Pediatric Bentall procedure for giant ascending aortic aneurysm in Loeys–Dietz syndrome

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ABSTRACT

Ascending aortic aneurysm is very rare in children, and is usually seen in patients with underlying connective tissue disorders such as Marfans and Ehler–Danlos syndrome. Loeys–Dietz syndrome (LDS) is less commonly seen as a cause of ascending aortic aneurysms in children. In this case report, we describe pediatric Bentall procedure, which we successfully performed to a child with LDS (Type I) with giant ascending aortic enlargement and significant aortic regurgitation.

Keywords: Aortic aneurysm, Bentall procedure, Loeys–Dietz syndrome

INTRODUCTION

Loeys–Dietz syndrome (LDS) is a connective tissue disease[1,2] characterized by arterial tortuosity, hypertelorism, and bifid uvula or cleft palate. LDS is an autosomal dominant condition resulting from mutations in the transforming growth factor (TGF), TGF beta receptor (TGFBR) 1 and 2, SMAD-3, and TGFB ligand genes.[2]

LDS affects tissues and organs throughout the body, especially the great arteries. Rapidly progressive aortic aneurysm is characteristic of prominent LDS, which necessitates close monitoring of these patients because of high mortality. Although the young age of the child poses risks, early prophylactic surgery is very important to prevent dramatic aortic complications and to obtain a good long-term prognosis.[3] With this report, we describe a case, in whom we successfully performed the Bentall procedure in a child with LDS (Type I) with giant ascending aortic enlargement and severe aortic regurgitation.

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CASE REPORT

A 5-year-old girl with the diagnosis of LDS (Type I) was admitted to our hospital with a complaint of fatigue. Physical examination revealed hypertelorism, malar hypoplasia, and bifid uvula [Figure 1]. Gene sequencing revealed a heterozygous missense mutation of the TGFBR1 gene. This confirmed the patient's diagnosis of LDS Type 1. Transthoracic echocardiography (TTE) revealed dilated aortic root and ascending aorta with aorto-annular dilatation and severe aortic regurgitation. Contrast-enhanced computed tomography of the aorta showed the presence of a giant dilatation involving the aortic root and extending into the ascending aorta [Figure 2a]. In cross-sectional measurements, the ascending aorta was 6.9 cm × 6.8 cm at its widest point [Figure 2b]. The decision was made to offer the Bentall procedure for the child.

A conventional median sternotomy was done. A giant ascending aortic aneurysm involving the aortic root was observed intraoperatively [Figure 3a]. Cardiopulmonary

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bypass was established by cannulation of the distal aorta and superior and inferior vena cava. Under moderate hypothermia, the aorta was cross-clamped and an aortotomy was performed. Direct osteal cardioplegia was administered and subsequently repeated at regular intervals. The extremely dilated aortic annulus was felt to be a contraindication to a valve-sparing aortic root replacement procedure. Coronary buttons were prepared by resecting the ascending aorta, and the aorta was suspended from three commissures [Figure 3b]. A 22-mm Dacron composite graft prepared with a 21-mm mechanical valve (St. Jude Medical Inc., Minnesota, MN, USA) was chosen. Multiple interrupted pledgetted mattress sutures (2-0 polyester) were taken at the annulus and then passed through the sewing ring of a composite valve conduit. The distal end of the graft was shaped appropriately, and the distal aorta was anastomosed with multiple interrupted pledgetted mattress sutures (2-0 polyester). Both coronary button implants were then anastomosed to both sides of the aorta [Figure 3c]. The operation was terminated by controlling the bleeding, and the sternum was closed. The cardiopulmonary bypass time was 164 min, and the aortic cross-clamp time was 115 min. Postoperative TTE showed normal prosthetic aortic valve and ascending aorta. The child was discharged on the 8th postoperative day with adequate oral anticoagulation therapy.

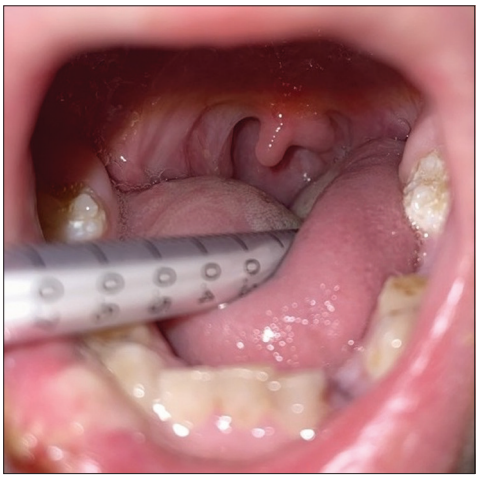

Figure 1: Bifid uvula image, which is common in patients with LDS: LDS: Loeys–Dietz syndrome

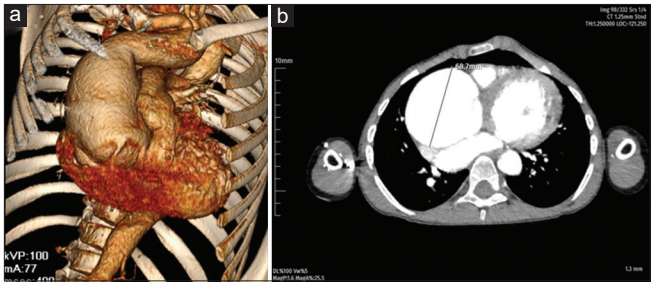

Figure 2: (a) 3D image of the contrast-enhanced tomography, (b) cross-sectional measurement of the diameter of the giant ascending aorta. 3D: Three-dimensional

DISCUSSION

Currently, the Bentall procedure is a widely used surgery for pathologies of the aortic root and ascending aorta.[4] Although it is widely used in adult cardiac surgery, the application of this procedure in the pediatric age group is extremely rare. When we review the literature, Marfan syndrome is the most common genetic cause for severe aortic diseases such as dilatation or dissection in the pediatric age group.[5]

LDS has several typical phenotypic features including, severe craniofacial pathologies (malar hypoplasia, retrognathia, cleft palate or bifid uvula, and hypertelorism), aortic root aneurysms, diffuse vascular aneurysm with arterial tortuosity and dissection, arachnodactyly, pectus deformity, scoliosis, joint laxity, and growth retardation.[6] When the prophylactic surgery phase is reached at the end of a sufficient follow-up period due to ascending aortic aneurysm, patients should be evaluated with a multidisciplinary approach with a joint council decision. When it comes to prophylactic surgery in the treatment approach, valve-sparing aortic replacement has become a safe and reliable alternative as the gold standard.[7] However, the ascending aortoplasty method is an undesirable strategy due to the high probability of recurrence. However, for the valve-sparing

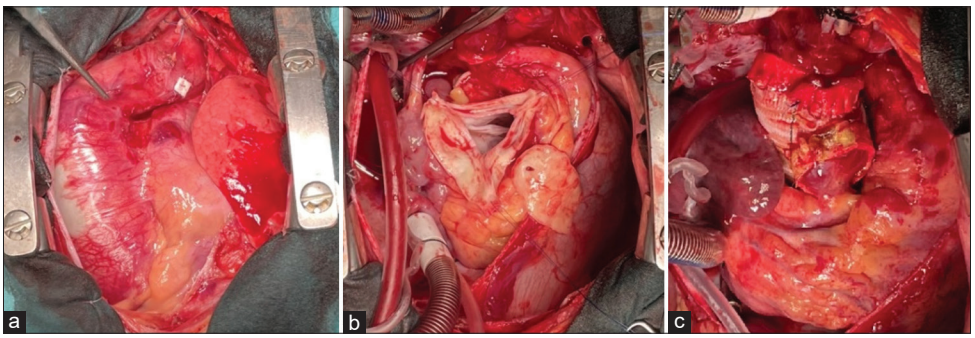

Figure 3: (a) Intraoperative giant ascending aortic aneurysm image, (b) after resection of the ascending aorta and separation of the coronary buttons, (c) after the operation is completed with a composite graft

surgical approach, the aortic root and valve morphology should be suitable for this method. In severely enlarged aortic root or impaired valve morphology, both valve and root replacement may be required. In our case, although we initially thought of performing valve-sparing surgery, we decided to replace the valve and root together due to the advanced dilatation of the intraoperative aortic annulus.

One of the most important steps of the Bentall procedure is the implantation of the coronary buttons. It is important to adopt a reliable strategy, especially in the pediatric age group, given that coronary arteries are quite thin and small. As we have presented in our case, wide resection of coronary buttons with a diameter of approximately 1 cm and covering the graft on the aortic root is a very safe and recommended method in the pediatric group. Thanks to this method, the risk of stenosis in the coronary ostia is minimized. This technique is more suitable, especially for children. Since LDS may cause the risks of dilatation and early dissection of the aorta with the same features as Marfan syndrome, the surgical intervention to be performed should be maximally curative. With our experience and short-term results, we think that the approach we use is a safe and effective method.

CONCLUSION

A life-threatening aortic aneurysm can occur in very rare syndromic diseases such as LDS in children. We believe that a challenging surgery such as the Bentall procedure is safe, effective, and successful in pediatric case groups where surgery is inevitable.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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