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Aortic Dissecting Aneurysm with a Bicuspid Aortic Valve in Turner's Syndrome: Report of a Case

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Abstract
We herein report the performance of a successful, modified Bentall procedure and a total arch replacement for a Stanford type A chronic aortic dissection and a bicuspid aortic valve in Turner's syndrome (TS). The patient was a 45-year-old woman with 45, XO karyotype TS, who had had a history of hypertension since the age of 20. She had also been diagnosed as having a dilatation of the ascending aorta and a bicuspid aortic valve 3 years earlier. The patient became aware of back pain 6 months prior to the current admission, and was diagnosed as having a Stanford type A chronic aortic dissection and a bicuspid aortic valve with mild aortic regurgitation. One of the greatest concerns in TS is the risk of aortic dissection. Regarding the operation, aortic root replacement is one of the options for a bicuspid aortic valve so as to avoid high-risk surgical procedures in TS.

Key words Turner's syndrome · Cardiac anomaly · Bicuspid aortic valve · Aortic dissection

Introduction

Turner's syndrome (TS) or monosomy X results from a complete or partial monosomy in the X chromosome. This is a relatively common chromosomal disorder, affecting approximately 1 in 3000 live female births. It is well known that various cardiovascular disorders such as coarctation of the aorta, bicuspid aortic valve, and anomalous pulmonary venous return can be recognized in TS. Although rare, one of the greatest concerns in TS is the risk of aortic dissection, which affects 1%–2% of this population.[1,2] Recently, we treated a patient with TS exhibiting a chronic Stanford type A aortic dissection and a bicuspid aortic valve with mild aortic regurgitation (AR).

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Case Report

The patient was a 45-year-old woman with 45, XO karyotype TS diagnosed at age 15. She had had a history of hypertension since the age of 20 and dilatation of the ascending aorta (42 mm) as well as a bicuspid aortic valve that had been diagnosed 3 years earlier. The patient became aware of back pain 6 months before the current admission. However, she had not sought any examination prior to admission to another hospital. An X-ray examination showed a widened mediastinum, and she was diagnosed as having a Stanford type A chronic aortic dissection on chest computed tomography (CT).

The patient was 144 cm in height. Physical examination demonstrated a pigeon chest and infantile nipples. However, there was no finding of a webbed neck.

The chest and abdominal CT demonstrated a Stanford type A aortic dissection with a large entry from the ascending to proximal aortic arch (Fig. 1). A double-barrel aorta was demonstrated in the bilateral common iliac arteries. Neither coarctation of the aorta nor renal anomalies were detected. The diameter of the aortic annulus, sinus of Valsalva, sino-tubular junction, and ascending aorta was 20 mm, 31 mm, 31 mm, and 57 mm, respectively. Transthoracic echocardiography (TTE) demonstrated a bicuspid aortic valve with mild AR. Magnetic resonance angiography demonstrated a partial anomalous pulmonary vein return (PAPVR) (right upper pulmonary vein returned to the superior vena cava) (Fig. 2). The diagnosis was a chronic Stanford type A aortic dissection, a bicuspid aortic valve with mild AR, and PAPVR. Regarding PAPVR, no Q_p/Q_s measurement was performed.

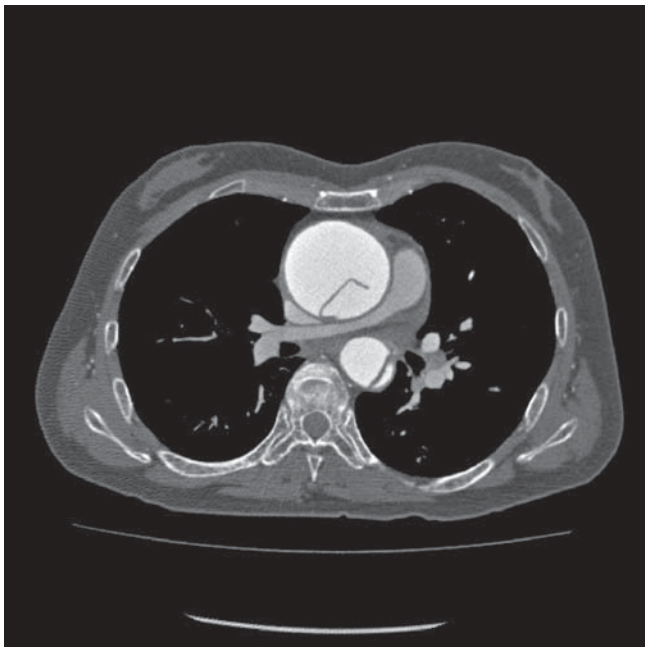

Fig. 1. Preoperative computed tomography showed a Stanford type A chronic aortic dissection with a large entry in the ascending aorta

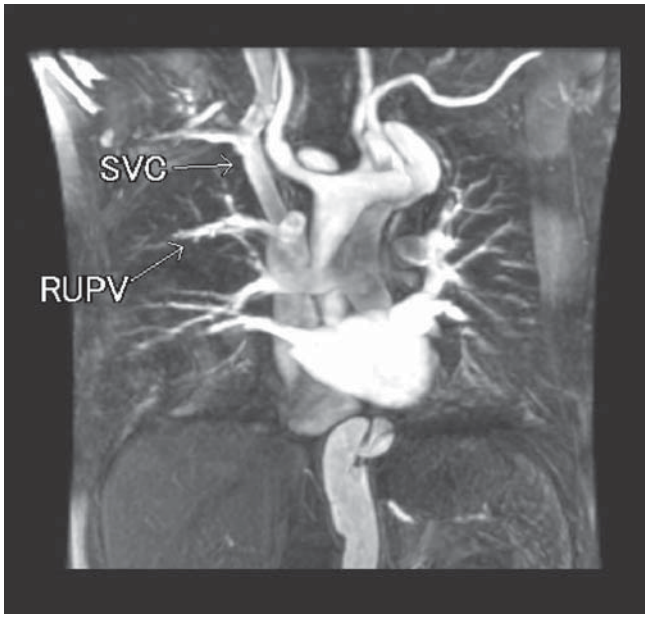

Fig. 2. Magnetic resonance angiography showed the right upper pulmonary vein to return to the superior vena cava

A median sternotomy was performed. Cardiopulmonary bypass (CPB) was initiated with arterial cannulae placed in the femoral and axillary arteries and venous cannulae in the superior and inferior vena cava. The esophageal temperature was maintained at 20°C for deep hypothermic circulatory arrest (DHCA) with antegrade selective cerebral perfusion. A horizontal incision was made in the ascending aorta and an intimal tear from the ascending and proximal aortic arch was found. After distal anastomosis using the elephant trunk technique, systemic antegrade perfusion was started. The left subclavian artery, left common carotid artery, and brachiocephalic artery were all anastomosed to the limbs of the graft. The aortic valve was examined, and the raphe was found between the right and left coronary cusps (Fig. 3). We performed a modified Bentall procedure and each graft was anastomosed end-to-end. The DHCA time was 71 min and the CPB time was 398 min.

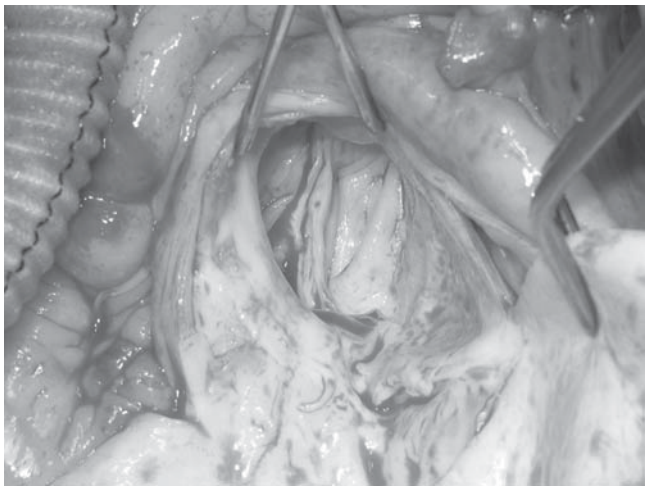

Fig. 3. The surgical findings showed a bicuspid aortic valve. Raphe existed between the right and left coronary cusps

The postoperative course was uneventful, and the pathological findings showed no cystic medial necrosis in the aortic wall. The patient was discharged on the 24th postoperative day, and the systolic blood pressure was kept under 120 mmHg with a beta-blocker and calcium antagonist.

Discussion

Since Hirose et al.[3] reviewed the literature on 32 cases of TS with aortic dissection, there have been five additional case reports listed on PubMed.[4–8] To our knowledge, this is the first case to undergo a successful combined total arch replacement for a Stanford type A chronic aortic dissection using the modified Bentall procedure for bicuspid aortic valve in TS.

A bicuspid aortic valve is one of the most common cardiac anomalies in TS. A bicuspid aortic valve is also associated with abnormalities of the aorta, such as dilatation, coarctation, cystic medial necrosis, and dissec-

tion.[9,10] In this case, AR was mild and the left ventricular (LV) dimension was not dilatated (LV diastolic dimension/systolic dimension = 40/23 mm). There may be some controversy regarding the need to perform a root replacement for this condition. Regarding Marfan's syndrome, a recent report suggested that either a one-stage total or a subtotal aortic replacement for the treatment of extensive aortic disease is feasible.[11] Hu et al.[11] also suggested that it can eliminate the risk of a remnant aortic aneurysm rupture in staged total aortic replacement including the aortic root. In addition, after repairing a coarctation of the aorta with a bicuspid aortic valve at 5 years of age, AR worsened at the age of 14. Then aortic valve replacement was performed but the patient died on the fifth postoperative day.[12] Meanwhile, regarding aortic root surgery, Schäfers et al.[13] reported remodeling the aortic root and reconstructing the bicuspid valve for a regurgitant bicuspid aortic valve and the dilatation of the proximal aorta. They concluded that the combined application of valve reconstruction and root remodeling is feasible while also leading to good early results. In this case, preoperative TTE revealed the bicuspid aortic valve to not be calcified, and the existence of a typical valve prolapse in the raphe was also confirmed. We previously reported aortic valve-sparing procedures and the results for a bicuspid aortic valve.[14] On the basis of the condition of the bicuspid aortic valve view, the valve-sparing procedure was possible in this case. However, the long-term results are still unknown, especially for genetic abnormalities. Therefore, we performed a modified Bentall procedure so as to avoid a high-risk re-operation. We also considered a dilatation of the descending aorta (double-barrel type) in the future. The elephant trunk procedure is thus considered to be an important surgical option in this case.

A routine cardiovascular evaluation to prevent premature cardiac death has been recommended for patients with TS.[1,2] Echocardiography is the mainstay of diagnostic aortic dilatation; however, studies have shown that magnetic resonance imaging (MRI) can detect dilatations missed on echocardiography. The current guidelines for the management of patients with TS recommend echocardiography and MRI assessment in patients suspected to have cardiovascular manifestations.[15]

Prophylactic beta-blockers or calcium antagonists have been used in Marfan's syndrome to prevent a progression of aortic dilatation.[16] However, there has not yet been any evidence reported in women with TS. The natural history of aortic root dilatation remains unknown, but the risk of an aortic dissection or rupture may be as high as 60%. Therefore, Elsheikh et al.[17] emphasize the importance of regular surveillance of the aortic root diameter in all women with TS and that hypertension should be aggressively treated when present, in order to minimize the risk of a potentially fatal aortic dissection.

In this case, the patient was diagnosed as having a bicuspid aortic valve and dilatation of the ascending aorta 3 years earlier. However, she had not received adequate medical follow-up with anti-hypertensive drugs and TTE, chest CT, or MRI. Fortunately, the patient could be salvaged by surgery. In addition, we have to keep in mind that most such patients diagnosed with an aortic dissection tend to be symptomatic, although asymptomatic dissections have also been reported.[18] Interestingly, Lie[19] also reported that six of the ten patients who died of a rupture of an aortic dissection had not experienced any pain. In our case, the symptoms of back pain were slight. Therefore, all patients with TS should routinely undergo TTE, chest CT, or MRI examinations. Regarding surgery, we also suggest that elective surgical intervention should be considered for patients with not only a dilatation of the aorta but also a bicuspid aortic valve.

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