

Case Report

Ruptured Sinus of Valsalva Aneurysm and Bicuspid Aortic Valve with Coarctation of Aorta in Young Male

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ABSTRACT

Coarctation of aorta and sinus of Valsalva aneurysm are frequently silent congenital cardiac defects that their diagnosis and management might be delayed. Coincidence of these cardiac defects is unusual. Here, we present a patient with bicuspid valve with coarctation of aorta and ruptured non-coronary sinus of Valsalva aneurysm leading to aorto-right atrial shunt and refractory heart failure.

Key words: Bicuspid Aortic Valve, Coarctation of Aorta, Transthoracic echocardiography.

INTRODUCTION

Sinus of Valsalva aneurysm (SVA) is usually referred to as a rare congenital anomaly. Its origin may be either acquired or congenital. In general, congenital SVA is clinically silent but may vary from a mild, asymptomatic dilatation to symptomatic presentations related to the compression of adjacent structures or intracardiac shunting caused by rupture of the SVA into the right side of the heart.

Nearly 65-85% of SVAs emerge from the right sinus of Valsalva and the most common complication is rupture into the right atrium or ventricle. However, the potential risk of rupture, cardiac failure, stroke, and sudden death has to consider surgical repair of unruptured aneurysms even if they are asymptomatic or incidentally detected, reporting a generally low early surgical and long term mortality. [1]

Coarctation of aorta (CoA) is a relatively common abnormality that occurs in approximately 6-8% of patients with

congenital heart disease. [2] The diagnosis of CoA may be missed unless an index of suspicion is maintained, and diagnosis is often delayed until the patient develops symptoms. Early diagnosis is possible in the first years of life in symptomatic patients whereas it might be delayed in asymptomatic patients. Untreated CoA might cause severe hypertension, intracranial bleeding, aortic aneurysm formation, and even aortic rupture. Additionally, infective endocarditis, early atherosclerosis due to intimal proliferation and degeneration in coronary arteries may be seen as well.

Association of coarctation of aorta and bicuspid aortic valve is common, but associated ruptures of sinus of Valsalva with these anomalies are unusual. We came across the patient with these three defects together.

CASE REPORT

A 25-year-old male, with no previous comorbidities and significant

family history, was admitted for abrupt onset of dyspnoea (New York Heart Association IV) over the previous two weeks. On examination, he was afebrile with a blood pressure of upper limb 150/70 mmHg and lower limb blood pressure of 130/70 respectively, with a heart rate of 90 bpm and signs of right heart failure. Jugular Venous Pressure (JVP) was elevated 13 cm of H₂O above sternal angle, and prominent 'v' wave. A continuous murmur was audible at the third and fourth intercostal space on the left sternal border. There were rales at both lung bases with pulsatile liver. The radial and femoral pulses were felt with slight delay. No pretibial oedema was observed, nor were there any remarkable laboratory findings. Chest radiography showed cardiac enlargement with mild congestion of the pulmonary vasculature with prominent rib notching. Electrocardiography indicated sinus rhythm without any sign of hypertrophy. Transthoracic echocardiography showed a non-coronary cusp ruptured into the RA, bicuspid aortic valve [Fig 1, 2]. Left ventricular (LV) dimensions and ejection fraction were normal. Doppler analysis highlighted a left-to-right high-speed shunt flow from the non-coronary SVA to the right atrium. A severe aortic isthmus coarctation was also documented by aortogram and confirmed by CT Angio. No evidence of infective endocarditis or coexistence of associated sub aortic membranes was identified.

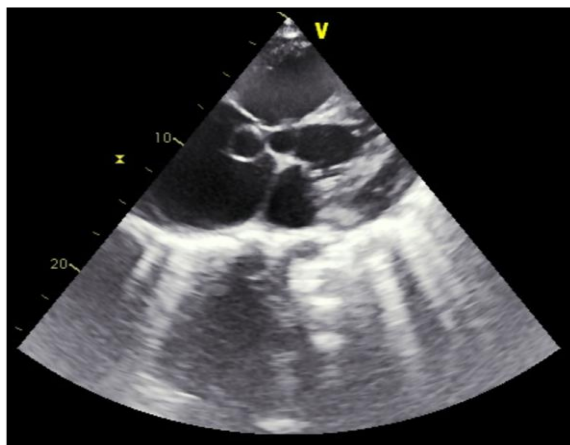


Figure 1: Transthoracic echocardiogram showing bicuspid aortic valve and noncoronary sinus of Valsalva aneurysm.

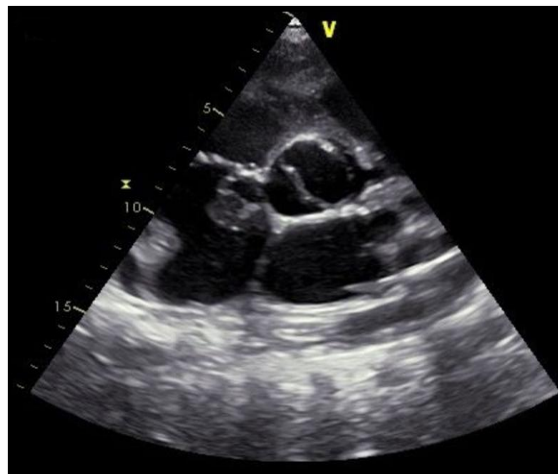


Figure 2: Transthoracic echocardiography showing a noncoronary cusp ruptured into the RA, bicuspid aortic valve.



Figure 3: Chest computed tomography (CT)-scan showing the aortic isthmus coarctation.

The patient was started with oxygen, iv diuretics, inotropes and other supportive measures. The Chest computed tomography (CT) was performed which confirmed the aortic isthmus coarctation with extensive intercostal and scapular collateral arteries [Fig. 3]. In view of persistent heart failure patient was advised surgery. Preoperatively patient decompensates and succumbed.

DISCUSSION

Coexisting lesions are common in patients with congenital ruptured SVA. [3,4] Sub arterial and perimembranous ventricular septal defects occur in Western patients with an incidence of 30-50%. Aortic regurgitation in ruptured SVA is also a commonly associated lesion, with 33.6% in the Asian group and 32.7% in the Western group. Bicuspid aortic valve occurs with an

incidence of 10-20%.^[4] Coexisting aortic coarctation was documented in 4% (3/86) of SVAs.^[3]

The association of ruptured SVA to aortic coarctation is exceptional and reported at a rate of only 1/57 of ruptured SVA.^[3] Only few cases have been described so far.^[5-7] Over a cohort of 580 corrected cases of aortic coarctation, Manganas et al have reported that 23 patients required operation after coarctation repair.^[6] Only one patient has been re-operated for a ruptured SVA (1/580 patients with aortic coarctation). In our case report, the aortic coarctation and bicuspid aorta were discovered by chance at adulthood after the rupture of the SVA.

For the correction of combined forms of aortic coarctation and SVA with or without rupture, different techniques have been employed. In the one-stage repair, simultaneous correction of both lesions through a median sternotomy was performed.^[8,9] The two-stage repair can be performed through a combination of lateral thoracotomy and median sternotomy. Hybrid approach was also employed with the combination of the interventional and the surgical approaches. The non-surgical treatment method of the aortic coarctation shown to offer a substantial benefit, transforming a sophisticated and risky surgical procedure into one of the common practice.^[10]

Surgery used to be the mainstay of treatment of ruptured SVA, however, in past few years several reports of transcatheter closure of ruptured SVA have come to light.^[7] Interventional treatment can be proposed to treat the two lesions with a device closure with a duct occluder device for the ruptured of SVA and percutaneous stent implantation for the aortic coarctation. Recently, Kerkar et al have reported the mid-term results of transcatheter closure using the first-generation Amplatzer duct occluder in patients with ruptured SVA who have no associated ventricular septal defect or aortic regurgitation.^[7] One patient had co-existing aortic coarctation. The rate of the procedure

success was 90% with encouraging short and mid-term outcomes.

Association of ruptured SVA to aortic coarctation is rare. Management of this complex association is based on early diagnosis and treatment. Hybrid approach or totally interventional approach may be advised based on clinical scenario.

Conflict of interest: All authors declare that they have no conflict of interest.

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