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CASE REPORT
Coronary heart disease

Giant unruptured aneurysm of the left coronary sinus of Valsalva presenting as acute coronary syndrome: a case report

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Background Sinus of Valsalva aneurysm (SVA) is a rare cardiac anomaly that can be congenital or acquired. Unruptured aneurysms may be asymptomatic but can present as malignant arrhythmias, acute chest pain, and even sudden cardiac death. Both ruptured and unruptured SVAs may have fatal complications, thus prompt diagnosis and surgery is critical.

Case summary We report a successful surgical repair of an unruptured aneurysm of the left sinus of Valsalva presenting as acute coronary syndrome. Coronary angiography (CAG) and cardiac multislice computed tomography (CT) revealed a large unruptured aneurysm of the left sinus of Valsalva. Surgical repair was performed by resection of the aneurysm, aortic valve replacement with composite graft, and coronary artery bypass.

Discussion Aneurysm of the left sinus of Valsalva is an extremely rare condition. Correct diagnosis can be done by echocardiography, CAG, or CT. Non-ruptured aneurysms should be surgically repaired if they are associated with significant symptoms or if their size enlarge rapidly.

Keywords Left sinus of Valsalva • Unruptured aneurysm • Acute coronary syndrome • Case report

Learning points

- A sinus of Valsalva aneurysm is a rare differential diagnosis to remember in patients presenting with chest pain with or without acute coronary syndromes.
- Coronary angiography and multislice computed tomography can provide exact information about complex structures, leading to successful surgical management.
- Non-ruptured aneurysms should be surgically repaired if they are associated with significant symptoms or their size enlarge rapidly.

Introduction

Sinus of Valsalva aneurysm (SVA) is an enlargement of the aortic root between the aortic valve annulus and the sinutubular junction. Either congenital or acquired, SVAs frequently originate from the right sinus of Valsalva (65–85%) and less frequently from the non-coronary (10–30%) and left sinuses (<5%).[1] An unruptured aneurysm is typically asymptomatic. However, compression of the coronary arteries by SVAs can cause myocardial ischaemia and chest pain.[2] The condition can be potentially fatal due to rupture; thus, prompt diagnosis and surgical management are critical.

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Timeline

Time	Event
Day 1	Patient presents to general practitioner with a 14-day history of chest pain and dyspnoea
	Electrocardiogram reveals newly acquired atrial fibrillation and non-specific ST wave changes
	Patient is field triaged to a heart centre for acute coronary angiography (CAG)
	Angiography shows large contrast-filled cavity
	Computed tomography (CT) scan confirms giant unruptured aneurysm of the left sinus of Valsalva
	Transoesophageal echocardiography demonstrated no involvement of the aortic valve and a left ventricular ejection fraction (LVEF) of 35%
Day 2	Detailed cardiac CT angiography is performed but is insufficient regarding information about coronary arteries
Day 7	Additional CAG shows significant circumflex artery stenosis
Day 8	Multidisciplinary conference: The patient is offered surgery with patch repair and coronary artery bypass when clinical performance has improved
Day 28	Operation: Patch repair is technically not possible. The defect is repaired with a mechanical composite graft and coronary artery bypass
Day 35	Discharged without any complications
1 month post-surgery	Echocardiography shows unchanged LVEF of 35%
1.5 years post-surgery	No symptoms and repeat echocardiogram shows an improved LVEF of 55%

Case presentation

A 76-year-old man with no family history of cardiovascular disease who was an active smoker and diagnosed with hypertension, hypercholesterolaemia, and diabetes (type 2) presented with intermittent chest pain and increasing dyspnoea on exertion throughout 14 days. As the chest pain and dyspnoea intensified, he contacted his general practitioner. An electrocardiogram revealed newly acquired atrial fibrillation and non-specific ST wave changes in V1–V2 (*Figure 1*). In light of on-going severe chest pain and pre-hospital elevated high-sensitive troponin T-level at 231 ng/L, the patient was field triaged directly to the heart centre for emergency coronary angiography (CAG), thus bypassing the local hospital.

Upon arrival to the catheterization laboratory, the patient was without chest pain and haemodynamically stable with a systolic blood pressure of 120 mmHg and heart rate of 130 b.p.m. Chest auscultation did not reveal any obvious abnormal heart murmurs. Bedside transthoracic echocardiography showed global hypokinesia with a left ventricular ejection fraction (LVEF) of 35%. ECG showed the reported atrial fibrillation but no further differences were detected.

The CAG revealed a large contrast-filled cavity (*Figure 2*). Angiography was halted at this stage, and the patient underwent an urgent computed tomography (CT) of the aorta, which confirmed the presence of a giant unruptured aneurysm of the left sinus of Valsalva measuring 3.5 × 6 cm (*Figure 3*).

Transoesophageal echocardiography demonstrated bi-atrial dilation, normal function of the aortic valve without annular enlargement. Global hypokinaesia persisted with LVEF of 35%. Cardiac contrast-enhanced CT angiography showed aneurysmal compression of the left anterior descending (LAD) artery and the circumflex artery (Cx). However, due to atrial fibrillation, the image quality was too poor to visualize the coronary arteries sufficiently (*Figure 4*). The patient continued to have prominent chest pain on exertion, and therefore additional CAG was performed. This showed significant stenosis of the Cx (Supplementary material online, *Video S1*).

Aortic surgery was performed on standard cardiopulmonary bypass and under cardioplegic arrest with moderate hypothermia. A 6 cm false aneurysm was found arising from the left sinus of Valsalva and with close relation to the LAD artery. Surgical correction with patch repair was technically impossible. A biological composite graft was found to be too big for the area of replacement; thus, a mechanical composite graft (SJM^{TM} Masters HP Series Valved Graft 27VAVGJ-515) was chosen to repair the defect. A coronary artery bypass graft was performed with end-to-end anastomosis with the 1st obtuse marginal artery, using the left radial artery as conduit. Cardiopulmonary bypass weaning was uncomplicated. Post-operative transoesophageal echocardiography showed unchanged LVEF of 35% and well-functioning prosthetic valve. Eighteen months after surgery, the patient was asymptomatic, and transthoracic echocardiography showed that LVEF had improved to 55%.

Discussion

Sinus of Valsalva aneurysm can be either congenital or acquired.[3] Congenital SVAs comprise up to 3.5% of all congenital heart defects and have been linked to connective tissue diseases, such as Marfan's syndrome and Ehlers–Danlos syndrome.[4] Patients diagnosed with a bicuspid aortic valve or ventricular septal defects may also be more likely to develop SVA.[5] Acquired SVAs are often associated with infectious aetiologies, including syphilis, bacterial endocarditis, and tuberculosis.[6,7]

An SVA usually remains clinically silent until it ruptures; nevertheless, when an unruptured SVA increases to a certain level, it takes on a variety of clinical presentations such as palpitations, chest pain on exertion, and gradually worsening dyspnoea. Controversy still exists regarding the exact time of intervention in the case of unruptured aneurysms. However, current guidelines recommend surgical correction if sinuses exceed 5.5 cm or 4.5 cm in the setting of connective tissue disease. Repair should also be considered when the growth rate exceeds 0.5 cm/year.[8] Surgical repair is also required in cases of unruptured but symptomatic aneurysms causing ventricular outflow tract obstruction, coronary ostia obstruction, or malignant arrhythmias. Ruptured SVAs have a clear indication for surgical intervention.

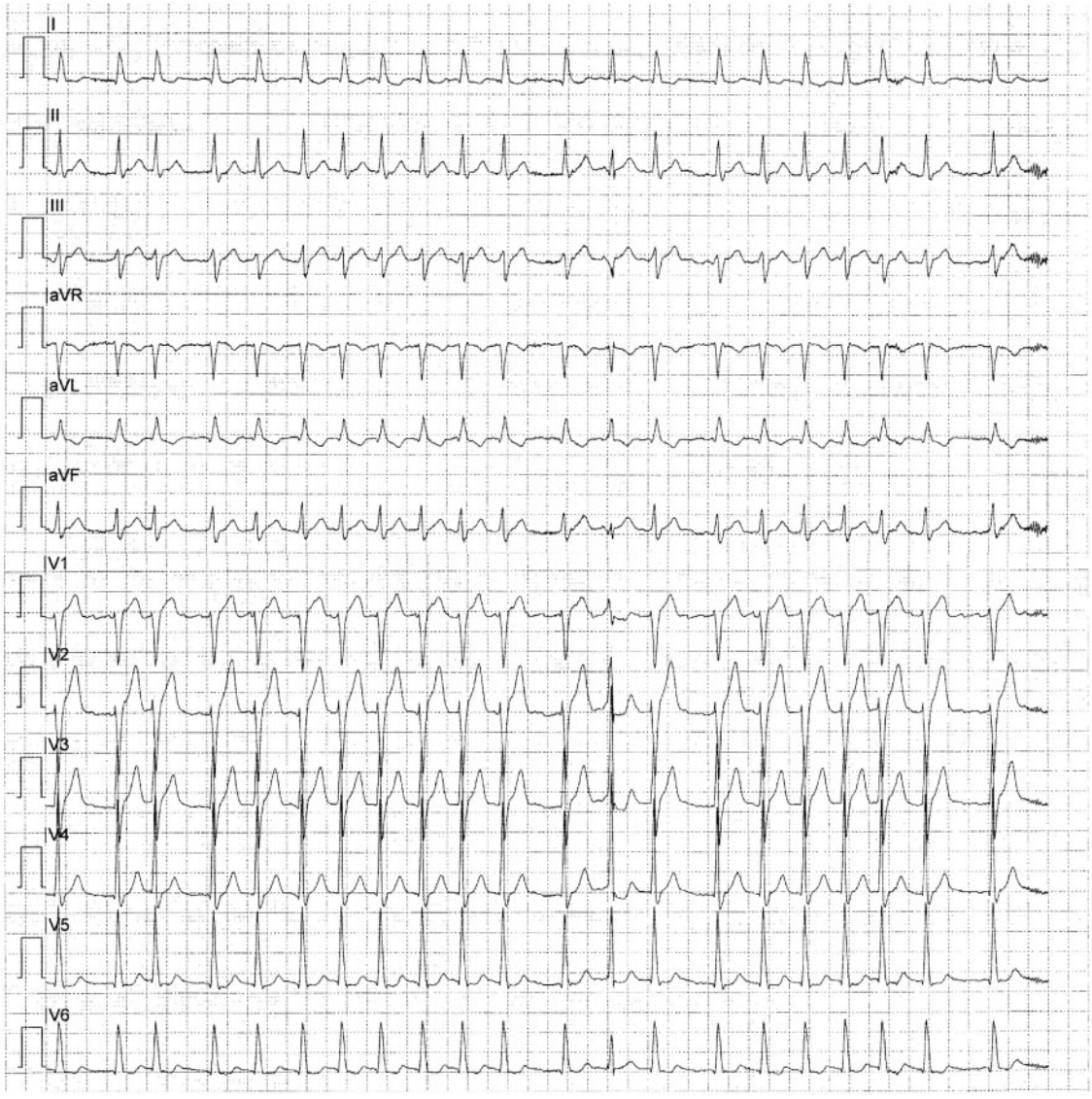

Figure 1 Electrocardiogram taken in the ambulance revealed newly acquired atrial fibrillation and non-specific ST wave changes in V1–V2.

Transthoracic and transoesophageal echocardiogram have traditionally been the diagnostic method of choice; however, in recent years CT has gained ground as a first-line technique. In our case, the patient was admitted to a high-volume cardiac centre with access to acute percutaneous coronary intervention, advanced cardiovascular imaging, and cardiac surgery. The multidisciplinary approach allowed prompt diagnosis, proper treatment, and a favourable outcome.

Conclusion

A left SVA is an extremely rare condition,[9,10] and symptoms typically occur between 30 and 45 years of age.[5] We report a successful correction of an unruptured left SVA with late presentation in a 76-year-old man.

Lead author biography

Sivagowry Rasalingam Mørk is a medical doctor at the Department of Cardiology, Aarhus University Hospital in Denmark. She obtained her marster's degree in medicine from Aarhus University in 2013. She is currently a Ph.D. student under the supervision of MD and DMSc Christian Juhl Terkelsen. Her primary research interest center on out-of-hospital cardiac arrest.

Supplementary material

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

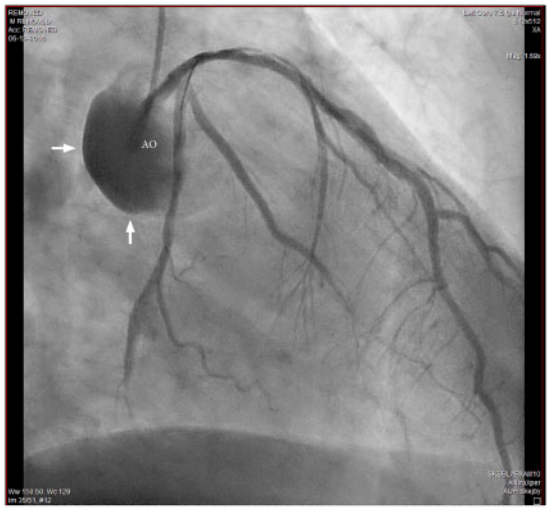

Figure 2 Acute coronary angiography. Contrast dye can be seen outlining the large aneurysmal dilatation of the aorta at the left sinus of Valsalva (white arrows). AO, aorta.

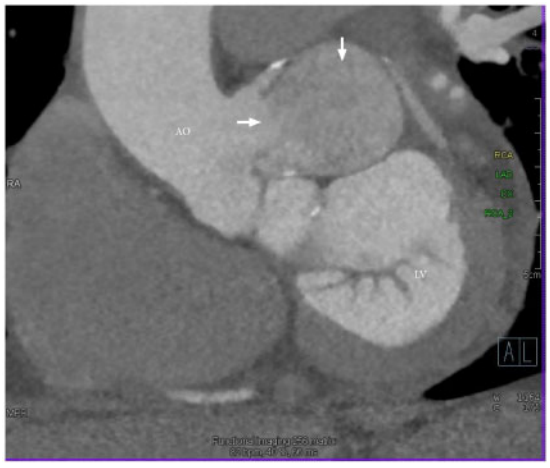

Figure 3 Computed tomography (CT) of the heart with contrast medium showing a 3.5 × 6 cm mass arising from the left coronary sinus (white arrows). AO, aorta; LV, left ventricle.

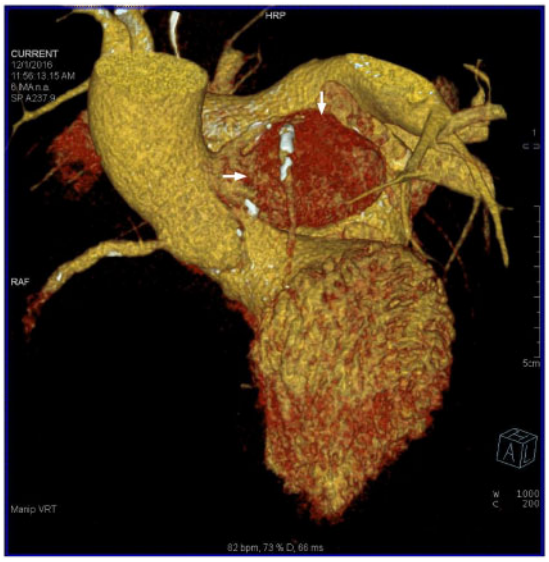

Figure 4 Computed tomography of the heart in 3D reconstruction. White arrows show the left Sinus of Valsalva aneurysm.

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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.

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