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Libman-Sacks Endocarditis with Unusual Large Size Vegetation Involving the Mitral Valve

Amjad Bani Hani, MD,[1] Mahmoud Abu-abeeleh, MD,[1] Murad M. Al Kharabsheh, MD,[1] Lubaba Qabba'ah, MD[2]

[1]The University of Jordan, Jordon University Hospital, Amman, Jordan; [2]The Islamic Hospital, Amman, Jordan

ABSTRACT

Antiphospholipid syndrome (APS) is an autoimmune hypercoagulable disorder characterized by thrombophilia, vascular thrombosis, and recurrent abortions associated with persistent antiphospholipid antibodies. APS may exist in its primary form, or more commonly is found to be associated with variety of rheumatic disorders, such as systemic lupus erythematosus. Cardiac involvement is not an uncommon complication in primary antiphospholipid patients. Libman-Sacks lesions are typically small, sessile, and wart-like, varying in size from 1-4 mm. Here we present an unusual case of a 37 year-old pregnant woman who suffered from heart failure associated with primary antiphospholipid syndrome and Libman-Sacks endocarditis, with large vegetations involving the mitral valve. The patient underwent mitral valve replacement with a mechanical prosthesis.

INTRODUCTION

Libman-Sacks endocarditis (LSE), or verrucous aseptic thrombotic endocarditis, is a commonly encountered disorder in patients with systemic lupus erythematosus (SLE) [Moaref 2010]. It has been reported in approximately 50% of fatal cases of SLE at autopsy. Vegetations mostly involve the mitral and aortic valves, but in some cases vegetations may involve all cardiac valves in addition to other endocardial surfaces. Another condition that Libman-Sacks lesions have been associated with is primary or secondary antiphospholipid syndrome (APS) [Cervera 2004; Long 2008; Roldan 2008].

Valvular involvement is commonly seen in primary APS. This may be in the form of global thickening, localized thickening involving the proximal or middle portion of the leaflet, or irregular nodules and/or vegetations of the edge of the valve, known as Libman-Sacks endocarditis. As a result, severe valvular dysfunction (regurgitation, stenosis) occurs [Cervera 2004; Long 2008; Roldan 2008]. Libman-Sacks lesions are typically small, sessile, and wart-like, varying in size from 1-4 mm [Moaref 2010]. There are scarce data in the literature describing large Libman-Sacks vegetations.

Clinically, lesions are usually silent unless significant valvular dysfunction occurs, which may result in cardiac failure [Nesher 1997]. Other possible clinical consequences of Libman-Sacks lesions are systemic emboli and septic endocarditis that can potentially result in a variety of clinical manifestations such as neurologic signs [Roldan 2013]. Transesophageal echocardiography is often required for diagnosis [Roldan 2008]. Treatment of Libman-Sacks endocarditis usually aims at treating secondary complications, such as mitral regurgitation or stenosis, septic emboli, and neurologic sequelae [Bouma 2010].

CASE REPORT

A 37 year-old patient who was 36 weeks pregnant presented with a one month history of hemoptysis, shortness of breath (NYHA IV), orthopnea, and paroxysmal nocturnal dyspnea. Transient loss of vision in her left eye was also reported a few days prior to presentation. Her past medical history was significant for multiple abortions. On examination, the patient was found to have pale mucus membranes, increased jugular venous pressure (JVP), basal crackles, gallop rhythm and apical pan-systolic murmur that radiated to the axillary region (Grade 2). Computed tomography scanning showed no evidence of brain infarction. Results of immunologic, autoantibody, and hypercoagulability profiles were consistent with primary antiphos-pholipids (Lupus Anticoagulant: PTT-LA > 99.3 sec, (range ≤ 40 sec) in the absence

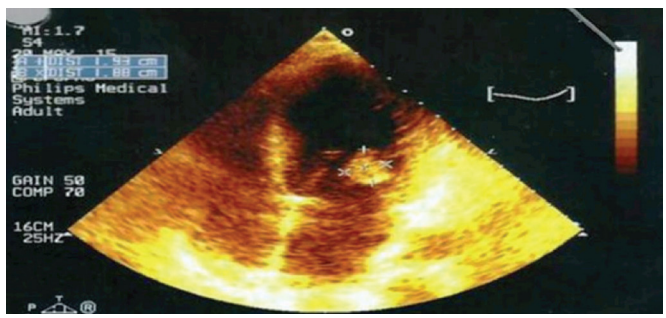

Figure 1. Transthoracic echocardiography showing a 1.9 x 2cm mass located on the posterior mitral valve leaflet.

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Correspondence: Amjad Bani Hani, MD, Assistant Professor of Cardiac Surgery and Intensive Care, The University of Jordan, Jordon University Hospital, P.O. Box 13046, Amman 11942, Jordan; 962-6-5353666; (e-mail: amjadbh@yahoo.com).

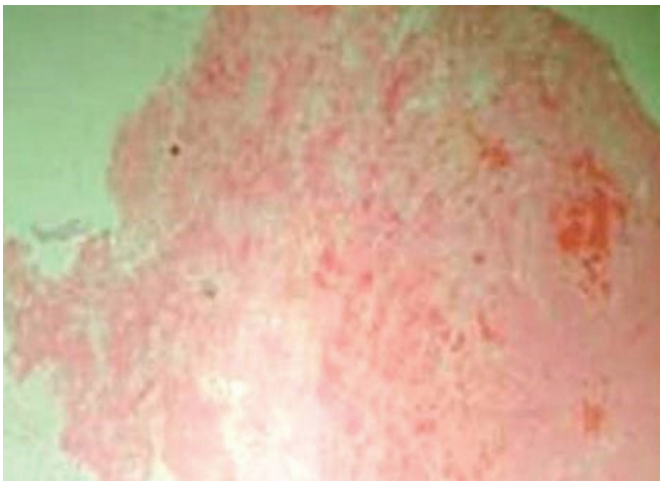

Figure 2. H&E stained section of the mitral valve showing an organized thrombus and granulation tissue consistent with valve vegetation and mild edema (4x).

of SLE markers). Chest radiograph showed bilateral pulmonary edema. Transthoracic echocardiography revealed a 1.9 x 2 cm mass on the posterior mitral leaflet, causing severe mitral regurgitation and ejection fraction of 65% with normal left ventricular size. (Figure 1).

The patient underwent an uneventful planned cesarean section and delivered a normal baby. However, she was still complaining of dyspnea (NYHA III) and her echocardiography showed the same findings. Subsequently a decision was made to intervene and the patient underwent conventional mitral valve replacement (ATS mechanical valve, size 27).

The vegetation, P2, part of P1, part of P3 of the posterior mitral leaflet (PML), and the anterior mitral leaflet (AML) were excised. Part of the subvalvular apparatus with part of P1 and P3 were preserved. The patient made an uneventful recovery and was discharged home. Histological evaluation of the removed valve showed organized nodules of fibrin thrombus and granulation tissue consistent with Libman-Sacks vegetation in addition to mild edema in the tissues of the valve beneath the vegetation. (Figure 2).

Cultures of blood and the surgically removed valve lesion revealed no bacterial growth. Postoperative transesophageal echocardiography showed that the mitral valve prosthesis was well sealed. The postoperative course was uneventful and the patient was discharged home.

DISCUSSION

Almost one third of patients with primary APS suffer from various heart valvular diseases (HVDs), which include global thickening, localized thickening involving the proximal or middle portion of the leaflet, or irregular nodules and/or vegetations of the edge of the valve, known as Libman-Sacks, which are usually small and rarely present as large size lesions. [Cervera 2004; Long 2008; Roldan 2008].

Although typically mild and asymptomatic, Libman-Sacks endocarditis can lead to serious complications, including embolic events, severe valvular dysfunction requiring surgery, and superimposed bacterial endocarditis [Bouma 2010].

Libman-Sacks endocarditis usually involves the left heart valves, mainly the mitral valve, whereas tricuspid valve involvement is rare. [Zhixuan 2015]. The majority of patients are diagnosed incidentally, with typical slow progression usually causing mild regurgitation.

However, a subset of patients develops significant valvular insufficiency, or to a lesser extent, stenosis, which can lead to symptomatic heart failure and require surgical intervention [Lee 2009]. Rarely, Libman-Sacks lesions have been known to affect prosthetic valves enough to interfere with function [Hoffer 2000].

The risk of embolic phenomena is increased in Libman-Sacks endocarditis [Lee 2009; Bouma 2010]. A study by Roldan et al. found that SLE patients with cerebrovascular disease were 2-3 times more likely to be found with positive lupus anticoagulant antibodies and valvular disease than in those without. As a result, Libman-Sacks endocarditis is considered an independ-ent risk factor for cerebrovascular disease [Roldan 2013]. Coronary artery embolization has been reported in a patient with aortic valve lesion in the setting of normal coronary arteries [Pritzker 1980; Tanyanan 2011].

Although the valvular lesions of Libman-Sacks endocarditis themselves are not bacterial in origin, structural abnormalities can predispose individuals to developing overlying infective endocarditis, even in those with healed lesions. Antibiotic prophylaxis may be a considered in those patients. [Lee 2009].

There are scarce data describing large valvular Libman-Sacks vegetations. Libman-Sacks endocarditis is typically asymptomatic unless the lesions progress to more severe valvular dysfunction or result in systemic embolic events [Roldan 2013]. In this patient, the valvular lesion was unusually large (1.9 x 2 cm in diameter). The large valvular lesion in this case resulted in mitral regurgitation and consequently led to heart failure and severe pulmonary edema. In addition, emboli originating from the valvular lesions are believed to be the cause of vision loss.

Libman-Sacks lesions are typically small, sessile, and wart-like, varying in size from 1 - 4 mm [Moaref 2010]. The size of the lesion in this case was relatively large (1.9 x 2 cm) which required surgical removal and valve replacement with a mechanical valve (ATS size 27). We did not attempt to repair the mitral valve due to the large size of the vegetation involving a large area of the PML, and fear of recurrence. Medical treatment usually includes the administra-tion of systemic anticoagulants to prevent embolization, corticosteroid therapy for autoimmune diseases, such as lupus erythematosus, in addition to surgical removal of the lesions and valve replacement using prosthetic implants [Bouma 2010].

In summary, this report describes an unusual case of large size vegetations due to Libman-Sacks endocarditis and primary antiphospholipid syndrome in a pregnant woman who presented with heart failure and transient vision loss. The patient delivered a healthy baby via cesarean section and made an uneventful recovery following successful surgical removal of the lesion. The mitral valve was successfully replaced using a prosthetic implant.

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