Minimally invasive cardiac surgery for a young woman with Marfan syndrome and mitral regurgitation

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Abstract The case involved a 26-year-old woman with Marfan syndrome (MFS) and severe mitral valve regurgitation who hoped to bear a child. Anticipating future surgery to treat cardiovascular disease via a median sternotomy, we performed mitral annuloplasty via a right anterior thoracotomy. Mitral valve repair for mitral valve regurgitation via a right anterior thoracotomy is one of the most beneficial procedures for patients with MFS.

Key words Marfan syndrome · Mitral regurgitation · Minimally invasive cardiac surgery · Right thoracotomy

Introduction

Marfan syndrome (MFS) is the most common heritable connective tissue disorder with multisystem manifestations.1 The cardiovascular effects are life threatening. Currently, however, these patients have achieved prolonged survival thanks to advances in medication and surgical treatment.

The surgical procedure used to treat patients with MFS should be determined in light of future surgery to treat other cardiovascular disease and the patient’s quality of life after surgery. Described here is a case of a young woman with MFS and severe mitral regurgitation.

Case report

A 26-year-old woman with MFS and severe mitral regurgitation hoped to bear a child. Her height was 164.5 cm, her weight was 55 kg, and her body surface area (BSA) was 1.52 m². Her mother had undergone total arch replacement and abdominal aortic replacement and her brother had undergone total arch replacement. Transthoracic echocardiography examination revealed moderate to severe mitral regurgitation associated with annular dilatation (Fig. 1), an ejection fraction of 64%, and a sinus of Valsalva diameter of 43.5 mm. In this case, we were planning to perform mitral annuloplasty through a right anterior thoracotomy.

The patient was positioned with her right side elevated 40°. The right chest was opened through an anterior thoracotomy smaller than 10 cm in the fourth intercostal space. The lung was retracted inferiorly to expose the heart. The operative field was flooded with CO₂. The pericardium was opened 2 cm anterior to the phrenic nerve. A cardiopulmonary bypass (CPB) was established with venous drainage through the superior vena cava with a 34 Fr. cannula (INKC-R2-34; Toyobo, Japan) and the right femoral vein with a 34 Fr. cannula (VFEM018; Edwards Lifesciences) and arterial return through the right femoral artery with a 24 Fr. cannula (HK 46 SH-91 V-J; MAQUET, Germany). The venting catheter was inserted from the right superior pulmonary vein into the left atrium. After the ascending aorta was clamped with the Cosgrove Flex Clamp, a cold blood cardioplegic solution was infused at a rectal temperature...
of 30°C. Mitral annuloplasty was performed with an annuloplasty ring (Cosgrove, 30 mm) through the right side left atriotomy on the interatrial groove.

Weaning from CPB was uneventful. The total operating time was 245 min, with aortic clamp time of 33 min and CPB time of 73 min. The postoperative course was uneventful, and transthoracic echocardiography revealed no mitral insufficiency (Fig. 2).

**Discussion**

MFS is the most common connective tissue disorder with multisystem manifestations. Its cardiovascular manifestations include aneurysmal dilatation of the ascending aorta, aortic valvular regurgitation, aortic dissection, floppy mitral valve with mitral valve prolapse, mitral valvular regurgitation, and mitral annular dilatation. Mitral valve dysfunction occurs in 80% of patients, and 12.5% of patients (1 in 8) has moderate to severe mitral regurgitation by the age of 30.

In the current case, the surgical procedure had been selected in light of two considerations: (1) the patient hoped to bear a child (precluding use of warfarin); and (2) the patient had a high risk of additional surgery to treat aortic disease in the future.

Mitral valve surgery for a young woman who hopes to bear a child comes down to a decision between mitral valve replacement using a bioprosthetic valve or mitral valve repair. Recently, long-term outcomes of mitral valve repair for adults with MFS have been reported to be consistent. Thus, mitral valve repair was chosen in the current case.

In addition, women with MFS have an increased risk of aortic dissection as a result of pregnancy. In particular, patients with an aortic root 40 mm or larger, with a rapidly growing aortic root, or with dissection of the ascending aorta have a 10% probability of potential aortic dissection. In the current case, the sinus of Valsalva was dilated to 43.5 mm, and the patient was also deemed to have a high likelihood of additional surgery, given her family history, for problems such as aortic dissection. Thus, we have adopted a policy of performing mitral surgery via a right thoracotomy for patients facing future cardiovascular surgery with a median sternotomy, which makes future surgery safer and quicker, albeit only slightly. In addition, outcomes of mitral valve surgery via a right thoracotomy are consistent, and such outcomes are also considered achievable in patients with MFS.

Thus, mitral valve repair was performed in the current case via a right thoracotomy to treat mitral regurgitation in a young woman with MFS who hoped to become pregnant. This was considered the best treatment to provide a satisfactory quality of life and in light of the need for future surgery.

Patients with MFS have now achieved long-term survival thanks to improvements in medication and surgical techniques. Thus, formulating a treatment strategy considering the patient’s quality of life and need for additional surgery is crucial.

**Conclusion**

Mitral annuloplasty was performed via a right thoracotomy in a young woman with MFS who hoped to