

Case Report

Multiple ruptured aneurysm of left sinus of valsalva: A rare entity

Anju Sarupria, Poonam Malhotra Kapoor, Usha Kiran, Milind Hote¹

Departments of Cardiac Anaesthesia, and ¹CTVS, CN Centre, All India Institute of Medical Sciences, New Delhi, India

ABSTRACT

Aneurysm of sinus of Valsalva is a rare congenital cardiac defect that can present with myriad signs and symptoms ranging from trivial to catastrophic events like cardiogenic shock and death. As clinical examination is not entirely reliable and the patient can sometimes be so ill as to preclude cardiac catheterization, echocardiography has become the definitive investigative tool not only to define and diagnose the lesion but also to quantify its severity. The following is a case report of multiple aneurysms of the left aortic sinus of Valsalva rupturing into the left ventricle. Diagnosis is made on multi plane transesophageal echocardiography and color Doppler regarding precise identification of structural anomalies and shunt locations for perioperative assessment and definitive treatment is surgical repair.

Received: 10-05-10
Accepted: 28-08-10

Key words: Aneurysm, sinus of Valsalva, transesophageal echocardiography

INTRODUCTION

Aneurysm of sinus of Valsalva (SVA) is a rare congenital cardiac defect producing various clinical signs and symptoms. As clinical examination is not entirely reliable and the patient can sometimes be so ill as to preclude cardiac catheterization, echocardiography has become the definitive investigative tool not only to define and diagnose the lesion but also to quantify its severity. SVA is comparatively rare. When present, it is usually in either the right (65%–85%) or in the noncoronary (10%–30%) sinus, rarely in the left (<5%).^[1] The defect is typically single and starts as a blind pouch from a localized site in one of the aortic sinuses but rarely multiple sinuses may be affected.^[2] The following is a case report of multiple aneurysms of the left aortic sinus of Valsalva rupturing into the left ventricle.

Definitive diagnosis was made on multi plane transesophageal echocardiography (TEE) and color Doppler regarding precise identification of structural anomalies and shunt locations for perioperative assessment and treatment is surgical repair.

CASE REPORT

A 16-year boy presented with a short history of shortness of breath and chest pain for 5 days. Chest pain was intense radiating to scapular region associated with vomiting. Patient was evaluated at a local hospital; and was administered medication for pain relief. He was referred to tertiary care hospital. On examination, he was dyspneic at rest. He had raised jugular venous pulse and pedal edema. His pulse rate was 120/min regular, blood pressure (BP) was 140/20 mmHg, chest auscultation revealed bilateral crepitation with harsh end diastolic murmur in aortic area all laboratory investigations were within normal limit. Chest X-ray revealed cardiomegaly with dilated right atrium, left atrium, and left ventricle. Two-dimensional transthoracic echocardiography revealed severe aortic regurgitation with dilated aortic annulus (4.5 cm) and ruptured sinus of Valsalva into the left ventricular cavity and double barrel aorta with a false lumen. Chest tomography scan revealed dilated aortic annulus with dilated sinus of Valsalva mainly noncoronary and right coronary sinus. Left coronary sinus was normal. The patient was taken for

Access this article online

Website:
www.annals.in

PMID:

DOI:
10.4103/0971-9784.74400

Quick Response Code:



Address for correspondence: Dr. Poonam Malhotra Kapoor, Department of Cardiac Anaesthesia, CN Centre, All India Institute of Medical Sciences, Ansari Nagar, New Delhi 110 029, India. E-mail: drpoonamaliims@gmail.com

emergency surgery presumed diagnosed as a case of aortic dissection. However, intraoperative TEE, revealed the true picture. Color Doppler revealed severe aortic regurgitation. Non coronary sinus, ascending aorta and origin of coronary artery was normal [Figures 1 and 2]. Patient underwent aortic valve replacement (AVR) with St Jude valve with gortex patch closure of sinus of Valsalva aneurysm on cardiopulmonary bypass. Following AVR subsequent TEE examination revealed residual sinus of Valsalva aneurysm extending in aorto-mitral window still rupturing into left ventricular cavity [Figures 3 and 4]. Closure of this remaining aneurysm was performed via the left atrium. Total cardiopulmonary and aortic cross clamp time was (58 + 28) and (50 + 22) sec respectively. Postoperatively, the patient had an uneventful recovery, and he was well on his most recent follow-up.

DISCUSSION

John Thurnam first described sinus of Valsalva

aneurysm (SVA) in 1840. Hope further described it in 1939. Aneurysms of left aortic sinus of Valsalva rupturing into left ventricle are relatively rare. In a series of 49 cases reported by Sawyers *et al.* [3] 34 arose from the right coronary sinus, 13 from the posterior (non-coronary) sinus and only 2 from the left. SVA is caused by dilation, usually of a single sinus of Valsalva, from a separation between the aortic media and the annulus fibrosus. A deficiency of normal elastic tissue and abnormal development of the bulbus cordis have been associated with the development of SVA due to the high pressure at the root of the aorta.

Associated structural defects include supracristal or perimembranous ventricular septal defect (30%–60%), bicuspid aortic valve (15%–20%) and aortic regurgitation (44%–50%). Two-dimensional transthoracic echocardiography (TTE) may detect as many as 75% of all patients with sinus of Valsalva aneurysm (SVA). Usually TEE or MRI is needed to confirm the diagnosis

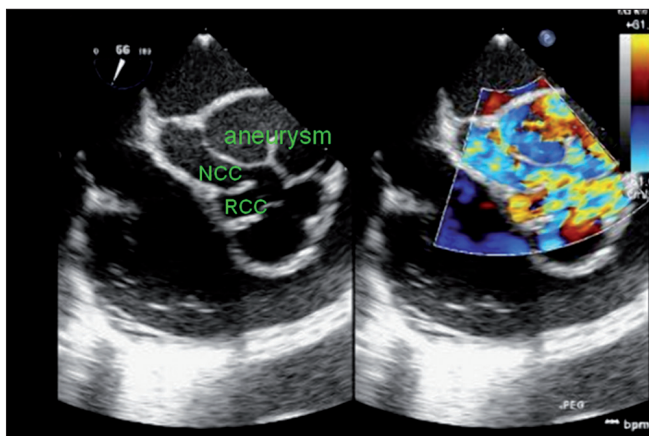


Figure 1: Mid esophageal aortic short axis view showing aneurysm arising from left coronary sinus, normal noncoronary sinus and dilated right coronary sinus
NCC: non coronary cusp; RCC: right coronary cusp

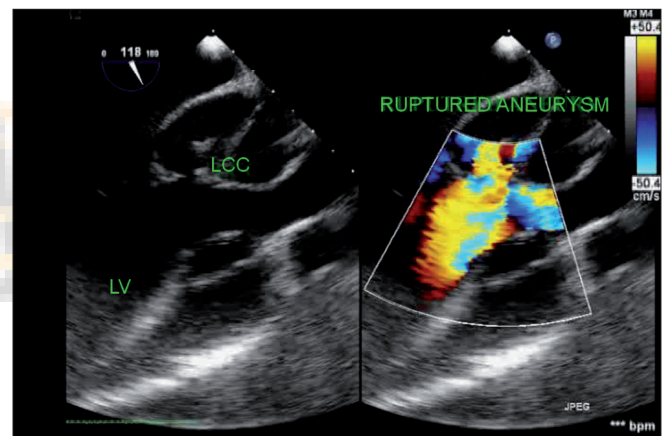


Figure 2: Mid-esophageal aortic long axis color Doppler view showing ruptured aneurysm from left coronary sinus into left ventricular cavity. LCC: left coronary cusp; LV: left ventricle

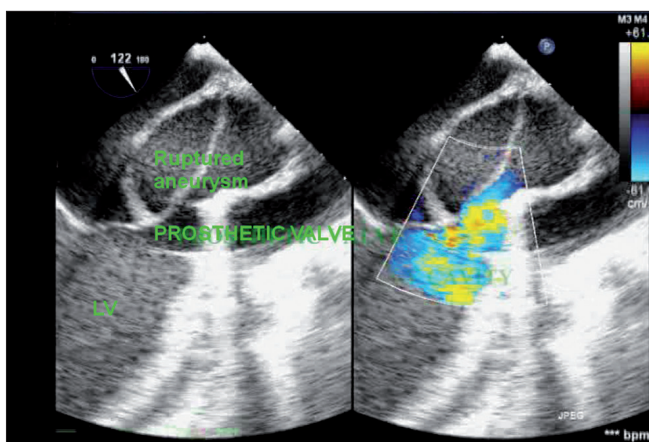


Figure 3: Post-aortic valve replacement mid-esophageal aortic short axis view showing prosthetic valve with residual aneurysm

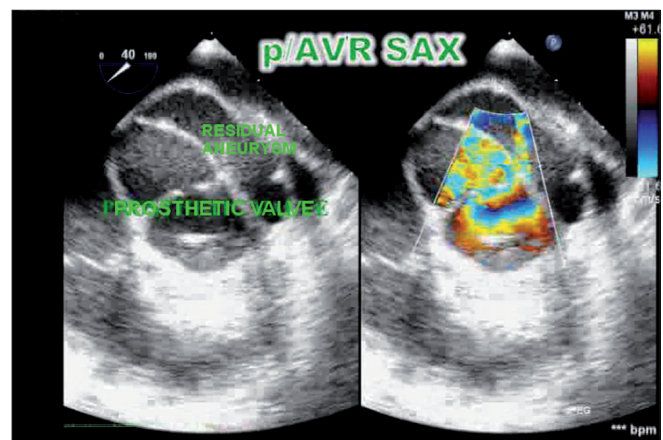


Figure 4: Post-aortic valve replacement mid-esophageal aortic long axis color Doppler view showing prosthetic valve with residual ruptured aneurysm into left ventricular cavity. LV: left ventricle

and for perioperative assessment. Transesophageal echocardiographic examination in addition to transthoracic echocardiography provides more powerful information about SVAs and coexistent cardiac malformations. This may be additional value for the cardiac surgeon planning resection of the lesion.^[3] Multiplane transesophageal echocardiography (TEE) provides conclusive information regarding the origin and size of the aneurysm and presence of thrombotic material and allows precise identification of structural anomalies and shunt locations for perioperative assessment.^[1,4,5] Prognosis is poor with progressive aneurysmal dilatation or rupture unless early surgery is done.

CONCLUSION

Thus the role of anesthetist as perioperative transesophageal echocardiographer is very crucial for

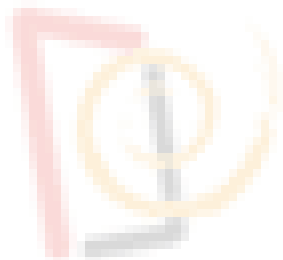
the accurate diagnosis and optimal management of patient with SVA.

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Cite this article as: Sarupria A, Kapoor PM, Kiran U, Hote M. Multiple ruptured aneurysm of left sinus of Valsalva: A rare entity. *Ann Card Anaesth* 2011;14:48-50.

Source of Support: Nil, **Conflict of Interest:** None declared.



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