Successful Replacement of Mitral Valve in a Patient with Antiphospholipid Syndrome

Selami Gürkan[1], Özcan Gür[1], Demet Özkaramanlı Gür[2], Cavidan Arar[3], Turan Ege[4]

[1] Namık Kemal University Faculty of Medicine, Department of Cardiovascular Surgery, Tekirdağ, Turkey
[2] Tekirdağ State Hospital, Clinic of Cardiology, Tekirdağ, Turkey
[3] Namık Kemal University Faculty of Medicine, Department of Anesthesiology and Reanimation, Tekirdağ, Turkey
[4] Trakya University Faculty of Medicine, Cardiovascular Surgery, Edirne, Turkey

ABSTRACT

We report a 51-year-old patient who was diagnosed with mitral insufficiency and was admitted to our hospital for surgical treatment. The patient was diagnosed with primary antiphospholipid syndrome (PAPS) before 4 years and had a history of thrombocytopenia. Mitral valve replacement was successfully performed with postoperative strict anticoagulant therapy, and the postoperative course was uneventful. Cardiac surgery in APS patients has been associated with high morbidity and mortality. Therefore, it is very important to initiate anticoagulant therapy immediately after the operation to prevent thrombosis.

Key Words: Antiphospholipid syndrome; mitral valve; thrombocytopenia

Antifosfolipid Sendromlu Bir Hastada Başarılı Mitral Kapak Replasmanı

ÖZET

Elli bir yaşında mitral yetmezlik tanısı ile cerrahi tedavi için yatırılan bir hastayı sunduk. Dört yıl önce primer antifosfolipid tanısı almış ve trombositopeni hikayesi mevcut. Postoperatif titiz antikoagülan tedavi ile mitral kapak başarılı bir şekilde replase edildi ve postoperatif dönem olaysız geçti. Antifosfolipid sendromlu hastalarda kardiyak cerrahide yüksek mortalite ve morbidite oranları bildirilmiştir. Postoperatif dönemde hemen antikoagülan tedavinin başlanması trombozun önlenmesi açısından çok önemlidir.

Anahtar Kelimeler: Antifosfolipid sendromu; mitral kapak; trombositopeni

INTRODUCTION

Primary antiphospholipid syndrome (PAPS) is defined by the presence of antiphospholipid antibodies, venous or arterial thrombosis, recurrent fetal abortion, and thrombocytopenia in the absence of systemic lupus erythematosus (SLE) or any other disease. In fact, these antibodies have been found in approximately 5% of the healthy population[1]. Various studies have described a valvulopathy incidence rate of 35%-82% using echocardiography in this population[2,3]. The most frequent valvular pathology includes an irregular thickening of the valve leaflets from the deposition of immune complexes. This process can affect both the mitral and aortic valves[4,5].

CASE REPORT

A 51 year old woman was referred for the surgical treatment of severe rheumatic mitral valve regurgitation. She had been suffering from dyspnea for 2 years. PAPS was first diagnosed before 4 years, with a clinical evidence of thrombocytopenia and increased serum levels of anticardiolipin antibodies. She was treated with diuretics and vasodilators for the past 2 years. The patient also had a 2-year history of chronic obstructive pulmonary disease and atrial fibrillation; however, there was no history of deep venous thrombosis or pulmonary embolism.

Recently, she began to suffer from severe exertional dyspnea and orthopnea. Echocardiogram revealed a reduced left ventricular function (EF= 40%), severe left ventricular hypertrophy, fibrotic mitral leaflets with a dilated left atrium, and severe mitral regurgitation. Coronary angiography revealed normal results. Respiratory function analyses revealed chronic obstructive pulmonary disease.

The blood laboratory results were as follows: hemoglobin, 12.9 g/dL; hematocrit, 40.2%; platelet, 53.000/mm^3; prothrombin time, 11.8 s; international normalized ratio (INR), 1.08;

Correspondence

Selami Gürkan

E-mail: gurkancem@yahoo.com
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activated partial thromboplastin time, 49.6 s; ESR, 28 mm/h; fibrinogen, 510 mg/dL (normal 200-400); lactate dehydrogenase, 309 IU/L (normal 100-190); and CRP, 18.64 mg/dL (normal= 5). Antiphospholipid IgG antibody levels were 140.73 GPL-U/m (normal range 15-20). Serum electrophoresis revealed the following: alpha-1, 5.06 g/L (normal range 2%-5%); alpha-2, 15.09 g/L (normal range 8%-13%); beta, 10.04 g/L (normal range 7%-14%); gamma, 23.69 g/L (normal range 12%-19%); and albumin, 46.13 g/L (normal range 50%-60%).

After consultation with a hematologist, she was treated with 20 mg methyl prednisolone for 2 weeks. Platelet count before the operation was 110.000/mm^3.

Mitral valve replacement was done through a conventional median sternotomy with mild hypothermic cardiopulmonary bypass (CPB) using antegrade blood cardioplegia in a standard fashion. Heparin was used as per standard protocol during CPB, and activated clotting time was kept over 400 s and measured every 30 min. A 29 mm St-Jude mitral valve was implanted. Ekstracorporeal circulation was discontinued after 95 min with an aortic-cross clamp time of 86 min. Heparin (150 mg) was notralized with 200 mg protamine sulfate. Despite adequate hemostasis, 3 U of packed RBC, 3 U of fresh frozen plasma, and 3 U of platelets were transfused. The platelet count was 110.000/mm^3 at the skin incision and 84.000/mm^3 at discharge.

The patient was transported to the intensive care unit, and hematologic consult recommended post-CABG care with regard to anticoagulation. This consisted of oral warfarin, 20 mg of methyl prednisolone, and tinzaparin sodium that was started on postoperative day (POD) 1. Chest tube drainage produced 750 mL and removed on POD 2. No hemostatic and thromboembolic problems were encountered after the surgery, and her postoperative course was uneventful. She was discharged on POD 12 and was prescribed 7.5 mg oral warfarin and 20 mg methyl prednisolone per day.

DISCUSSION

The recently revised Sapporo criteria require thrombosis-related manifestations and laboratory evidence of APL antibodies[6]. Cardiac surgical patients with APS are a high-risk group. In all patients with APS, the risk of thrombotic complications needs primary management, and the other major management issue is the monitoring and maintenance of adequate anticoagulation during cardiac surgery. Several case reports have described intra- and postoperative mortality during valve replacement in patients with APS[2,7,8].

Despite all treatment strategies, postoperative thrombotic complications can occur in these patients. Thrombocytopenia, excessive anticoagulation, antiprothrombin anticors, and lack of coagulation factors can lead to postoperative bleeding[9].

Heparin reversal with protamine remains controversial. Reversal of protamine differs between centers. A recent study exposed 52% complete reversals and 38% one-half reversals with protamine through 91% responding centers[10]. In our case, we used this one-half reversal dose strategy.

Anticoagulation after cardiac surgery remains controversial. Thrombosis risk is decreased in patients with 3-3.5 INR rates when compared to those with 2-3 INR rates[11]. We kept the INR levels over 3 to prevent postoperative thrombosis. Aggressive anticoagulation can prevent complications, and the risk of high morbidity and mortality remains high. There have been no large studies with newer anticoagulants such as direct thrombin inhibitors (lepuridin, argatroban, etc.) and factor X-A inhibitors (fondoparinux).

A careful follow-up including the close monitoring of the anticoagulation therapy is mandatory in PAPS. It is very important to initiate anticoagulant therapy immediately after the operation to prevent thrombosis.

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