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Case Report

Severe mitral valve regurgitation secondary to Libman-Sacks endocarditis

Michelle Mizrahi[1*], Silvia Jimenez-Becerra[2*], Eric Rivera-Navarrete[3*], Mario Gomez-Sanchez[4*]

[1]General Physician, [2]Attending Pathologist, [3]Cardiac Surgery Resident, [4]Attending Cardiovascular Surgeon, National Institute of Cardiology "Ignacio Chavez", Juan Badiano 1, 5to piso, Col. Seccion XVI, Tlalpan, CP 14080 Mexico City, Mexico. [*]Equal contributors.

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Abstract: Although uncommon, Libman-Sack endocarditis is the most characteristic cardiac manifestation of Systemic Lupus Erythematosus (SLE). It forms vegetations made of inflammatory tissue on the cardiac valves, leading them to malfunction. Here we present a case of a young woman who presented with severe mitral valve regurgitation in need for a valve replacement. Integral evaluation of the patient revealed the diagnosis of SLE, which was aggressively treated in an outpatient setting with immunosuppressive therapy. Only after achieving medical stabilization of the underlying disease, she was able to undergo surgical mitral valve replacement. After the surgery, the patient no longer suffered from mitral regurgitation, and with a mechanical prosthesis in place, the risk of Libman-Sacks endocarditis recurrence is thought to be minimal.

Keywords: Systemic lupus erythematosus, Libman-Sacks, nonbacterial endocarditis, cardiac surgery

Introduction

Libman-Sacks (LS) endocarditis, described since 1924, is the most characteristic cardiac manifestation in SLE, although it is not the most common [1-4].

Its prevalence has diminished since the introduction of corticosteroid agents in the treatment of SLE [4]. Currently, the reported prevalence of these lesions in patients suffering from SLE is one in ten [2], nevertheless, its clinical impact is much less common (20%), and for this reason, valvular surgery is necessary only in few occasions [3, 5].

This form of nonbacterial thrombotic endocarditis forms sterile vegetations on the cardiac valves, which have great thrombogenic potential [1, 2].

The treatment of the SLE endocarditis or LS endocarditis should be focused on the management of the subjacent disease. In patients suffering from severe valvular disfunction, the surgical guidelines for cardiac valvular disease should be followed [2, 5]. If a valvular replacement is necessary due to LS endocarditis, the implantation of a mechanical valve is usually recommended to avoid recurrence of the disease on biological tissue (autologous or prosthetic) [3, 6].

Case report

A 36-year-old female with past medical history of familial hypercholesterolemia, peripartum myocardiopathy, and Antiphospholipid syndrome (APS), presented at our institution with heart failure.

Upon arrival, the patient was dyspneic, she referred significant deterioration from her normal functional status, and a recent episode of generalized seizures. On physical evaluation, a grade II/IV holosystolic murmur with mesosystolic reinforcement, suggestive of mitral regurgitation, was found. The patient was admitted for a further evaluation.

An echocardiographic evaluation confirmed the suspected diagnosis of severe mitral regurgitation, with a clear indication for surgical intervention. However, interstitial pneumonitis and high

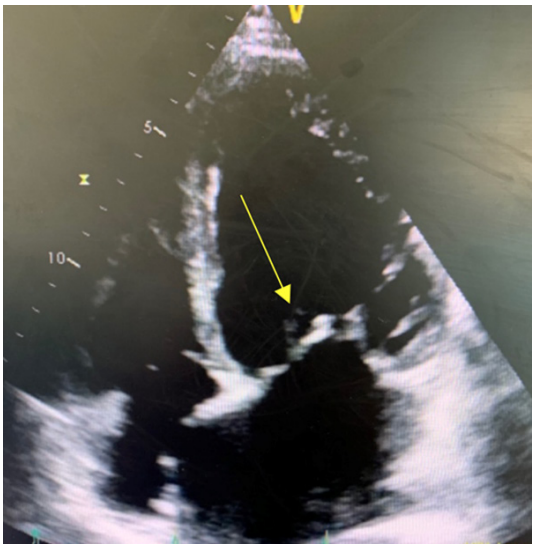

Figure 1. Apical 4-chamber view TTE. Yellow arrow showing a thickened and irregular mitral valve leaflets. (TTE = transthoracic echocardiography).

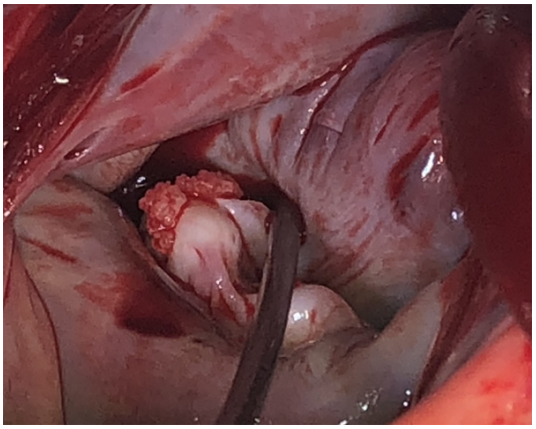

Figure 2. Multiple granular vegetations on the auricular side of the mitral valve.

titers of anti-phospholipid antibodies were also found; leading to a more comprehensive evaluation. The patient was diagnosed with SLE with neurological, pulmonary and probably cardiac activity; prompting the decision to first stabilize the autoimmune process, and only then consider surgery. So, the patient was discharged home with immunosuppressive treatment.

A month after starting medical management, no SLE activity was found, and the patient was re-admitted for re-evaluation by the surgical team. A transthoracic echocardiography (TTE) at that moment showed thickening of the mitral valve (**Figure 1**) with adequate opening and a deficit in leaflet coaptation, conditioning severe

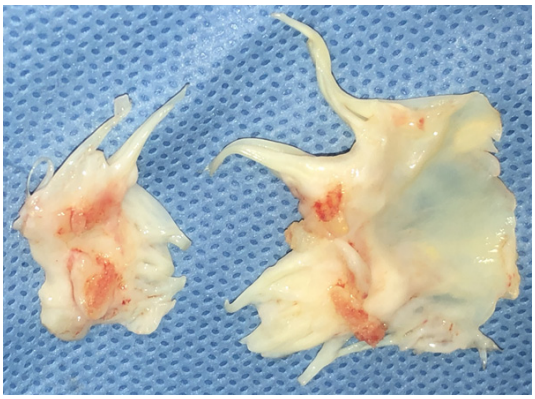

Figure 3. Macroscopic appearance of the posterior (left) and anterior (right) leaflets of the mitral valve. Notice the presence of fibrinous-hemorrhagic vegetations on the edges, closing line and circling the Chordae Tendineae, which are thickened and merged.

mitral regurgitation with dense holosystolic flow, Coanda effect and a regurgitation fraction of 69%; there was no evidence of involvement of the sub-valvular apparatus, nor affection of the rest of the cardiac valves. The left ventricle (LV) was dilated with systolic and diastolic diameters of 47 mm and 63 mm respectively, an indexed end-diastolic volume of 115 mL/m^2, and a biplane Simpson ejection fraction (EF) of 55%. The right ventricle (RV) was also dilated with the following measures: diameter at the base of 41 mm, the mid-right-ventricular diameter of 28 mm and length of 84 mm. The left atrium showed severe dilation with an indexed volume of 138 mL/m^2, while the right atrium size remained unaffected with an indexed volume of 22 mL/m^2. The estimated pulmonary artery systolic pressure (PASP) was 51 mmHg. A coronary artery angiotomography was also performed reveling a Calcium Score of 0; and an MRI scan also documented considerable dilation of the left side heart cavities, LV eccentric hypertrophy, thickening of the mitral valve borders with severe dysfunction of the valve, and bilateral ventricular dysfunction (LVEF 52% and RVEF 46%).

A consensus was made with the patient for surgical intervention with cardiopulmonary bypass support through total median sternotomy. Direct visualization of the heart showed severe cardiomegaly with a particularly grown left atrium. Following atriotomy and mitral valve exposure, a dilated mitral annulus and a fibrous looking mitral valve were found. The anterior leaflet of the mitral valve was thickened in its borders, the posterior leaflet was retracted, and both leaflets had several granular and

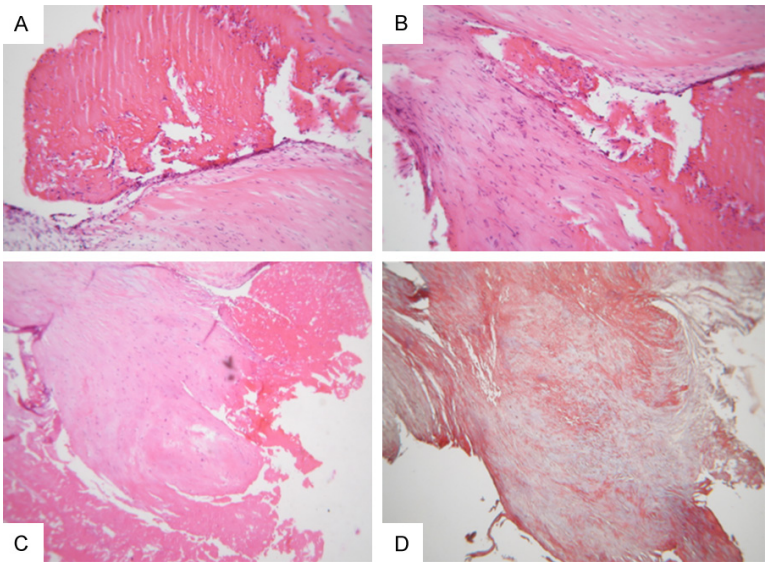

Figure 4. Valvular histopathology. A. HE 10×. Fibrinous-thrombotic and aseptic vegetations adhered to the atrial surface of the mitral valve. B. HE 10×. Fibrinous-thrombotic and aseptic vegetations adhered to the ventricular surface of the mitral valve. C. HE 10×. Fibrinous-thrombotic and aseptic vegetations covering the mitral subvalvular apparatus. D. Trichome stain (Masson) 10×. Distortion of the trilaminar valve architecture due to fibrosclerotic changes. HE: hematoxylin and eosin stain.

medium-sized vegetations adhered to their surface (**Figure 2**). Although significant inflammation of the sub-valvular apparatus was found, we were able to preserve it after removing both valvular leaflets (**Figure 3**), and a mechanical prosthesis (St Jude Masters 31 mm) was implanted. Transoperative transeshopagic echocardiography (TEE) discarded periprosthetic leaks and confirmed proper functioning of the mechanical prosthesis.

The vegetations found on both sides of the mitral valve and the involvement of the subvalvular apparatus suggested the diagnosis of LS endocarditis, which was later confirmed by the histopathological analysis of the valve.

As mentioned above, the histopathological analysis of the native valve (**Figure 4**) showed aseptic fibrinous-thrombotic vegetations, compatible with Libman-Sacks endocarditis [2, 4, 5].

The patient's hospitalization course remained uneventful until her discharge home 7 days after her surgery. One month later, the patient indicated significant improvement from her previous status and return to her regular activities. 10 months after the surgery, the patient remains asymptomatic.

Discussion

The origin of cardiac valvular lesions in the setting of SLE, is highly related to the presence of anti-phospholipid antibodies [7, 8], the duration of SLE, and its activity [2, 3]. The onset of the lesions has been linked to endothelial damage in the setting of a hypercoagulability state, which promotes the deposition of platelet thrombi and inflammatory molecules on the cardiac valves [2].

LS endocarditis is a form of NBTE caused, in the majority of the cases, by advanced malignancy (80%), followed in frequency by SLE and other inflammatory conditions [3].

Although mitral and aortic affection is the most common presentation, it presents as granular deposits on the surface of any cardiac valve. The valvular damage consists of diffuse thickening of the leaflets leading to valvular dysfunction, especially in the form of insufficiency [2, 4], as the case presented above.

No laboratory test that orients nor confirms the diagnosis of LS endocarditis is available. Thus, echocardiography is the method of choice for its initial evaluation [2, 3]. The reported incidence of these lesions by TTE is 6-18%, reaching 43-74% with TEE [3, 8]. Nevertheless, only 20% of these lesions are clinically significant. The most characteristic ultrasonographic findings of LS endocarditis are: irregular valvular borders, heterogeneous echogenicity, and non-independently moving mases [2].

In addition to controlling the underlying disease [5], it is recommended that those patients with severe lupus valvulopathy requiring surgical corrective intervention, undergo valvular replacement by a mechanical prosthesis. This is due to the fact that biological prosthesis and homografts are likely to develop lupus valvulitis [3, 6] and potentially require further and early reintervention.

In this case, it was decided upon mitral valvular replacement by a mechanical prosthesis, being

that the underlying disease and the sub-valvular apparatus severe inflammation, conditioned bad prognostic factor for valvular repair.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Michelle Mizrahi, General Physician, National Institute of Cardiology, Juan Badiano 1, 5to piso, Col. Seccion XVI, Tlalpan, CP 14080 Mexico City, Mexico. Tel: +52 5523-007650; E-mail: mich28m@gmail.com

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