

# An Interesting Case of Libman-Sacks Endocarditis

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## Background

- Libman-Sacks Endocarditis (LSE) OR Nonbacterial thrombotic endocarditis (NBTE) is found in 0.9 - 1.6% of autopsies without sex predilection, most common in 4th-8th decades of life
- LSE is more likely in hypercoagulable patients, especially malignancy and Systemic Lupus Erythematosus (SLE) but less commonly by sepsis and antiphospholipid syndrome
- Mostly cases are asymptomatic, found incidentally during echo but merit interest as they can lead to investigations for causes of vegetations - discovery requires strong clinical suspicion in patients without systemic infection but with risk factors
- Treatment focuses on underlying condition which may include anticoagulation, recommended to be indefinitely due to high risk of embolization
- The condition has a poor prognosis

## Objective

Showcase a presentation of LSE, the subsequent workup by means of which to increase understanding of optimizing workup, minimizing unnecessary intervention and managing potential consequences/ underlying conditions

## Case Presentation

- 55yo female patient with PMH antiphospholipid syndrome on Xarelto, untreated SLE (due to allergy) presented from cardiologist following mitral valve vegetation seen on echo. Patient also had brain CT, MRI ordered due to nonspecific episodes of dizziness/vertigo/blurred vision. Office carotid dopplers negative.
- Blood cultures drawn. ED gave single dose meropenem out of concern from story for endocarditis. Troponinemia. Normal C3, C4, anti-ds DNA unlikely lupus flare.
- Workup for endocarditis with ID + cards consulted.
- Vitals within normal limits, afebrile, normal CBC → antibiotics were not continued
- Cultures returned negative.
- TEE performed: Moderately thickened leaflets. 1.2cmx. 3cm filamentous vegetation on atrial surface of posterior mitral valve leaflet. Trace mitral mild stenosis. Mild mitral regurgitation. Otherwise normal. In combination with clinical history suggestive of nonbacterial endocarditis
- DC Xarelto, start Lovenox on discharge. Follow up with rheumatology scheduled to address underlying condition

## Discussion

- What anticoagulation for Libman-Sacks endocarditis? Medical treatment for patients with LS endocarditis is not well established, partially due to the scarcity of information on the natural progression of the disease in untreated patients. Recommendations regarding management originate from expert opinion and small observational case studies rather than randomized trials. Therapeutic dose subcutaneous low molecular weight (LMW) heparin or intravenous unfractionated heparin should be used, rather than factor Xa inhibitor (these agents should be avoided for antiphospholipid syndrome).
- Although no formal comparison between heparin and warfarin has been reported in patients with non-bacterial thrombotic endocarditis, older studies suggest that warfarin is less effective than heparin in reducing the rate of recurrent embolization. For patients with newly diagnosed APS presenting with their first thrombosis, gold standard is Warfarin with an INR goal of 2 to 3.
- When to do Surgery? The indications for surgery (vegetation excision or valve replacement) are the same as for infective endocarditis (eg, heart failure, acute valve rupture) but reports suggest that prevention of recurrent embolization is the most common reason for surgery. In contrast to infective endocarditis where complete removal of infected tissue is important, preservation of the valve may be possible in some cases of NBTE
- LS endocarditis is not a common condition, but when it presents, it often correlates with high morbidity and mortality. The healthcare team, including the pharmacists and nurses, should be aware that the vegetations can embolize not only to the brain but also to the extremities and intestine. If this is not recognized, then it can result in ischemia and necrosis of the organ involved.
- The healthcare team should incorporate multiple specialties. Cardiology can assist with the assessment of the nature and severity of valvular disease and heart failure management. A cardiac surgeon may be helpful if valve replacement is required. A rheumatologist can optimize lupus. Infectious disease to address potential sepsis or secondary infective endocarditis. Neurologist for cerebral vascular complications

## Conclusion

Libman-Sacks endocarditis refers to sterile vegetations on previously normal heart valves. The valves most often affected are mitral and aortic and the size of vegetations range from microscopic to large. The pathophysiology appears to be endothelial injury in a hypercoagulable state so they are typically seen in pts with systemic lupus erythematosus, antiphospholipid antibody syndrome and patients with cancer.

- Libman-Sacks is typically asymptomatic and often found incidentally while evaluating for other cardiac pathology. Lesions rarely lead to significant valvular dysfunction.
- Pts may present with signs of stroke or TIA from a cerebrovascular embolic event.
- Diagnosis can be made on echocardiography. Other potential etiologies must be excluded through workup such as infective endocarditis. TEE has greater sensitivity and specificity than TTE.
- The optimal treatment needs further investigation because recommendations are based on small observational studies rather than randomized trials.
- The underlying disease SLE and APS in our case needs to be adequately treated as well as anticoagulation. Warfarin with a goal INR 2-3 remains the gold standard for treatment of APS.
- There is no formal comparison between heparin/LMWH and warfarin in Libman-Sacks Endocarditis, but older studies suggest that warfarin is less effective in reducing rates of embolization. Factor Xa inhibitors should be avoided.
- Surgery reserved for significant valvular dysfunction and according to established guideline of valvular heart disease.
- Scheduled echocardiography every 3 to 6 months may be beneficial to monitor the progression or resolution of the disease.

## References

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