



Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Surgical management of Libman-Sacks mitral valve endocarditis

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ARTICLE INFO

Article history:

Received 15 December 2020

Received in revised form 1 January 2021

Accepted 1 January 2021

Available online 16 January 2021

Keywords:

Case report

Libman sacks endocarditis

Mechanical mitral valve replacement

Systemic lupus erythematosus

Valvular re-intervention

Prevention

ABSTRACT

INTRODUCTION AND IMPORTANCE: Libman-Sacks endocarditis is a non-bacterial form of thrombotic endocarditis that is associated with Systemic Lupus erythematosus (SLE) syndrome.

CASE PRESENTATION: A 32 years old male with SLE and Antiphospholipid syndrome, presented with shortness of breath, intermittent chest pain and pyrexia. He had an embolic cerebral infarct a year previously with recurrent seizures. Echocardiogram showed severe Mitral regurgitation with a fixed posterior leaflet and Mobile structures on the leaflet tips. He underwent mechanical Mitral valve replacement. Vegetation was found on valve leaflets. Blood and valve tissue Cultures were sterile. Valve histology showed scarring and active inflammatory changes but no bacterial or fungal colonies.

CLINICAL DISCUSSION: In such case a durable valve repair cannot be guaranteed and bioprosthetic valve replacement is prone to early structural degeneration requiring re intervention especially in young patients. Mechanical valve has capacity to withstand the destructive inflammatory milieu of SLE and chronic renal failure.

CONCLUSION: A mechanical valve replacement is a reasonable choice in a young patient with SLE in the setting of antiphospholipid syndrome to reduce valvular re-intervention rate.

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1. Introduction

Libman-Sacks endocarditis is a non-bacterial form of thrombotic endocarditis that damages the heart valves, because of the trauma mitral leaflet coaptation inducing injury and exposure of phospholipid on leaflets edges. This is why the vegetation typically occurs at the leaflet edges. Valve surgery is indicated in the presence of large vegetation, significant valvular malfunction, recurrent thrombotic events or heart failure. Antiphospholipid syndrome a common associate of SLE also influences management strategies as it may require lifelong anticoagulation. Early failure of mitral valve repair and tissue valve degeneration has been reported in Libman-Sacks endocarditis despite optimal medical treatment of SLE. The search for the optimal treatment of Libman-Sacks endocarditis is still on going. Our case report is from a university hospital which is a reference heart valve Centre. The report demonstrates that Libman-Sacks mitral valve endocarditis can be treated with mechanical mitral valve replacement with a good outcome. This case has been reported in line with the SCARE criteria [6].

2. Case presentation

A 32 years old Mediterranean male, presented with worsening shortness of breath and intermittent chest pain and pyrexia associated with a grade 4 apical systolic murmur of mitral regurgitation. He has a previous history of embolic cerebral infarct. Trans-esophageal echocardiogram (TOE) (Fig. 1), showed severe mitral regurgitation due to retracted leaflets with vegetation, suspicious for infective endocarditis. Multiple blood cultures did not isolate any pathogen. Diagnostic angiogram showed unobstructed coronary arteries. The patient was taken to mitral valve surgery and because cultures were sterile, a diagnosis of Libman-Sacks endocarditis was considered.

The leaflets had vegetation's typical of Libman-Sacks endocarditis (Fig. 2). The patient had a Mechanical Mitral valve replacement performed by a senior mitral valve specialist cardiac surgeon. The operation and recovery was uncomplicated.

Histopathology (Fig. 3) showed valve leaflets covered by fibrinous exudate with neutrophils, lymphocytes and histiocytes, but no bacterial or fungal colonies.

These appearances are in keeping with Libman-Sacks endocarditis.

At 12 months follow-up he was clinically stable and echocardiogram showed good prosthetic mitral valve and ventricular function.

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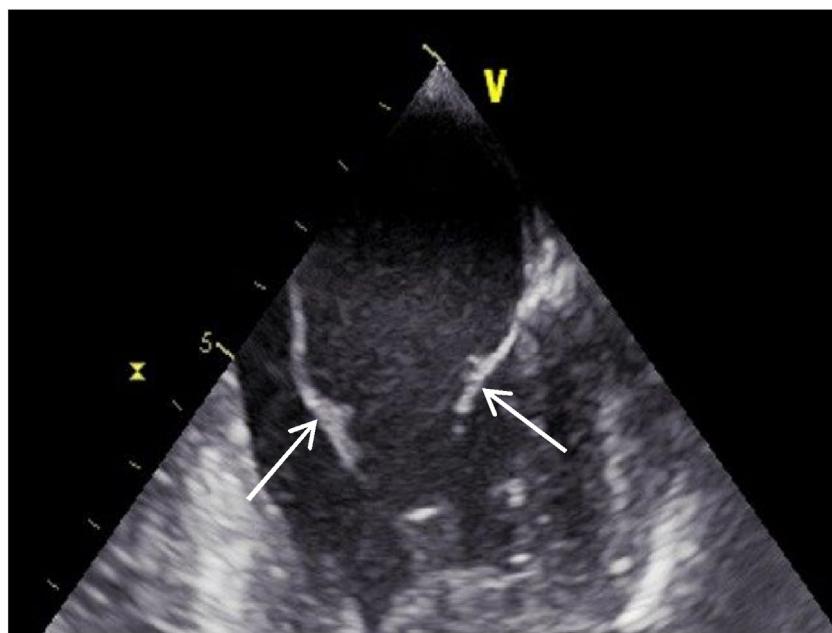


Fig. 1. TOE showing mobile vegetation on tip of mitral leaflet. (Arrows points to vegetations).

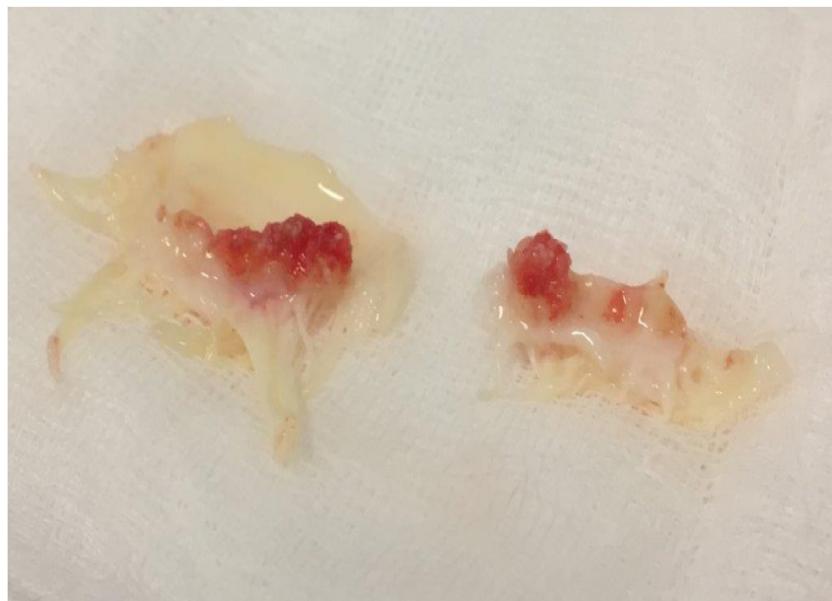


Fig. 2. Excised Mitral valve vegetation.

3. Discussion

Up to 61% of SLE patients, have valvular vegetation and regurgitation [1].

The indications for surgery are severe valvular dysfunction, large vegetation and recurrent embolization despite therapeutic anti-coagulation. Some reports suggest that native valve repair does not alter the progression of valve thickening and calcification, and that replacement is ultimately necessary [2]. Additionally, it has been reported that while corticosteroid therapy improves overall survival of patients with SLE, their use may cause shrinking and scarring of heart valves [3]. It has also been noted that porcine bio-prosthetic valves can be affected by valvulitis with perforation in the cusps in patients with SLE requiring replacement. Furthermore,

it has been reported that SLE-associated renal failure can accelerate native and porcine bio-prosthetic valvular degenerative calcification and stenosis due to derangements in calcium and phosphate homeostasis [4,5].

Mechanical valve was favored based on the patient's young age and the existing need for lifelong anticoagulation recommended in patients with an antiphospholipid Syndrome coagulopathy [5].

As medical management has improved the survivability of patients with SLE, more patients can be expected to survive long enough for severe valve damage to develop, and more people will need valve replacement in the future and subsequent re-intervention in younger patients for recurrent valve disease or structural tissue valve degeneration.

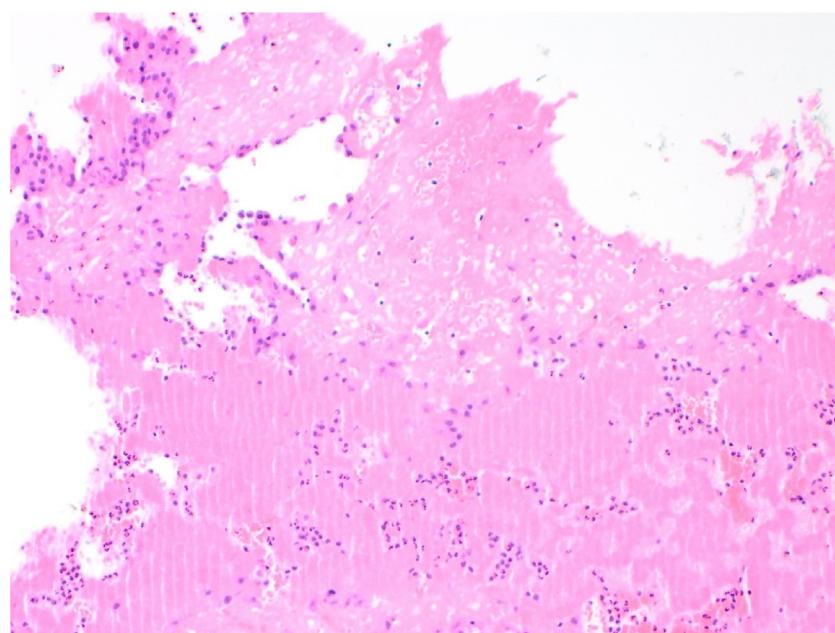


Fig. 3. Histological of Excised Valve showing fibrosis and inflammatory reaction.

4. Conclusion

A mechanical valve replacement is a reasonable treatment choice in a young patient with Libman–Sacks mitral endocarditis.

Declaration of Competing Interest

No conflict of interest declared by all authors.

Funding

No any Sources of funding to declare for this report.

Ethical approval

The study is exempt from ethical approval in our institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author's contribution

Youssef Abouelela: study concept or design, data collection, data analysis or interpretation, writing the paper, and approved final copy.

Philemon Gukop: study concept or design, data collection, data analysis or interpretation, writing the paper, and approved final copy.

Steve Livesey: study concept or design, data collection, data analysis or interpretation, writing the paper, and approved final copy.

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Registration of research studies

Not Applicable.

Guarantor

Mr Steve Livesey, Philemon Gukop.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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