CASE REPORT

Unruptured congenital aneurysm of the left sinus of Valsalva in an adult with complex left heart malformations

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A 26 year old man who presented with the first signs of right heart failure was found to have a large congenital aneurysm of the aortic sinus of Valsalva and of the left coronary sinus. These were combined with left heart anomalies in the form of a bicuspid aortic valve, a rare variant of a persistent left superior vena cava with blood flow from the left atrium through the brachiocephalic vein into the superior vena cava and a kink in the aortic arch. An aortic coarctation had been corrected with a patch 12 years earlier. Although the aneurysm was not perforated and there were no clinical signs of infarction, the aneurysm was resected prophylactically and the left coronary artery was reinserted through a bypass with a Gore-Tex conduit. The case is noteworthy because this diagnosis is very rare and its early treatment may prevent several complications. The clinical features, treatment, and outcome are discussed.

Congenital aneurysm of the left coronary sinus of Valsalva is rare.¹ More often congenital aneurysms of the non-coronary or the right coronary sinus are described in the literature. The distribution has been stated to be approximately 70% for the right coronary sinus, 29% for the non-coronary sinus, and 1% for the left coronary sinus.^{2,3} An aneurysm of the right coronary sinus can penetrate the atrium, the membranous ventricular septum, or the conus septum and can obstruct the right outflow tract.¹ More often we see the acquired form, which occurs spontaneously after chest trauma or bacterial endocarditis.⁴ The onset is usually acute with no history of heart disease. We report on a young man with left heart anomalies and an unruptured sinus of Valsalva aneurysm of the left coronary sinus.

CASE REPORT

A 26 year old man, who had full time employment, presented with right heart failure 12 years after the surgical correction of an aortic coarctation. During this 12 year period he had had an uneventful course without follow up. Echocardiography showed a kink, without gradient, in the aortic isthmus, a bicuspid aortic valve without stenosis or severe regurgitation, and an atypical persistent left superior vena cava with a left to right shunt from the left atrium through the left brachiocephalic vein and the right superior cava vein in to the right atrium. Oxygen saturation in the superior cava vein was 94%. The dilated ostium of the left anterior descending artery arose from an aneurysm of the left sinus of Valsalva (fig 1). The diameter of the aneurysm was $2.7 \times 3.0 \times 2.5$ cm and there were no signs of perforation. Magnetic resonance imaging confirmed these findings. On the ECG we saw no signs of infarction and no arrhythmia.

Surgery

The aneurysm was repaired using standard cardiopulmonary bypass with moderate hypothermia and cold crystalloid cardioplegia for myocardial preservation. After ligation of the left superior vena cava, an incision through the aorta enabled us to view the aortic root with the large thin walled aneurysm in the left coronary sinus. The coronary artery exited from the dorsal wall of the aneurysm. After the aneurysm had been opened the coronary artery was severed and reinserted into the ascending aorta through a bypass with a Gore-Tex conduit 8 mm in width and 40 mm in length (fig 2). The wall defect in the ascending aorta was closed using a Vascutech patch (Sulzer Medica, Hamburg, Germany) before reanastomosis of the aorta.

Immunohistology of the resected aneurysm showed smooth muscle fibres in irregular slices. There was no intima or elastic fibres.

The postoperative course was uneventful and the patient was discharged from hospital on the 12th day. Three years

Figure 1 Angiography: (A) Aortic root injection shows the bicuspid aortic valve with mild regurgitation and the aneurysm (arrow). (B) Direct injection into the aneurysm shows its extent (arrow), and confirms its unruptured nature and the origin of the left coronary artery (broken arrow); asc.ao, ascending aorta.

Figure 2 Relation of the aneurysm of the left coronary sinus of Valsalva to the left coronary artery (LCA) and the bicuspid aortic valve. (A) Before repair. (B) After resection and repair with Gore-Tex bypass graft (BG).

after surgery, he continued to be in good condition on follow up examination. The coronary artery angiography performed three years postoperatively showed the best possible results—no stenosis, good ejection fraction, and good flow in the coronary artery prosthetic graft.

DISCUSSION

An aneurysm of the left sinus of Valsalva is extremely rare. Edwards and Burchell⁷ found a lack of continuity of the aortic media between a congenital sinus of Valsalva aneurysm and the aortic valve ring. Our immunohistology confirmed this pathogenic aspect.

An unruptured congenital aneurysm of a sinus of Valsalva does not usually cause symptoms but if it remains unresected it can result in various risks, such as right ventricular outflow tract obstruction,⁶ coronary artery occlusion⁷ or compression,⁸ aortic regurgitation,⁹ complete heart block,¹⁰ and resistant ventricular tachycardia.¹⁰

Shahrabani and Jairaj⁷ reported on a patient with cerebrovascular embolism caused by an intraluminal thrombus that displaced the right coronary artery ostium.

In addition to the above, there is the risk of spontaneous rupture, especially of the left coronary sinus, which carries a very high mortality rate due to cardiac tamponade.

In the present case, cardiac surgery had to be performed because of an immense left to right shunt produced by the left superior vena cava. We decided to use extracorporeal circulation for the repair of the aneurysm of the sinus of Valsalva, which was undertaken in the same session because of the danger of stroke or congestive heart failure caused by infarction, which was considered to be a greater risk than the surgical risk of correction.

Early prophylactic surgical treatment of the aneurysm makes the operation simple and prevents the development of complications, such as spontaneous rupture, thrombosis of the aneurysm with subsequent closure of the coronary artery, emboli of the cerebral arteries, or kinking of the coronary arteries. On follow up examination three years postoperatively there was no stenosis in the left coronary artery prosthetic graft and good flow through it.

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