An Extreme Case of Bioprosthetic Valve Thrombosis in a Patient With Systemic Lupus Erythematous and Antiphospholipid Antibody Syndrome

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A young woman presented with increasing fatigue, dyspnea, light-headedness, and intermittent chest pain. She had a history of systemic lupus erythematosus, antiphospholipid antibody syndrome, and Libman-Sacks endocarditis, resulting in a bioprosthetic aortic valve replacement 6 months before presentation. A bioprosthetic valve was chosen at that time over a mechanical valve because of inability to consistently maintain therapeutic anticoagulation levels with warfarin. Transthoracic echocardiography was performed and showed severe aortic valve prosthetic stenosis with a mean gradient of 69 mm Hg and thickened leaflets (Figure A) with reduced mobility, consistent with bioprosthetic valve thrombosis (BPVT). On review of echocardiographic studies, the immediate postimplantation gradient on intraoperative transesophageal echocardiography was 10 mm Hg. The mean gradient was 25 mm Hg on predischarge transthoracic echocardiography and 31 mm Hg at 2 weeks of follow-up. However, this increase in gradient was attributed to high flow, with the formal diagnosis of BPVT subsequently being made only 5 months later, implying significant diagnostic delay (Figure B). Aggressive anticoagulation with heparin followed by thrombolytic therapy was initiated and resulted in a decrease of gradients to a minimum level of 36 mm Hg. Given inability to maintain therapeutic levels with warfarin, dalteparin was continued. Despite continued anticoagulation, gradients started to rise again, and her symptoms persisted. The patient eventually underwent valve replacement with a mechanical prosthesis 12 months later. The explanted valve showed clear evidence of thrombus on both the aortic (Figure C) and ventricular (Figure D) sides of the prosthesis. It is important to have a high index of suspicion for BPVT in the context of increasing mean gradients, especially in patients at high risk, as closer surveillance may facilitate earlier diagnosis.

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FIGURE. An extreme case of bioprosthetic valve thrombosis (BPVT) in a patient with lupus and anti-phospholipid antibody syndrome. A, Transthoracic echocardiogram, parasternal long-axis view, shows thickened leaflets of aortic valve (arrows). B, Progression of mean gradient of bioprosthetic valve from 25 mm Hg before dismissal to 69 mm Hg on formal diagnosis of bioprosthetic valve thrombosis. Note the significant initial increase in gradient (31 mm Hg) at 2 weeks of follow-up. Clear evidence of thrombus on both the aortic (C) and ventricular (D) sides of the explanted prosthesis. LA, left atrium; LV, left ventricle; RV, right ventricle.