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

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A 23 year old male patient was admitted with a two week history of congestive heart failure. An echocardiogram revealed an aneurysm of the sinus of valsalva 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. To close the infundibulotomy 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 twelfth postoperative day.

Aortic valve insufficiency and a high output shunt causes an increase in 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

Introduction

Sinus 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) . These anomalies are thought to contribute to an increase in the size of the aneurysm and in the pathogenesis of rupture . Hence, after a diagnosis of SVAR is made, the aneurysm repair and the repair of the 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 our hospital with a two week history of congestive heart failure for which treatment had been commenced in another centre. In the previous center inotropic support had also been given. He was referred to our hospital for surgery following transthoracic echocardiogram (TTE) which revealed severe AI, a dubious VSD and pulmonary hypertension. Following admission to our hospital, his initial physical examination revealed that his signs of congestive heart failure had regressed. His functional capacity was class 3 according to the New York Heart Association (NYHA) Classification. There was an increase in cardiothoracic index on chest x-ray. A more detailed transesophageal echocardiogram displayed an SVA originating from the right sinus 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, the ejection fraction (EF) was decreased to 30% and the pulmonary artery systolic pressure was reported as 45mmHg. 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 valsalva revealed a windsock deformity, with a perforation at the tip, originating from the right sinus valsalva and extending to the right ventricular outflow tract. It was seen that the windsock deformity covered the 2x1cm sized doubly committed VSD (Fig. 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 valsalva was repaired from the inside. To close the infundibulotomy a continuous suture technique was carried out using the gluteraldehyde fixed pericardial patch and incorporating the VSD and the previously repaired right sinus valsalva (Fig. 2). Septal myectomy was perfomed starting from the nadir of the right coronary cusp up to the right and left coronary commissure extending approximately 1 cm towards the apex in order 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 valsalva. A Dacron graft was used to wrap the dilated ascending aorta. The tricuspid valve was not repaired since 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 cardio-pulmonary bypass (CPB) was achieved with the help of an epicardial pacemaker (due to atrio-ventricular block), adrenalin (0.1 mcg/kg/min) and dopamine (5 mcg/kg/min). The patient was transferred to the intensive care unit (ICU) where normal sinus rhythm was achieved on the same day. On the first postoperative day he developed a single episode of ventricular fibrillation for which he received resuscitation for one minute and defibrillated. Normal sinus rhythm was achieved without any haemodynamic or neurological sequelae. He was followed up in the ICU for five days and then transferred to the ward on a dopamine infusion of 5 mcg/kg/min. His follow up was unproblematic and he was discharged on the 12th postoperative day. TTE performed in the 1st postoperative month revealed that the end diastolic diameter of the left ventricle decreased from 9 cm to 7 cm. However, the 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 New York Heart Association Classification 3,5. Congestive heart failure (CHF) may not be seen 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 RSVA is reported as 3.6% and the actuarial survival as 93.4 ± 3.7% at 10 years and 87.1 ± 5.6% at 15 years 6. VSD is the commonest congenital anomaly seen to coexist with SVA. According to some series, its incidence is between 45 and 50%. The commonest type to accompany a VSD is subarterial 1,3,7. The incidence of coexisting AI and VSD is up to 40% 3,4,5,7. Our patient had a subarterial VSD and severe AI together with SVAR. The pathology and autopsy findings of those with SVA have long previously revealed the discontinuity of the medial layer.
between the sinus valsalva and the aortic valve tissue\textsuperscript{8}. Later studies lead to a better understanding that due to the close proximity of the VSD to the afore mentioned region, the weak annulus is displaced towards the right ventricle externally and distally as a result of the Venturi effect. This eventually results in an increase in the diameter of the aneurysm and increases the severity of the AI\textsuperscript{2}.

Severe AI and a high output shunt soon causes an increase in the ventricular diameter, severe impairment of the EF, severe congestive heart failure 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 centers have reported both their short and long-term results\textsuperscript{1,3,9}. Long-term surgical repair of SVARs can be performed with low mortality and low morbidity\textsuperscript{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\textsuperscript{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\textsuperscript{3,9}. In this case, we took a double sided approach by opening both 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 valsalva from the inside and then using a pericardial patch and incorporating the VSD into the repair.

Another controversial issue is how to approach the AI. It has been shown that the prognosis of the long term follow up is related to the presence of preoperative AI\textsuperscript{12} and the presence of AI at the time of discharge is a risk factor for deterioration of AI during the long-term follow up\textsuperscript{11}. 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 severe coaptation impairment. Yoshihisa et. al reported that patients with SVAR undergoing valve repair due to valvular insufficiency will develop further impairment of valvular insufficiency at long term follow-up if there is also a VSD\textsuperscript{13}. We preferred to replace the aortic valve with a mechanical valve due to the presence of mixomatous degeneration, impaired coaptation and severe insufficiency of the native valve in order 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, a good approach which is worth considering is the simultaneous repair of the aneurysm and concomitant VSD using a pericardial or synthetic patch. Coexisting AI, if present, should be evaluated carefully to minimize the possibility of surgical revision in the long term.

**Conflict of Interest**

The authors declare that there are no conflicts of interest.
References


Figure legends

Fig. 1. The arrow indicates the doubly committed VSD through the infundibulotomy.
Fig. 2. The arrow indicates the infundibulotomy. The pericardial patch incorporated the VSD and the previously repaired right sinus valsalva.