

Is it really ruptured sinus of valsalva? The crucial role of comprehensive transesophageal echocardiography in clinical decision-making

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INTRODUCTION

Aneurysms of the sinuses of valsalva are rare congenital defects resulting from aortic wall weakness at the junction of annulus fibrosa and the aortic media at any of the 3 cusps.^[1] With reference to patients from eastern countries, the most common site of origin is right coronary sinus (65–85%) with nearly all rupturing into right chambers.^[1] Thus, an aneurysm of the right sinus of valsalva rupturing into the left ventricle is a very rare condition if at all accurately diagnosed.

There is literature available where ruptured sinus of valsalva aneurysm (RSOVA) could only be diagnosed upon transesophageal echocardiography (TEE) and was misdiagnosed on transthoracic echocardiography (TTE) and cardiac catheterization.^[2,3] However, this report is about a hemodynamically stable patient who was preoperatively identified as suffering from RSOVA, whereas the actual pathology was different and was only discovered on intraoperative TEE and influenced the surgical technique and cardiopulmonary bypass (CPB) conduct, improving the therapeutic outcome.

CASE HISTORY

A 49-year-old New York Heart Association grade III male patient presented with orthopnea and grade III dyspnea. His electrocardiogram showed left axis deviation and evidence of left

ventricular (LV) hypertrophy. The chest X-ray except for cardiomegaly with LV apex was inconclusive. Upon TTE, patient was diagnosed with RSOVA draining into the left ventricle. Cardiac catheterization and angiography further showed normal coronaries, right coronary cusp (RCC) aneurysm with rupture into left ventricle causing severe regurgitation [Figure 1], normal left ventricle function, absence of any ventricular septal defect (VSD), no gradient across left ventricle and aorta and $Q_p/Q_s = 1$. Patient was scheduled for RSOVA repair with aortic valve repair/replacement.

Following anesthetic induction, intraoperative multiplane, color Doppler, two-dimensional and three-dimensional (3D) - TEE examination was performed (iE33™; Phillips Medical Systems, Andover, MA, USA). While mid esophageal right ventricular inflow-outflow view revealed bicuspid aortic valve (BAV) with normally co-apting leaflets and created suspicion of perforated leaflet [Figure 2 and Video 1]; we observed an eccentric jet of aortic regurgitation upon color flow doppler in mid esophageal aortic valve long axis view with normal coronary sinuses which made us suspicious of the preoperative diagnosis of RSOV. Upon close observation in both mid esophageal aortic valve long axis view [Figure 3 and Video 2] and mid esophageal 5 chamber view [Figure 4] we witnessed a left-to-right shunt flow across VSD restricted by septal tricuspid leaflet. Furthermore, on mid esophageal 5 chamber view, the shunt flow

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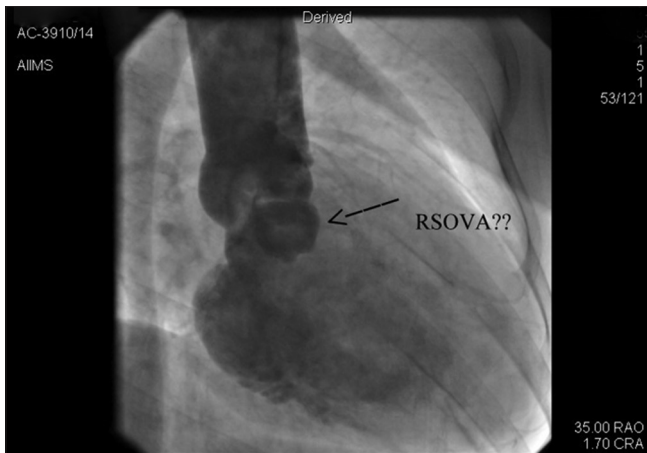


Figure 1: Root angiogram showing suspected rupture of sinus of valsalva aneurysm (arrow)

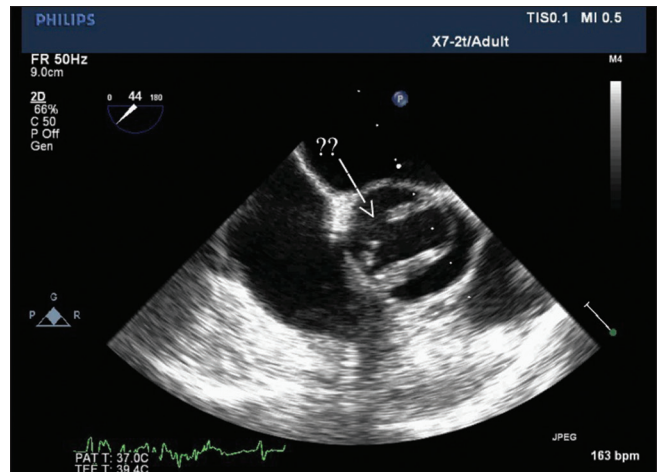


Figure 2: Mid esophageal right ventricular inflow-outflow view showing bicuspid aortic valve with normally co-apting leaflets and suspected perforation (arrow)

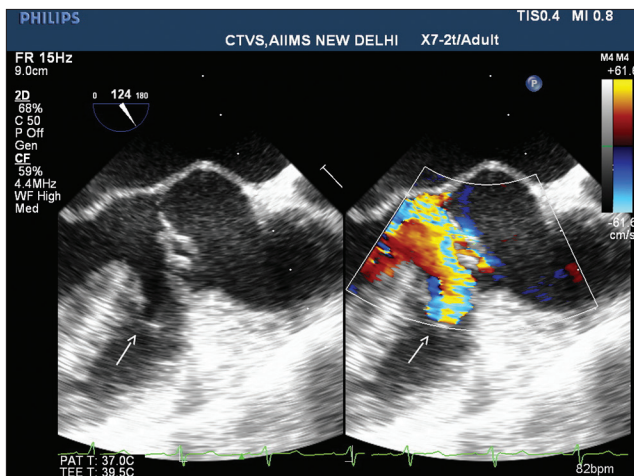


Figure 3: Mid esophageal aortic valve long axis view (color compare) showing left-to-right shunt flow across ventricular septal defect restricted by septal tricuspid leaflet (arrow)

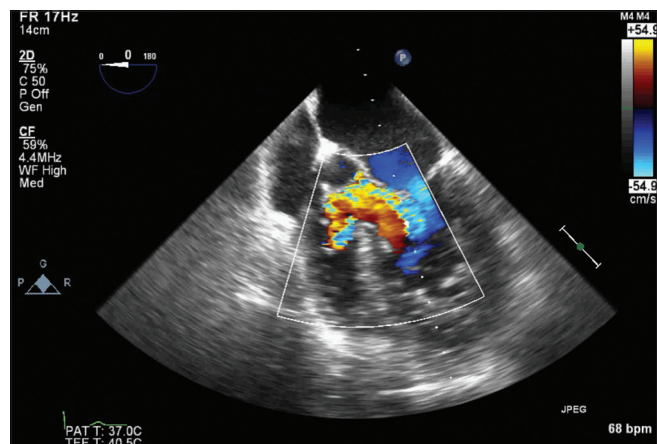


Figure 4: Mid esophageal 5 chamber view showing left-to-right shunt flow across ventricular septal defect restricted by septal tricuspid leaflet resembling a windsock deformity

looked like a windsock deformity [Figure 4] which was masquerading as ruptured RCC aneurysm in the opinion of a cardiologist.

OBSERVATIONS

The 3D examination of the aortic valve disclosed a perforated posterior coronary cusp [Figure 5] corroborating our finding. We conveyed the information to the surgical team, who then instead of dual-stage venous cannulation switched to bicaval venous cannulation for CPB in order to be prepared for trans-right atrial VSD closure, in case trans-aortic VSD closure was not amenable. The surgeons finally substantiated our finding of 3D TEE when they removed the perforated posterior coronary cusp en-bloc [Figure 6]. Following rewarming, patient was weaned off CPB successfully, with minimum inotropic

support [Figure 7a, b, and Video 3]. The patient was discharged on 5th postoperative day following an uneventful postoperative period.

DISCUSSION

Ruptured sinus of valsalva aneurysm often presents a diagnostic dilemma owing to its varied clinical presentations.^[4] Differential diagnosis of RSOVA from other acyanotic heart diseases accompanied by a continuous murmur includes-patent ductus arteriosus, pulmonary arteriovenous fistula, coronary arteriovenous fistula, VSD combined with aortic regurgitation and aortopulmonary window. Therefore, accurate preoperative diagnosis is essential to determine the urgency of surgery and technique of surgical, anesthetic and CPB approach and should be reaffirmed intraoperatively by TEE.

In surgical procedures involving the left side of the heart, dual-stage venous cannulation is usually the preferred method because it ensures the best decompression of the right heart and is fast and easy.^[5] Whereas, whenever

right atriotomy is required such as in the presence of a left-to-right communication, bicaval communication is mandatory.^[5] Dual stage venous cannulation, in presence of a left-to-right communication can entrain air upon opening the aorta, and thereby result in inadequate venous drainage or result in unnoticed continuous air lock in venous line, which can be catastrophic.^[5] Therefore, it is always crucial to identify any shunt flow present if at all and consequently modify the venous drainage technique for safe and uneventful CPB performance. A comprehensive intraoperative TEE evaluation in our case thus enabled us in accurately identifying a missed diagnosis and helped us guide our surgical colleagues to make necessary changes in surgical steps.

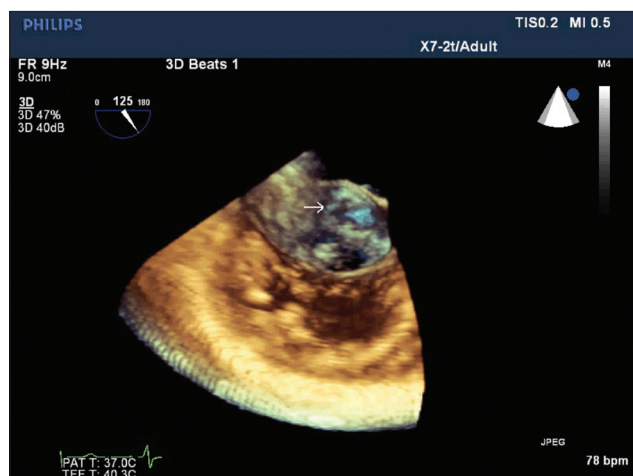


Figure 5: Three-dimensional examination (as seen from the left ventricle) of the aortic valve revealing a perforated posterior coronary cusp (arrow)

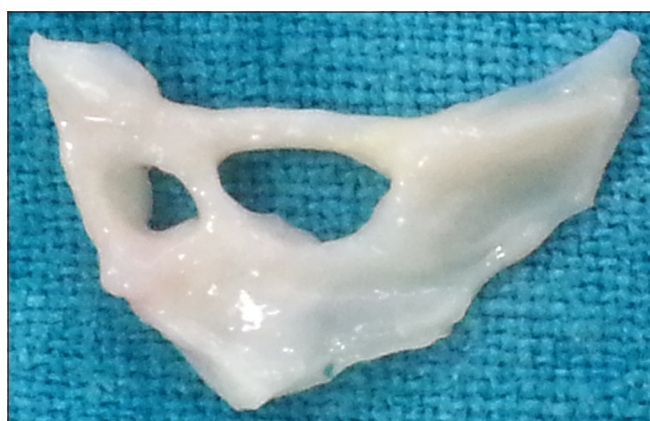


Figure 6: En-bloc removed posterior perforated coronary cusp

In the era of evidence-based medicine, with increasing awareness about health care provisions and legal rights, cardiac anesthesiologists are integral to clinical decision-making and therapeutic outcome. Over the years, TEE has fast emerged not only as an essential monitoring instrument but also as a therapeutic tool for a cardiac anesthesiologist, influencing the perioperative course and results. Although TEE exam for suspected RSOV finds no mention in the latest 2010 published American Society of Anesthesiologists guidelines for TEE, there is enough scientific literature available that emphasizes impact of perioperative multiplane, color Doppler TEE over TTE and angiography in improving surgical, anesthetic, and intensive care management of patients suspected of RSOVA.^[2,3,6-8] Garrido Martín *et al.* even concluded that TEE is the most accurate tool in the preoperative evaluation of RSOVA to right chambers.^[8] Furthermore, with real-time imaging at high temporal and spatial resolution, 3D advancements of TEE allow delineation of

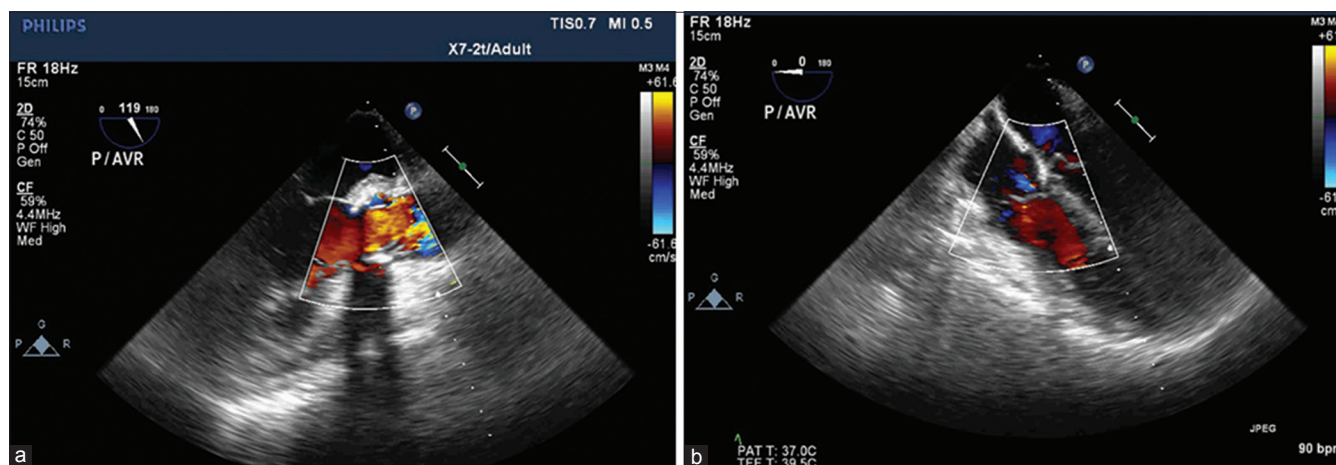


Figure 7: (a) Post aortic valve replacement (AVR) - mid esophageal aortic long axis view. (b) Post AVR - mid esophageal 4 chamber view

precise cardiac anatomy which leaves no room for error and has been reported to provide both diagnostic and procedure advantages in interventional cases.^[1,9]

CONCLUSION

To conclude whenever a rare pathology such as RSOVA draining into LV is encountered, other differential diagnosis including commonly associated lesions such as BAV and VSD should be suspected and must be thoroughly evaluated with intraoperative TEE.

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