Title

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Authors

Tabrizi, Roxana

Shamsa, Kamran

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CLINICAL VIGNETTE

Case Study: Systemic Lupus Erythematosus and Antiphospholipid Syndrome in Valvular Heart Disease

Roxana Tabrizi[1] and Kamran Shamsa[2], M.D.

[1]University of California, Los Angeles; Los Angeles, CA 90095
[2] University of California, Los Angeles; David Geffen School of Medicine; Department of Cardiology; 100 UCLA Medical Plaza, Suite 630 West; Los Angeles, CA 90095

Correspondence:
Kamran Shamsa, M.D.
100 UCLA Medical Plaza, Suite 630 West
Los Angeles, CA 90095
(310) 825- 9011

Background

Systemic lupus erythematosus (SLE) is an autoimmune disorder that is characterized by the production of autoantibodies and immune complexes that direct against a wide variety of a patient's self antigens and leads to inflammation and multi-organ damage.[1,2] The most frequent findings in patients with SLE are inflammation, immune complex deposition, and vasculopathies.[3] It is significant to note a high rate of cardiac involvement, which is estimated to be as high as 50% of SLE patients.[4] One of the most challenging cardiac manifestations of SLE is valvular involvement.[5] Based on transesophageal echocardiogram (TEE) studies, Fluture et al[6] reported that in patients with SLE, thickening of the heart valves is encountered more frequently (51-52%) than valve masses/vegetations (34-43%), with the mitral valve being the most commonly affected valve.[6,7] About 10% of SLE patients develop significant valvular disease, with need for valve surgery in many of these cases.[8] We present a complex case of mitral valve involvement in a patient with SLE and Antiphospholipid Syndrome (APS), and the choice between mechanical versus bioprosthetic valve replacement options.

Case Presentation

The patient has a significant past medical history of SLE and APS, with hypercoagulable state (history of deep venous thrombosis and pulmonary embolism), on chronic anticoagulation therapy. At the age of 20, she was found to have severe mitral regurgitation with a mass noted on her native mitral valve. She underwent mitral valve repair with annuloplasty and closure of a patent foramen ovale. The following year she experienced hemiparesis from an embolic cerebrovascular accident (CVA), which was attributed to a large mitral valve thrombus. She underwent mitral valve replacement (MVR) with a 27 mm Carpentier-Edwards bovine prosthesis. A bioprosthetic valve was chosen due to patient's high thrombotic risk and development of valvular thrombus while on therapeutic warfarin, making mechanical valve prosthesis less desirable. Due to treatment failure on warfarin, she was maintained on of daily injections of Fondaparinux.

She had done well for three years after bioprosthetic MVR but presents with symptoms of worsening shortness of breath with exertion. The patient underwent transthoracic echocardiogram that revealed severe prosthetic mitral valve stenosis with transmitral mean pressure gradient of 22 mmHg. For reference, mean gradient > 10 mmHg is considered severe mitral stenosis (Figure 1). TEE revealed decreased mobility of the bioprosthetic posterior leaflet and severe stenosis (Figure 2, Figure 3), as well as severe pulmonary hypertension with systolic pulmonary artery pressure of 57 mmHg). All findings were consistent with severe and symptomatic prosthetic mitral valve stenosis.

Given the early failure of the bioprosthetic valve, a mechanical valve was chosen for redo valve surgery. She underwent successful lateral thoracotomy with a 27 mm St. Jude Medical mechanical MVR. Intraoperatively, the bioprosthetic valve was noted to be severely stenotic with small orifice (Figure 4). Due to patient's hypercoagulable state and failure with warfarin therapy, treatment with Fondaparinux was continued with the addition of low dose Aspirin.

Discussion

This case highlights the effects of the combination of SLE and APS, resulting in severe valvular heart lesions, estimated 300% higher prevalence with degeneration and early failure of bioprosthetic valve.[9] In patients with SLE and APS, the presence of antibodies against cardiolipin can lead to the deposition of immunoglobulins and complement, causing an increase in inflammation and destruction of tissue, which is felt to be the cause of the degeneration of the valve (native or bioprosthetic).[10] The hypercoagulable state of the patient can cause deposition of fibrin and platelets on the valve with immune complex formation, leading to further damage and inflammation.[11-14]

For the general population needing mitral valve replacement, deciding on mechanical versus bioprosthetic valve is challenging. Kaneko et al[15] found the 10 year survival rate for MVR in a propensity matched group of patients under the age of 65 was 57.6% for bioprosthetic valves and 69.2% for mechanical valves. Although there are some cases of MVR in patients with SLE or APS, there are very few case reports discussing the group of patients with both SLE and APS, in which the outcome of MVR is significantly worse.[13,16-17] Our case highlights the many challenges faced in this patient population.

Conclusion

SLE is an autoimmune disease that affects every organ of the body, including the heart. The most significant cardiac involvement is the valvular dysfunction and degeneration due to the inflammatory response from SLE.[18] The prevalence of valvular dysfunction is far greater in patients with both SLE and APS, with significant portion of these patients eventually requiring valve replacement surgery. Both patients and physicians must work together to choose the optimal valve replacement—a bioprosthetic or mechanical valve. The occurrence of early valvulitis and premature degeneration of bioprosthetic valves must be weighed against the risk of mechanical prosthetics' higher risk of thrombosis in patients that are already hypercoagulable.[10] In patients with SLE (with or without APS), mechanical prosthetic valves may be the best option since they will minimize the need for repeat operations due to early failure of bioprosthetic valves brought on by heightened immune response.[18]

In the case presented here, the patient developed early failure of bioprosthetic valve requiring a repeat surgery with mechanical MVR thought to be due to her underlying disease of SLE and APS. Although initial consideration of the bioprosthetic valve seemed to be the safer option for a patient with a hypercoagulable state, a mechanical prosthetic valve in hindsight may have been a more suitable initial choice to withstand the immune complexes and inflammatory response in patients with SLE and APS that so often result in early bioprosthetic valve failure. In the future, it would be beneficial to compare the survival rates of patients with both SLE and APS that have undergone valvular replacement with mechanical versus bioprosthetic valves.

Figure Legends

Figure 1: Transthoracic echocardiogram with a mean pressure gradient of 22 mmHg

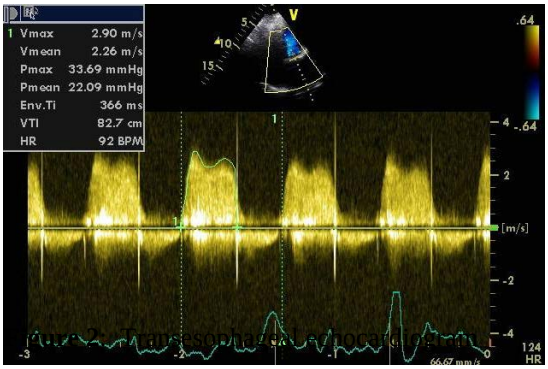


Figure 2: Transesophageal echocardiogram

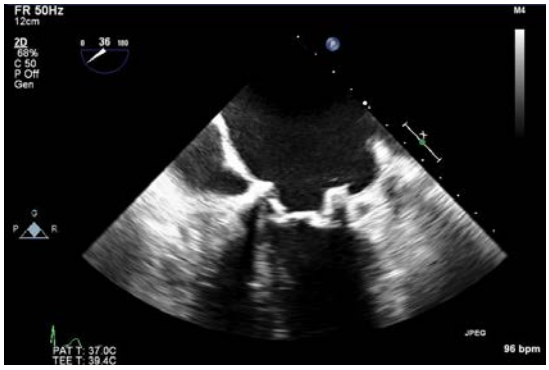


Figure 3: Transesophageal echocardiogram with color doppler

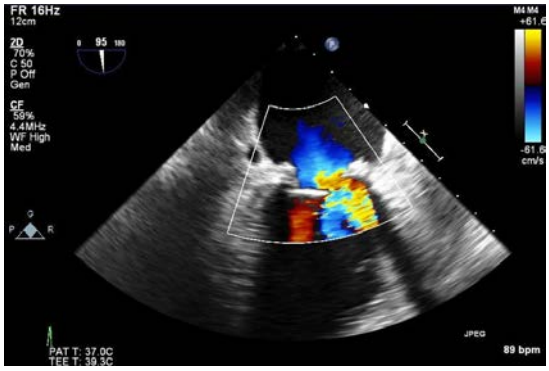


Figure 4A: Photograph of explanted stenotic mitral valve bioprosthesis (originally a 27 mm bovine mitral valve bioprosthesis), ventricular view

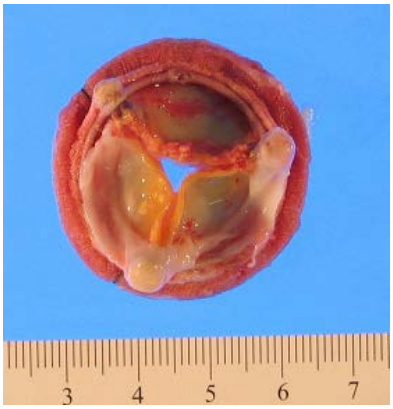
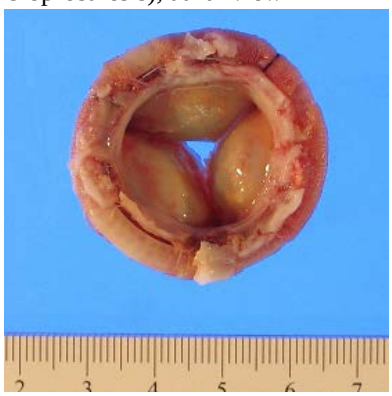


Figure 4B: Photograph of explanted stenotic mitral valve bioprosthesis (originally a 27 mm bovine mitral valve bioprosthesis), atrial view



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