Repair of a Unique Sinus of Valsalva Defect in an Infant

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Abstract

Sinus of Valsalva aneurysm is a rare disease characterized by the partial elevation of the aortic root. Few reports are available on the surgical treatment for infants. We report the repair of an extremely rare case of a sinus of Valsalva defect with a ventricular septal defect and right ventricular outflow tract stenosis in an infant. It was not a sinus of Valsalva aneurysm, but it exhibited abnormal partial bulging of the aortic root and forming an aneurysm-like cavity within the right ventricular myocardium. We performed direct closure of the sinus of Valsalva aneurysm-like cavities and intracardiac repair in two stages. Three years after total repair, the patient remained healthy and asymptomatic.

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Running head: Sinus of Valsalva Defect

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Abstract

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Introduction

Sinus of Valsalva aneurysm (SVA) is a rare condition characterized by partial bulging of the aortic root [1,2]. We report an extremely rare case of a sinus of Valsalva defect repaired in infancy and a deformity that exhibited abnormal partial bulging of the aortic root and formed SVA-like cavities within the right ventricular myocardium. This study is exempt under the Institutional Review Board at our institution. Consent was obtained from her parents.

Case Report

A one-day-old infant girl was transferred to our institution because of dyspnea and a heart murmur. She was neonatally diagnosed with a ventricular septal defect (VSD), an atrial septal defect (ASD), patent ductus arteriosus (PDA), persistent left superior vena cava, right ventricular outflow tract (RVOT) stenosis (RVOTS), and a suspected SVA on echocardiography. She had no family history of connective tissue disease.

Preoperative echocardiography revealed SVA-like cavities protruding toward the RVOT (Figure 1A, B), as well as RVOTS (2.5 m/s), and a large VSD (14.6 mm). Preoperative cardiac catheterization revealed the SVA-like cavities to be large. Preoperative contrast computed tomography revealed large SVA-like cavities protruding from the right coronary sinus (RCS) toward the RVOT and small SVA-like cavities below the non-coronary sinus (NCS).

At 2 months old (4.9 kg), the infant underwent pulmonary artery banding (PAB) and PDA ligation. At 1 year old (7.7 kg), she underwent total repair, detailed below. Under cardiopulmonary bypass, the ascending aorta was transected above the sinotubular junction. The walls of the right coronary and non-coronary sinuses of Valsalva were deficient, forming aneurysm-like cavities with trabeculation (Figure 2A, B), connected behind the commissure between the right and non-coronary cusps (Figure 2C). Thus, we diagnosed the infant with a sinus of Valsalva defect rather than an SVA. Anatomically, the transition between the aortic media and annulus was unclear; we believed that suturing would injure the leaflets and distort the aortic annulus, leading to aortic valve deformity. Therefore, patch closure of the sinus of Valsalva defect was abandoned. Direct closure of the aneurysm-like cavities was performed using a pledgeted 5-0 polyvinylidene fluoride (PVDF) suture (Figure 2D). A 0.4-mm expanded polytetrafluoroethylene patch was applied to the VSD using pledgeted 5-0 PVDF sutures. The SVA-like cavities protruding into the RVOT was closed with horizontal mattress suturing using a pledgeted 5-0 PVDF suture. After debanding, reconstruction of the pulmonary artery was performed using an autologous pericardial patch.

The postoperative course was uneventful. Postoperative echocardiography revealed normal aortic valvular function with trivial aortic regurgitation and no residual VSD or RVOTS (Figure 3). Three years after total repair, the patient remained healthy and asymptomatic, with grade I aortic regurgitation.

Discussion

SVA is characterized by a lack of continuity between the aortic media and annulus [3]. Congenital SVA is often concomitant with other congenital heart defects [4]. An unruptured SVA may develop pathologies such as RVOTS and aortic regurgitation[5]. The pathogenesis of this case was considered a large deficiency in fetal sinus of Valsalva wall development, leading to the exertion of aortic pressure on the right ventricular myocardium, and bulging of the latter into the RVOT, forming SVA-like cavities in the sinus of Valsalva. Our patient presented with heart failure symptoms due to VSD and PDA. We performed PAB and PDA ligation when she was 2 months old. We performed direct closure of the SVA-like cavities and intracardiac repair in two stages, as the procedure is risky in the early infancy owing to tissue fragility. We know of no reports of similar sinus of Valsalva defects in an infant. Following satisfactory surgical outcomes, long-term follow-up will be required.

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Legends

Figure 1. Pre-total-repair examination

A, Long- and B, short-axis views of transthoracic echocardiography reveal a large sinus of Valsalva aneurysm-like cavities protruding from the right coronary sinus toward the right ventricular outflow tract. Ao, aorta; LA, left atrium; LV, left ventricle

Figure 2. Intraoperative image

A, B, The defects of the walls of the right coronary and partial non-coronary sinuses of Valsalva, and a myocardial component and trabeculation in the sinus of Valsalva. C, Intraoperative schema. D, Post-direct closure of sinus of Valsalva aneurysm-like cavities.

Figure 3. Post-total-repair examination

A, Short-axis view of transthoracic echocardiography; the right ventricular outflow tract stenosis was released. B, Long-axis view of transthoracic echocardiography revealed trivial aortic insufficiency. Ao, aorta

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