Successful aortic reimplantation in a three-year-old child with Marfan syndrome

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Abstract

Aortic root dilatation is rare in children, and is often secondary to Marfan syndrome (MFS). We experienced a case of a three-year-old boy (92 cm, 12 kg) with MFS presenting with progressive dilatation of aortic root. We electively performed a valve-sparing aortic root replacement using a 24-mm Gelweave Valsalva graft. Although the patient required a mitral valve repair due to infective endocarditis postoperatively, the recovery from the second surgery was uneventful. This case is one of the youngest children of valve-sparing aortic root replacement in the literature.

Keywords: Aortic root; Aneurysm; Marfan syndrome

1. Introduction

The prognosis of Marfan syndrome (MFS) is primarily determined by cardiovascular complications, such as rupture of the aortic root and aortic dissection. To avoid these complications, valve-sparing root replacement has recently become an optional treatment for patients with MFS [1, 2]. In children with MFS, however, few operative outcomes on an aortic valve-sparing surgery has been reported, because aortic root aneurysm is extremely rare in children <10 years of age [3]. Herein, we present a case of a successful valve-sparing root replacement in a three-year-old boy with MFS.

2. Case report

A three-year-old boy was referred to our hospital presenting with progressive dilatation of the aortic root (5 mm per month). He was born by normal delivery and diagnosed with infantile MFS based on specific physical manifestations, and had no familial history of MFS. On admission, he was an active child who was 92 cm tall and weighed 12 kg. An echocardiogram revealed extremely enlarged Valsalva sinuses of 40 mm in diameter with a competent aortic valve (Fig. 1a), which was also observed by computed tomography (Fig. 1b). He was then scheduled to undergo a prophylactic operation.

On opening the pericardium, a large aortic root aneurysm appeared (Fig. 2a). After establishing cardiopulmonary bypass, cardioplegic arrest was obtained in a retrograde fashion. The aortic sinuses were excised and coronary arteries were widely mobilized creating coronary buttons. The aortic root was dissected out circumferentially to the level of basal ring. After assessment of aortic valves, including thickness and coaptation of each leaflet, we decided to perform a valve-sparing procedure (Fig. 2b). Since aortic annulus was of 20 mm in diameter, we used a 24-mm Valsalva graft (Gelweave, Termo, Tokyo, Japan).

Fig. 1. (a) Preoperative echocardiogram showing extensively enlarged sinus Valsalva (40 mm in diameter) and competent aortic valve. (b) A preoperative chest computed tomography also demonstrates marked dilatation of the sinus Valsalva, and funnel chest.
Fourteen horizontal mattress sutures were placed at the level of sub-annular position, and the preserved valve was placed inside the graft. After careful consideration commissural suspension was decided upon to obtain optimal coaptation of the leaflets, and the remnant wall of sinuses was then fixed against the graft (Fig. 2c). The valve coaptation was tested by saline infusion under direct vision and confirmed by cardioplegia injection through an occlusion balloon placed in the graft (Fig. 2d). The coronary buttons were then attached in a standard fashion. The distal end of the graft was plicated to approximate the distal size of the aorta. The cardiopulmonary bypass was uneventfully weaned off with low-dose catecholamine support and the immediate transesophageal echocardiogram showed no sign of aortic regurgitation.

He was extubated on postoperative day 4 and started oral feeding. Although he had a high fever associated with MRSA, his infective status subsided after administration of Vancomycin over a four-week period. However, severe mitral regurgitation originating from the base of the anterior leaflet suddenly emerged, and the patient was subjected to a re-do operation. A slit-like tear in the subannular position just beneath the left coronary sinus was identified and internally closed by a couple of 5-0 Prolene sutures buttressed with pericardium. Weaning from cardiopulmonary bypass was uneventful, as was the postoperative course. He was discharged on postoperative day 44 in active status. During his one-year outpatient follow-up, an echocardiogram showed no sign of heart failure and proper aortic and mitral leaflet coaptation (Fig. 2e).

3. Discussion

The standard surgical treatment for dilated aortic root is replacement using a composite valve graft or homograft. Valve-sparing aortic root replacement has become an attractive alternative option eliminating the long-term risk of thromboembolism and oral anticoagulation even in the patients with MFS. The Valsalva graft, Dacron conduit with prefashioned sinus of Valsalva, was also introduced by DePaulis and has shown satisfactory mid-term results in MFS patients.

In the present case, we performed a valve-sparing root replacement using this commercially available graft. Our strategy to treat this patient was as follows. First, he had progressively dilating sinus Valsalva and completely intact aortic valves. Second, the aortic orifice of 20 mm was large enough to provide a chance of lifetime evasion from valve replacement. Also, the 24-mm graft size (available minimum size) was appropriate for the 20-mm aortic orifice. Regarding mitral regurgitation, the subannular tear may be attributable to the subannular stitches to fragile connective tissue of MFS patient and subsequent local inflammation due to unexpected endocarditis.

In conclusion, we performed a valve-sparing root replacement using a 24 mm-Valsalva graft on three-year-old boy with MFS. Although an additional operation possibly due to infective endocarditis was required, the patient fully recovered. A few reports are available on a valve-sparing surgery for children with a dilated aortic root. To our knowledge, this reports a successful surgical case of the reimplantation procedure for the youngest child with MFS in the current literature.

References