left parasternal window showing an amorphous image of the irregular surface on the ventricular surface of the posterior mitral valve leaflet without independent mobile echoes.



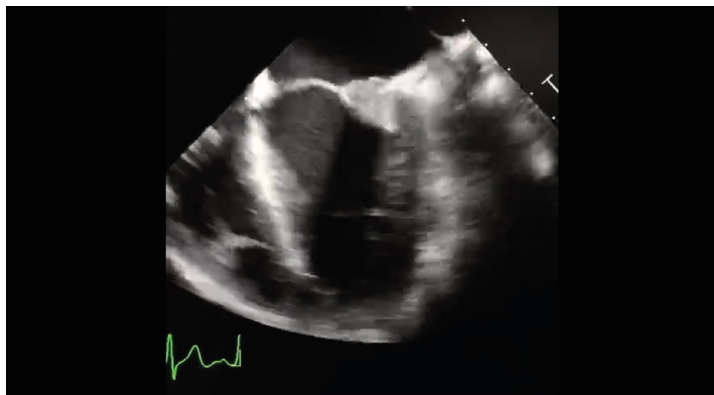
Figure 2 – A: Apical window, three chambers showing amorphous image measuring 1.29×0.92 cm on the posterior mitral valve leaflet. B: Long-axis parasternal window showing important mitral regurgitation occupying almost the entire left atrium.



Video 2 – Long-axis transthoracic echocardiography of the left parasternal window showing significant mitral valve reflux directed to the posterior wall of the left atrium.



Video 3 – Two-chamber transesophageal echocardiography (approximately 90°) demonstrating posterior mitral valve leaflet thickening.



Video 4 – Four-chamber transesophageal echocardiography (approximately 0°) showing a homogeneous echogenic, sessile, well-delimited image on the ventricular face of the posterior leaflet.

Case Report

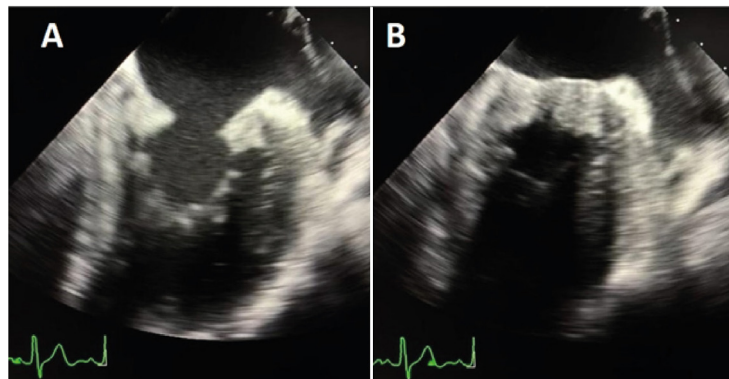


Figure 3 – Two-chamber transesophageal echocardiography at 90° demonstrating diffuse thickening of mitral valve leaflets in diastole (A) and systole (B).

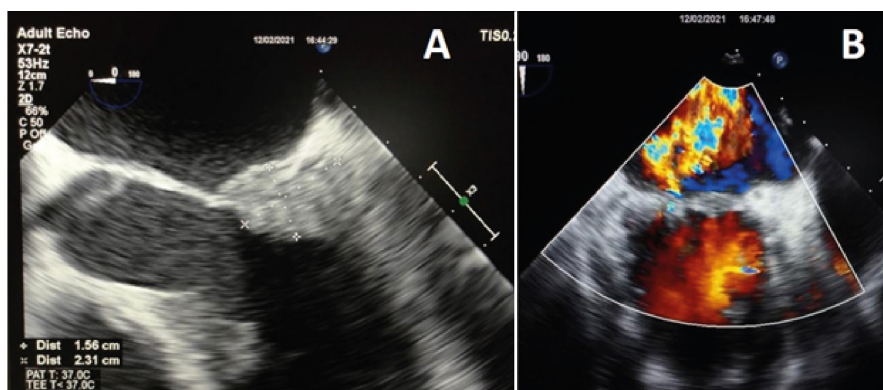


Figure 4 – Transesophageal echocardiography A: four-chamber view at 0° showing a sessile image in the posterior mitral valve leaflet measuring 1.56 × 2.31 cm. B: two-chamber view at 90° demonstrating moderate to severe reflux toward the posterior wall of the left atrium.

neoplasms, SLE, and APS. It is correlated with disease activity and duration and its prevalence increases in the overlap of these pathologies.¹ Studies do not show a sex predilection; however, SLE and APS, causes associated with LSE, occur five- to nine-fold more often in women of childbearing age than in men and are more prevalent in Black and Hispanic women.^{2,3}

Patients are usually asymptomatic; findings are often incidental in the investigations of other cardiac conditions by imaging or on postmortem examination. The most common clinical presentations are secondary to valve embolism, such as stroke, transient ischemic attacks, or symptoms of peripheral embolization in the lower limbs. They also include symptoms related to the activity of underlying diseases, such as fever, malar rash, renal, hematological, or immunological changes, among others.¹

High clinical suspicion is required for the diagnosis of NBTE. No laboratory tests can confirm this diagnosis. Patients with suspected NBTE should be screened for blood cell count, metabolic profile, and blood cultures for the differentiation from other etiologies such as IE. Hypercoagulability should be investigated, including lupus anticoagulant and

antiphospholipid antibodies. Three blood culture samples should be tested before starting any empirical antibiotic therapy. The absence of typical or atypical endocarditis-causing agents may strengthen the NBTE suspicion.¹

However, the primary screening test for LSE is echocardiography. Transesophageal echocardiography has greater sensitivity and specificity than transthoracic echocardiography. Irregular borders, heterogeneous echogenicity, and the absence of independent mobile echoes are characteristic of the masses (verrucous vegetation) found in the heart valves and endocardium. These are usually small sessile masses, but they can be larger than 10 mm. The basal and middle portion of the mitral and aortic valves are the most affected. Diffuse or focal thickening of the mitral and aortic valves may be seen. The involved valves may present regurgitation.¹ Vegetations are more frequent on the left side, with two thirds of cases involving the mitral valve, mainly on the ventricular face, and up to a quarter involving the aortic valve and less commonly both valves. Echocardiography cannot distinguish vegetations from thrombi but can provide diagnostic clues for lesion characterization.¹

NBTE treatment is based on expert positioning and small observational studies. Underlying disease control, anticoagulation in patients with major embolic phenomena, and serial echocardiograms are recommended for monitoring valve lesions. Surgical indications are the same as for IE (i.e., heart failure, acute valve rupture), but reports suggest that recurrent embolization prevention is the most common indication for surgery. In contrast to IE, which requires complete removal of the infected tissue, valve preservation may be possible in some NBTE cases.¹

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Authors' contributions

Research conception and design: Vedana Filho AA, Alves MSL; Data collection: Vedana Filho AA, Mattar TA; Manuscript writing: Vedana Filho AA, Bittencourt VR; Critical review of the manuscript: Alves MSL, Botelho FS.

Conflict of interest

The authors have declared that they have no conflict of interest.