Case Report

Aortic Valve Replacement and Graft Replacement of the Ascending Aorta Using Deep Hypothermic Circulatory Arrest in a Patient with Myelodysplastic Syndrome

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A 73-year-old woman with a 10-year history of myelodysplastic syndrome (MDS) had severe aortic regurgitation (AR) and an ascending thoracic aortic aneurysm (TAA) with a maximum diameter of 55 mm. By retrograde cerebral perfusion (RCP) in the patient under deep hypothermic circulatory arrest (DHCA), we replaced the ascending aorta graft and aortic valve. After surgery, we periodically administered granulocyte colony-stimulating factor (GCSF) with platelet aggregation. On postoperative day 20, the patient had a duodenal ulcer. On postoperative day 22, she had a subarachnoid hemorrhage, which was treated, nonoperatively, with a hemostatic agent. On postoperative day 126, she was discharged without sequelae, and 1.5 years after the surgery, she has had neither heart failure nor deterioration of MDS.

Key words: heart surgery, myelodysplastic syndrome, aortic valve replacement, thoracic aortic aneurysm, deep hypothermic circulatory arrest

Introduction

Myelodysplastic syndrome (MDS) is a clinical condition characterized by pancytopenia caused by dysplasia of bone marrow. The patient with MDS has risks of bleeding and infection, and cardiac surgery using cardiopulmonary bypass (CPB) increases those risks. In this paper, we report a rare case of cardiac surgery in a patient with MDS requiring deep hypothermic circulatory arrest (DHCA), which has higher risk than ordinary CPB.

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Case

A 73-year-old woman with a 10-year history of MDS with subtype of refractory anemia with excess blasts (RAEB) had been treated with methenolone. In July 2007, she developed moderate exertional dyspnea (New York Heart Association (NYHA) Functional Capacity Class III) and was referred to the Department of Cardiovascular Surgery, Miki City Hospital, Hyogo, Japan. Physical examination revealed a Levine grade of 3/6 for "to and fro murmurs" at the left sternal border. Laboratory investigations revealed pancytopenia: white blood cell (WBC) count, 3700 /mm^3; red blood cells (RBC), 251 $\times$ 10^4/mm^3; Hb, 8.4g/dl; Ht, 22.3%; and platelets, 4.3 $\times$ 10^4/mm^3, and no peripheral blood myeloblasts. Other hematologic values were normal. Transthoracic echocardiography showed a LVDd/Ds of 61/46 mm, FS 25, LVEF 55%, grade 3/4 aortic, 2/4 mitral, 3/4 tricuspid regurgitation, and normal aortic annulus, 21 mm. (**Fig. 1**) Computed tomography (CT) showed a thoracic aortic

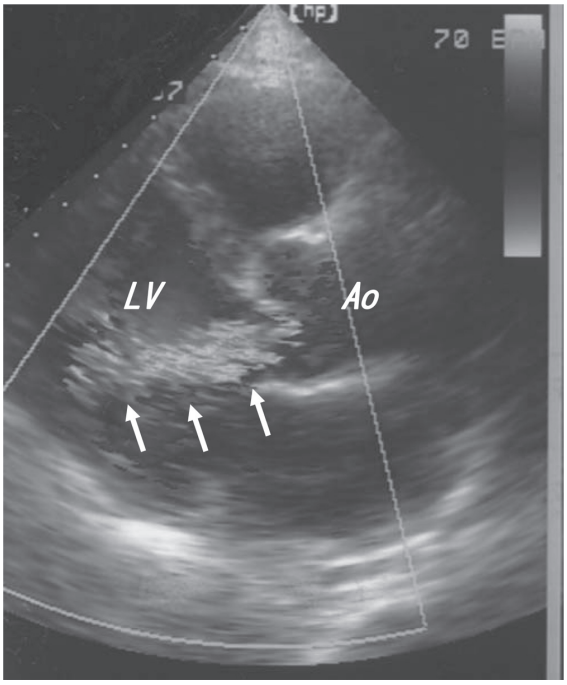

Fig. 1 Transthoracic echocardiography showing a tricuspid aortic valve with the grade 3/4 aortic regurgitation (arrows), normal aortic annulus with the 21mm.
Ao, aorta; LV, left ventricle

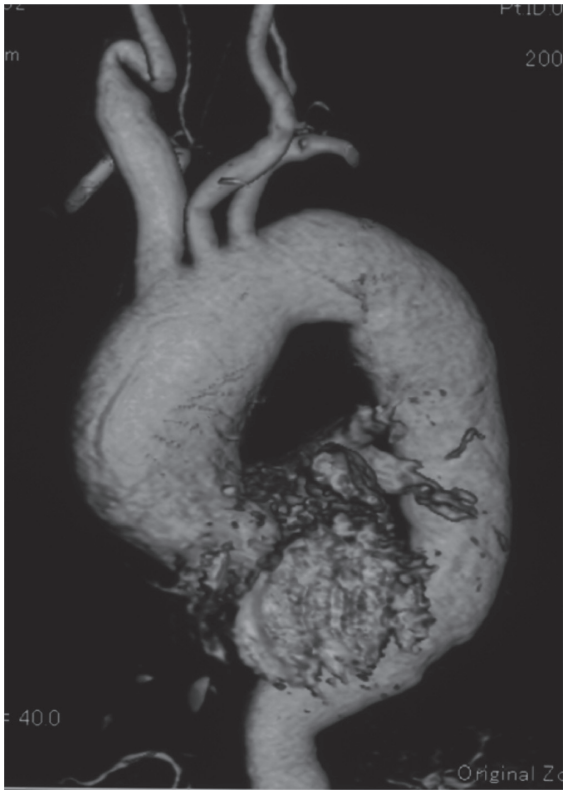

Fig. 2 Third dimension computed tomography (3DCT) showing a thoracic aortic aneurysm (TAA) from the sino tubular junction (STJ) to the proximal arch with a maximum diameter of 55 mm.

aneurysm (TAA) from the sino tubular junction (STJ) to the proximal arch with a maximum diameter of 55 mm (**Fig. 2**). Coronary angiography showed a normal coronary and aortography revealed a dilatation of the aorta and grade 3/4 aortic regurgitation. Cardiac catheterization revealed a right arterial pressure, 4/3 mm Hg (mean 2); a right ventricular pressure, 24/2 (4); a pulmonary artery pressure, 21/5 (11); and a pulmonary wedge pressure, 7/4 (12). One day before surgery, 100 μg of granulocyte colony-stimulating factor (GCSF) was administered. We planned a graft replacement of the ascending aorta and the aortic valve replacement.

The operation was performed with a standard CPB of an arterial cannula in the ascending aorta just proximal to the innominate artery and 2 venous cannulas in the superior and inferior vena cavae, using full anticoagulation with heparin. In the median sternotomy, we aspirated bone marrow. The ascending aortic aneurysm was dilated just proximal to the innominate artery at the diameter of 55 mm. Immediately after the cooling, we induced ventricular fibrillation (VF), cross-clamped the aneurysm that involved the ascending aorta, and delivered cold blood cardioplegia in a continuous retrograde fashion. After 11 minutes of cooling and the tympanic temperature reached 17.8 °C, we started DHCA and retrograde cerebral perfusion. We opened the ascending aorta in which the aneurysmal wall was not calcified. Distal anastomosis of the 28-mm Dacron prosthesis (Gelweave®, Sulzer Vascutek, Refrewshire, Scotland) were performed just proximal to the innominate artery. CPB was restarted after clamping of the graft and during rewarming. We identified the prolapse of noncoronary cusp on the tricuspid aortic valve and replaced aortic valve with 21-mm aortic bioprosthesis (Mosaic® valve; Medtronic, inc. Minneapolis, MN) in the supraannular position, anastomosed proximal aorta with the graft at the site of STJ, and performed tricuspid valve annuloplasty (DeVega procedure). She was easily weaned from cardiopulmonary bypass. The operative time was 194 minutes, cardiopulmonary bypass time, 144 minutes; the aortic cross clamp time, 111 minutes; and circulatory arrest time including RCP, 22 minutes. 14 units of RBC, 10, fresh, frozen plasma (FFP), and 40, platelet concentration (PC) were transfused, and surgical hemostasis could be easily

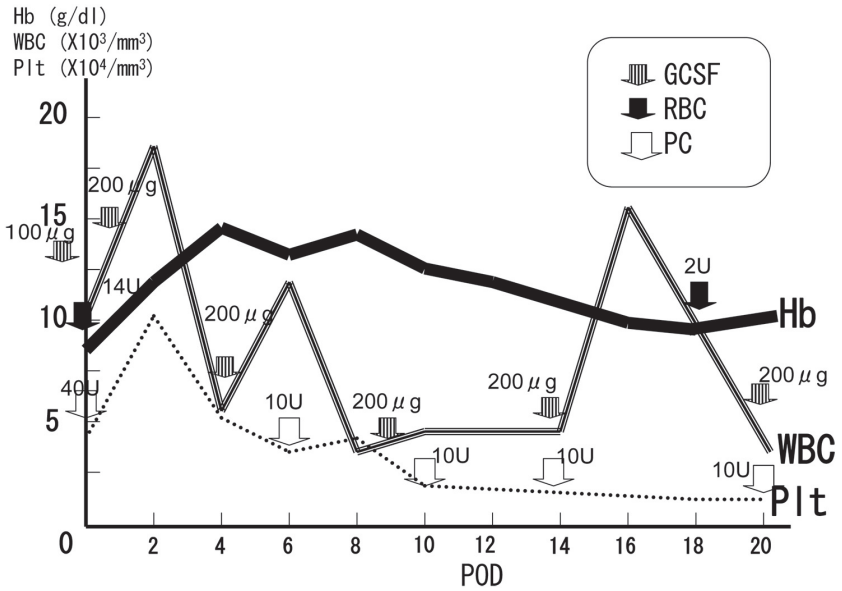

Fig. 3 Perioperative clinical course
Hb, hemoglobin; WBC, white blood cell; RBC, red blood cell transfusion; Plt, platelet transfusion; GSCF, granulocyte colony stimulating factor; POD, postoperative day

obtained with the aid of the usual pharmacological treatment.

Postoperatively, the pathology revealed no specific findings in the aneurysmal wall. The intraoperative marrow aspiration was compatible with the diagnosis with MDS with a subtype of RAEB, according to the FAB classification. After operation GCSF and platelet concentration were administered for several times (**Fig. 3**).

The postoperative course was eventful. On postoperative day 20 even with the postoperative medication of the proton pump inhibitor (PPI), the patient suffered from a duodenal ulcer. On postoperative day 22, the patient had a sudden, severe headache, which was diagnosed as a subarachnoid hemorrhage. Therefore, the patient was immediately transferred to the neurosurgery hospital for nonoperative treatment of bleeding with a hemostatic agent. On postoperative day 55, she recovered and was transferred to Miki City Hospital for rehabilitation. On postoperative day 126 she was discharged without sequelae. Now, 1.5 years after the operation, she is doing well and has had neither heart failure nor deterioration of MDS.

Discussion

MDS is a clonal hemopathy characterized by ineffective hematopoiesis cellular dysfunction.[1] MDS is classified by the French-American-British Cooperative Group into 5 subtypes: refractory anemia (RA), RA with ringed sideroblasts (RARS), RA with excess blast (RAEB), RAEB transformation (RAEBIT), chronic myelomonocytic leukemia (CMML).

It was difficult for us to decide whether surgery was indicated in this case because the first onset of MDS was 10 years ago. The life expectancy of patients with RAEB is not long, a 5-year survival rate of about 20%.[2, 3] The operation was relatively complex, since the patient had valve disease with thoracic aneurysm requiring DHCA. Even if the patient did survive the operation, the primary MDS might get worse. Although her MDS had been stable for these 10 years, heart failure was imminent. Therefore, we planned to operate so as to reduce the chance of her heart failing and thus extend her life.

About 10 cases of heart surgery in patients with MDS have been previously reported, and most were due to an isolated CABG or a single valve replacement.[4–6] Only 3 cases, including the case described here, were RAEB; the others were RA, with a better prognosis and a 5-year survival of about 50 %.[2, 3] Only one report[7] of a complex operation requiring DHCA, was for TAA of ascending and AR, where the bleeding tendency was so severe and re-exploration to control the surgical bleeding was performed. The risk of a bleeding complication was greater with DHCA and extended CPB than with ordinary CPB.

In most cases, the GCSF was administered continuously and was effective; however, in our case, the WBC count decreased sharply at the start, so we administered the GCSF periodically.

The general problems associated with MDS are bleeding and infection. Postoperative bleeding was controlled by keeping the PC above $2 \times 10^4/mm^3$ by PC transfusion; however, the patient did have cerebral bleeding It is commonly thought that a PC count of $1.5 \times 10^4/mm^3$ would initiate bleeding diathesis, though it is probable that during surgery bleeding diathesis can be initialed at a higher PC count. In this case, the perioperative bleeding tendency was worse than what we had expected. Because we immediately found the subarachnoid bleeding and transferred her to the neurosurgery hospital, she could recover completely. As for the next problem of the infection, we could overcome it with the adequate use of antibiotics. However, the patient also had a duodenal ulcer which required treatment with a PPI or H_2 blocker, and after she had taken the PPI the pancytopenia worsened. Fortunately, we could finally treat the ulcer with rebamipide (Mucosta®).

Conclusion

With the careful management of several complications, we were able to perform cardiac surgery with DHCA safely in a high-risk patient with MDS.

References

1) Noel P, Solberg LA Jr. Myelodysplastic syndromes. Pathogenesis, diagnosis and treatment. Crit Rev Oncol Hematol 1992; **12**: 193–215.
2) Lee JH, Lee JH, Shin YR, Lee JS, Kim WK, et al. Application of different prognostic scoring systems and comparison of the FAB and WHO classifications in Korean patients with myelodysplastic syndrome. Leukemia 2003; **17**: 305–13.
3) Malcovati L, Porta MG, Pascutto C, Invernizzi R, Boni M, et al. Prognostic factors and life expectancy in myelodysplastic syndromes classified according to WHO criteria: a basis for clinical decision making. J Clin Oncol 2005; **23**: 7594–603.
4) Miyagi Y, Yamauchi S, Suzuki S, Kitagawa A, Masaki Y, et al. Investigation of coronary artery bypass grafting for a patient with myelodysplastic syndrome. Ann Thorac Cardiovasc Surg 2001; **7**: 250–3.
5) Kurisu K, Ochiai Y, Kajiwara T, Tominaga R. Successful aortic valve replacement in severe pancytopenia related to myelodysplastic syndrome. Jpn J Thorac Cardiovasc Surg 2003; **51**: 77–80.
6) Tanaka K, Kanemitsu S, Tanaka J, Suzuki H, Kinoshita T. Successful replacement of aortic valve using a stentless porcine valve in a patient with myelodysplastic syndrome. Ann Thorac Cardiovasc Surg 2008; **14**: 123–5.
7) Hasegawa T, Tsukube T, Higuma T, Okita Y. Successful replacement of aortic root with valve-sparing technique and proximal arch in a patient with myelodysplastic syndrome. Jpn J Thorac Cardiovasc Surg 2003; **51**: 322–5.