Successful Replacement of Aortic Root with Valve-Sparing Technique and Proximal Arch in a Patient with Myelodysplastic Syndrome

Cardiac surgery in a patient with myelodysplastic syndrome (MDS) increases the risk of bleeding and infection. Here we report a case of a 70-year-old man with MDS who underwent successful replacement of the aortic root with the valve-sparing technique and proximal arch for aneurysmal dilatation from the aortic root to ascending aorta with moderate aortic valve regurgitation. Perioperatively, a transfusion of red blood cells and an infusion of a granulocyte colony-stimulating factor were required for his serious erythrocytopenia and leukocytopenia. Bleeding tendency was so severe that re-exploration to control postoperative surgical bleeding was performed and a large amount of blood cells were transfused. There was no infection on the postoperative course. Perioperative management for cardiac surgery in patients with MDS must be carefully programmed by a co-operative team consisting of cardiovascular surgeons and hematologists. (Jpn J Thorac Cardiovasc Surg 2003; 51: 322–325)

Key words: cardiac surgery, myelodysplastic syndrome, valve-sparing technique, granulocyte colony-stimulating factor

Tomomi Hasegawa, MD, Takuro Tsukube, MD, Tomonori Higuma, MD, and Yutaka Okita, MD.

Patients with hematologic disorders who require surgical correction are at risk of bleeding and infection. Cardiac operations requiring cardiopulmonary bypass are especially high risk because of the necessity of full heparinization and the destructive effects on all blood components. Myelodysplastic syndrome (MDS) is a group of hematologic disorders characterized by abnormal bone marrow and peripheral blood morphology, various and sometimes progressive cytopenias, functional abnormality of blood cells and a propensity for transformation into acute myelogenous leukemia.

In this case report, we describe a patient with serious erythrocytopenia and leukocytopenia caused by MDS, who underwent successful replacement of the aortic root with the valve-sparing technique and proximal arch.

Case

A 70-year-old man with a 50-year history of MDS was referred to our hospital for a cardiac operation because of annuloaortic ectasia and an aneurysm of the ascending aorta. He had been followed up with no medication for MDS because of asymptomatic progress. On admission, physical examination revealed a heart rate of 62/min, blood pressure of 130/62 mmHg and respiratory rate of 18/min. On cardiac auscultation, a to and fro murmur of Levine II/VI intensity was audible at the apex. Laboratory investigations showed the following results: red blood cell (RBC) count 141 ×10^4 per cubic millimeter (mm^3), hemoglobin 4.7 g/dl, hematocrit 15.3%, white blood cell count (WBC) 2,100/mm^3, platelet count 22.4×10^4/mm^3, 1% peripheral blood myeloblasts. Other hematologic parameters were normal including no bleeding diathesis. Results
of bone marrow aspiration revealed 1.6% myeloblasts, and less than 1% ringed sideroblasts. A chromosomal analysis showed karyotypic abnormalities: 46 X, -Y, +1, der(1; 7) (q10; p10) + mar1. These findings were compatible with a diagnosis of MDS with a subtype of refractory anemia. Echocardiography, computed tomography (CT), and aortography confirmed the aneurysmal dilatation from the aortic root to the ascending aorta and moderate aortic valve regurgitation (Fig. 1). Preoperatively, 12 units of RBC were transfused and 100 g of a granulocyte colony-stimulating factor (G-CSF) was administrated.

On December 22, 2000, the patient underwent an elective operation. After full anticoagulation with heparin, cardiopulmonary bypass (CPB) was established by cannulating the distal ascending aorta and the vena cava. The ascending aorta was cross-clamped, and myocardial protection was achieved with repetitive doses of cold blood cardioplegia in an antegrade and retrograde fashion. Annuloaortic ectasia was confirmed, however, the aortic cusps were essentially normal. The maximal diameter of the ascending aorta was 55mm and the size of the annulus was 24mm. Therefore aortic root replacement with a valve-sparing technique was performed. A 26-mm woven double-velour Dacron graft (Intergard Collagen Impregnated Woven Dacron Graft, Intervascular Inc., Tampa, FL) was chosen according to the formula suggested by David. The graft was sewn in a subaortic horizontal plane and aortic cusps were reimplanted into the graft using a running technique. Reanastomosis of coronary ostia to the graft was carried out in a button fashion. Deep hypothermia (17°C tympanic temperature) with circulatory arrest and retrograde cerebral perfusion were performed for replacement of the proximal arch using another 26-mm woven double-velour Dacron graft. The proximal end of the Dacron graft was anastomosed to the graft that was used for aortic root replacement by an end-to-end fashion. The aortic cross-clamp time was 174 min, circulatory arrest time 17 min, and total CPB time 237 min. After the end of CPB, the heparin was neutralized by intravenous administration of protamine sulfate to approximate an activated clotting time to 110–120 seconds of the baseline. The complete blood count at the end of CPB exhibited a hemoglobin of 10 g/dl, a WBC count of 3,100/mm³, and a platelet count of 13.6×10⁴/mm³. Despite adequate surgical hemostasis and pharmacological treatment, massive transfusion of RBCs, fresh frozen plasma (FFP), and units of platelet concentrate (PC) were given because of constant bleeding. The postoperative course was complicated with a cardiac tamponade, which was cor-

![Fig. 1.](image)