Mitral Valve Prosthesis Dehiscence with Severe Regurgitation and Pseudoaneurysm in a Young Woman with Recurrent Nonbacterial Thrombotic Endocarditis

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INTRODUCTION

Nonbacterial thrombotic endocarditis (NBTE) is a noninfectious cause of valvular vegetations. Although NBTE is most commonly associated with malignancy, it is also seen in patients with hypercoagulable states such as burns, sepsis, and hemolytic anemias and, most important, in autoimmune diseases such as systemic lupus erythematosus (SLE) and antiphospholipid syndrome (APS).[1] The exact pathogenesis of NBTE is not well understood but is thought to be secondary to endothelial damage, with subsequent deposition of platelets and inflammatory cells, ultimately forming thrombi mixed with strands of fibrin and immune complexes.[2] Surgical intervention is necessary in those with persistent severe valvular insufficiency and/or recurrent thromboembolic events. We report an unusual case of a young woman with SLE who presented in fulminant heart failure in the setting of NBTE with associated mitral valve prosthesis dehiscence, severe perivalvular regurgitation, and annular pseudoaneurysm whose clinical course was complicated by recurrent left atrial thrombus.

CASE PRESENTATION

A 31-year-old woman with a history of SLE, stage IV lupus nephritis, prior stroke, and intravenous drug use presented with cardiogenic shock and mental status changes. Her surgical history was significant for three prior mitral valve replacements over 1 year because of recurrent NBTE despite aggressive immunosuppression and anticoagulation. Examination demonstrated a drowsy patient with elevated jugular venous pressure and a prominent v wave. Her body temperature was 36.3°C, blood pressure 95/66 mm Hg, and heart rate 94 beats/min. Pertinent laboratory findings included hemoglobin of 10.3 g/dL, a white blood cell count of 18.9 $\times$ 10^9/L, a platelet count of 64 $\times$ 10^9/L, an international normalized ratio of 3.6, a creatinine level of 1.7 mg/dL, and drug studies positive for narcotics and cannabis. Results of antiphospholipid serology performed at her local facility were reported as normal. Her outside blood cultures remained negative, but they were collected after initiation of intravenous antibiotics for an initial diagnosis of pneumonia. Because of her history of drug abuse, immunosuppression for lupus, and presence of a prosthetic valve, infectious endocarditis was seriously considered. As such, the infectious disease team advised ongoing broad-spectrum intravenous antibiotics, blood cultures with extended cultures, and an extensive series of serologic studies assessing for *Coxiella, Legionella, Chlamydia, Bartonella, Brucella, Tropheryma whipplei,* human immunodeficiency virus, and hepatitis. Additionally, operative cultures were sent for Gram staining, anaerobic and aerobic cultures, fungal smear and culture, acid-fast smear, mycobacterial culture, pathologic examination, and, if indicated, 16s recombinant ribonucleic acid testing. Despite this comprehensive evaluation, no culprit organisms were identified.

Transthoracic echocardiography (TTE) showed a dehisced mitral valve prosthesis displaced to the mid atrium with severe mitral regurgitation (Videos 1 and 2). The intervalvular fibrosa was completely dehisced, with an associated cavity (Videos 3 and 4). There were two dominant jets of periprosthetic regurgitation (Videos 5 and 6). Three-dimensional imaging was used to better appreciate the defect (Videos 7 and 8). Preoperative transesophageal echocardiography (TEE) further elucidated the posterior annulus dehiscence, loculated cavity, and perivalvular regurgitation.

The patient and family were counseled regarding the high-risk nature of the planned procedure but decided to proceed because of the patient's poor clinical state, young age, and anticipated death without an attempted intervention. She underwent a fourth sternotomy. The mitral valve prosthesis was dehisced along the majority of the posterior annulus and sitting high in the left atrium, close to the pulmonary veins, with the true mitral annulus displaced apically (Figures 1 and 2). A large cavity at the intervalvular fibrosa with multiloculations was debrided; surgical pathology showed bland fibrin thrombus and fibrous tissue with mild chronic inflammation. It was negative for microorganisms consistent with NBTE. Given the patient's surgical history, severe destruction of the valves and the intervalvular fibrosa, the surgical team conducted an extensive reconstruction of the fibrous skeleton of the heart and atria. The mitral valve annulus was reconstructed integrating a 33-mm bioprosthesis in the anatomic mitral annular position. A large patch of bovine pericardium was then used to reconstruct the left atrial dome and the interatrial septal incision.

Postbypass TEE showed a well-seated mitral valve bioprosthesis with a mean gradient of 3 mm Hg and no regurgitation. The patient required extracorporeal membrane oxygenation in the setting of a left ventricular ejection fraction of 25% with sluggish flow in the left

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VIDEO HIGHLIGHTS

Video 1: Parasternal long-axis view on TTE demonstrating a prosthetic valve sewn into the left atrium above the valve annulus. The valve has notable rocking motion consistent with dehiscence.

Video 2: Parasternal long-axis view on TTE with color Doppler showing severe mitral regurgitation.

Video 3: TTE, parasternal at the level of the aortic valve, slightly off axis, demonstrating the mitral valve prosthesis dehiscence.

Video 4: Color Doppler at the level of the aortic valve, slightly off axis, on transthoracic echocardiographic parasternal imaging demonstrating the mitral valve prosthesis dehiscence. The mitral regurgitation is not as well visualized, because of the lack of jet alignment.

Video 5: Color Doppler at the apical long-axis window showing two dominant jets of periprosthetic regurgitation: one medial near the aortic-mitral junction and the second lateral.

Video 6: Color Doppler at the apical two-chamber view, slightly off axis, showing two dominant jets of periprosthetic regurgitation: one medial near the aortic-mitral junction and the second lateral.

Video 7: Three-dimensional color Doppler TTE demonstrating multiple jets of periprosthetic regurgitation at the site of dehiscence.

Video 8: Three-dimensional TTE demonstrating the prosthetic mitral valve dehiscence.

Video 9: Two-dimensional TEE of the long axis demonstrating poor mobility of the mitral valve and sluggish blood flow within the left atrium.

Video 10: Two-dimensional biplane TEE demonstrating thrombus formation in the left atrium adjacent to the mitral valve.

Video 11: Intraoperative TEE demonstrating sluggish flow and evolving thrombus in the left atrium.

Video 12: Intraoperative TEE at 94 degrees again showing the large, evolving thrombus in the left atrium.

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atrium (Video 9). In the immediate postoperative period, she required multiple blood product transfusions. At postoperative day 5, TEE in the intensive care unit showed layered thrombus in the left atrium adjacent to the mitral valve (Video 10). The patient underwent surgical clot removal; however, while in the operating room, the thrombus reaccumulated (Videos 11 and 12). Extracorporeal membrane oxygenation support was withdrawn after discussion with the family, and the patient died. Autopsy was declined.

DISCUSSION

SLE is an autoimmune disorder resulting in multiple-organ inflammatory damage. Cardiac manifestations of SLE have become more apparent in recent decades, with improvement in diagnostic techniques, specifically echocardiography. One of the most important cardiac manifestations is NBTE. First described in 1924 by Libman and Sacks,[3] NBTE's postmortem prevalence was 0.9% to 1.6%. However, prevalence can be as high as 6% to 11% on TTE and up to 43% on TEE in patients with SLE.[4] Although our patient reportedly had negative serologic findings for APS at her local facility, valvular abnormalities are seen up to 35% in SLE without APS (compared with 48% in SLE with APS).[5]

In the presence of a pseudoaneurysm in this patient with many risk factors, it is possible that a concurrent unidentified organism contributed to the severity of her situation. Unfortunately, broad-spectrum antibiotics did not lead to improvement, and her significant comorbidities including lupus, and blood dyscrasia, contributed to the many challenges encountered. However, an organism was not identified at any point in our patient's case or in her history upon review of outside records, and hence NBTE was the most likely diagnosis.

NBTE is often mild and asymptomatic.[6] However, serious complications might arise with fulminant presentations including thromboembolic events such as stroke and transient ischemic attack, superimposed bacterial endocarditis, and valvular regurgitation and/or stenosis.[7] Our case importantly highlights dehiscence and pseudoaneurysm as potential complications not otherwise reported in the literature to our knowledge. One may hypothesize that the formation of the pseudoaneurysm was secondary to prior surgical procedures or prior infectious endocarditis instead of NBTE; we think that this is certainly possible but could not identify an organism. Regardless, this complex case is informative regarding imaging approaches and management challenges.

As in our case, the most commonly affected valve in NBTE is the mitral valve, constituting >60% of reported cases, followed by the aortic valve with regurgitation.[8,9] Valvular surgery is required when there is severe symptomatic valvular dysfunction or recurrent thromboembolic events. Although data are limited, repair is generally considered the preferred method.[10]

Unfortunately, our patient had dehiscence of the mitral valve into the left atrium. Although this large defect was appreciated on TTE, TEE facilitated high-resolution interrogation of the mitral valve prosthesis dehiscence and perivalvular regurgitation and allowed thorough assessment of pseudoaneurysm. Color Doppler aided in assessment for channel communications. The three-dimensional imaging guided detailed assessment and mechanism of valve dehiscence and was critical for preoperative decision-making.

As seen in this patient, postoperative morbidity and mortality are predictably high, given the extensive destruction of valvular and cardiac tissues. Despite undergoing a technically successful procedure, her situation was complicated by recurrent left atrial thrombus in the setting of severe cardiac dysfunction, an abnormal left atrium, and a hypercoagulable state.

CONCLUSION

This case demonstrates an extreme presentation of NBTE. Several key aspects can be learned from this case. First, NBTE is a cardiac manifestation of SLE, and although mild in many cases, it may present with severe valvular and cardiac dysfunction. In our case, NBTE also presented with severe mitral valve dehiscence and pseudoaneurysm, although it is important to note that prior surgical procedures and/or infectious endocarditis (despite the lack of an identified organism) could have contributed to the severity of her presentation. Preoperative imaging with TTE was essential in understanding the underlying anatomy and developing a successful surgical plan. TEE was

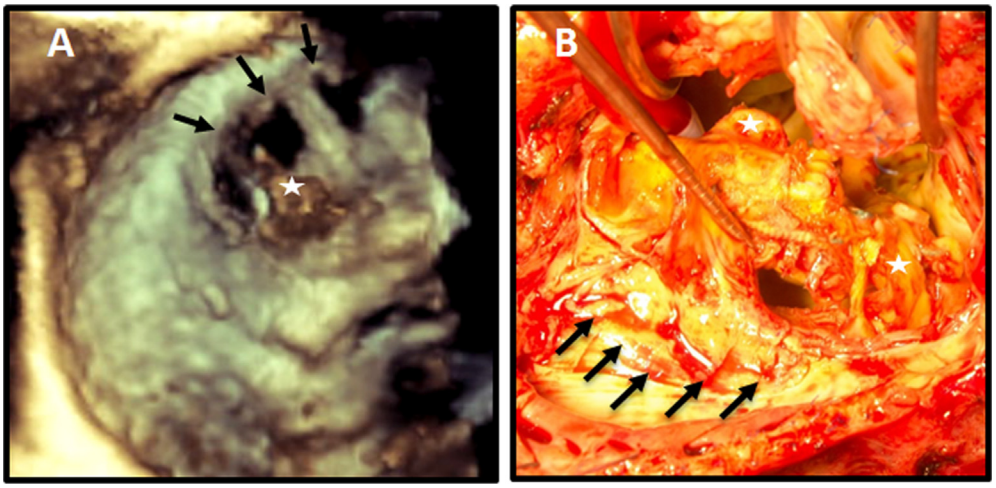

Figure 1 **(A)** Preoperative three-dimensional echocardiography demonstrating dehiscence of the mitral valve prosthesis (*arrows*) with thickening of the valve leaflets (*star*). **(B)** Intraoperative photograph showing dehiscence (*arrows*) along the majority of the posterior annulus of the mitral valve and multiple vegetations (*stars*) on the bioprosthetic valve leaflets.

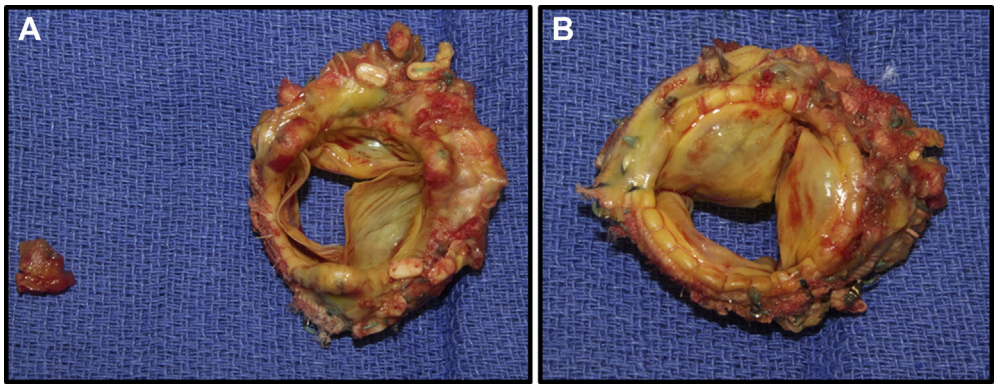

Figure 2 Postoperative photograph of explanted mitral valve prosthesis demonstrating leaflet destruction on the ventricular **(A)** and atrial **(B)** sides correlating to preoperative clinical evaluation and echocardiographic data.

particularly helpful for providing high-resolution images with color Doppler and three-dimensional imaging. Finally, TTE and TEE were useful to monitor for postoperative complications, which were predictably high in this patient.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2019.09.005.

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