Mitral Valve Libman–Sacks Endocarditis Visualized by Real Time Three-Dimensional Transesophageal Echocardiography

Hersh Shroff, B.S., Ricardo Benenstein, M.D., Robin Freedberg, M.D., Sydney Mehl, M.D., and Muhamed Saric, M.D., Ph.D.

Leon H. Charney Division of Cardiology, New York University Langone Medical Center, New York, New York

Libman–Sacks endocarditis (LSE) is a common manifestation of valve disease in antiphospholipid syndrome. Mitral valve LSE is characterized by verrucous vegetations on the atrial surfaces of valve leaflets. In this report, mitral valve LSE was visualized by real time 3D transesophageal echocardiography (TEE). 3D TEE provides a unique en face view of the mitral valve akin to a surgical or autopsy view that allows for an accurate determination of the size, shape, and location of the vegetations. (Echocardiography 2012;29:E100-E101)

Key words: 3D TEE, echocardiography, nonbacterial endocarditis, Libman–Sacks

Case Report:

A 68-year-old woman with presumed transient ischemic attack was referred for transesophageal echocardiography (TEE) to rule out a cardiac source of emboli. Her medical history was notable for myelodysplastic syndrome and lupusanticoagulant-associated antiphospholipid syndrome (APS).

Two-dimensional (2D) TEE revealed vertucous masses on the atrial surfaces of both mitral valve (MV) leaflet tips. The masses measured approximately 5×5 mm each. They had a "kissing lesion" appearance upon coaptation of the mitral valvular leaflets. There was mild to moderate mitral regurgitation as well (Fig. 1 and movie clip S1).

A real time, three-dimensional TEE (RT-3D TEE) was performed for further evaluation, and a shortaxis, en face view of the MV from the left atrial perspective was obtained. The 3D TEE revealed mound-like protuberances in the region of A2 and P2 MV scallops along their coaptation surfaces (Fig. 2; movie clips S2 and S3).

Repeated blood cultures were negative. Furthermore, there were no clinical or laboratory findings suggestive of an infective process, a rheumatologic disorder or malignancy. Given the clinical scenario, the echocardiographic findings were consistent with the diagnosis of Libman–Sacks nonbacterial thrombotic endocarditis in the setting of primary (idiopathic) APS.

Discussion:

The most common cardiac presentation of APS is valvular heart disease.¹ Approximately one-third of patients with primary APS develop valvular disease.^{2,3} Valvular findings are even more prevalent in patients with systemic lupus erythematosus-associated secondary APS.⁴

Libman–Sacks endocarditis (LSE) is a common manifestation of valve disease in APS. LSE was initially described in patients with systemic lupus erythematosus but has since been extended to include other forms of nonbacterial thrombotic endocarditides, including those associated with APS.⁵

LSE is commonly characterized by leaflet thickening or sterile, verrucous vegetations that occur most often on left-sided valves, with an approximately equal frequency of aortic and mitral involvement.⁵ These sterile vegetations usually occur along the mitral leaflet or aortic cusp closure line. LSE lesions can be clinically asymptomatic or can be associated with valvular dysfunction and embolism.⁶ In a series of patients with LSE of the MV, about one-half of patients had some degree of mitral regurgitation. In three-quarters of such patients, mitral regurgitation was only mild. Embolic risk is directly correlated with the size of LSE vegetations.⁷

²D echocardiography is the standard modality for antemortem diagnosis of LSE⁸ but offers only cross-sectional views of the MV. In contrast, 3D TEE provides an en face view of the MV—akin to a surgical or autopsy view—that allows for a more accurate determination of the size, shape, and location of the vegetation.

The differential diagnosis of the 3D TEE MV findings may include bacterial or fungal vegetations, Lambl's excrescences, and MV prolapse. Unlike the verrucous vegetations, which

Address for correspondence and reprint requests: Muhamed Saric, M.D., Ph.D., Noninvasive Cardiology Laboratory, New York University Medical Center, 560 First Avenue, New York, NY 10016. Fax: (212) 263-8461; E-mail: muhamed.saric@nyumc.org

Figure 1. Libman–Sacks endocarditis on 2D TEE. 2D TEE obtained at 129 degrees. **A.** Libman–Sacks vegetations (arrows) on the atrial surface of the mitral leaflet tips seen during diastole. **B.** During systole, the vegetations on the two leaflets give the appearance of a "kissing lesion" (arrow). LA = left atrium; LV = left ventricle; LVOT = left ventricular outflow tract.





Figure 2. Libman–Sacks endocarditis on 3D TEE. Real time 3D TEE zoom en face view of the mitral valve seen from the left atrial perspective. Mitral valve Libman–Sacks endocarditis appears as mound-like protuberances (arrows) on the tips of the A2 scallop of the anterior leaflet and the P2 scallop of the posterior leaflet. AV = aortic valve; AML = anterior mitral leaflet; PML = posterior mitral leaflet.

usually have smooth and gelatinous appearance, the bacterial or fungal vegetations tend to be shaggy with irregular borders and excrescences. Although Lambl's excrescences also tend to occur near valvular closure lines, they usually do not appear as masses; they are typically thin and threadlike. In MV prolapse, the leaflets balloon out into the left atrium and thus may appear as masses.

To our knowledge, this is the first description of MV LSE using RT-3D TEE.

References

- 1. Tenedios F, Erkan D, Lockshin MD: Cardiac involvement in the antiphospholipid syndrome. *Lupus* 2005;14:691–696.
- Vianna JL, Khamashta MA, Ordi-Ros J, et al: Comparison of the primary and secondary antiphospholipid syndrome: A European multicenter study of 114 patients. *Am J Med* 1994;96:3–9.
- Brenner B, Blumenfeld Z, Markiewicz W, et al: Cardiac involvement in patients with primary antiphospholipid syndrome. J Am Coll Cardiol 1991;18:931–936.

- Cervera R, Khamashta MA, Font J, et al: High prevalence of significant heart valve lesions in patients with the 'primary' antiphospholipid syndrome. *Lupus* 1991;1:43–47.
- Reisner S, Brenner B, Haim N, et al: Echocardiography in nonbacterial thrombotic endocarditis: From autopsy to clinical entity. J Am Soc Echocardiog 2000;13:876–881.
- Hojnik M, George J, Ziporen L, et al: Heart valve involvement (Libman-Sacks endocarditis) in the antiphospholipid syndrome. *Circulation* 1996;93:1579–1587.
- Roldan CA, Qualls CR, Sopko KS, et al: Transthoracic versus transesophageal echocardiography for detection of Libman-Sacks endocarditis: A randomized controlled study. J Rheumatol 2008;35:224–229.
- Murphy RT, Garcia MJ: Role of echocardiography in diagnosis and management of endocarditis. *Curr Infect Dis Rep* 2005;7:257–263.

Supporting Information

Additional Supporting Information may be found in the online version of this article:

Movie clip S1. Gray scale and color Doppler 2D TEE obtained at 129 degrees demonstrates Libman–Sacks vegetations (arrow) on the atrial side of the mitral valve leaflets (A2 and P2 scallops) as well as mild to moderate mitral regurgitation. LA = left atrium; LV = left ventricle; LVOT = left ventricular outflow tract; AV = aortic valve.

Movie clip S2. Real time 3D TEE zoom en face view of the mitral valve seen from the left atrial perspective demonstrates mound-like Libman– Sacks vegetations (arrow) on anterior and posterior mitral valve leaflets (A2 and P2 scallops). AV = aortic valve; AML = anterior mitral leaflet; PML = posterior mitral leaflet.

Movie clip S3. Postprocessed real time 3D TEE zoom en face view of the mitral valve seen from the left atrial perspective shows Libman–Sacks vegetations from various angles during systole.

Please note: Wiley-Blackwell are not responsible for the content or functionality of any supporting materials supplied by the authors. Any queries (other than missing material) should be directed to the corresponding author for the article.