Surgical Repair of a Ruptured Sinus of Valsalva Aneurysm Complicated by Ventricular Septal Defect and Aortic Valve Insufficiency

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ABSTRACT

A 23-year-old male patient was admitted with a 2-week history of congestive heart failure. An echocardiogram revealed an aneurysm originating from the right sinus of Valsalva with a small perforated area on the edge, a ventricular septal defect and severe aortic valve insufficiency. A decision for surgery was made. The right sinus of Valsalva was repaired from the inside. The infundibulotomy was closed using a continuous suturing technique and pericardial patch was used incorporating the ventricular septal defect and the previously repaired right sinus of Valsalva. The aortic valve was replaced with a mechanical prosthesis. Follow-up was unproblematic, and he was discharged on the 12th postoperative day. Aortic valve insufficiency and a high output shunt cause an increase in the ventricular diameter, impairment of the ejection fraction, congestive heart failure and resultant pulmonary pathology. Ventricular enlargement or impaired ejection fraction does not necessarily deem these patients inoperable.

Key Words: Sinus of Valsalva; rupture; aortic valve insufficiency; congestive heart failure

Ventriküler Septal Defekt ve Aort Kapak Yetmezliği ile Komplike olan Rüptüre Valsalva Sinüsü Anevrizmasına Cerrahi Yaklaşım

ÖZET

Yirmi üç yaşındaki erkek hasta 2 haftalık konjestif kalp yetmezliği öyküsü ile yatırıldı. Ekokardiyografide, sağ valsalva sinüsünden köken alan anevrizma ile ventriküler septal defekt ve ciddi aort kapak yetmezliği saptandı ve cerrahiye karar verildi. Sağ valsalva sinüsü içeriden onarıldı. Kontinü dikiş tekniği ile perikard yaması kullanılarak, ventriküler septal defekt ve daha önce onarılan sağ valsalva sinüsü de dahil edilecek şekilde infundibulotomi onarıldı. Aortik pozisyona mekanik kapak takıldı. Postop takipte sıkıntısı olmayan hasta 12. günde taburcu edildi. Aort kapak yetmezliği ve yüksek debili şant; ventrikül çapında artışa, ejeksiyon fraksiyonunda bozulmaya, konjestif kalp yetmezliğine ve bunun neden olduğu pulmoner semptomlara yol açar. Ventrikül genişlemesi ya da düşük ejeksiyon fraksiyonu, bu hastalarda inoperabilite nedenleri değildir.

Anahtar Kelimeler: Valsalva sinüsü; rüptür; aort kapak yetmezliği; konjestif kalp yetmezliği

INTRODUCTION

Sinus of Valsalva aneurysm rupture (SVAR) is a rare but life-threatening pathology, and most patients are symptomatic at the time of diagnosis. It is mostly accompanied by a ventricular septal defect (VSD) and aortic valve insufficiency (AI)[1]. These anomalies contribute to an increase in the size of the aneurysm and in the pathogenesis of rupture[2]. Hence, after a diagnosis of SVAR, repair of the aneurysm and concomitant anomalies should be performed simultaneously and without delay. Here we present a surgical approach to a symptomatic case of SVAR and concomitant doubly committed VSD and severe AI.

CASE REPORT

A 23-year-old male patient was admitted to Yüksek İhtisas Training and Research Hospital, Ankara, with a 2-week history of congestive heart failure (CHF) for which treatment had been commenced in another centre. In the previous centre, inotropic support had also been provided. He was referred to our hospital for surgery following transthoracic echocardiogram which revealed severe AI, a dubious VSD and pulmonary hypertension. Following admission to our hospital, his initial physical examination revealed that his signs of CHF had regressed. His functional capacity was class 3 according to the New York Heart

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Association (NYHA) Classification. There was an increase in the cardiothoracic index on chest X-Ray. A more detailed transesophageal echocardiogram displayed SVA originating from the right sinus of Valsalva prolapsing into the right ventricular outflow tract with a small perforation at its edge. The aneurysmal pouch reached the level of the pulmonary valve. There was a doubly committed VSD and severe AI, and the aortic diameter at the level of the sinus of Valsalva was 4.2 cm. The end diastolic diameter of the left ventricle was 9 cm, ejection fraction (EF) was decreased to 30% and pulmonary artery systolic pressure was reported as 45 mmHg. Additionally, there was second degree tricuspid insufficiency and thickening of the interventricular septum (1.3 cm). Considering all the findings, a decision for surgery was made. A standard oblique aortotomy incision extending to the non-coronary sinus and a longitudinal right ventricular infundibular incision was made. Inspection of the sinus of Valsalva revealed a windsock deformity, with a perforation at the tip, originating from the right sinus of Valsalva and extending to the right ventricular outflow tract. The windsock deformity covered the 2 × 1 cm-sized doubly committed VSD (Figure 1). The aortic valve was mixomatous with an impaired coaptation. Besides this, there was thickening of the interventricular septum causing stenosis of the left ventricular outflow tract. The weak part of the windsock was excised and opened. The right sinus of Valsalva from the inside. The infundibulotomy was closed by a continuous suturing technique using a gluteraldehyde-fixed pericardial patch and incorporating VSD and the previously repaired right sinus of Valsalva (Figure 2). Septal myectomy was performed starting from the nadir of the right coronary cusp up to the right and left coronary commissure extending approximately 1 cm towards the apex to prevent any possible limitation of the prosthetic aortic valve by the thick septum. The aortic valve was excised and replaced with a 23# mechanical valve prosthesis in such a way that the teflon supported sutures were passed through the patch at the level of the annulus at the right sinus of Valsalva. A Dacron graft was

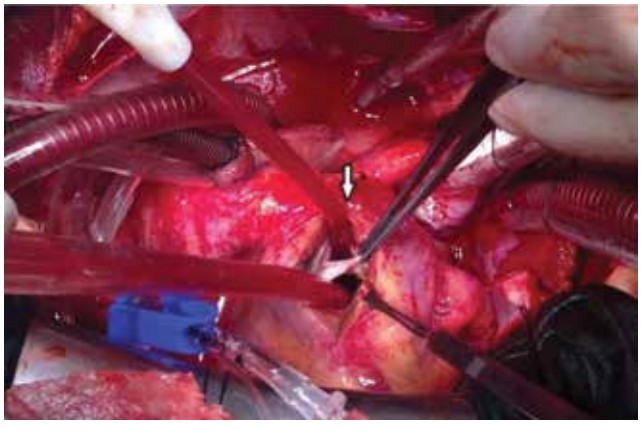

Figure 1. The arrow indicates the doubly committed VSD through the infundibulotomy.

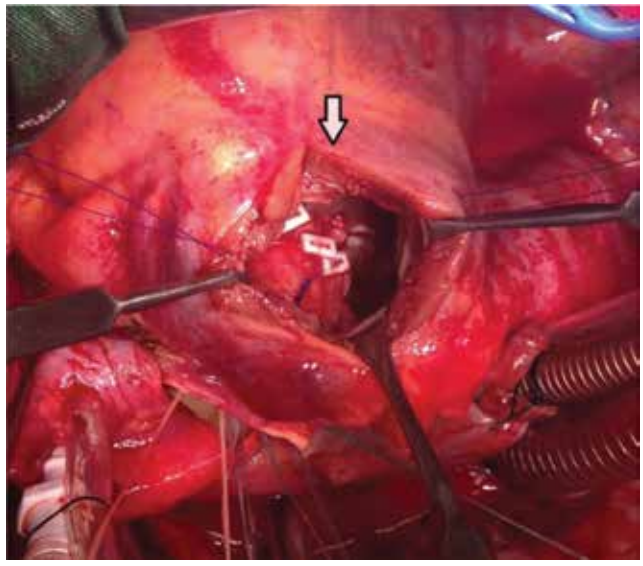

Figure 2. The arrow indicates the infundibulotomy. The pericardial patch incorporated VSD and the previously repaired right sinus of Valsalva.

used to wrap the dilated ascending aorta. The tricuspid valve was not repaired because it was deemed to be competent during right atrial exploration. Primary closure of the incisions was followed by removal of air and the cross-clamp. Weaning off the cardiopulmonary bypass was achieved with the help of an epicardial pacemaker (used for the atrioventricular block), adrenalin (0.1 µg/kg/min) and dopamine (5 µg/kg/min). The patient was transferred to the intensive care unit where normal sinus rhythm was achieved on the same day. On the 1st postoperative day, he developed a single episode of ventricular fibrillation for which he underwent resuscitation for 1 min and he was defibrillated. Normal sinus rhythm was achieved without any haemodynamic or neurological sequelae. He was followed up in the intensive care unit for 5 days and was then transferred to the ward on a dopamine infusion of 5 µg/kg/min. His follow-up was unproblematic, and he was discharged on the 12th postoperative day. Transthoracic echocardiogram performed in the 1th postoperative month revealed that the end diastolic diameter of the left ventricle decreased from 9 cm to 7 cm. However, EF had not changed postoperatively but remained at 35%.

DISCUSSION

The incidence of VSA in patients undergoing open heart surgery is 0.4%-0.6%[1,3,4]. It is usually diagnosed during the third and fourth decades of life[1,3]. More than 80% of patients are asymptomatic with a functional capacity of 3 or 4 according to the NYHA Classification[3,5]. CHF may not be observed at the time of diagnosis[5]. In this case, the patient's CHF had already regressed prior to admission to our hospital, and the prominent symptom was impairment of functional capacity.

In-hospital mortality following surgery of SVAR is reported to be 3.6%, and the actuarial survival is 93.4% ± 3.7% at 10

years and 87.1% ± 5.6% at 15 years[6]. VSD is the commonest congenital anomaly observed to coexist with SVA. According to some series, its incidence is between 45% and 50%. The commonest type of VSD is subarterial[1,3,7]. The incidence of coexisting AI and VSD is up to 40%[3,4,5,7]. Our patient had subarterial VSD and severe AI together with SVAR. The pathology and autopsy findings of those with SVA have previously revealed the discontinuity of the medial layer between the sinus of Valsalva and aortic valve tissue[8]. Later studies lead to a better understanding that because of the close proximity of VSD to the aforementioned region, the weak annulus is displaced towards the right ventricle externally and distally because of the Venturi effect. This eventually results in an increase in the diameter of the aneurysm and increases the severity of AI[2].

Severe AI and a high output shunt soon cause an increase in the ventricular diameter, severe impairment of the EF, severe CHF and resultant pulmonary findings. Greater than expected ventricular enlargement or impaired EF does not necessarily mean that these patients are inoperable. Therefore, open surgical repair is obligatory following the diagnosis of SVAR. Many centres have reported both their short- and long-term results[1,3,9]. Long-term surgical repair of SVARs can be performed with low mortality and morbidity[10]. Techniques used and their results are still a matter of debate. It has been shown that primary repair of SVAR by suture is a risk factor for early deterioration of the untouched AI[11]. Prior opinions considered the opening of a single area during repair to be sufficient. However, it is contemporarily believed that opening the areas from which the aneurysm originates and the aneurysm ruptures into will assist in better understanding of the pathology and more accurate repair[3,9]. In this case, we used a double-sided approach by opening the aorta and the infundibulum. Having resected the weak part of the windsock, we attempted to prevent a possible recurrence by primarily repairing the right sinus of Valsalva from the inside and then using a pericardial patch and incorporating VSD into the repair.

Another controversial issue is how to approach AI. It has been shown that the prognosis of long-term follow-up is related to the presence of preoperative AI and the presence of AI at the time of discharge[11,12]. Repair or replacement of the incompetent aortic valve may be useful in preventing a surgical revision at a later date. Repair may not be useful in patients with severe valve degeneration and coaptation impairment. Yoshihisa et. al reported that patients with SVAR undergoing repair of the valve because of valvular insufficiency will develop further impairment in the long-term if they also have VSD[13]. We preferred to replace the aortic valve with a mechanical valve because of the presence of mixomatous degeneration, impaired coaptation and severe insufficiency of the native valve to prevent a possible AI in the long-term.

CONCLUSION

SVAR is a disease which causes rapid deterioration in the patient's clinical status. Surgery should be planned as soon as possible. According to our current knowledge and experiences, simultaneous repair of the aneurysm and concomitant VSD using a pericardial or synthetic patch is a good approach worth considering. Coexisting AI, if present, should be carefully evaluated to minimise the possibility of surgical revision in the long-term.

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