

Ruptured Sinus of Valsalva Aneurysm Mimicking a Flail Tricuspid Leaflet

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Citation

J Andrade, M Nicolae, K Ramanathan. *Ruptured Sinus of Valsalva Aneurysm Mimicking a Flail Tricuspid Leaflet*. The Internet Journal of Cardiology. 2008 Volume 7 Number 1.

Abstract

Sinus of Valsalva aneurysms are uncommon clinical entities that are associated with other congenital abnormalities. They most often they remain asymptomatic until abruptly presenting in adolescence and adulthood with acute chest pain and heart failure secondary to rupture. With prompt diagnosis and management the long-term prognosis remains excellent. We present below a case of acute rupture of a long sinus of Valsalva aneurysm into the right atrium. The length of the "windsock" as well as its close proximity to the tricuspid valve led to unique diagnostic and therapeutic issues as outlined below.

CASE REPORT

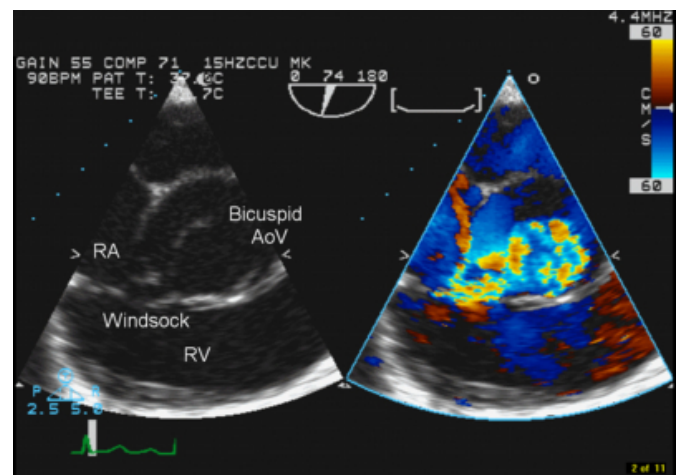
A 24-year-old woman with known bicuspid aortic valve and prior aortic coarctation end-to-end anastomotic repair (9 years of age), presented to a community hospital with a 24-hour history of unrelenting central chest pain and progressive right-sided heart failure. Initial 2D transthoracic echocardiogram suggested RV dilatation with flail tricuspid leaflet and a left to right shunt of undetermined origin and termination. As a result of the diagnostic uncertainty the patient was urgently transferred to our tertiary care center for further evaluation. Upon arrival she was hemodynamically stable but remained profoundly symptomatic with evidence of biventricular congestive cardiac failure. Physical examination revealed symmetric blood pressures of 150/70 and a heart rate of 100 bpm. Clinically there was evidence of RV and LV failure as well as a new continuous grade 4 murmur throughout the precordium but maximal at the left sternal edge in the 5th intercostal space. ECG revealed sinus tachycardia with a first degree AV block that was unchanged from previous examination. Chest X-ray revealed pulmonary congestion but was otherwise unremarkable.

Urgent transthoracic and trans-esophageal echocardiograms revealed normal biventricular systolic function. Arising from 3mm above the attachment of the aortic valve was a long fistulous "windsock" connection that terminated in right atrium at the level of the anterior leaflet of the tricuspid valve. Occasionally the "windsock" would prolapse through the tricuspid valve into the right ventricle giving the appearance of an extraneous echo target on transthoracic

imaging. Doppler interrogation revealed continuous flow from aorta to right atrium through the aneurysm with a peak gradient of 136 mmHg observed during diastole. (Figure A).

Figure 1

Figure A

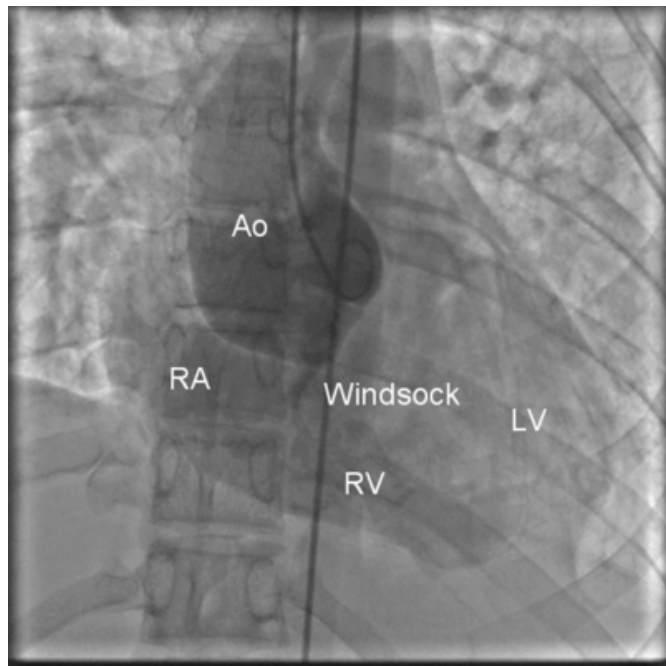


As she was known to have a membranous ventricular septal defect a decision was made to pursue cardiac catheterization to achieve diagnostic certainty, as at this time there was some debate surrounding the origin of her fistulous connection. Right-sided pressures were elevated with a right atrial pressure of 12 mmHg, pulmonary artery pressure of 47/31 mmHg with a mean of 37 mmHg, a pulmonary wedge pressure of 20 mmHg and a left ventricular end-diastolic pressure is 24 mmHg. There was a significant 14% rise in oxygen saturation between the right atrial and vena caval oxygen saturations. A single injection

was done at the aortic root which demonstrated a communication between the origin of the right sinus of Valsalva into the right atrium (RA), with eventual opacification of the right atrium, right ventricle (RV) and pulmonary arteries. (Figure B) There was no gradient at the coarctation repair site.

Figure 2

Figure B



Unfortunately, the location of the defect did not lend itself to closure with a percutaneous device. Due to the close proximity of the fistulous connection to the tricuspid valvular apparatus there was significant concern that implantation would interfere with the tricuspid valvular apparatus.

Due to her increasing symptoms and biventricular failure she was referred for emergency surgical repair. At cardiac surgery she was found to have an 8 mm diameter ruptured sinus of Valsalva aneurysm with classic windsock appearance. It was again noted to be overlying the anterolateral border of the tricuspid valve. The aneurysmal portion of the sinus of Valsalva was resected and oversewn with a 1.2 cm Dacron patch. The patient had an unremarkable recovery period and has done well since her procedure. Follow-up echocardiogram demonstrated only mild-moderate tricuspid regurgitation.

DISCUSSION

Sinus of Valsalva aneurysms are uncommon clinical entities but are known to be associated with up to 0.5-1.5% of

patients. They develop as a result of a separation of the aortic media of the sinus from the ventricular fibrous structures leading to a progressive protrusion into the low-pressure chamber due to the pressure differential. Most often the aneurysm begins in the right coronary sinus (80%) and terminates in the right ventricle (72%) but up to 15% of cases originate in the non-coronary sinus, which usually leads to a termination in the RA. (1).

Sinus of Valsalva aneurysms can be congenital or acquired as a result of injury, infection, or be related to Marfan syndrome or Behcet disease. Up to 47% of patients with sinus of Valsalva aneurysms are found to have an associated congenital defect. In one series an associated ventricular septal defect was most common (49.3%), followed by aortic insufficiency (23.4%), tricuspid regurgitation, infundibular pulmonary stenosis, patent ductus arteriosus and patent foramen ovale (each 1.9%). An association with bicuspid aortic valves and aortic coarctation has also been described. (2-3)

Although the aortic media deficiency is congenital most cases do not present until the third-fourth decade when, in 34% of cases, the asymptomatic aneurysm spontaneously ruptures. (4-5) This abrupt increase in left to right flow results in the classic clinical presentation of acute onset chest pain, palpitations and biventricular heart failure as well as a new loud continuous murmur with thrill throughout the precordium.

Chest radiography and ECG are usually nonspecific. Echocardiography is diagnostic in 65% however there can be some difficulty in distinguishing Valsalva aneurysms from coronary fistula. In this case cardiac catheterization with hemodynamic assessment and aortic root injection is confirmatory(4-5). Once diagnosed, medical therapy is merely a means of stabilization until definitive therapy can be undertaken.

Traditionally definitive management was provided by open-heart surgery with fistula resection and aortic reinforcement. This procedure has been associated with a less than 5% perioperative mortality and excellent long-term outcomes (>95% 20 year survival) with only minimal complications (1-2% residual leak, <1% aortic insufficiency or paravalvular leak)(3-9).

In recent years attempts have been made to avoid open-heart surgery through and close the defect through percutaneous insertion of an intravenous occluded device. The first case

series was published in 2004 by Arora et al. who reported the results from 8 male patients in NYHA class III-IV heart failure. (10) The size of the defect size ranged from 7-12 mm in diameter and had calculated Qp/Qs ratios of 2–3.5. There was some heterogeneity in the devices used where 2 patients received Rashkind umbrella devices and 6 patients received Amplatzer occluders. Of the eight only one patient required re-intervention and eventual surgical repair due to right ventricular outflow tract obstruction obstruction, residual shunting and unremitting hemolysis. Over 2-96 months of follow-up there was no device embolisation, no infective endocarditis and no aortic regurgitation. At the end of the series 6 patients remained asymptomatic with only one developing progressive heart failure.

More recently, Zhao reported the results of a case series of 10 patients (4 men and 6 women) aged from 7 years to 69 years who were all treated with implantation of an Amplatzer duct occluder via a femoral vein approach. (11) Defect size ranged from 2-10 mm (average 6.2 mm) with calculated Qp/Qs ratios of 1.19-2.7 (average 1.86). Half of the cases terminated in the RV and half terminated in the RA. In all cases the immediate, and 3 month results were excellent with no reported complications.

CONCLUSION

Sinus of Valsalva aneurysms represent a rare clinical entity for which the treatment is in constant evolution. Although uncommon the diagnosis can usually be made from a combination of the clinical examination, non-invasive echocardiography and cardiac catheterisation. Although open heart surgery remains the mainstay of management, early results are promising regarding the feasibility of

percutaneous closure.

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