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BEGINNER

CASE REPORT: CLINICAL CASE

Cardiogenic Shock and Mitral Valve Chord Rupture

A Rare Presentation of Libman-Sacks Endocarditis

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ABSTRACT

Distinguishing Libman-Sacks endocarditis from other valvular heart disease etiologies has important implications for management. We present a case of a 23-year-old man who presented in extremis with fever and cardiogenic shock caused by Libman-Sacks endocarditis with associated mitral valve chord rupture. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2020;2:1988–91) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 23-year-old undomiciled man presented to the emergency department with 3 h of acute-onset dyspnea. Over the past 6 weeks he reported subjective fever, diaphoresis, weight loss, arthralgias, pleuritic chest pain, and frothy urine. Two months ago he completed treatment with amoxicillin-clavulanate for streptococcal pharyngitis and denied injected drug use.

LEARNING OBJECTIVES

- To highlight how Libman-Sacks endocarditis can mimic infective endocarditis.
- To describe a severe, rare presentation of this uncommon clinical entity.

On physical examination, the patient was tachypneic, using accessory muscles of respiration. The heart rate was 143 beats/min, blood pressure was 109/57 mm Hg, the oxygen saturation was 95% on 4 l/min oxygen by nasal cannula, and the temperature was 39.4°C. Jugular venous distention, a II/VI decrescendo systolic murmur at the left lower sternal border, a blowing III/VI holosystolic murmur at the apex radiating to the axilla, and S3 were noted. A Janeway lesion was present on the plantar surface of the left second toe. The extremities were cold, with trace pretibial edema. The lungs had bilateral rales and pulses were intact. There was no hepatosplenomegaly.

PAST MEDICAL HISTORY

The patient had no known medical or surgical history.

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INVESTIGATIONS

Chest radiography revealed diffuse pulmonary edema. An electrocardiogram showed sinus tachycardia. The white blood cell count was 11.7 × 10^3/μl with 81% neutrophil predominance. The hemoglobin was 8.9 g/dl and the platelet count was 226 × 10^3/μl. Serum creatinine peaked at 2.18 mg/dl with subnephrotic proteinuria and hematuria on urinalysis. A serum arterial lactate level peaked at 3.7 mmol/l. A ferritin level and transferrin saturation were elevated at 1,718 ng/ml and 64%, respectively. Troponin levels were normal. The erythrocyte sedimentation rate was elevated at 139 mm/HR and C-reactive protein was elevated (7 mg/l). Testing for acquired immunodeficiency, tick-borne illnesses, tuberculosis, and cultures for bacteria, fungi, and HACEK organisms were negative. A rheumatologic work-up revealed a positive antinuclear antibody, positive double-stranded DNA titer (>1:10), positive Smith antibodies (>8.0 antibody index), and negative antineutrophil cytoplasmic antibody serologies. Complement levels were low, including C3 (35 mg/dl), C4 (6 mg/dl), and complement CH50 (<10). An antiphospholipid syndrome (APLS) evaluation was negative.

Transthoracic echocardiography (**Figure 1**, Video 1) obtained the day of presentation revealed normal biventricular function and posterior mitral valve leaflet thickening with severe mitral regurgitation and mild to moderate aortic insufficiency. Transesophageal echocardiography (**Figure 2**, Video 2) obtained 5 days later demonstrated restricted mitral valve leaflets with poor leaflet coaptation. Mitral valve chord rupture was noted.

DIFFERENTIAL DIAGNOSIS

The constellation of heart failure, fever, murmur, and echo findings raised concern for infective endocarditis (IE). Despite meeting modified Duke criteria for definite IE, the patient did not defervesce on empiric antibiotics and cultures remained negative. Myocarditis or myopericarditis were considered as was subvalvular apparatus pathology. Systemic lupus erythematosus (SLE) with nephritis and Libman-Sacks endocarditis (LSE) were likewise considered in context of fever, arthralgia, and foamy urine. Recent streptococcal pharyngitis raised consideration of rheumatic fever and rheumatic valvular disease.

MANAGEMENT

Noninvasive positive pressure ventilation was started concomitantly with intravenous furosemide. Cultures were obtained and empiric vancomycin and ceftriaxone were started. Three days after transesophageal echocardiography, the patient underwent mitral valve replacement for cardiogenic shock (size 27-mm Epic biological valve), aortic valve repair (left coronary cusp-noncoronary cusp commissural closure), and tricuspid valve repair (posterior DeVega annuloplasty). Intraoperatively, there was an inflamed mitral valve with a retracted posterior leaflet, mitral valve chord rupture, and subvalvular destruction. The aortic valve demonstrated diffuse thickening and loss of integrity of the left coronary cusp and noncoronary cusp. Pericardial adhesions were noted.

ABBREVIATIONS AND ACRONYMS

APLS = antiphospholipid syndrome

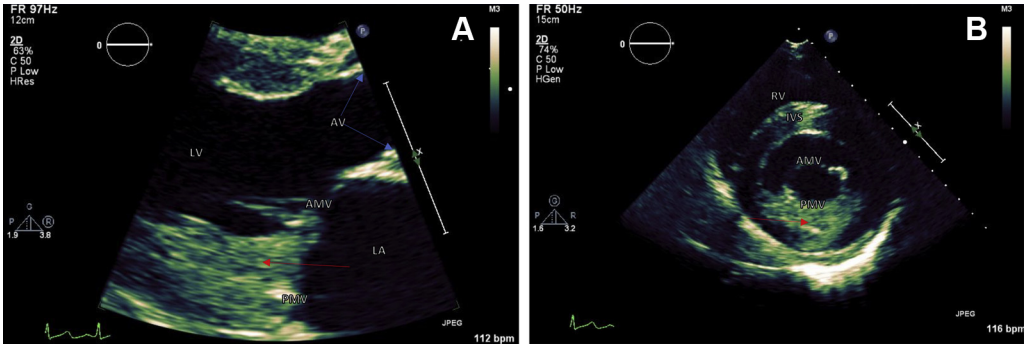
HACEK = *Haemophilus, Aggregatibacter, Cardiobacterium, Eikenella, Kingella*

IE = infective endocarditis

LSE = Libman-Sacks endocarditis

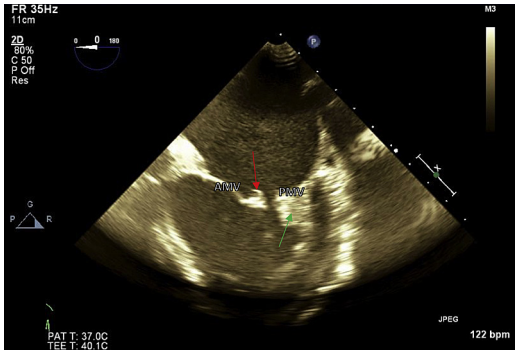
SLE = systemic lupus erythematosus

FIGURE 1 Transthoracic Echocardiography of the Mitral Valve



(A) Parasternal long-axis view demonstrating a thickened posterior mitral valve leaflet and a collection on the ventricular surface of the posterior leaflet **(red arrow)**. Aortic valve, **blue arrows**. **(B)** Parasternal short axis at the level of the mitral valve, demonstrating leaflet thickening and perivalvular collection **(red arrow)**. AMV = anterior mitral valve; AV = aortic valve; IVS = interventricular septum; LA = left atrium; LV = left ventricle; PMV = posterior mitral valve; RV = right ventricle.

FIGURE 2 Transesophageal Echocardiography of the Mitral Valve



Ruptured mitral valve chord **(red arrow)**, poor leaflet coaptation, and posterior subvalvular thickening on the ventricular surface of the posterior mitral valve leaflet **(green arrow)**. Abbreviations as in Figure 1.

Renal biopsy demonstrated diffuse proliferative lupus nephritis and thrombotic microangiopathy. Antibiotics were discontinued because cultures and tissue were negative for infectious organisms. Aspirin and immunosuppression were initiated, followed by coumadin on discharge.

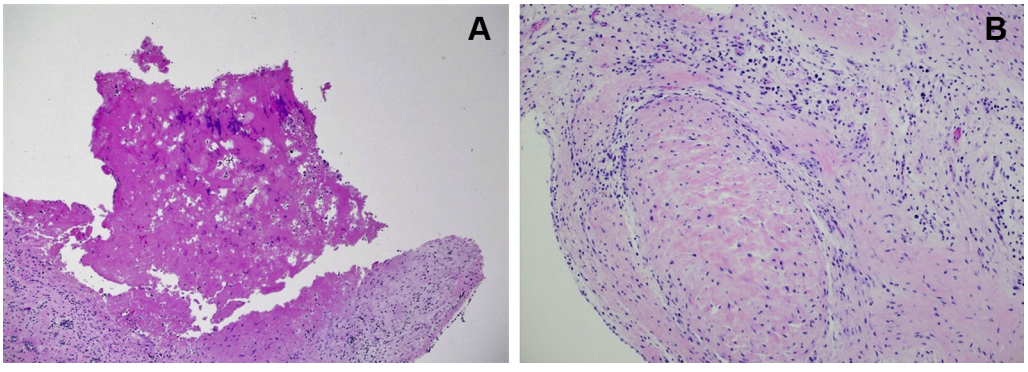
DISCUSSION

This case demonstrates a dramatic presentation of LSE manifesting as cardiogenic shock because of acute valvular insufficiency from mitral valve chord rupture with poor leaflet coaptation in the context of concomitant aortic valve destruction. LSE is a form of non-IE, also known as marantic, verrucous, or nonbacterial thrombotic endocarditis, and is the result of sterile fibrin and platelet accumulation involving the cardiac valves, chordae tendineae, or endocardium. Endothelial injury is thought to be the inciting pathophysiologic insult resulting in platelet deposition and migration of inflammatory cells to exposed subendothelial connective tissue (1). LSE must be differentiated from IE. This patient met criteria for "definite" IE by modified Duke criteria, which is approximately 80% sensitive and specific (2). The persistent fever, negative cultures, and failure to improve with appropriate empiric antibiotics favored a noninfective etiology. Antibiotics were discontinued on clinical improvement after surgery.

LSE is associated with SLE and APLS, yet has been described in APLS without SLE and with malignancies (3). LSE is most often found postmortem, with incident autopsy series findings ranging from 0.9% to 1.6%. Rates are estimated to be higher in patients with underlying malignancy, particularly with adenocarcinomas. LSE most frequently involves the aortic and mitral valves (4,5).

LSE frequently presents with thromboembolism, and up to 43% of patients with SLE demonstrate valvular vegetations on initial echocardiographic evaluation. Vegetations are associated with increased mortality and may involve isolated basal, middle, or tip portions of leaflets on both valve surfaces (Figure 3); however, diffuse thickening of the aortic valve cusps or mitral valve leaflets is a distinctive finding in these patients (1,6). Valvular disease does not relate temporally to the clinical features of lupus, and coexisting infective and Libman-Sacks

FIGURE 3 Mitral Valve Surgical Pathology



(A) Atrial side of the mitral valve showing a sterile vegetation fragment primarily composed of fibrin-like material and lacking acute inflammation or conspicuous microorganisms (hematoxylin & eosin, original magnification ×40). **(B)** Ventricular side of the mitral valve showing a chronic, mononuclear inflammatory infiltrate percolating through the stroma between muscular insertions of the chordae tendinae (hematoxylin & eosin, original magnification ×100).

vegetations have been described. Valvular regurgitation is not usually progressive and valve stenosis is unlikely to develop (7).

Management of LSE focuses on treatment of the underlying disease and anticoagulation for thromboembolic events. Surgery is reserved for patients with symptomatic disease or recurrent thromboembolism (8). Guideline recommendations for antithrombotic therapy in patients with LSE without evidence of thromboembolism or concomitant APLS are limited, with a Class IIc recommendation in patients with signs of systemic or pulmonary emboli (9). This patient had evidence of thrombotic microangiopathy and was therefore anticoagulated in the absence of APLS. A bioprosthetic valve was chosen over a mechanical mitral valve because of concern regarding compliance with anticoagulation.

FOLLOW-UP

The patient was maintained on coumadin, prednisone, mycophenolate mofetil, and hydroxychloroquine. Cardiovascular symptoms resolved after surgery. The blood pressure on discharge was 140/96 mm Hg and heart rate was 62 beats/min.

CONCLUSIONS

LSE presenting as acute heart failure and hemodynamic decompensation with mitral valve chord rupture is rare. LSE can mimic IE and diagnosing this noninfectious etiology is paramount to guiding further management.

AUTHOR RELATIONSHIP WITH INDUSTRY

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS aortic valve, endocarditis, mitral valve

APPENDIX For supplemental videos, please see the online version of this paper.