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## CASE REPORT

# Aortic valve replacement for Libman-Sacks endocarditis

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## SUMMARY

A 24-year-old man with systemic lupus erythematosus and antiphospholipid syndrome complicated by lupus nephritis presented with acute limb ischaemia secondary to an embolus. Following embolectomy, the patient underwent a transthoracic echocardiogram which revealed a large vegetation on all three cusps of the aortic valve. The patient was taken for an urgent aortic valve replacement with a mechanical valve. Cultures of one cusp remained sterile. Histopathological examination of the remaining two cusps revealed sterile fibrin-rich thrombotic vegetations characteristic of non-bacterial thrombotic endocarditis.

## BACKGROUND

Libman-Sacks (verrucous) endocarditis is a form of non-bacterial thrombotic endocarditis (NBTE) that causes damage to heart valves in the setting of systemic lupus erythematosus (SLE). Surgical valve replacement is necessary in patients with large vegetations, valvular insufficiency and recurrent thromboembolic events. We present a case of a 24-year-old man with SLE and antiphospholipid syndrome complicated by lupus nephritis and NBTE who was treated with aortic valve replacement.

## CASE PRESENTATION

A 24-year-old man with a medical history of SLE, stage IV lupus nephritis, antiphospholipid syndrome (aPL) and recurrent deep vein thromboses (DVTs) requiring life-long anticoagulation presented to the emergency department after the acute onset of a cool, painful, numb right leg and foot. The patient's home medications included low-dose prednisone, hydroxychloroquine and mycophenolate for SLE as well as rivaroxaban after multiple DVTs. On physical examination, the patient had both a grade II/VI systolic and diastolic murmur and did not have palpable or Doppler ultrasound-detectable pulses in the right foot. Arterial ultrasonography revealed an acute occlusion in the distal common femoral artery, and the patient was taken to the operating room for an emergent embolectomy for acute limb ischaemia. On admission, the prothrombin and partial thromboplastin times were prolonged, and the International Normalised Ratio was 1.8. Rivaroxaban was stopped and the patient was placed on heparin. Histopathological examination revealed a recent, laminated fibrin-rich thrombus.

The patient was stable following embolectomy and underwent a transthoracic echocardiogram

which revealed a 1.4×1.7 cm mass on his aortic valve, with moderate aortic regurgitation and an ejection fraction of 62%. The bubble study was negative for evidence of a patent foramen ovale. Workup for endocarditis showed no signs of an infective process, as there was no associated fever, constitutional symptoms and physical examination findings and blood cultures were negative. As further workup, the patient had a real-time three-dimensional transesophageal echocardiogram, which revealed large, irregular, mobile vegetations on all three cusps of the aortic valve (figure 1). There was also moderate-to-severe aortic regurgitation. These findings were highly suspicious for Libman-Sacks endocarditis. Therefore, the patient was taken for an urgent aortic valve replacement.

## TREATMENT

Following full sternotomy, cardiopulmonary bypass was established via aortic and right atrial cannulae. Following cross-clamping, cold antegrade cardioplegia was applied for cardioprotection. Exposure of the aortic valve revealed verrucose thickening of the aortic leaflets with large vegetations involving the three aortic leaflets. The valve could not be preserved due to insufficient coaptation of the leaflets. Following excision of the leaflets, a 21 mm St. Jude mechanical heart valve was implanted. The patient was weaned off bypass on minimal inotropic support after 119 min of bypass time with 89 min of cross-clamp time. Postoperatively, the patient was extubated within 24 hours and made an uncomplicated recovery. He was bridged to therapeutic warfarin with heparin.

## OUTCOME AND FOLLOW-UP

One excised valve cusp was sent for culture, but no bacterial or fungal growth was observed. Histopathological examination of the remaining two cusps revealed features that are characteristic of NBTE, including abundant fibrin-rich thrombotic vegetations containing essentially no inflammation. The vegetations were adherent to the aortic surface of the cusps along the closing edge, with preservation of cusp architecture. No perforations or destruction of cusp tissue was identified, and tissue Gram and Gomori-methenamine silver stains were negative for bacterial and fungal organisms, respectively, further supporting a diagnosis of NBTE (figure 2).

The patient is doing well 1 year post-aortic valve replacement and has not had any thromboembolic events.



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**Figure 1** Real-time three-dimensional transesophageal echocardiogram image of the aortic valve showing large, irregular vegetations on all three cusps.

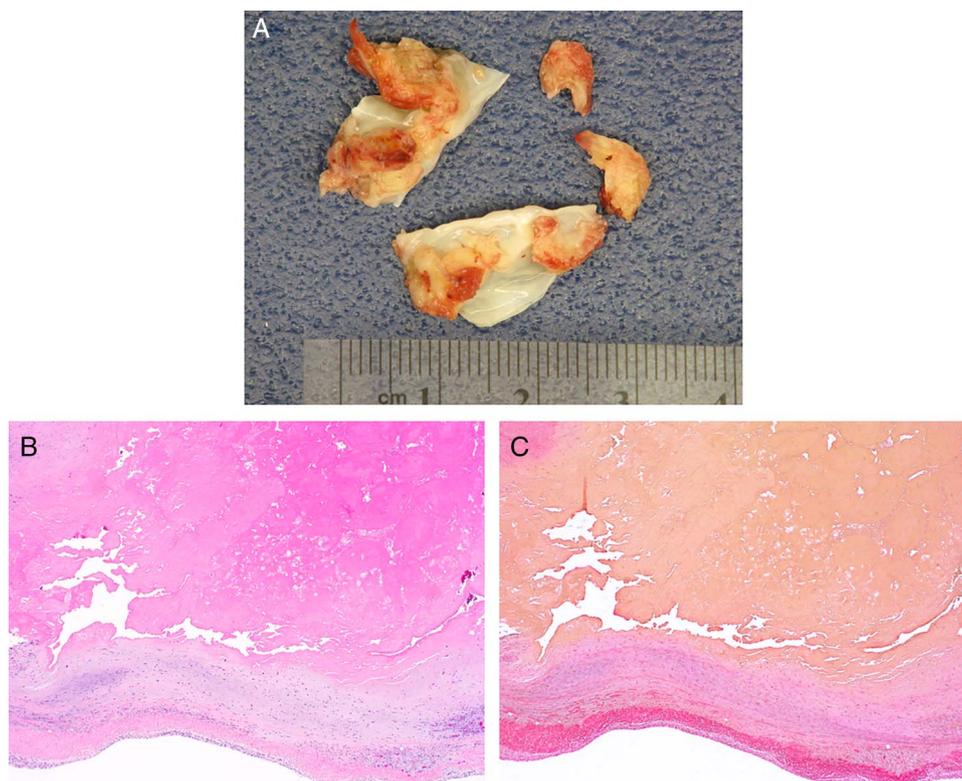
## DISCUSSION

Valvular abnormalities are common in SLE, occurring in up to 61% of patients evaluated by transesophageal echocardiography. Valvular thickening is the most common echocardiographic abnormality observed, followed by valvular vegetations and regurgitation.<sup>1</sup> In patients with aPL, the prevalence of mitral valve vegetations and mitral regurgitation is significantly higher than in patients with SLE without aPL.

The vegetations of Libman-Sacks can affect all four valves and usually accumulate at the valve edge or on both surfaces. These

lesions consist of immune complexes, monocytes, fibrin and platelet aggregates and can have varying degrees of fibrosis, granulation tissue, calcification and necrosis. These lesions are typically friable and can fragment and embolise, particularly when they are large. Healing of the lesion causes fibrotic scarring and further deformity of affected valve components, resulting in insufficiency. Valvular lesions can also predispose to infective endocarditis.

Unlike infective endocarditis where the valve needs to be excised to remove all infected tissue, repair and preservation of the valve is possible in selected patients with NBTE and may eliminate the need for anticoagulation. While data on surgical outcomes for patients with Libman-Sacks endocarditis receiving valve replacement have been limited to a few case reports and small case series and indications for surgery have not been sufficiently studied, severe valvular dysfunction, large vegetations and recurrent embolisation, despite therapeutic anticoagulation, are clear indications for surgical intervention. Case reports suggest that native valve repair does not alter the progression of valve thickening and calcification, and that replacement is ultimately necessary.<sup>2</sup> 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.<sup>3</sup> It has also been noted that porcine bioprosthetic 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 bioprosthetic valvular degenerative calcification and stenosis due to derangements in calcium and phosphate homeostasis.<sup>4 5</sup>



**Figure 2** (A) Grossly, the excised aortic valve cusps show pink-red, bulky vegetations adherent to their ventricular surfaces along the lines of closure, without perforations or cusp thickening (one cusp was submitted separately for culture). (B and C) Histopathological examination reveals features consistent with NBTE, including fibrin-rich thrombus with essentially no inflammatory component (upper two-thirds of images) on the surface of the cusp (lower third), with preservation of cusp architecture and no evidence of infection. (B: H&E stain; C: Verhoeff Van-Gieson stain; original magnification 40 $\times$ ). NBTE, non-bacterial thrombotic endocarditis.

In our patient, selection of mechanical valve was based on the patient's age and the existing unavoidable need for anticoagulation. Lifelong anticoagulation is recommended in patients with an aPL coagulopathy.<sup>5</sup>

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.

### Learning points

- ▶ Patients with systemic lupus erythematosus (SLE) commonly develop valvular vegetations of non-bacterial origin.
- ▶ Valves should be repaired or replaced in patients who develop symptomatic embolisms of cardiac origin.
- ▶ If repair of the valve is not possible, a mechanical valve is a good choice in a young patient with SLE in the setting of antiphospholipid syndrome as lifelong anticoagulation is necessary for the coagulopathy.

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contributed critically important intellectual content. All authors contributed substantially to the work. All authors discussed the results and implications and commented on the manuscript at all stages.

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