

A Unique Presentation of Libman-Sacks: Non-Bacterial Tricuspid Valve Endocarditis

Introduction

- Systemic Lupus Erythematosus (SLE) is a chronic autoimmune condition affecting many organ systems.
- Cardiac manifestations are common including valvular disease, but not routinely screened. [3]
- Mitral and aortic valves are most commonly affected in Libman Sacks endocarditis. [2,5]
- Tricuspid valvular changes are the rarest echocardiographic findings. [6]
- Valvular involvement is important to identify particularly in hypercoagulable states.

Case Description

- 63-year-old female with SLE, recent thromboembolic disease on apixaban presented with chest discomfort and shortness of breath.

Table 1 - Findings on Presentation

Vital Signs	
Heart Rate	117 beats/min
SpO2	87% on room air
Lab Results and Imaging	
Troponin	3339 ng/L
EKG	Sinus tachycardia, right axis deviation, without acute T-ST changes
CXR	Without acute processes

- Cardiology was consulted. TTE and TEE obtained, see Table 3. Hypercoagulability panels obtained, see Table 2.
- Infectious Disease consulted to rule out infectious etiologies. Repeat blood cultures remained negative.
- Patient transferred to another facility for AngioVac procedure and pathologic interpretation, see Table 4.

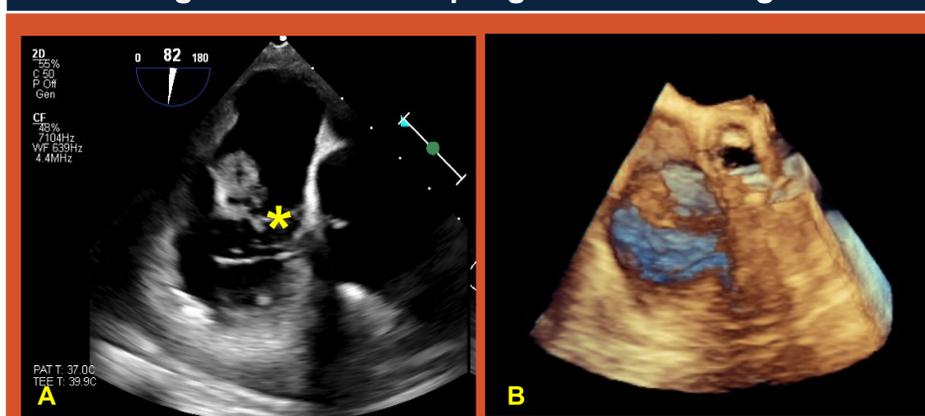
This research was supported (in whole or in part) by HCA Healthcare and/or an HCA Healthcare affiliated entity. The views expressed in this publication represent those of the author(s) and do not necessarily represent the official views of HCA Healthcare or any of its affiliated entities.

Labs and Figures

Table 2 - Immunology Findings

Hypercoagulability Antibodies	Results		
Anti-Cardiolipin Ab	IgG <1.6	IgM <1.5	IgA < 2.0
Anti- Beta 2 Glycoprotein I Ab	IgG <9	IgM <9	IgA <9
Lupus Anticoagulant Test	Negative		

Figure 1 – Transesophageal Echocardiogram



A) Midesophageal View showing vegetation on tricuspid valve (*)
B) 3D view of vegetation on tricuspid valve

Table 3 - Transesophageal Echocardiogram Findings

	11/07/2022	12/22/2022
Tricuspid Valve	<ul style="list-style-type: none"> • Normal valve structure • Normal leaflet separation • No evidence for vegetation • No evidence for stenosis • No regurgitation 	<ul style="list-style-type: none"> • Degenerative tricuspid valve with possible leaflet perforation and associated sessile mass • Severe tricuspid valve regurgitation

Table 4 - Pathology Findings

Gross Description
<ul style="list-style-type: none"> • Multiple fragments of tan soft tissue • 1.5 x 0.4 x 0.2cm
Tricuspid Vegetation
<ul style="list-style-type: none"> • Organizing thrombus with interspersed inflammatory cells (predominantly macrophages with occasional acute inflammatory cells) • Negative for neoplasia

Discussion

- Prevalence of Libman-Sacks endocarditis is unknown with rates estimated from 6% to 74%. [1,3]
- Nonbacterial thrombotic endocarditis is associated with antiphospholipid syndrome with a suggested prevalence of 33%. [3]
- Endothelial injury from circulating cytokines, immunoglobulins, and complements result in the deposition of platelet thrombi and inflammatory molecules is a proposed mechanism. [2,5]
- Echocardiography is a sensitive and specific test to detect cardiac abnormalities, which is recommended for patients with antiphospholipid antibodies. [4]
- Unfortunately, the patient expired before the full laboratory and pathologic evaluation could be completed.

Conclusion

- Libman Sacks tricuspid valve endocarditis in treatment compliant SLE with negative antiphospholipid antibodies is uncommon.
- There may be a role for routine cardiac screening regardless of clinical status.
- More research is needed to better understand the pathophysiology of Libman Sacks endocarditis and role of anticoagulants in treatment.

References

Thank you to HCA Trinity Hospital faculty and staff for their contribution and support.

- Miner JJ, Kim AH. Cardiac manifestations of systemic lupus erythematosus. *Rheum Dis Clin North Am.* 2014 Feb;40(1):51-60. doi: 10.1016/j.rdc.2013.10.003. PMID: 24268009. [1]
- Ibrahim AM, Siddique MS. Libman Sacks Endocarditis. [Updated 2022 Sep 5]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK532864/> [2]
- Yoo BW, Lee SW, Song JJ, Park YB, Jung SM. Clinical characteristics and long-term outcomes of Libman-Sacks endocarditis in patients with systemic lupus erythematosus. *Lupus.* 2020 Aug;29(9):1115-1120. [PubMed] [3]
- Doria A, Iaccarino L, Sarzi-Puttini P, Atzeni F, Turriel M, Petri M. Cardiac involvement in systemic lupus erythematosus. *Lupus.* 2005;14(9):683-686. doi:10.1191/0961203305lu2200oa [4]
- Flutere A, Chaudhari S, Frishman WH. Valvular heart disease and systemic lupus erythematosus: therapeutic implications. *Heart Disease (Hagerstown, Md.).* 2003 Sep-Oct;5(5):349-353. DOI: 10.1097/01.hdx.0000089834.97829.5e. PMID: 14503933. [5]
- Moaref, A. R., Affi, S., Rezaian, S., & Rezaian, G. R. (2010). Isolated Tricuspid Valve Libman-Sacks Endocarditis and Valvular Stenosis: Unusual Manifestations of Systemic Lupus Erythematosus. *Journal of the American Society of Echocardiography*, 23(3), 341.e3-341.e5. <https://doi.org/10.1016/j.echo.2009.09.004> [6]