

## Cardiac Surgery in a Patient with Idiopathic Aplastic Anemia: A Case Report

Major surgery in a patient with pancytopenia might be associated with increased surgical risks, especially for bleeding and infection. A 66-yr-old man was admitted to the hospital due to shortness of breath. His dyspnea was classified by the New York Heart Association (NYHA) as functional class III. Prior to admission, he had a 5-yr history of medical management for idiopathic aplastic anemia. The severity of aplastic anemia of the patient was graded as non-severe aplastic anemia. Echocardiography revealed reduced left ventricular function and severe aortic valve regurgitation (grade IV) with left ventricular end diastolic dimension measuring 87 mm. Because of dyspnea and echocardiographically documented aortic valve insufficiency, the patient underwent elective aortic valve replacement. Although extracorporeal circulation for valve operations might be associated with aggravation of impaired blood cell function, the patient recovered from surgery uneventfully. Here, we report a successful cardiac surgery with extracorporeal cardiopulmonary bypass in a patient with severe aortic valve insufficiency and concomitant idiopathic aplastic anemia.

**Key Words :** Cardiac Surgical Procedures; Anemia, Aplastic

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### INTRODUCTION

Malignant hematologic disorders (multiple myeloma, myelodysplasia, chronic lymphocytic leukemia, and Hodgkin's and non-Hodgkin's lymphoma) and idiopathic aplastic anemia that occur in the population with an incidence of 0.5-5/100,000 increase the surgical operative risk due to coagulation defects, changes of blood viscosity, immunosuppression, and bone marrow insufficiency (1). Pancytopenia associated with idiopathic aplastic anemia may pose an increased risk for postoperative bleeding and infection (2). Here, we report a patient with severe aortic valve insufficiency undergoing bioprosthetic aortic valve replacement who suffered from idiopathic aplastic anemia.

### CASE REPORT

A 66-yr-old man was admitted to the hospital because of shortness of breath. His dyspnea was classified by the New York Heart Association (NYHA) as functional class III. The patient's history included treatment for severe aplastic anemia with anti-thymocyte globulin and prednisone for 5 yr prior to admission. Cyclosporin therapy for aplastic anemia was declined because of reimbursement issues. Chest reont-

genography showed cardiomegaly. Echocardiography revealed reduced left ventricular function and severe aortic valve regurgitation (grade IV) with left ventricular end diastolic dimension measuring 87 mm. Coronary angiography revealed no significant coronary artery stenosis. The presence of severe dyspnea and echocardiographically documented severe aortic valve insufficiency led us to schedule a surgery for elective aortic valve replacement.

On admission, laboratory testing showed leukocytopenia (white blood cells 1,900/ $\mu$ L), anemia (hemoglobin 6.7 mM/L), and thrombocytopenia (platelets 34,000/ $\mu$ L). The differential blood count revealed 42.9% neutrophils (50-70%), 42.3% lymphocytes (25-40%), 6.9% monocytes (2-8%), and 4.3% eosinophils (2-4%).

Preoperative preparation included two weeks of granulocyte colony-stimulating factor (Neupogen™ 300  $\mu$ g, Amgen<sup>c</sup>, subcutaneously three times a week), transfusion of four units of packed red blood cells (RBCs), and ten units of platelet concentrations (PCs) one day before the surgical procedure resulting in a hemoglobin level of 9.8 mM/L, a leukocyte level of 10,300/ $\mu$ L, and a platelet level of 120,000/ $\mu$ L preoperatively.

The patient received aortic valve replacement with a 21-mm bioprosthetic aortic valve (Carpentier-Edwards™, Edwards Lifesciences, Irvine, CA, U.S.A.) by a standard medi-

**Table 1.** Summary of perioperative laboratory findings

	Admission	POD 1	POD 2	POD 3	Discharge
WBC ( $\times 10^3/\mu\text{L}$ )	1.90	9.40	7.50	5.90	4.30
Hb (g/dL)	6.7	10.9	11.9	12.3	6.2
PLT ( $\times 10^3/\mu\text{L}$ )	34	113	48	46	34
PT (INR)	1.35	1.78	1.41	1.34	1.36

WBC, white blood cells; Hb, hemoglobin; PLT, platelets; PT, prothrombin time; INR, international normalization ratio; POD, postoperative day.

an sternotomy; 2 milrione units of aprotinin were added to the cardiopulmonary bypass circuit priming solution. The time to extubation was 17 hr. On the second day after surgery, atrial fibrillation deveolped and was successfully converted with intravenous amiodarone. Antibiotic prophylaxis was given using ceftriaxon (Rocephin™, Roche, Basel, Swiss) 2 g intravenously over 6 days. The total amount of drainage via a mediastinal chest tubes was 785 mL. The patient was discharged from the intensive care unit 4 days after surgery and was discharged from the hospital postoperative on day 11 with a leukocyte count 4,300/ $\mu\text{L}$ , hemoglobin of 6.2 mM/L, and a platelet count of 62,000/ $\mu\text{L}$ . After the surgery, the patient received 5 units of RBCs and 20 units of PCs until discharge. Perioperative laboratory findings are summarized in Table 1. During the 6 months of follow-up, the patient did well with a functional class of NYHA class I. The chest radiography at 3 months post surgery showed a decreased cardiac size.

## DISCUSSION

According to the definition of the severity of aplastic anemia (5), the patient's aplastic anemia could be categorized as non-severe aplastic anemia. Regardless of the severity of aplastic anemia, pancytopenia itself still poses an increased surgical risk.

As described by Christiansen et al. (1), two questions have to be considered when cardiac surgery is indicated in patients with a hematologic disorder causing pancytopenia.

Given the increased operative risk and the potential benefit with regard to the life expectancy and quality of life, aggressive surgical treatment has to be compared with conservative treatment and the natural course of both, the hematologic disorder and the cardiac disease. Our patient suffered from severe dyspnea on exertion; the patient had stable idiopathic aplastic anemia and was under hematological supervision. Progressive deterioration of ventricular function was documented on serial echocardiographic examinations. Both symptomatic aortic regurgitation and impaired left ventricular function led us to the decision to perform aortic valve replacement for this patient.

Another important issue for this specific subset of patients

is perioperative management to decrease morbidity and mortality (1). Idiopathic aplastic anemia is characterized by pancytopenia. Potential complications are caused by a decreased number or impaired function of blood cells; this condition is aggravated by the cell damaging properties of extracorporeal circulation. In addition to meticulous surgical hemostasis, substitution of various blood products is important to reduce the risk of bleeding complications. Total amount of the blood product transfused to this patient is regarded to be acceptable considering the higher risk of bleeding in these specific subset of patients.

Aprotinin has been shown to decrease blood loss and reduce the use of blood products by at least 30%, in open heart surgery (4). Thoracic drainage in our patient was 785 mL, which was similar to that of other patients who required cardiac surgery with a variety of malignant hematological disorders (1, 3).

Another important aspect of perioperative care in these patients is the increased risk for infections. It is of great importance to perform surgery in aseptic conditions and to avoid transmission of pathogenic microorganisms during the perioperative period. Our patient was discharged to the intensive care unit prepared for an immunocompromised patient. G-CSF was administered perioperatively for leukopenia as described previously (3). Furthermore, we did not observe any complication associated with G-CSF usage.

Anticoagulation therapy after bioprosthetic valve replacement is well established. The patient received warfarin sodium 2 days after surgery, and INR was maintained between 1.5 to 2.0. It was continued by 3 months after surgery. During follow up, we have noticed neither thrombotic nor hemorrhagic complications related to the anticoagulation therapy.

Finally, consistent with the report by Karsten et al. (3), cardiac surgery with extracorporeal circulation is feasible in patients with idiopathic aplastic anemia, and it was not associated with excessive complications that might be expected in a patient with this disorder.

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