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A Woman with Systemic Lupus Erythematosus and Odd Valvular Presentation: A Case Report

Authors' Contribution:
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Data Collection B
Statistical Analysis C
Data Interpretation D
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Patient: Female, 35
Final Diagnosis: Lupus induced valvular stenosis
Symptoms: Fever
Medication: —
Clinical Procedure: Cardiac surgery
Specialty: Cardiology

Objective: Unusual clinical course

Background: Systemic lupus erythematosus (SLE) is a systemic disease with various cardiac and non-cardiac presentations. We present the case of a young woman with odd presentation of SLE mistakenly identified as a valve abscess that was scheduled for surgery.

Case Report: This 35-year-old woman presented with rapid progression of aortic stenosis, and the transesophageal echocardiography report showed a misdiagnosed aortic web (congenital) and aortic wall abscess. She was scheduled for surgery as a case of subacute bacterial endocarditis (SBE) and aortic abscess, despite lack of fever.

Conclusions: Cardiovascular involvement should be considered in any SLE patient, especially those with high SLE scores, even with negative antiphospholipid antibody. Cardiovascular involvement may be odd and misleading in some cases, which may warrant especial attention and experienced caregivers for clinical reasoning and proper management.

MeSH Keywords: Aortic Stenosis, Subvalvular • Cardiac Valve Annuloplasty • Endocarditis, Subacute Bacterial • Lupus Vasculitis, Central Nervous System

Abbreviations: **SLE** – systemic lupus erythematosus; **SBE** – subacute bacterial endocarditis; **AVR** – aortic valve replacement; **AS** – aortic stenosis; **TEE** – transesophageal echocardiography; **AR** – aortic regurgitation

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Background

Systemic lupus erythematosus (SLE) is one of the common autoimmune disorders which are typically reported more in women of reproductive age, with a prevalence rate of 50 patients among 100 000 people. This disorder has multiorgan involvement, including the skin, kidney, joints, central nervous system, and cardiopulmonary systems [1]. Among these clinical signs, cardiovascular manifestations are the most important because they have serious effects on mortality [2].

The most common cardiac presentation of SLE is pericarditis, with valvular involvement, myocarditis, and atherosclerosis reported less frequently. In recent research, atherosclerosis has been reported to be an important cause of death in younger patients [3].

Doppler echocardiography is used for detecting valvular complications, especially when it is performed transesophageally. SLE presentations include valve thickening and dysfunction, and bacterial and sterile vegetation. Some studies have focused on the relationship between anti-phospholipid antibodies and valvular manifestations, but their exact pathophysiologic pathway is unclear [4].

The most common primary valvular involvement is mitral regurgitation, while pulmonary artery hypertension usually results in involvement of the tricuspid and pulmonary valves. These complications may need surgical treatment, especially in severe cases when using corticosteroid or immunosuppressive agents does not improve the valvular lesions, or when infective endocarditis is suspected [4,5].

Libman-sacks endocarditis (LSE) is a well-known valvular disease in SLE. LSE was defined as a "non-infectious, verrucous, progressive endocarditis", which consists of fibrin and immune-complex depositions [4]. The most catastrophic manifestation is embolic events, especially to the brain. There is a strong relationship between antiphospholipid antibody elevation and LSE, and the best diagnostic tool is echocardiography, especially when performed transesophageally [4–6].

In this case presentation, a woman with SLE was evaluated for suspicious infective endocarditis and was misdiagnosed with subvalvular discrete aortic stenosis based on the echocardiography study, while the final diagnosis was inflammatory valvular involvement. To the best of our knowledge, this is the first such case report.

Case Report

A 35-year-old woman with known SLE and hypertension, was evaluated for valvular abnormality in the aorta root by transesophageal echo (TEE). The patient had had SLE for 11 years, and she started proper medical treatment approximately 3 years ago because of poor compliance in the early years of diagnosis. She had proteinuria due to lupus nephritis, which was controlled by immunosuppressive agents. She used prednisolone 15 mg daily, Cellcept (mycophenolate) 500 mg PO BID, bisoprolol 2.5 mg PO QD, and losartan 25 mg PO TID before admission, and ceftriaxone 1 g IV Q12h and Vancomycin 1 g IV Q12h started for her on the first admission day due to suspicious infective endocarditis.

She also had had mild aortic valve stenosis (AS) for 6 years, which had progressed to moderate stenosis in recent investigations. In the last follow up, echocardiography showed that the AS was severe and the patient was a candidate for cardiac valve surgery.

On the admission day, she had stable hemodynamic vital signs and was afebrile. In heart auscultation, a systolic III/IV murmur at the left sternal border with diastolic murmur at the right sternal intercostal space was detected. Physical examinations of the lung, abdomen, and nervous system were normal. The laboratory data did not show any complications or active disease, and evaluations for infective endocarditis, 3 blood cultures, and antiphospholipid antibody were negative, and the SLE disease activity index (SLEDAI) was 16 (Table 1).

Leukocytosis was found, but there was no thrombocytopenia, and creatinine showed no increase. In the three-dimensional echocardiography, severe subvalvular AS was reported. Thickening of the posterior aortic root (0.6×1.8 cm) was highly suggestive of aortic root abscess (Figure 1A, 1B). The tricuspid aortic valve with fused, and severely thickened leaflets associated with moderate AR (Aortic valve Regurgitation) were detected, strongly suggesting the presence of endocarditis. Additional imaging with PET-CT or angio-IRM was not available. The patient was sent to the OR as a case of possible misdiagnosed subaortic web and infective endocarditis and aortic stenosis, and the therapy proposed by the heart team was aortic valve replacement (AVR) and Bental.

However, it became clear during surgery that the patient had subaortic thickening of the aortic wall with protrusion to the aortic lumen, causing compression of the valve and stenosis. The surgeon decided to resect the subaortic stenosis and the thick tissue beneath the cusps and to repair the aortic valve without AVR or aortic root replacement. No evidence of subaortic web or congenital stenosis was seen during the operation. Although the aortic valve was involved, the main pathology was at the aortic wall, with a mass obstructing the aortic orifice and involving the aortic valve, and these findings did not match LSE. The consistency of the mass was much more matched to chronic inflammation and fibrosis without any

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Table 1. Laboratory findings of the patient.

Value	Measurement	Value	Measurement
WBC	11000/microliter	BUN	19 mg/dl
Hb	12.2 gr/dl	Creatinine	0.8 mg/dl
Plt	251×10^3/microliter	Na	136 mEq/L
INR	1.07	K	4 mmol/L
Blood culture (×3)	Negative	2ME test	Negative
VDRL	Negative	Wright	Negative
ESR	61 mm/h	CRP	32 mg/l
U/A	Proteinuria: Trace Bacteriuria: Negative	Anti-Ro	Negative
B2-glycoprotein (ELISA)	8 (Negative)	Anti-smith Ab	Negative
Anti-scl70	Negative	Anti-U1RNP	Negative
Anti-lupus anticoagulant	30 (20–39)	Antiphospholipid Ab	10 (Negative)
ANA test	15.9 ng/dl (higher than normal range: <10 ng/dl)	dsDNA antibody	21 ng/dl (normal range: <18 ng/dl)
C3	1.41 (0.8–1.6 mg/dl)	C4	0.34 (0.16–0.48 mg/dl)

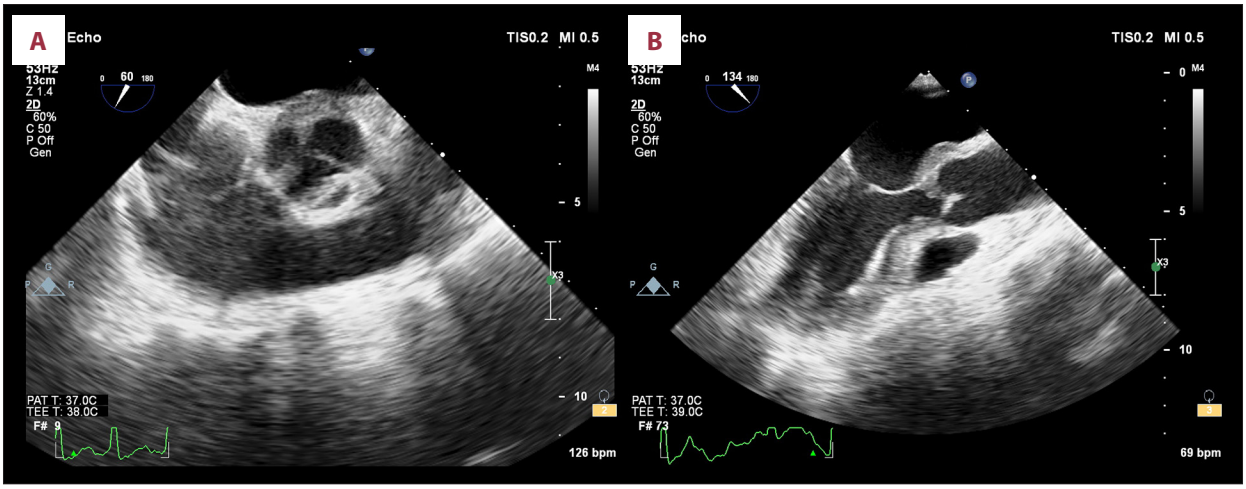

Figure 1. (**A, B**) Echocardiographic features of the patient, which were mistakenly identified as SBE.

evidence of abscess or infection. Finally, the specimen sent for pathology was reported to be chronic inflammation and fibrosis without any evidence of infective endocarditis. Microbiologic studies, including different cultures and staining, were negative.

After surgery, her leukocytosis was improved and she was put on IV hydrocortisone, after which her condition quickly stabilized, and she was then placed on her routine immunosuppressive therapies, and her medications were prednisolone 10 mg daily, Cellcept 500 mg PO BID, and Bisoprolol 2.5 mg PO QD.

Discussion

The most specific valvular involvement of SLE is Libman-Sacks endocarditis (LSE), which usually damages the left-side cardiac valves [7]. In a color Doppler echocardiographic investigation by Moyssakis et al. among 342 lupus patients, LSE was recognized in 38 cases (24 mitral, 13 aortic, and 1 tricuspid valve); 34% of the patients had aortic valve involvement, 11 had mild regurgitation, 8 had mild stenosis, and 1 had mild tricuspid regurgitation. They concluded that Libman-Sacks verrucous lesions were reported in 1 of 10 patients suffering

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from SLE, and the lesions were associated with disease duration, activity, and antiphospholipid Ab [8].

Kini et al. designed a cross-sectional investigation of cardiac involvement in SLE. An echocardiographic abnormality was noted in 50% of the patients. They concluded that screening echocardiography should be done, especially at presentation, disease flare-up, and in the presence of cardiac manifestations [9].

Chen et al. found that patients with SLE developed combined valvular changes and also reported an increase in the left atrial diameter and the left ventricular mass index. They concluded that SLE is related to significant changes in cardiac function [10].

To the best of our knowledge, this odd presentation of SLE as subaortic mass and aortitis has not been reported before. This should be considered in addition to the more common Libman-Sacks endocarditis, and properly diagnosed and taken into account before cardiac surgery, as they change the therapeutic strategy. However, there are few case reports of SLE-induced aortitis, which can be considered as an additional proof for this kind of presentation with SLE, in addition to the more common LSE [11].

The present case report and similar observations show that cardiovascular events, especially valvular involvement, can be a catastrophic complication of SLE. These lesions may be the leading cause of cardiac surgery and increase in mortality. Routine physical examination, proper follow up, and suitable treatment protocols can improve the diagnosis and decrease the morbidity and mortality of these patients.

In our case, it was important to first manage the life-threatening complications of endocarditis or severe valvular damage, and then to address the less important manifestations like Libman-Sacks endocarditis. SLE scores can predict cardiovascular involvement. Any patient with an SLE score of more than 10 should be evaluated for cardiovascular problems [1,10,12].

Conclusions

Cardiovascular involvement of SLE is very wide and may include aortitis and subacute aortic stenosis, with negative antiphospholipid antibodies. Cardiovascular presentations should be considered and diagnosed properly, especially in patients with high SLE disease scores. Valvular involvement of SLE patients are diverse; they sometime mimic SBE and can be misdiagnosed if not properly studied.

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Conflict of interest

None.

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