Mitral Valve Repair in a Patient with Myelodysplastic Syndrome

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Open heart operations for patients with myelodysplastic syndrome (MDS) are associated with infective and bleeding complications. We report a 67-year-old woman with rheumatic, severe mitral regurgitation and mitral stenosis associated with MDS who underwent a mitral valve (MV) repair. Commissurotomy was performed in the anterior commissure. Autologous pericardial patch treated with glutaraldehyde solution was prepared. The anterior leaflet was completely detached from the posterior to the anterior commissure. The anterior leaflet was augmented by autologous pericardial patch treated with glutaraldehyde solution and three pairs of artificial chordae were implanted. Postoperative transesophageal echocardiography showed an increase in the MV orifice and less than trivial mitral regurgitation. Two years after the operation, the patient has normal sinus rhythm with no deterioration of the MV lesion by transthoracic echocardiography. Although the feasibility of MV repair is low in patients with restrictive pathology due to rheumatic disease, MV repair may be preferred in patients with MDS.

Keywords: mitral valve repair, rheumatic disease, myelodysplastic syndrome

Introduction

Myelodysplastic syndrome (MDS) is a set of oligoclonal disorders of hematopoietic stem cells characterized by ineffective hematopoiesis that manifests clinically as anemia, neutropenia and/or thrombocytopenia of varying severity. Patients with MDS have the risk of infection, hemorrhage and transformation due to acute myelogenous leukemia. The feasibility of mitral valve (MV) repair for rheumatic MV disease is low, but it may be a preferable surgical option to MV replacement in a patient with MDS because of potential risks of prosthetic valve endocarditis and severe hemorrhage associated with warfarin therapy.

Case Report

A 67-year-old female was admitted to our hospital with congestive heart failure. Three years before admission, she had been diagnosed as MDS with a subtype of refractory anemia by bone marrow examination in another hospital. Cyclosporine was administered for 2 years, which resulted in a partial remission, and afterwards, anabolic steroid (metenolone acetate) was administered. The electrocardiography revealed atrial fibrillation and a rapid ventricular response on admission. Echocardiography revealed a preserved left ventricular function (EF = 67%), left ventricular dilatation (end-diastolic dimension/end-systolic dimension = 56 mm/35 mm), severe calcification of posterior mitral leaflet and anterior commissure, severe mitral regurgitation (Fig. 1), mitral stenosis (mitral valve orifice area = 1.3 cm²), and severe tricuspid valve regurgitation (trans-valvular pressure...
gradient = 55mmHg). Coronary angiography was normal. Laboratory tests exhibited the following: leukocytes, 4600/μl, erythrocytes, 484 × 10^4/μl; hemoglobin, 11.9 g/dl; hematocrit, 35.9%; platelets, 30.7 × 10^4/μl; blast cells, 0%. Her International Prognostic score was low (score 0). She was scheduled for a Cox maze procedure with an MV operation.

After a median sternotomy, a large pericardial patch was harvested and treated with 1% glutaraldehyde solution for 1 minute. The MV was exposed with an incision just posterior to the interarterial groove. A modified Cox maze procedure was performed by a bipolar and monopolar radiofrequency device (Atricure, Century Medical Inc., Tokyo, Japan) prior to the MV procedure. The operative findings and procedures are demonstrated in Fig. 2. There was strong fusion and calcification in the anterior commissure. The anterior mitral leaflet (AML) was redundant on its rough zone, and its subvalvular

Fig. 1 Two distinct mitral regurgitation jets by apical four chamber view of trans-thoracic echocardiography.

Fig. 2 Operative findings: There was fusion on the anterior commissure and severe calcification on the P2/3 area with torn chordae (left side). Autologous pericardium was used for anterior mitral leaflet augmentation which was supported by 3 pairs of artificial chordae.
apparatus was fused. There was severe calcification and restriction in the P2 area and a quadrant leaflet resection was performed. Commissurotomy was performed in the anterior commissure. Size of annular ring prosthesis (Physio ring, Edwards Life-Sciences, Irvine, CA) was determined as 26mm according to the distance between two trigones. The pericardial patch was cut in the shape of a 26-mm prosthetic ring sizer. The AML was detached from the posterior to the anterior commissure, and a remnant of 2mm of leaflet tissue was left at the base of the AML. Chordal fusion and calcification of the AML were completely excised from inside and outside of the AML. The pericardial patch was fixed by a continuous 5-0 Prolene suture (AML augmentation). Implantation of three pairs of artificial chordae was used for the posteromedial and anterolateral papillary muscle as previously described. Tricuspid valve repair was performed using the Edwards MC3 annuloplasty ring (Edwards Life-Sciences, Irvine, CA). Postoperative transesophageal echocardiography revealed less than trivial mitral regurgitation and mild tricuspid regurgitation. The patient recovered to sinus rhythm. The postoperative course, including wound healing, was normal until the 7th postoperative day.

On the 8th postoperative day, the patient developed a fever (38°C) and leukocytosis with negative blood cultures. Mediastinitis was diagnosed by computed tomography, which showed fluid collection and air in the soft tissue. The patient underwent a resternotomy, debridement, and a substernal irrigation. Methicillin-resistant Staphylococcus epidermidis was isolated from the mediastinal fluid. The patient recovered and showed no clinical sign of re-infection after 14 days of intravenous vancomycin treatment. Postoperative transthoracic echocardiography revealed a preserved left ventricular function (EF = 61%), regression of left ventricular dimension (end-diastolic dimension/end-systolic dimension = 42 mm/29 mm), less than trivial mitral regurgitation, improved movement of the AML without mitral stenosis; pressure half-time = 130ms, peak/mean pressure gradient of the MV = 6.7 mmHg/3.3 mmHg, MV orifice area = 1.7 cm²), and mild tricuspid valve regurgitation (trans-valvular pressure gradient = 24 mmHg). The patient was discharged on the 30th postoperative day. On 2-year follow-up, the patient had clinically improved to New York Heart Association class 1, and she remained in sinus rhythm without deterioration of MV lesion by transthoracic echocardiography.

Discussion

The restrictive component of rheumatic mitral regurgitation remained a technically difficult lesion. Enlarging the AML by autologous pericardial patch and chordae replacement with ePTFE showed good durability. An autologous pericardium fixed with glutaraldehyde is widely used in MV repair for active infective endocarditis in our institute, and restriction or calcification has not been demonstrated by follow-up echocardiography. Combinations of chordae replacement, ring annuloplasty and AML augmentation may increase the feasibility of MV repair for rheumatic MV disease.

MDS is recognized as a preleukemic disorder. Refractory anemia, one of the five MDS subtypes, comprises about 25% of all patients with MDS. The median survival varies from years to a few months, and prognostic indicators are pancytopenia, serial cytogenic determination and an increase in the number of blasts.

Coagulation disorders, with or without thrombocytopenia, are common in patients with MDS. Thrombin-antithrombin complexes, which are associated with chronic coagulation activation, are higher in patients with MDS. Platelet dysfunction is common in patients with MDS, even in those with a normal platelet count. Risks of perioperative bleeding are associated with MDS with the use of extracorporeal circulation, and the patient had an apparent bleeding tendency during the operation, which required platelet substitution, in order to control coagulability.

Patients with MDS have high risks of infection during and after an open heart intervention. A high risk of wound infection after a cardiac operation in patients with hematologic malignancies has been reported previously. The present case also required reoperation due to mediastinitis caused by methicillin-resistant Staphylococcus epidermidis, 8 days after the operation. MV replacement with a biologic prosthesis was an alternative option, but patients with MDS may be at increased risk of developing prosthetic valve endocarditis. Dayyani et al. analyzed the cause of death (COD) in a cohort of 273 patients with low-risk MDS and demonstrated that the most common disease-related COD was infection (38%), transformation to acute myelogenous leukemia (15%) and hemorrhage (13%). A systematic review of the literature by Feringa et al. showed that MV repair is associated with lower, late recurrent endocarditis compared with a replacement (1.8% vs. 7.3%).

In conclusion, although the feasibility of MV repair is
low in patients with restrictive pathology due to rheumatic disease, MV repair may be preferred when patients have MDS.

References