Open Access

Annals of Surgical Case Reports & Images

Volume 1 | Case Report

Mitral valve surgery in libman sacks endocarditis: A case report

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Abstract

Libman Sacks endocarditis is a cardiac manifestation of erythematous systemic lupus which often goes unnoticed but can lead to valvular surgery in certain complications, we report the case of a patient with systemic lupus erythematosus and Libman-Sacks endocarditis that progressed rapidly to severe mitral insufficiency that needed surgery.

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Received: Jun 27, 2024

Accepted: Jul 26, 2024

Published Online: Aug 02, 2024

Journal: Annals of Surgical Case Reports & Images

Online edition: https://annscri.org

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Cite this article: Rhissassi J, Briki J, Idrissa M, Maaroufi I, Bouhdadi H, et al. Mitral valve surgery in libman sacks endocarditis: A case report. Ann Surg Case Rep Images. 2024; 1(4): 1040.

Keywords: Endocarditis; Lupus; Mitral valve insufficiency.

Introduction

First described in 1924 by Libman Sacks [1], and characterized by verrucous lesions of the heart valves and the presence of small aseptic vegetations, Libman Sacks Endocarditis (LSE) is a characteristic cardiac manifestation of Systemic Lupus Erythematosus (SLE) [2]. 61% of patients with SLE have valvular involvement [3].

In their series of 69 lupus patients, Roldan et al. found a 43% prevalence of Libman-Sacks endocarditis with systematic Trans Esophageal Echocardiography (TEE) [4]. In contrast, other authors have found a prevalence of no more than 20% in lupus patients [5], a difference that may be explained by the fact that TEE is not routinely performed.

We report the case of a 52-year-old patient with SLE complicated by lupus nephritis who underwent mitral valve surgery.

Clinical case

A 52-year-old patient with SLE, complicated by lupus nephritis on prednisone, was diagnosed with dyspnea stages II and III of NYHA.

The clinical examination was done on a conscious patient, well oriented in time and space, with a general condition corresponding to stage 2 of the WHO Performance Status hemodynamically stable: blood pressure (BP)=110/60 mmHg/ heart rate (HR)=85 bpm/ saturation (SpO₂)=97% on room air.

On auscultation, the cardiovascular examination revealed regular heart sounds as well as a systolic murmur in the mitral foci. Peripheral pulses, which are ample and bouncy, are perceived symmetrically.

Electrocardiogram demonstrated sinus tachycardia without other associated anomalies. Chest x-ray at admission showed mild cardiomegaly and bilateral basilar interstitial lung pattern. In view of this clinical picture, Transthoracic Echocardiography (TTE) was performed, which confirmed the significant mitral insufficiency and large mitral vegetation (Figure 1).

A TOE was used to complete the assessment in front of the MI and mitral vegetation (Figure 2).

Biological blood tests showed C reactive protein at 85, white blood cells 15000, and cytobacteriological examination of urine returned positive for Esherishia Coli. Blood cultures were negatives.



Figure 1: Trans thoracic echocardiography.

TTE:

- Vegetation measuring 18mm, on the atrial side of the anterior leaflet of the mitral valve responsible for a perforation at this level.
- On Doppler: Presence of a severe mitral leak, PISA 18 mm, on an 8-mm perforation at A3 level
- Tele diastolic diameter of the left ventricle = 69mm, tele systolic diameter = 46mm, left ventricle ejection fraction 65%
- Tricuspid insufficiency grade II to III, pulmonary systolic pressure = 82mmHg



Figure 2: Trans esophageal echocardiography.

TOE :

- The anterior leaflet of the mitral valve describes a rounded neocavity measuring 12mm*10mm, with thickened contours, hyper echogenic, and which appears ruptured with color passage
- 2 small elements appended to the neocavity on its ventricular side measuring 8mm, 5mm.
- Presence of 2 apparently significant mitral valve flows, one of which is at the level of the valve coaptation and the other, which appears more significant, is at the level of the ruptured neocavity described by the anterior leaflet of the mitral valve.

On the basis of this diagnosis, the patient was admitted to the operating theatre for surgical management.

Under general anesthesia, a vertical median sternotomy was performed, followed by the installation of a extracorporeal circulation, between aortic cannulation and cannulation of both vena cava, cardiac arrest occurred after administration of crystalloid cardioplegia via the anterograde route. Exposure of the mitral valve through the opening of the right atrium, and the septum through the foramen ovale, revealing large vegetation raising suspicion of a degeriated abscess suspended on the atrial side of the anterior leaflet of the mitral valve between A2, A3, with a large perforation at this level

A careful and enlarged vegectomy was performed, and a mitral plasty was carried out, which was unsuccessful due to the fragility of the valve leaflets. Hence the decision to replace the mitral valve with a CARBOMEDICS n31 mechanical prosthesis after resection of the anterior leaflet, and the conservation of the posterior leaflet and the subvalvular apparatus.

Tricuspid annulopasty was performed with an EDWARDS n°32 ring.

Exit from bypass under positive inotropic drugs, after 87 min of bypass time and 62 min of clamping time.

The patient was extubated at H10, and the postoperative course was straightforward.

Patient anticoagulated with heparin and vitamin K antagonist.



Figure 3: Trans thoracic echocardiography.

Black arrow: mitral valve Blue arrow: mitral valve perforation Red arrow: mitral valve vegetation

Results and follow-up

- Bacteriological examination of the vegetation was negative, while histological examination revealed a totally destroyed valve wall, replaced by eosinophilic fibrinous material showing multiple foci of calcification on an inflammatory background in favor of infectious endocarditis in the course of organization.
- Haematological blood test for Lupus Anticoagulant (LA) was positive.
- A follow-up TTE after 1 month showed a mechanical prosthesis in a well-functioning mitral position, with a mean gradient of 5 mmHg and no leakage, with an ejection fraction of 50%.

Discussion

Libman Sacks endocarditis, a form of Non-Bacterial Thrombotic Endocarditis (NBTE), is a well-known cardiovascular manifestation of SLE, with valvular thickening, vegetation formation and regurgitation being the main pathological features [3].

When medical management fails to control symptoms or prevent complications, surgical intervention becomes necessary.

The decision between Mitral Valve Replacement (MVR) and mitral valve repair is influenced by multiple clinical, anatomical and patient-related factors.

Lesions mainly affect the mitral and aortic valves, but all four valves and the entire endocardial surface can be affected [6].

TTE is the examination of choice for diagnosing SLE, with the incidence of valvular heart disease varying between 18-50% [5,7,8] and increases to 74% if TEE is performed [9].

Valvular lesions are characterized microscopically by fibrin deposits, an infiltrate of mononuclear inflammatory cells, fibrosis, neovessels and sometimes immunoglobulin and complement deposits [10,11].

The presence of Antiphospholipid Antibody Syndrome (APAS), found in our case by a positive LA blood test, is associated with a high prevalence of mitral vegetation, suggesting, according to studies by Khamashta MA et al, Gabrielli F et al, a pathogenic involvement of antiphospholipid antibodies.

Mitral valve surgery in SLE

- No consensus has yet been reached on this question, and data are limited to a small number of case reports and series of a few cases, so indications are not well studied. Surgical treatment is certainly indicated in cases of large vegetation, severe Mitral Insufficiency (MI) and recurrent thromboembolic events, even under medical treatment.
- Some authors, Hakim JP and al., Chauvaud SM and al, suggest that mitral valve replacement leads to better results than plasty [14,15], arguing that by the evolution of valve calcification and fibrosis in lupus and APAS leads to rapid recurrence of MI after plasty, necessitating further surgery. In addition, it has been reported that corticosteroid therapy indicated in the treatment of SLE can lead to narrowing of the heart valves [16], while the presence of APAS indicates the need for lifelong anticoagulant therapy [17].
- In specific cases, Bouma et al [18] conclude that conservative surgical treatment is justified, particularly in young patients, if LES and/or APAS are stabilized on immuno-suppressive therapy, and if intraoperative macroscopic examination shows suitable valve leaflet anatomy, with localized lesions, and mitral plasty appears feasible. And above all, in young women wishing to become pregnant, to avoid the need for anticoagulant treatment.
- In our case, despite being a young patient with localized lesions in the anterior mitral valve, and thin flexible leaflets, a mitral valve repair was envisaged for reconstruction of the valve perforation and consolidation of the plasty. However, the fragility of the infiltrated and scarred tissue meant that the mitral valve had to be replaced by a mechanical prosthesis.

Conclusion

Early, multidisciplinary management of SLE can prevent its complications, which can be serious. In the event of surgical indication, the choice between mitral valve repair or replacement must meet clinical, anatomical and patient-related criteria.

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