

Ruptured right sinus of valsalva aneurysm with bicuspid aortic valve

Case Report

Abdul R. Hakeem*, Telal O. Mudawi, William L. Morrison

Department of Cardiology, Liverpool Heart and Chest Hospital, Liverpool L14 3PE, United Kingdom

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Abstract: The sinus of Valsalva aneurysm is a relatively rare condition that may be congenital or acquired. We present a case of a young man who was admitted to the emergency department with acute dyspnoea whilst playing football. After investigation, the diagnosis of a ruptured right sinus of Valsalva aneurysm into the right atrium was made. The patient also had a bicuspid aortic valve. Percutaneous approach to close the fistula was unsuccessful and the patient underwent surgical closure of the fistula with replacement of his bicuspid aortic valve.

Keywords: Sinus of Valsalva aneurysm • Transthoracic Echocardiography • Transoesophageal Echocardiography • Fistula closure devices

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1. Case Report

A 23-year old male presented to the emergency department of a district general hospital with sudden onset of breathlessness whilst playing football. There was no preceding history of any chest pain or trauma. There was no previous cardiac history. A provisional diagnosis of ruptured sinus of Valsalva was made on the basis of clinical features and transthoracic echocardiography. The patient was then transferred to our tertiary cardiac centre for further management.

At presentation, he was icteric with a sinus tachycardia of 130 beats per minute and a blood pressure of 110/50. There was a loud rasping continuous machinery murmur and a right ventricular gallop rhythm. There was epigastric tenderness and the liver edge was palpable and pulsatile. Blood results revealed normal full blood count, electrolytes and renal function. There was metabolic acidosis with a calculated base-excess of -8. Liver function was deranged with Gamma GT = 279 IU/L, Alkaline Phosphatase = 269 IU/L, Alanine Transaminase = 78 IU/L and total Bilirubin = 67 IU/L. Chest x-ray showed mild cardiomegaly and a small right-sided pleural effusion. A 12-lead ECG showed P-pulmonale and voltage criteria of left ventricular hypertrophy

(LVH). A Transoesophageal Echocardiogram (TOE) demonstrated a sinus of Valsalva fistula with rupture into the right atrium at the base of the tricuspid valve. The right heart was dilated and the aortic valve was bicuspid but not stenosed or regurgitant.

Cardiac catheterisation revealed an elevated right atrial pressure of 26 mmHg, pulmonary arterial pressure of 38/20 mmHg and aortic pressure of 120/60 mmHg. There was a step up in the Oxygen saturation at the right atrium with a Qp:Qs ratio of 4.6:1. Aortogram showed a fistula extending from the right coronary cusp into the right side of the heart (Figure 1). The neck of the fistula measured between 11 and 14 mm. A CT scan of the thorax confirmed the findings and measured the distance between the fistula and the right coronary artery to be around 12 mm.

After liaising with the cardiac surgeons, it was decided to close the fistula percutaneously under fluoroscopy and TOE guidance. The fistula was crossed with a probe and an arterio-venous loop was created. The neck of the fistula measured around 12-13 mm. A 14 mm AGA PDA occluder was found to be too small as it pulled through the defect. A 16 mm occluder did initially sit in a good position, but upon releasing the device from the delivery cable, its position became partially twisted and flow through the fistula was seen on colour Doppler. Further

* E-mail: drhabdulrahman@yahoo.com

attempts were abandoned as the fistula appeared too large for percutaneous closure and it was then decided to proceed for surgery to excise the fistula.

During surgery the Amplatzer device was removed. The aortic root was completely transected at the level of the sino-tubular junction and was closed with mattress sutures. Tricuspid valve leaflets were repaired and the bicuspid aortic valve was replaced with a mechanical device. The decision to replace the valve during the operation had to be weighed against the implications of long-term anticoagulant therapy in a young patient. While leaving the native bicuspid non-stenotic valve was certainly a valid option that may be supported by many cardiologists and cardiac surgeons, our surgical colleagues felt that the risk of post-operative severe aortic regurgitation [1,2] was significant enough to justify replacing the valve with a mechanical device.

The patient had a prolonged and complicated post-operative recovery. He initially had an episode of tamponade requiring re-exploration and removal of clots from the pericardial cavity. He then developed acute renal failure requiring continuous haemo-filtration followed by bilateral pneumonia causing septicaemia. He also developed left-sided radiculopathy of C5 and C6 nerve roots that was treated by physiotherapy. He eventually fully regained his renal function and responded well to antibiotic therapy. Post-operative transthoracic echo showed only mild aortic and tricuspid regurgitation. He was discharged back to his local district hospital to continue an intensive physiotherapy programme, and to remain on long-term anticoagulant therapy for his mechanical aortic valve.

2. Discussion

Rupture of sinus of Valsalva aneurysm is a rare entity and its association with bicuspid aortic valve is much rarer with only 4 previous reported cases of such a combination. The minimum age of such presentation was 20 years as reported by Hanai *et al.* in May 1998 [1]. Azakie *et al* studied 34 patients with congenital ruptured sinus of Valsalva aneurysms (RSVA) where the mean age was found to be 31.6 years [2]. Our patient was 23 years old at presentation and had no previous known cardiac history.

Sinuses of Valsalva are three localized bulgings in the aortic root opposite the aortic valve cusps. The right and left sinuses give origin to the respective right and left coronary arteries and the third one is the non-coronary sinus, which lies opposite the non-coronary aortic cusp. Patients with bicuspid aortic valves have only two sinuses and therefore it is presumed that there

is excessive pressure on each coronary sinus than in a normal valve with three sinuses [3]. This causes subsequent aneurysmal enlargement of the sinuses [4] and in certain cases may rupture into any cardiac chamber, such as the right heart chambers [5], the interventricular septum [6] or the pericardial space [7].

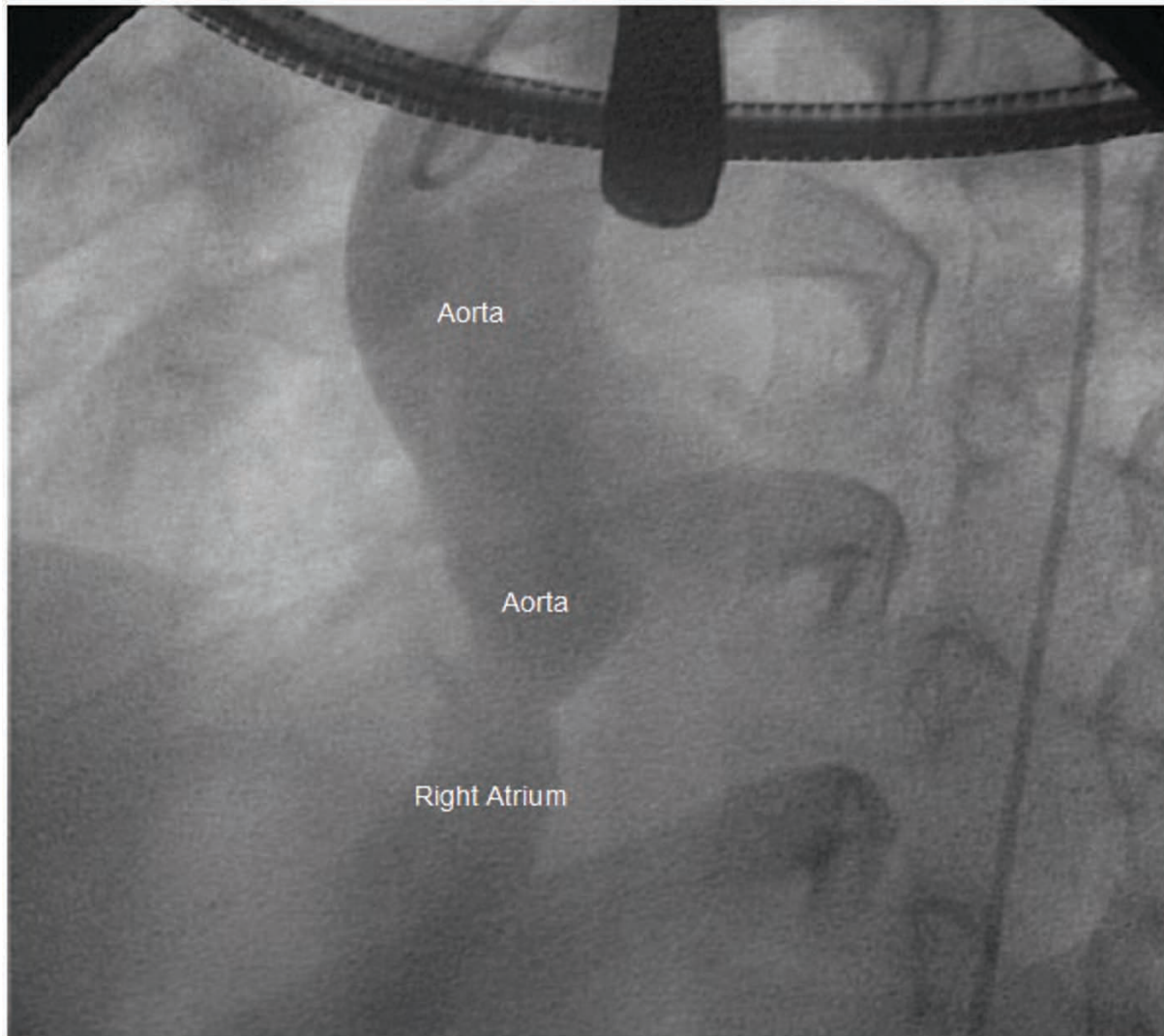
Sinus of Valsalva aneurysms may be congenital, associated with aortic valve anomalies and VSD, or it may be acquired [8]. Causes of the latter include conditions that affect the aortic root such as Syphilis [9], Marfan's Syndrome [10], Infective Endocarditis, Ankylosing Spondylitis [11], Rheumatoid Arthritis [12] or trauma, all of which may lead to weakness of the media layer followed by generalized dilatation of one or more sinuses, usually the right one [13].

Echocardiography, both transthoracic and transoesophageal are the two most important diagnostic tools [14]. Along with them, cardiac catheterisation, CT angiography and magnetic resonance imaging (MRI) are useful to both confirm the diagnosis and plan for suitable surgical or percutaneous management [15].

An algorithm for managing patients with sinus of Valsalva aneurysms, ruptured or un-ruptured, was formulated by Vural *et al.* [16]; they used a simple and functional classification of the therapeutic approach based on the clinical picture and the echocardiographic findings. It states that surgical intervention is required for both ruptured aneurysms and symptomatic unruptured ones. For unruptured asymptomatic aneurysms, surgical treatment becomes necessary if the aneurysm size exceeds 50% of the average size of the other two normal Valsalva sinuses or if the aneurysm size is seen to be increasing on consecutive echocardiographs. The algorithm also recommended surgery if there is compression of the adjacent tissues.

Percutaneous closure of the fistula using devices like Amplatzer occluders can be attempted, but its effectiveness in such rupture cases still needs answering [17]. Early surgical intervention is generally encouraged in view of the low peri-operative risk, satisfactory correction of the fistula and excellent prognosis with subsequent normal life expectancy [18,19]. Surgical repair of such fistulas include direct or patch closure of the aortic defect, while protecting the aortic valve and origins of the coronary arteries [20]. Although it currently has a limited value, percutaneous trans-catheter fistula repair may, with advancing technology, supersede surgery in becoming the treatment of choice in the near future.

Figure 1. The aortogram showing contrast dye filling the right atrium through the ruptured sinus of Valsalva fistulous connection.



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