

Use of an On-X Prosthetic Valve In A 42-Year Old Female With Antiphospholipid Syndrome

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ABSTRACT

Antiphospholipid syndrome is a rare autoimmune disease with a hypercoagulable state causing vascular thrombosis. We present the case of a 42-year old female who underwent mitral valve replacement with a mechanical valve 15 months ago. The postoperative course was uneventful, and echocardiography performed 14 months postoperatively showed good valve function. The patient developed sudden dyspnea 15 months postoperatively and was referred to our hospital. Echocardiography revealed mitral stenosis with stuck leaflets. Emergent re-mitral valve replacement was successfully performed using an On-X valve (On-X Life Technologies, Austin, TX, USA). The patient tested positive for antiphospholipid antibodies. Antiphospholipid syndrome should be considered when valve dysfunction occurs suddenly in relatively young female patients. The On-X valve may be considered as a therapeutic option in patients with antiphospholipid syndrome because of its low anticoagulation intensity.

INTRODUCTION

Antiphospholipid antibody syndrome (APS) is an autoimmune disease that leads to thromboembolic events and pregnancy morbidity related to the presence of antiphospholipid antibodies [Miyakis 2006]. Herein, we report on a patient with APS who underwent successful re-mitral valve replacement with an On-X valve (On-X Life Technologies, Austin, TX, USA) after the initial mechanical valve became stuck due to rapid acute thrombus formation.

CASE REPORT

A 42-year old female underwent mitral valve replacement with a mechanical valve (25 mm; St. Jude Medical Inc., St. Paul, MN, USA) because of mitral valve regurgitation. Histological analysis of valve specimens revealed non-inflammatory lesions with fibrin deposits, vascular proliferation, and calcification without superficial thrombosis or vegetation. The patient had a history of stroke 7 months before the first

operation, but no history of pregnancy termination. The postoperative course was uneventful, and the patient was discharged with warfarin that was controlled at our outpatient clinic by a cardiologist at an international normalized ratio (INR) of 2.0-2.5, which was within the target ratio of Japanese guidelines for valvular heart disease (recommendation within 2.0-3.0 after mitral valve replacement). Echocardiography performed 14 months postoperatively showed normal valve function without severe mitral valve stenosis (mean pressure gradient was 5.5 mmHg). One month after the follow-up echocardiogram, the patient developed sudden dyspnea. Echocardiography revealed mitral stenosis with stuck leaflets; the mean pressure gradient was 28 mmHg. Blood pressure was 90/67 mmHg with sinus rhythm at admission; however, the patient's hemodynamic condition gradually worsened. The patient was then intubated, and we commenced percutaneous cardiopulmonary support and intra-aortic balloon pumping. Urgent mitral valve re-replacement was planned.

Median re-sternotomy was performed. The adhesion between the heart and pericardium was dissected, and cardiopulmonary bypass was performed with ascending aortic and bicaval venous cannulations. The aorta was clamped and cardiac arrest was obtained with cardioplegia. The mitral valve was observed via the superior trans-septal approach; the mechanical valve was thrombosed and both leaflets were stuck, but there was no pannus formation around it (Figure 1). The thrombosed valve was removed, and a 23 mm On-X valve (On-X Life Technologies) was implanted. Aortic cross-clamp

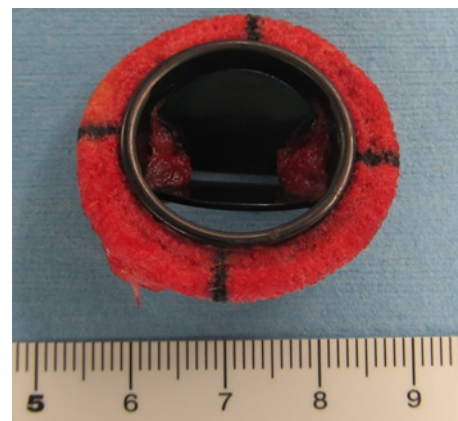


Figure 1. Mechanical valve with thrombus removed at the second operation.

Received March 26, 2017; received in revised form July 31, 2017; accepted August 19, 2017.

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time was 98 min and operation time was 262 min. The patient was hemodynamically stable intra- and post-operatively.

Intra-aortic balloon pumping was removed on postoperative day 1. Intravenous heparin administration that was started at surgery was replaced by warfarin after extubation. Hematological investigation revealed elevated serum anticardiolipin β 2-glycoprotein immunoglobulin G antibody three times. INR was maintained at around 2.5 and there have been no thromboembolic events in the 30 months after the second operation.

DISCUSSION

APS is characterized by venous or arterial thromboses and obstetric morbidity associated with persistent antiphospholipid antibodies [Miyakis 2006]. Heart valve disease reportedly occurs frequently in APS patients. A meta-analysis of 23 studies reported a three-fold higher risk for any heart valve lesion in systemic lupus erythematosus patients with antiphospholipid antibodies than in those without [Zuily 2011], and stroke and valvulopathy have been reported in up to 77.4% of APS patients [Erdogan 2005]. In the present case, the history of stroke and mitral valve regurgitation should have led to investigation of antiphospholipid antibodies at the first operation to perform strict postoperative control of anticoagulation.

Valve selection in APS patients is still controversial. Tissue valves are often used in APS patients due to their hypercoagulable state. However, in cases like the present patient, warfarin should be recommended because of the history of stroke even if a tissue valve had been used at the second operation. Moreover, there is a risk of immunologic deterioration of tissue valves in APS patients [Chan 2010]. If a tissue valve had been used, we believe that a third operation of mitral valve replacement would not have been avoided. We used an On-X

mechanical valve for the second surgery in the present case. The On-X valve has greater thromboresistance because of its all-carbon manufacturing and improved flow dynamics. A previous study reported that the 5-year freedom from major thromboembolism was $97.7 \pm 0.9\%$ in patients who underwent mitral valve replacement with an On-X valve, although INR was maintained at 2.0-3.0 [Karkar 2015]. We believe that another type of mechanical valve should be used at the second operation. There is no evidence related to the safety of the On-X valve in patients with hypercoagulability. However, the On-X valve is a potential option in APS patients undergoing mitral valve replacement.

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