Ruptured Congenital Sinus of Valsalva Aneurysms

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ABSTRACT

Background and aim of the study: Rupture of the sinus of Valsalva is rare, and there is a higher incidence of such rupture in Oriental countries than elsewhere. The objective of this study is to present the pathologic features and the clinical outcome after surgery in such patients. Methods: Between 1980 and 2001, a total of 17 patients (15 males and 2 females) with ruptured aneurysm of sinus of Valsalva underwent surgical intervention at the Tri-Service General Hospital, Taiwan. Their age ranged from 22 to 59 years with a mean of 33.5 years. These surgical operations made up 0.51% of the total cardiac operations (3305) performed during this period. The medical records were retrospectively reviewed. Results: The origin of the ruptured aneurysm of the sinus of Valsalva was the right coronary sinus in 12 patients, the noncoronary sinus in 4, and the left coronary sinus in one patient. The aneurysms ruptured into the right ventricle in 12 patients, into the right atrium in 3 patients, and into the left ventricle in 2 patients. Associated cardiac anomalies were aortic regurgitation in eight patients, ventricular septal defect in seven, and coronary artery fistula in one patient. There was no early postoperative death and one patient underwent a second operation after aneurysm of the sinus of Valsalva (ASV) relapse. Conclusions: The operation for a ruptured aneurysm of sinus of Valsalva carries a low operative risk and results in excellent long-term survival after surgical treatment. (J Card Surg 2004;19:99-102)

Aneurysm of the sinus of Valsalva (ASV) is a rare cardiac lesion that may be acquired or congenital. Most patients with an unruptured ASV are asymptomatic; however, when it ruptures, symptoms ranging from angina to acute pulmonary edema and cardiac collapse may occur.1,2 Ruptured ASV is commonly diagnosed preoperatively. The combined use of various echocardiographical techniques has become more and more important in making the diagnosis although preoperative catheterization with routine aortic cineangiography should still be obligatory.3 Medical treatment of isolated, unruptured ASV has been recommended unequivocally.4 However, the prognosis for ruptured ASV is regarded as grave unless it is treated surgically.5,6

MATERIAL AND METHODS

From January 1980 to December 2001, 17 patients with ruptured ASV underwent cardiac surgical procedures at the Department of Cardiovascular Surgery, Tri-Service General Hospital, Taipei, Republic of China. The mean age of these 15 males and 2 females was 33.5 years (range: 22–59 years). (Fig. 1) These operations constituted 0.51% of the 3305 surgical procedures that utilized cardiopulmonary bypass (CPB) during this period. All ruptured ASV included in this study were congenital in origin. Ruptured ASVs from infective endocarditis or trauma were excluded from this series.

The clinical symptoms ranged from symptomless to frank heart failure. In more than half of the patients, the symptoms progressed insidiously so the date of rupture of ASV could not be identified. However, six patients with severe aortic regurgitation experienced a sudden onset of symptoms, including acute chest pain, dyspnea, and palpitation. A loud machine-like continuous murmur along with a strong thrill along the left sternal border, was the most common physical finding. All patients underwent cardiac catheterization to confirm the diagnosis before being operated upon. (Fig. 2)

The origin of the ruptured aneurysm of the sinus of Valsalva was the right coronary sinus in 12 patients, the noncoronary sinus in 4, and the left coronary sinus in one patient. The most common perforation was a fistula connecting the right coronary sinus and the right ventricle, which occurred in 11 patients. (Table 1) Isolated ASV occurred in five patients. Coexistent cardiac anomalies were present in 12 patients; of these patients had an associated ventricular septal defect (VSD), and 5 patients had type I VSD. Aortic valve regurgitation coexisted in eight patients and six of these were severe. In one patient, a coronary artery fistula was found and finally, complicated complete heart block was found in one patient. (Table 2)

Surgical procedures were performed under CPB with moderate systemic hypothermia. In our early cases, the aneurysm was approached through the chamber into which it ruptured. Recently, we used a combined approach to repair closure of the fistula. The aorta was
also opened to check the pathology of the aneurysm and the aortic cusps. After repairing the aneurysm and correction of any associated cardiac anomaly, the aortic valve was checked again to evaluate if there was a need for aortic valvuloplasty or valve replacement.

The protruding aneurysm was excised and the base of the aneurysm was closed with a Teflon buttress or knitted Dacron patch. Any VSD adjacent to the aneurysm was closed either by direct suture or with a separate patch depending on the size. Aortic valvuloplasty was performed in two patients with mild aortic regurgitation, and aortic valve replacement was done in six patients with severe aortic regurgitation. Direct repair of the VSD was done in one patient and patch repair was done in the other six patients.

RESULTS

There was no early postoperative death, but one late death. The patient was a 25-year-old male who had aneurysm from the right coronary sinus that ruptured into the right ventricle. Severe aortic regurgitation and liver function impairment were noted before operation. He received aortic valve replacement with a 23 mm prosthetic valve. Unfortunately, endocarditis complicated with sepsis developed and he died of multiple organ failure 6 months after the surgery.

ASV relapse occurred in one patient who was a 22-year-old male with associated type I VSD. Direct closure was done initially and then a secondary Dacron patch was performed on the third postoperative day. All the remaining 15 patients have survived. The mean follow-up period has been 11.5 years (range: 1–21 years). Twelve patients (75%) were found to be in the New York Heart Association (NYHA) functional class I and three patients were in the NYHA class II. One patient, who is now in NYHA class III, had associated moderate aortic regurgitation and this resulted in valvuloplasty during the operation. He had developed progressive aortic regurgitation in recent years and aortic valve replacement is now being considered. Except for this patient, there has been no recurrence of fistula between the coronary sinus and the cardiac chamber following repair of ruptured ASV in the presented series.

DISCUSSION

Ruptured ASV was first reported by Thurnam in 1840, in which he described the clinical features of an abnormal communication between the aorta and the pulmonary circulation. It is regarded as a rare condition and its incidence is only 0.09% in a large older autopsy series, but was in 0.14% to 0.23% in a Western surgical series, and 0.46% to 3.5% in a surgical series from the Orient. Ruptured ASV occurs rarely in infancy, and in our and other series, the majority of patients have surgery between the ages of 20 and 40 years.

In the past, the occurrence of these aneurysms was thought to be of syphilitic or bacterial origin.
TABLE 1

<table>
<thead>
<tr>
<th>Origin</th>
<th>Total (%)</th>
<th>Ruptured Chamber</th>
<th>Number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>RCS</td>
<td>12</td>
<td>RV</td>
<td>11 (64.7%)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>RA</td>
<td>1 (5.9%)</td>
</tr>
<tr>
<td>NCS</td>
<td>4</td>
<td>RA</td>
<td>2 (11.8)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>RV</td>
<td>1 (5.9%)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>LV</td>
<td>1 (5.9%)</td>
</tr>
<tr>
<td>LCS</td>
<td>1</td>
<td>LV</td>
<td>1 (5.9%)</td>
</tr>
</tbody>
</table>

RCS = right coronary sinus; NCS = non-coronary sinus; LCS = left coronary sinus; RV = right ventricle; RA = right atrium; LV = left ventricle.

However, it is now generally accepted that either a congenital lack of continuity between the aortic media and the annulus fibrosus, or a developmental structural defect in the aortic annulus itself can gradually give way under aortic pressure to form an aneurysm. Additionally, other disease processes involving the aortic root and sinuses can also be associated with aneurysms of the sinus of Valsalva. They include endocarditis, cystic medial necrosis, atherosclerosis, inherited connective tissue disorders, and trauma.

Rupture of ASV rarely occurs asymptptomatically. The presentations of ruptured ASV ranged from asymptptomatically to frank heart failure due to a hyperdynamic cardiac state. The infrequency of severe symptoms at the time of rupture in the majority of patients may be because most ruptures are initially small. More than 50% of patients are reported to have shown a gradual onset of complaints, including shortness of breath, fatigability, palpitations, and tachycardia. Usually, a continuous “to and fro” murmur best heard on the left side of the sternum is present. Radiological signs of pulmonary hypertension are frequently evident. Although the rupture usually causes pathological changes, electrocardiographic findings are highly inconsistent. However, complete heart block may occur suddenly (Case 1) from compression of the bundle of His, but this is exceedingly rare.

Ruptured ASV originates most frequently from the right coronary sinus (65–85%), less frequently from the noncoronary sinus (10–30%), and rarely from the left coronary sinus (<5%). It has been observed that if the noncoronary sinus is involved, most aneurysms eroded into the right atrium. In this series, four of the patients had ruptured noncoronary ASV and two of these aneurysms terminated in the right atrium.

TABLE 2

<table>
<thead>
<tr>
<th>Association condition</th>
<th>Number</th>
<th>Percent</th>
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<tbody>
<tr>
<td>VSD</td>
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<td>41.2%</td>
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<tr>
<td>Supracristal:5</td>
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<td></td>
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<tr>
<td>Infra:crstal:2</td>
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<td></td>
</tr>
<tr>
<td>AR</td>
<td>8</td>
<td>47%</td>
</tr>
<tr>
<td>Mild:1</td>
<td></td>
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</tr>
<tr>
<td>Moderate:1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Severe:6</td>
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<td></td>
</tr>
<tr>
<td>Coronary artery fistula</td>
<td>1</td>
<td>5.9%</td>
</tr>
</tbody>
</table>

VSD = ventricular septal defect; AR = aortic regurgitation.
REFERENCES


