Surgery for A Sinus of Valsalva Aneurysm After Radical Repair of the Aortic Coarctation Complex in A Young Adult with Congenital Bicuspid Aortic Valve: A Case Report

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Abstract

We describe a young adult who underwent surgery for sinus of Valsalva aneurysm and bicuspid aortic valve regurgitation, after undergoing radical repair of the aortic coarctation complex during childhood. A 22-year-old man had undergone radical repair for coarctation of the aorta and ventricular septal defect during childhood. He required surgical treatment of an enlarged ascending aorta and a sinus of Valsalva aneurysm with a congenital bicuspid aortic valve, as indicated using Computed Tomography (CT). Echocardiography revealed moderate regurgitation from the bicuspid aortic valve. We performed a repair of the sinus of Valsalva and replacement of the ascending aorta. Postoperatively, echocardiography revealed trivial aortic regurgitation, while CT indicated resolution of the enlarged ascending aorta and sinus of Valsalva aneurysm. The patient remains asymptomatic 1 year later. Valve sparing and aortic valve repair extensively benefit young patients exhibiting aortic root pathology following radical repair of congenital heart disease.

Keywords: Sinus of Valsalva aneurysm; Repair of the sinus of Valsalva; Aortic coarctation complex; Congenital bicuspid aortic valve

Abbreviations: CHD: Congenital Heart Disease; BAV: Bicuspid Aortic Valve; CoA: Aortic Coarctation; VSD: Ventricular Septal Defect; CT: Computed tomography

Introduction

Recently, surgical outcomes for Congenital Heart Disease (CHD) have improved. As patient who underwent radical repair for their CHD during childhood begin to age, an increasing number are now candidates for surgical treatment of another heart diseases during adulthood. Congenital Bicuspid Aortic Valve (BAV) coexisting with the Aortic Coarctation (CoA) complex is relatively common. Therefore, such patients are expected to develop aortic valve disease or aortic root disease during adulthood. We describe the surgical case of a young adult who developed sinus of a Valsalva aneurysm and BAV regurgitation after undergoing radical repair of the CoA complex during childhood.

Case Report

A 22-year-old man, who underwent radical repair for CoA with Ventricular Septal Defect (VSD) during childhood, was a candidate for surgical treatment of an enlarged ascending aorta and a sinus of Valsalva aneurysm associated with BAV. Computed Tomography (CT) imaging revealed an enlarged ascending aorta (55mm) and sinus of Valsalva aneurysm (58mm), which was particularly prominent in the right sinus of Valsalva of the BAV (Figure 1). An innominate artery and the left common carotid...
artery arose from common canal, while the right subclavian artery exhibited an aberrant origin. No stenotic lesions were identified at the site of CoA repair. Echocardiography revealed moderate regurgitation from the BAV, no aortic stenosis (peak pressure gradient: 10.7mmHg) and no residual shunt following the repair of VSD.

Figure 2: A: The ascending aorta enlarged eccentrically (arrow) and is shown with the right sinus of Valsalva aneurysm (arrow head). B: The aortic valve presents as a bicuspid valve due to fusion of the left-coronary cusp with the non-coronary cusp. The left, non-fused cusp is redundant (*) and a gap between the cusps exists at the right non-commissure (arrow). LCA: left coronary artery. C: The redundant cusps are checked with Frater’s stitch and corrected with cusp plication (*). The gap between the cusps at the right non-commissure is corrected with Cabrol’s stitch (arrow). D: Aortic root partial remodeling and ascending aorta replacement. The right coronary artery is reconstructed using a saphenous vein graft. SVG: saphenous vein graft.

Median sternotomy was performed. Dense adhesions had formed around the heart. Therefore, cardiopulmonary bypass was established via the right axillary artery, right femoral artery, and superior and inferior vena cava. The ascending aorta had enlarged eccentrically and an aneurysm of the right sinus of Valsalva was observed (Figure 2A). The aortic valve was bicuspid due to fusion of the left-coronary and non-coronary cusps (Figure 2B). Aspects of the aortic cusp were normal, and the aneurysm was only observed at the right sinus of Valsalva. Therefore, repair of the sinus of Valsalva was performed. The right sinus of Valsalva was resected and reconstructed with a linguiform knitted Dacron patch. The redundancy of cusps was assessed using Frater’s stitch and the required corrections were performed using cusp plication. The gap of the right-coronary cusp and non-coronary cusp at the commissure was corrected with Cabrol’s stitch (Figure 2C). The ascending aorta was replaced using an artificial vascular graft. Since the right coronary artery originating from the right aneurysmal Valsalva wall was running intramurally, the aortic wall around the right coronary ostium was fragile and creating a coronary button was impossible; hence, a saphenous vein graft was connected to the ascending aorta and right coronary ostium (Figure 2D). Postoperatively, echocardiography revealed trivial aortic regurgitation and no aortic stenosis (peak pressure gradient: 5.1mmHg), and CT indicated good blood flow through the right coronary artery as well as resolution of the enlarged ascending aorta and sinus of Valsalva aneurysm (Figure 3). There were no post-surgical events at one year of follow up.

Figure 3: Postoperative computed tomography scan indicates good blood flow through the saphenous vein graft to the right coronary artery (arrow), as well as resolution of the enlarged ascending aorta and sinus of Valsalva aneurysm.
Discussion

Improvements in cardiovascular surgery have contributed to enhanced CHD surgical outcomes. As results, more patients experience a healthy adolescence after undergoing radical repair of a CHD in childhood. Accordingly, there is an increasing number of patients who candidates for surgical treatment for other cardiac diseases.

CoA occurs in 5-8 % of all CHD [1]. The CoA complex is a combination of CoA malformation and other cardiac anomalies (commonly VSD). Additionally, congenital BAV is present in 50-70 % of CoA cases [1]. Although the CoA complex is treated surgically shortly after diagnosis, coexisting BAV is not usually treated surgically unless a functional disorder arises as a result. Persistent BAV can result in valve stenosis, regurgitation, and eccentric enlargement of the ascending aorta [2,3].

The patient in the case described herein was a rare young case of an enlarged ascending aorta and sinus of Valsalva aneurysm that coexisted with congenital BAV following the radical repair of the CoA complex during childhood. Some surgical cases of sinus of Valsalva aneurysm with BAV have been reported [4,5]. Since clinicians anticipate that patients who have undergone a radical repair of CHD will eventually require additional surgery, cases like the one described herein will increase in frequency. The long-term outcomes after conventional repair of CoA complex are acceptable; however, coexisting BAV is a significant risk factor for valve-related reinterventions [6].

Generally, the Bentall procedure with a composite graft, valvular reimplantation, and aortic remodeling are the surgical techniques of choice for resolving aortic root pathologies. The Bentall procedure using prosthetic valve has reliably good surgical results; however, concerns have arisen regarding complications associated with anticoagulant therapy for mechanical valves and the durability of bioprosthetic valves. Recently, the surgical results of valve sparing procedures [8] could be performed rather than a reimplantation procedure because it would require fewer suture lines.

Aortic valve repair is a difficult technique that requires experience and an ability to precisely evaluate the valve cusps; however, its importance is underscored by its ability to improve the surgical results of valve sparing procedures [8].

Conclusion

Although the surgeon and institution much determine the most suitable procedure for each patient, the procedures of valve sparing, and aortic valve repair confer extensive benefits for young patients who exhibit aortic root pathology following the radical repair of CHD.

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