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CASE REPORT
Cardiac imaging

A case report: a rare case of severe aortic incompetence

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Background Aneurysms of the sinus of Valsalva (SOV) are thin-walled outpouchings most commonly involving the right or non-coronary sinuses. Because they are asymptomatic, they are rarely discovered before they rupture and form an aorto-cardiac fistula. We present a rare case of unruptured aneurysm of the right coronary SOV burrowing into the interventricular septum with severe aortic incompetence and left ventricular dysfunction. To our knowledge, burrowing of the SOV aneurysm (SVA) into the interventricular septum and its large sac-like appearance has never been described using three dimensional (3D) echocardiography before.

Case summary A 37-year-old man presented to the cardiology outpatient department with complaints of dyspnoea and palpitations (New York Heart Association Class II–III) for the last 6 months. He was evaluated with transthoracic echocardiography which showed a large mobile sac-like structure with irregular borders bulging and prolapsing into the left ventricular cavity with each cardiac cycle along with severe aortic incompetence. On transoesophageal echocardiogram, the right coronary cusp showed malcoaptation with deformed aortic sinus causing severe aortic incompetence. Cardiac computed tomography showed sparing of right coronary artery at the origin. A diagnosis of SVA was made. The patient underwent aortic valve replacement along with partial resection of the aneurysm. The patient had an uneventful postoperative course. Follow-up echocardiography after 4 weeks showed well-seated aortic valve prosthesis with residual SVA. The ejection fraction decreased from 46–48% to 36–38%.

Discussion Comprehensive multimodality imaging can be used for management strategy of SVA.

Keywords Three dimensional (3D) echocardiography • Aortic regurgitation • Sinus of Valsalva aneurysm • Case report • Left ventricular dysfunction

Learning point
- Comprehensive multimodality imaging can help delineate management strategy in sinus of Valsalva aneurysm.

Introduction

Aneurysms of the sinus of Valsalva (SOV) are thin-walled outpouchings, most commonly involving the right or non-coronary sinuses. Because they are asymptomatic, they are rarely discovered before they rupture and form an aorto-cardiac fistula.[1] We present a rare case of an unruptured aneurysm of the right coronary SOV burrowing into the interventricular septum with severe aortic incompetence and left ventricular dysfunction.

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Timeline

Day 1	Patient presents to the outpatient department
Day2	Patient undergoes transthoracic echocardiography (TTE) (TTE shows sac-like mobile structure prolapsing in the left ventricular cavity)
Day 3	Patient undergoes transoesophageal echocardiogram (TOE) [TOE confirms diagnosis of sinus of Valsalva (SOV) aneurysm]
Day 4	Patient underwent aortic valve replacement and repair of SOV aneurysm
Day 14	Patient is discharged
Day 60	Patient's most recent follow-up

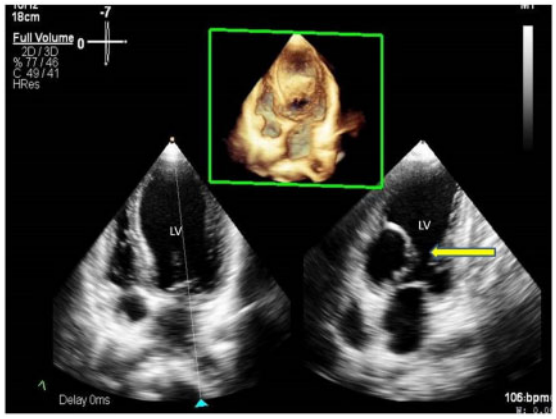

Figure 1 Transthoracic three-dimensional apical view showing large sac-like structure (unruptured sinus of Valsalva aneurysm) occupying left ventricle cavity posteriorly.

Case presentation

A 37-years-old man presented to the cardiology outpatient department with complaints of dyspnoea and palpitations (New York Heart Association Class II–III) for the last 6 months. There was no history of blunt chest trauma or long-standing fever. He did not have any significant past medical history.

On physical examination, the patient was alert, afebrile with mildly elevated jugular venous pulse. He had a normal body habitus. His blood pressure was 140/70 mmHg, while heart rate was 90 b.p.m. Chest examination revealed bilateral normal air entry with no added sounds. Cardiac examination showed apical displacement to the left 6th intercostal space, an ill sustained heave, left ventricle 3rd heart sound (S3) with early diastolic murmur (Grade IV/IV) over the left 2nd intercostal space. Abdominal and neurological examination was

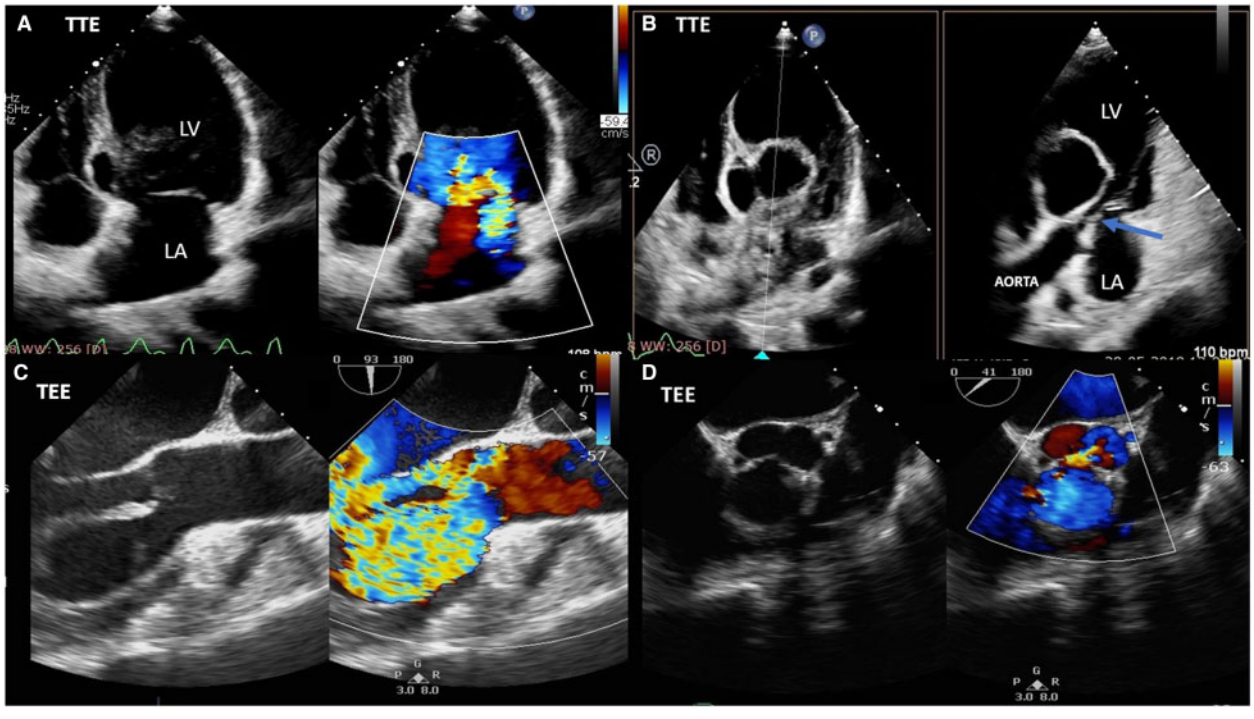

Figure 2 (*A*) Transthoracic echocardiography apical four-chamber view showing moderate mitral regurgitation. (*B*) Transthoracic echocardiography showing narrowing of left ventricular outflow tract lumen (blue arrow) in systole by prolapsing sinus of Valsalva aneurysm. (*C*) Transoesophageal echocardiography showing large diastolic torrential jet from aorta filling cavities of left ventricle and sinus of Valsalva aneurysm simultaneously. (*D*) Transoesophageal echocardiography short-axis view 45° showing sinus of Valsalva aneurysm bulging in systole.

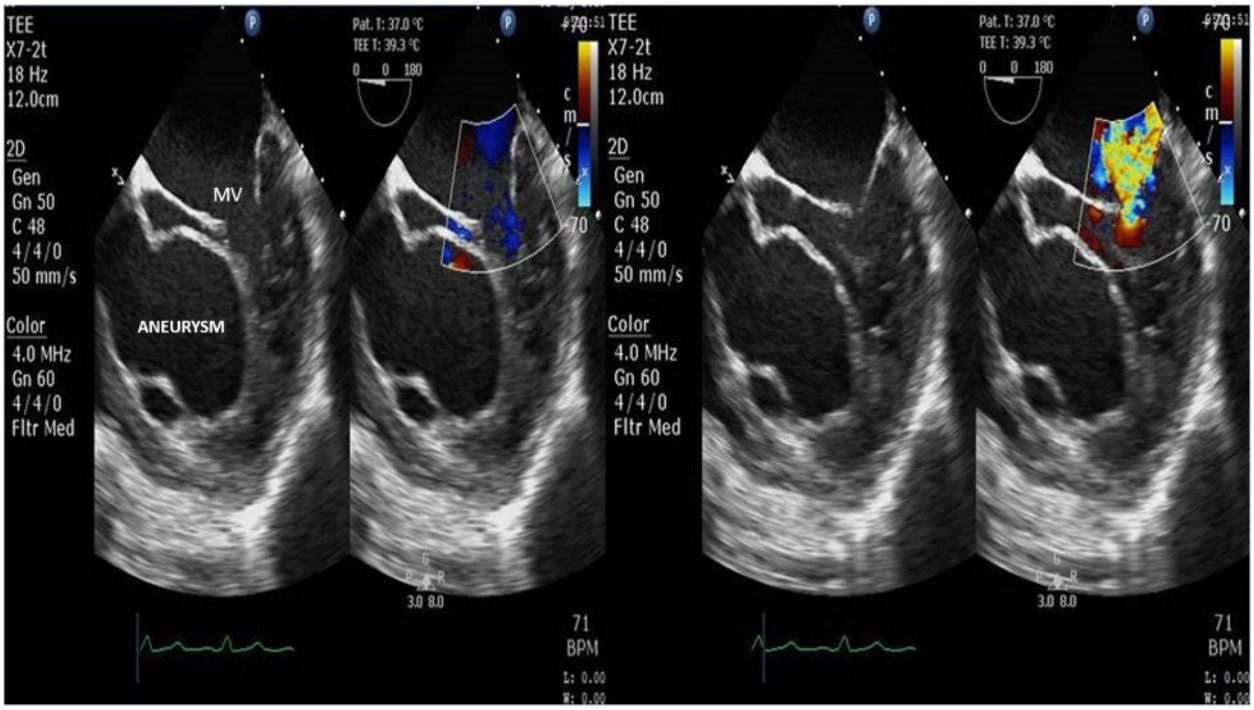

Figure 3 Transoesophageal echocardiography mid oesophageal level 0° view shows aneurysm sac and mitral regurgitation in the systolic frame. MV, mitral valve.

normal. Electrocardiogram showed sinus tachycardia with left bundle branch block. Chest X-ray showed cardiomegaly with prominent aortic shadow.

Transthoracic echocardiography showed a large mobile sac-like structure with irregular borders bulging and prolapsing into the left ventricular cavity with each cardiac cycle along with severe aortic incompetence. Differential diagnosis of Hydatid cyst of the interventricular septum, unruptured SOV or septal tricuspid valve aneurysm prolapsing into the left ventricle through a partially closed ventricular septal defect was kept. (*Figures 1* and *2*, Supplementary material online, *Video S1*). The left ventricle was moderately dilated with mild impairment in global contractility (ejection fraction 46–48%). The right ventricle and the right atrium were normal in dimensions. Right ventricular systolic pressure was 40 mmHg. On transoesophageal echocardiography, the right coronary cusp showed malcoaptation with deformed aortic sinuses causing severe aortic incompetence. There was no evidence of rupture of the aneurysm. There was also moderate central mitral incompetence and mild left atrial dilatation (*Figure 3*). Cardiac computed tomography showed sparing of right coronary artery at the origin (*Figure 4*).

At surgery, through a transaortic approach, the right aortic valve cusp was seen as thin and fibrotic. About 4.0 mm under the right coronary ostium, there was 5.0 × 2.0 cm opening of the aneurysm which led to a trilobulated cavity burrowing into the interventricular septum. The largest cavity measured 6.0 × 4.0 × 3.0 cm. The aortic side of the aneurysm opening was closed with a Dacron patch sutured with 6/0 prolene avoiding the conduction system area and preserving the right coronary ostium. The aneurysm of right SOV was partially resected and a Saint Jude Medical mechanical bileaflet aortic valve (# 23) was sutured to the aortic annulus with interrupted pledgeted sutures. The patient had an uneventful postoperative course. The patient was discharged on Nicoumalone 2 mg once daily, Ramipril 2.5 mg once daily, and Metroprolol 12.5 mg twice daily. Follow-up echocardiography after 4 weeks showed well-seated aortic valve prosthesis and the residual SOV aneurysm (SVA). The ejection fraction decreased from 46–48% to 36–38% (*Figure 5*). The dose of Ramipril was increased to 5 mg once daily and Metoprolol to 25 mg twice daily. Patient did not report any cardiovascular symptoms on follow-up.

Discussion

Sinus of Valsalva aneurysm is defined as the dilatation of one or more of the aortic sinuses located between the aortic valve annulus and the sinotubular junction. The prevalence is estimated to be 0.09% in the general population. Sinus of Valsalva aneurysms are commonly congenital and represent 0.1–3.5% of congenital heart defects. Sinus of Valsalva aneurysm usually originates from the right coronary sinus (70–90%), followed by the non-coronary sinus (10–25%) and left sinus (<5%).[1–3]

Unruptured aneurysms are asymptomatic and are incidentally detected during imaging workup of heart murmurs. Rarely, they may present with dyspnoea, palpitations, arrhythmias, or angina/chest pain.[4,5] Aortic regurgitation occurs in 30–50% of patients. Sinus of Valsalva aneurysms may be associated with several congenital cardiac abnormalities and cross-sectional imaging can be useful for their

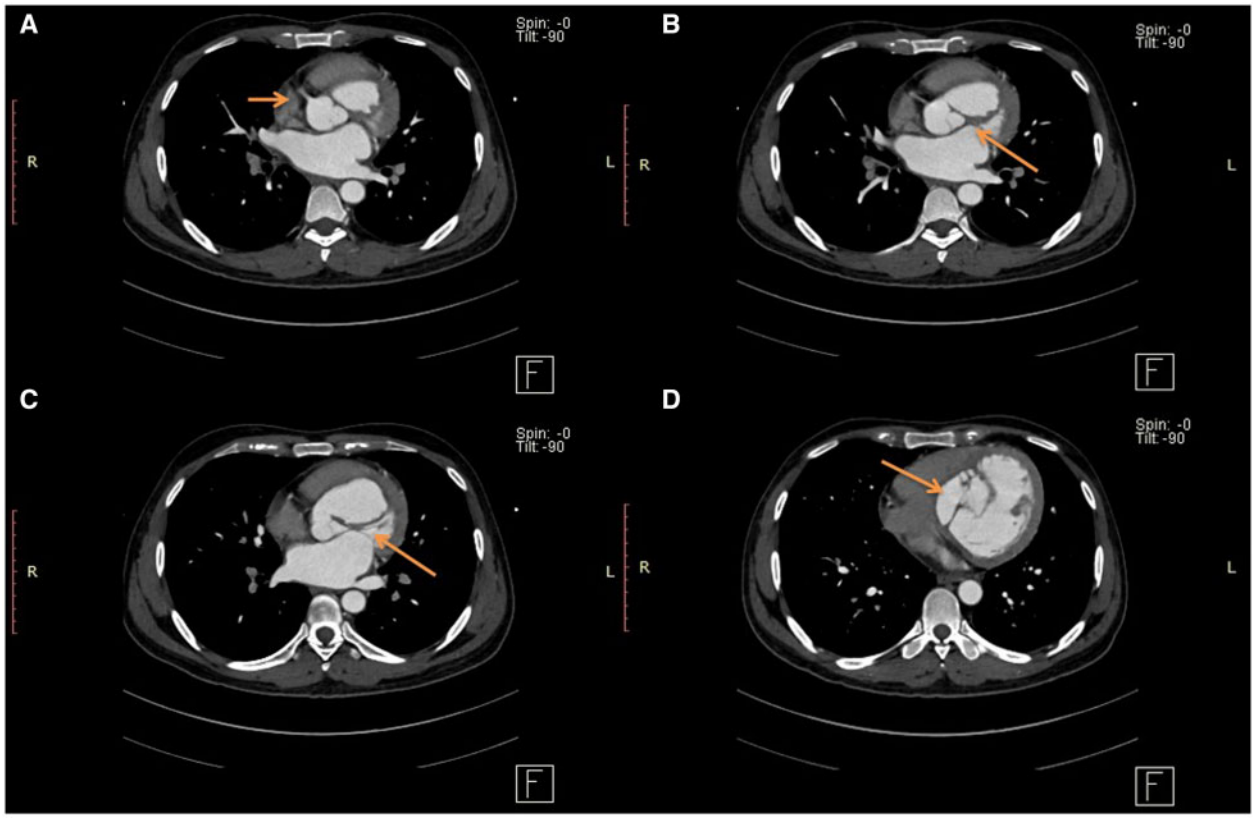

Figure 4 (*A*) Origin of right coronary artery. (*B*) Origin of sinus of Valsalva aneurysm 2 mm inferior to origin of right coronary artery. (*C*) Sinus of Valsalva extending into mid left ventricular cavity. (*D*) Sinus of Valsalva aneurysm sac in relation to interventricular septum.

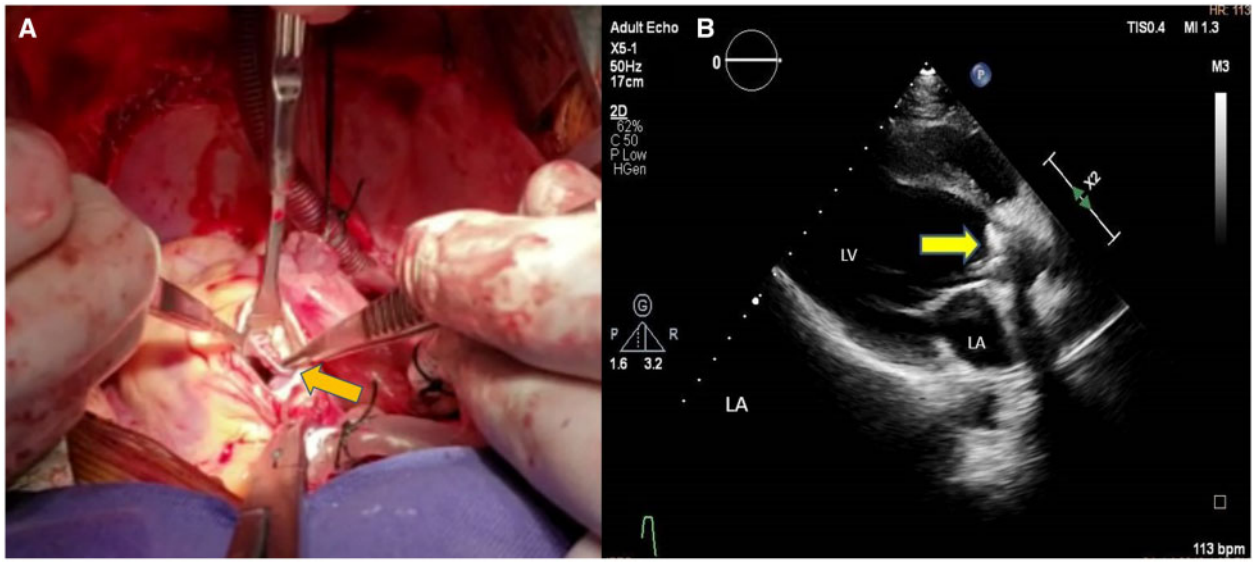

Figure 5 (*A*) Intraoperative picture showing prolapsed right aortic cusp. (*B*) Post-operative echocardiogram after 1 month showing well-seated prosthetic aortic valve.

assessment. Sinus of Valsalva aneurysms have been associated with bicuspid aortic valves in approximately 10% of cases, ventricular septal defects (30–60% of patients), aortic stenosis, infundibular pulmonary stenosis, left ventricular non-compaction, atrial septal defect, coronary anomalies, left-sided superior vena cava, atrial septal defect, patent ductus arteriosus, and hypertrophic obstructive cardiomyopathy.[6,7] Ruptured SVAs can lead to development of sudden onset dyspnoea and deterioration of cardiac function.

Congential sinus of Valsava aneusyms have been classified into four categories by Sakakibara and Konno. Our case does not fit the categories described in the classification.[8] To our knowledge, burrowing of the SVA into the interventricular septum and its large sac-like appearance has never been described using 3D echocardiography before. This rare entity could be managed successfully by surgeons only with aid of comprehensive multimodality imaging.

Lead author biography

Mandeep Singh Sondh is an MBBS Student at Hero DMC (Dayanand Medical College) Heart Institute, Ludhiana, Punjab.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

References

1. Takach TJ, Reul GJ, Duncan JM, Cooley DA, Livesay JJ, Ott DA, Frazier OH. Sinus of Valsalva aneurysm or fistula: management and outcome. *Ann Thorac Surg* 1999; **68**:1573–1577.
2. Tomar M, Radhakrishnan S, Kaushal S, Iyer KS. Unusual case of ruptured sinus of Valsalva: rupture into left ventricular cavity along with distortion of mitral valve requiring double valve replacement. *Images Paediatr Cardiol* 2009;**38**:1–6.
3. Otto C. *The Practice of Clinical Echocardiograph*, 3rd ed. 2007, p1305.
4. Vural KM, Sener E, Tasdemir O, Bayazit K. Approach to sinus of Valsalva aneurysms: a review of 53 cases. *Eur J Cardiothorac Surg* 2001;**20**:71–76.
5. Segab C, Davy JM, Schebule C, Bouchareine F, Dussarat G, Aigueperse J, et al. Atrioventricular block disclosing an isolated congenital aneurysm of the sinus of Valsalva, extending into the septum and not ruptured. *Arch Mal Coeur Vaiss* 1981;**74**:1233.
6. Dev V, Goswami KC, Shrivastava S, Bahl VK, Saxena A. Echocardiographic diagnosis of aneurysm of the sinus of Valsalva. *Am Heart J* 1993;**126**:930–936.
7. Unlu M, Ozeke O, Kara M, Yesillik S. Ruptured sinus of Valsalva aneurysm associated with noncompaction of the ventricular myocardium. *Eur J Echocardiogr* 2008;**9**:311–313.
8. Sakakibara S, Konno S. Congenital aneurysm of the sinus of Valsalva: anatomy and classification. *Am Heart J* 1962;**63**:405–424.