CASOS CLÍNICOS CASE REPORTS

RUPTURED SINUS OF VALSALVA INTO THE RIGHT VENTRICLE: A NEW MANAGEMENT STRATEGY

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Abstract

We present 2 cases presented to the emergency department with shortness of breath (SOB). Their preoperative echocardiographies showed ruptured right sinus of Valsalva (RSOV) into the right ventricle (RV). Ventricular septal defect (VSD) was diagnosed only intraoperatively.

CASE 1

A 23 years-old Pakistani male patient with history of cardiac murmur, presented to the emergency department (ED) with palpitation and progressive SOB

for few weeks. His vital signs were normal, continuous murmur was heard over the left sternal border with clear chest. Blood investigations showed high pro-BNP. There were cardiomegaly and prominent broncho-vascular markings by chest x-ray. Pre-operative 2D transthoracic



Figure 1

(A) Apical 4 chamber TTE view; marked RV and atrium enlargement. (B) Apical 5 chamber TTE view, the arrow is pointing at the RSOV (C) Parasternal long axis view (PLAX). (D) Colour Doppler flow across the ruptured sinus. RA: right atrium, LV: left ventricle, LA: left atrium, AV: aortic valve, AO: aorta.

echocardiography (TTE) showed dilated sinus of Valsalva; 4.2cm in diameter with ruptured right coronary sinus into the right ventricle outflow tract (RVOT), no VSD could be detected. Ejection fraction (EF) was 57%, severe tricuspid regurgitation and severe pulmonary hypertension; 104mmHg.

Intra-operative findings were the same. Additionally, there was a sub-aortic VSD. He underwent repair of the RSOV and the VSD using a tailored synthetic hemashield Dacron patch and repair of the tricuspid valve. The postoperative course was uneventful.

CASE 2

A 35 years-old previously healthy Indian male patient, presented to ED with SOB for two days, gradual onset and progressive course. Chest showed continuous murmur over the left sternal border, decrease air entry on lungs bases with crepitation. Blood investigations showed markedly elevated pro-BNP. Chest X ray showed increased broncho-vascular marking with bilateral mild pleural effusion. Preoperative TTE and

TEE showed ruptured aneurysm of right coronary sinus of valsalva into the RVOT, no VSD could be detected, EF was 55%.

A small sub-aortic VSD was discovered intra-operatively in addition to the RSOV. The VSD was repaired using pledgetted proline stitch after repairing the Valsalva defect using a double tailored hemashield Dacron patch. The patient had the same postoperative course as the 1st patient. the non-coronary sinus (23%), and rarely in the left coronary sinus. $^{\rm 2}$

It is frequently associated with VSD (30-60%), bicuspid aortic valve (15-20%) and aortic regurgitation (44-50%).

Rupture of the aneurysmal sac may occur spontaneously or be precipitated by exertion, blunt trauma, or cardiac catheterization. Rupture into RV is most common (60% to 90%), right atrium (10%) and left atrium (2% to 3%).

Aneurysm of the sinus of Valsalva in Asian patients compared with Western series is characterized by a higher incidence, more originating from the right coronary sinus (85.8% vs 67.9%), more rupture into the RV (72.5% vs 60%), a higher incidence of association with VSD (52.4% vs 37.5%), and lower incidence of association with bicuspid aortic valve (0.6% vs 7.8%), both Asian and Western patient series have similar incidence of combination with aortic regurgitation (33.6% vs 32.7%).³

TTE identifies sinus of valsalva aneurysm in most cases. TEE may be necessary in as many as 25% of cases. The ruptured aneurysm by echocardiography frequently has a "wind-sock" appearance; that is an elongated tubular structure expanding and collapsing with the cardiac cycle. Some cases of RSOV aneurysm reported missed VSD. 3D echocardiography is usually useful in precise anatomical delineation of RSOV particularly if it's associated with VSD.⁴ In the presence of coexistent VSD, the large shunt of RSOV overlaps VSD flow which may be difficult to recognize on 2D echo. Magnetic resonance imaging (MRI) allows an exact presentation of the anatomy, including areas that are difficult to assess, so it may be considered in stable patients.



Figure 2

A) Midesophageal lateral axis view at 120 degrees with color. B) 3D TTE PLAX showing the ruptured sinus.

DISCUSSION

Incidence of Sinus of Valsalva aneurysms is 0.1% to 3.5% of all congenital heart defects,¹ male preponderance (4:1), high in Asian populations. The aneurysm is found most commonly in the right coronary sinus (77%), In our cases we believe that VSD's were missed preoperatively because either the large aneurysmal sacs were covering the VSD or because of the overlap of the two shunts. In addition, in the first case right ventricular pressure was high approaching systemic pressure which probably reduced the shunt across the VSD. Early and aggressive intervention is recommended to prevent endocarditis or enlargement of the ruptured aneurysm; the long-term results are excellent after surgical repair,⁵ with actuarial survival rate is 95% at 20 years. If left untreated, patients die of heart failure or endocarditis within 1 year after the onset of symptoms.

CONCLUSION

RSOV aneurysm need vigilant preoperative assessment for coexisting cardiac defects, including TEE, with 3D reconstruction if available. MRI is an option if the patient is stable and can tolerate it. Surgical repair is still the modality of choice in many cases.

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