

Case Report

Congenital Malformations of the Aortic Root: Bicuspid Aortic Valve in Combination with Unruptured Aneurysm of the Left Sinus of Valsalva and Aberrant Left Coronary Artery

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Key words:

Bicuspid aortic valve, unruptured aneurysm of left sinus of Valsalva, left heart malformations.

We describe the case of a 53-year-old man with a severely stenotic bicuspid aortic valve combined with an unruptured aneurysm of the left sinus of Valsalva and an aberrant left coronary artery. The patient was successfully treated with aortic valve replacement and closure of the aneurysm. It is well known that patients with a bicuspid aortic valve have an increased incidence of other congenital anomalies, but the combination presented in our case is very rare.

An unruptured aneurysm of the left sinus of Valsalva is extremely rare (1%). It is usually associated with other congenital malformations of the heart. In our case there were three such congenital anomalies. A bicuspid aortic valve was combined with an aneurysm of the left sinus of Valsalva, as well as an aberrant left coronary artery origin with separate ostia for the left anterior descending and circumflex arteries.

Case presentation

A 53-year-old man was admitted to the neurology department after an episode of generalised seizures accompanied by loss of consciousness for a few seconds and no memory of the event. His previous medical history was clear, except for a well functioning bicuspid aortic valve with mild calcification that had been found during a routine transthoracic echocardiogram 10 years before.

The patient underwent all the appropri-

ate clinical and laboratory investigations, including brain scans with computed tomography and magnetic resonance imaging, but there was no obvious reason to explain the episode of seizures.

A new transthoracic echocardiogram confirmed the presence of a bicuspid aortic valve but this time the calcification was severe, causing aortic stenosis with a peak gradient of 96 mmHg, a mean gradient of 61 mmHg, as well as mild regurgitation. The left atrium was dilated, as was the ascending aorta (4.2 cm). The left ventricle was hypertrophic with good systolic function and an ejection fraction of 60%. There was no coarctation and pulmonary flow was normal.

The patient was put on antiepileptic medication and scheduled for a cardiac electrophysiological study. In the meantime he was discharged home in a very good condition. While at home the patient had a second episode of seizures exactly like the first one and he was promptly readmitted. The cardiac electrophysiological study revealed

Manuscript received:
January 30, 2008;
Accepted:
March 24, 2008.

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a prolonged HV interval, and a permanent pacemaker was implanted.

The severe calcification of the aortic valve, in combination with the prolonged HV time interval and the negative neurological tests, led us to suggest aortic valve replacement. During preoperative evaluation the patient had a coronary artery catheterisation that revealed normal coronary arteries without stenosis and a short or absent left main stem (Figure 1). The scheduled operation was performed under cardiopulmonary bypass, mild hypothermia, and with the perfusion of antegrade root and intracoronary crystalloid cardioplegia for myocardial protection. After the aortotomy the findings were as follows:

- Severely calcified bicuspid aortic valve with almost akinetic cusps.
- Aneurysm of the left sinus of Valsalva with an opening of 2.5 by 3 cm and a depth of 2 cm. The wall of the aneurysm was thin with no signs of rupture.
- Separate origins for the left anterior descending and circumflex arteries, which were adjacent to each other and over the opening of the aneurysm.

The calcified native valve was removed and replaced with a mechanical bileaflet valve (a size 23, St. Jude Regent aortic valve) which was placed in a supra-annular position. The same single mattress sutures were used to hold down the mechanical valve to the annulus, as well as obliterating the opening of the aneurysm. Thus, the 2-0 pledgeted sutures near the opening of the aneurysm were placed first at the aortic annulus, then the edges of the opening of the aneurysm, and finally at the suture cuff of the mechanical valve (Figures 2-4). The result was satisfactory.

The operation was complicated by bleeding from the aortotomy and we had to reinstate cardiopulmonary bypass in order to control it. In the intensive care

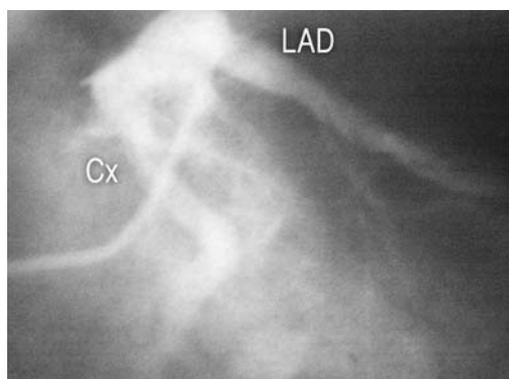


Figure 1. Coronary catheterisation. Cx – circumflex coronary artery; LAD – left anterior descending coronary artery.

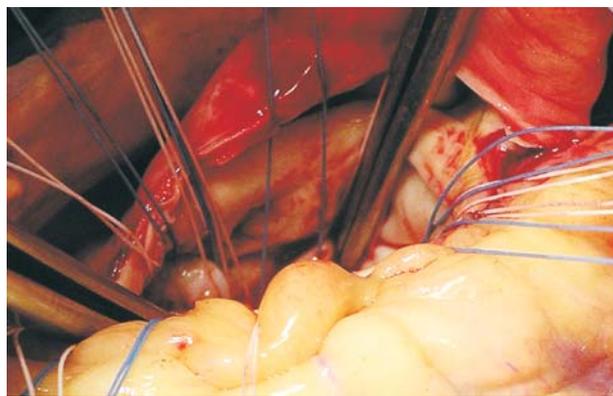


Figure 2. The placing of the sutures.

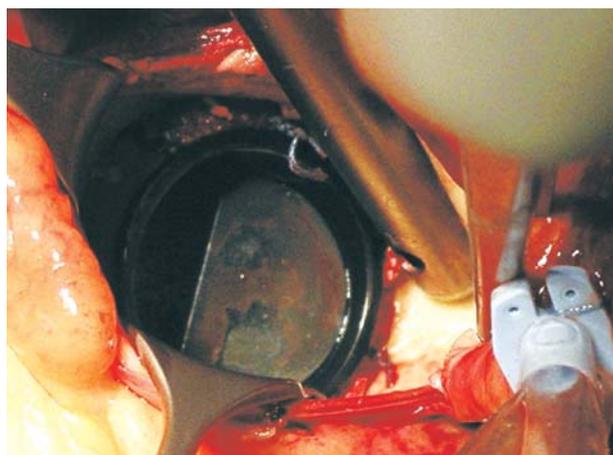


Figure 3. The result; the opening has been closed and the mechanical valve is in place.

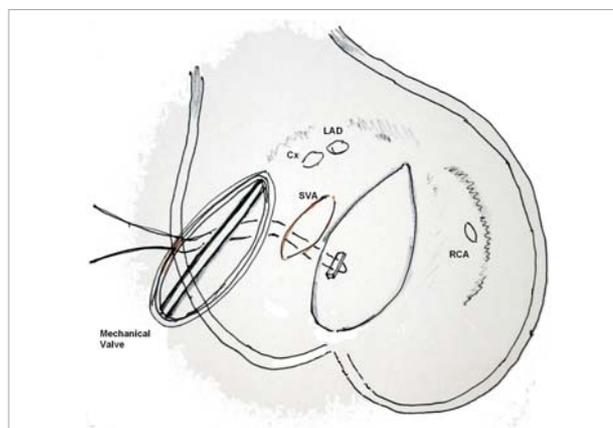


Figure 4. Drawing shows the surgical treatment: the oval opening of the aneurysm is closed by the same sutures that will hold the mechanical valve down. Cx – circumflex coronary artery; LAD – left anterior descending coronary artery; RCA – right coronary artery; SVA – opening of the aneurysm.

unit the patient had a bleeding diathesis. The amount of blood loss was 150-200 ml per hour. This time we administered recombinant factor VII (rFVIIa, Novo Seven[®], Novo Nordisk). The amount of blood loss diminished dramatically and after 6 hours the bleeding stopped. The rest of the postoperative course was uneventful. The patient was discharged from hospital on the 11th postoperative day. One and a half years later he is in perfect health.

Discussion

The association between congenitally abnormal bicuspid aortic valves and other congenital abnormalities is well established.¹⁻³ Most commonly, the condition is associated with aortic coarctation, hypoplastic left ventricle, reversal of dominance of coronary arteries, atrial septal defect, ventricular septal defect and dilated ascending aorta. In the case presented here, the congenital bicuspid aortic valve was associated with an unruptured congenital aneurysm of the left sinus of Valsalva and an aberrant left coronary artery origin.

Sinus of Valsalva aneurysms (SVA) are not frequent anomalies.⁴ The majority are congenital, caused by a defect in the continuity of the aortic media with the fibrous ring of the aortic valve.^{1,4-6} In contrast, acquired SVAs can be caused by bacterial endocarditis, syphilis, atherosclerosis or trauma.

Of congenital SVAs, those of the right coronary sinus are most common with a frequency of 94%, followed by those of the non-coronary sinus with a frequency of 5%. Left coronary sinus aneurysms are extremely rare and their frequency is only 1%.^{3,5} A congenital SVA can be associated with other congenital anomalies, such as ventricular septal defect,⁷ atrial septal defect, and bicuspid aortic valve. If not ruptured, SVAs can remain asymptomatic but if they are untreated they can cause complications depending on the anatomical site, such as right ventricular outflow tract obstruction, coronary artery occlusion or compression, aortic valve regurgitation, malignant arrhythmias (complete heart block, ventricular tachycardia), formation of emboli and infection (endocarditis). The most dramatic complication of SVA is abrupt rupture, either intracardiac or extracardiac. Rupture into the pericardium is even more dramatic because it can cause cardiac tamponade and death.^{3,6}

Because the risk of complications is high, SVAs must be treated immediately after the diagnosis is made. Surgical treatment is the treatment of choice.^{6,8} The technique is based on closing the opening of the

aneurysm. For this, the surgeon can choose either closure with a patch (most often Dacron), or the technique of direct closure using single mattress sutures with pledgets.⁹ In our case we chose to close the opening of the aneurysm with the same sutures we used to tie down the mechanical valve on to the aortic valve annulus. Therefore, we placed the sutures first at the aortic annulus, then at the edges of the opening of the aneurysm and finally at the suture cuff of the mechanical aortic valve. When we tied down the sutures the opening was obliterated by the aortic valve annulus and the pledget on the one side and the suture cuff of the mechanical valve on the other, thereby reinforcing the friable tissue of the aneurysm. The end result was quite good.

A review of the literature found no previous descriptions of this technique, although several surgeons chose the technique of direct closure with single pledgeted sutures.⁸

Bicuspid aortic valve is a congenital anomaly of the aortic valve with a frequency of 0.9 to 1.36% in the general population.² Its natural history is quick degeneration, thickening and calcification of the leaflets. Eventually, one third of all bicuspid valves will function normally, one third of them will result in aortic stenosis, and the final third will develop various degrees of stenosis and regurgitation.¹⁰

There is a significantly higher incidence of both a short left main coronary artery (<10 mm) and immediate bifurcation of the two major branches of the left coronary artery in patients with bicuspid aortic valves, compared with patients without evidence of bicuspid aortic valve disease. There have been a few reports concerning the combination of a bicuspid aortic valve and aberrant coronary artery origin as in our case.¹¹ These anatomical variations of the left coronary artery do not occur on a chance basis, and may be related to the developmental abnormalities responsible for bicuspid aortic valve disease.

The combination of bicuspid aortic valve, sinus of Valsalva aneurysm and aberrant left coronary artery origin may suggest that genetic mechanisms interfering with the histogenesis of the aorta and coronaries are responsible for such anomalies.¹¹

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