ESC European Society of Cardiology

European Heart Journal - Case Reports (2024) **8**, 1–6
https://doi.org/10.1093/ehjcr/ytae047

CASE REPORT
Coronary intervention

Giant left sinus of Valsalva aneurysm as a rare cause of acute myocardial infarction: a case report

Ghaith M. Maqableh [1,2]*, Mengshi Yuan[1], William Moody[1,3], Shanat Baig[1,3], and Sohail Q. Khan[1,3]

[1]Department of Interventional Cardiology, Queen Elizabeth Hospital, University Hospitals Birmingham NHS Foundation Trust, Mindelsohn Way, Birmingham B15 2TH, UK; [2]Cardiology Department, Faculty of Medicine, Al Balqa Applied University, Amman 19117, Jordan; and [3]Institute of Cardiovascular Sciences, University of Birmingham, Birmingham B15 2TT, UK

Received 2 September 2023; revised 9 January 2024; accepted 23 January 2024; online publish-ahead-of-print 25 January 2024

Background Sinus of Valsalva aneurysm (SVA) is a rare but potentially life-threatening condition. Acute myocardial infarction (MI) is a rare consequence of aneurysmal dilatation of one or more sinuses of Valsalva. We present a case of an unruptured and partially thrombosed left SVA, presenting as anterior MI and congestive heart failure.

Case summary A 55-year-old gentleman was admitted with pulmonary oedema and a late presenting ST-elevation MI with Q wave. After initial treatment on furosemide infusion, a coronary angiography showed significant stenosis in both his left main stem (LMS) and left anterior descending artery (LAD). This is likely a result of external compression, potentially from the enlarged left sinus of Valsalva. A subsequent transthoracic echocardiogram and transoesophageal echocardiogram (TOE) confirmed large SVA involving the left coronary cusp measured 9.9 cm compressing both LMS and LAD.

Discussion Left SVAs are rare and frequently asymptomatic, typically being identified incidentally. Due to the close proximity of the left coronary system, they can present with myocardial ischaemia due to extrinsic compression of the coronary system. We were able to perform a comprehensive multi-modality assessment of left SVA, which helped establish this unusual diagnosis and guide management. Transthoracic echocardiogram and TOE helped assess the SVA and demonstrated the thrombus *in situ*, aortic valve insufficiency, and cardiac function. The computed tomography scan aided in accurately defining the extent of the aneurysm and the extent of compression of the left coronary system and cardiac magnetic resonance scan was able to demonstrate viability in LAD and circumflex territory.

Keywords Sinuses of Valsalva aneurysm • Coronary angiogram • Echocardiogram • Myocardial infarction • Cardiac imaging • Case report

ESC curriculum 2.3 Cardiac magnetic resonance • 2.2 Echocardiography • 2.4 Cardiac computed tomography • 3.2 Acute coronary syndrome • 3.4 Coronary angiography

* Corresponding author. Tel: +44 7544185636, Email: ghaith.maqableh@uhb.nhs.uk; maqablehghaith1@gmail.com
Handling Editor: Dimitrios A. Vrachatis
Peer-reviewers: A. Shaheer Ahmed; Parag Bawaskar
Compliance Editor: Ralph Mark Louis Neijenhuis

Learning points

- While sinuses of Valsalva aneurysm (SVAs) are rare, it is important to be aware of their potential complications and to consider them in differential diagnosis when appropriate.
- Utilizing a combination of multi-modality imaging techniques, including transthoracic echocardiography, transoesophageal echocardiography, computed tomography, and cardiac magnetic resonance, facilitates precise diagnostic assessment of SVA and associated complications. Furthermore, these modalities help to guide the development of a comprehensive management strategy.

Introduction

Sinus of Valsalva aneurysm (SVA) represents a rare yet clinically significant cardiac condition, with an estimated prevalence of approximately 0.09% in the general population. Within the spectrum of congenital cardiac defects, SVAs account for between 0.1% and 3.5%. Gender-specific variations exist, as men exhibit a four-fold higher likelihood of being affected compared with women. Additionally, certain ethnic groups, particularly Asian populations, have a higher incidence of these anomalies.[1]

From an anatomical perspective, SVAs predominantly involve the right coronary sinus, followed by the non-coronary sinus, with the left coronary sinus being the least frequently affected. The anatomical location of the sinus aneurysm is a fundamental factor in the clinical course of SVA formation or rupture.

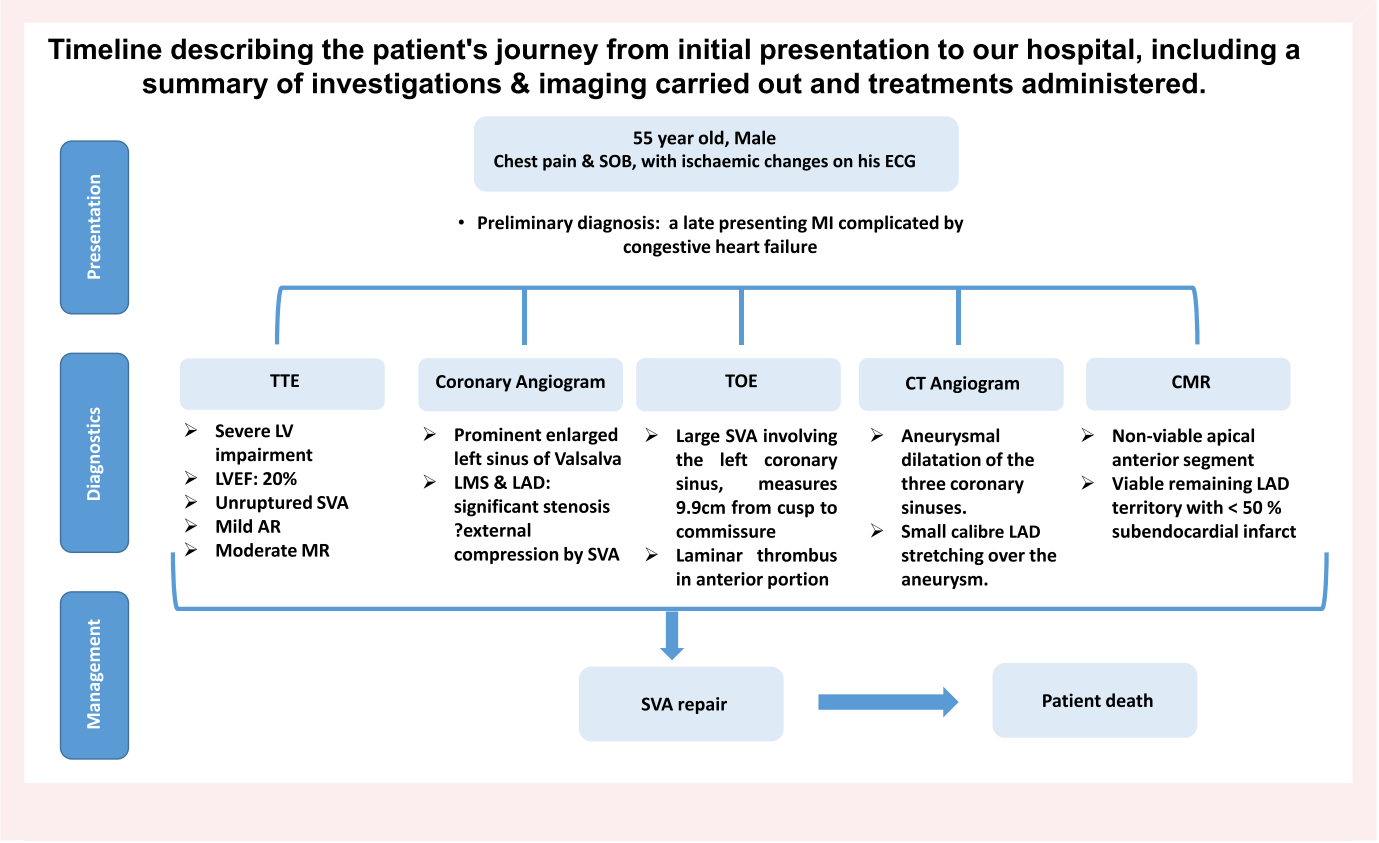
The clinical complexity of SVA is highlighted by its diverse manifestations and outcomes. Left untreated, ruptured SVAs are associated with a dismal prognosis, with a mere 1-year life expectancy.

Complications of both ruptured and non-ruptured SVAs involve aortic regurgitation in 30–50% of cases, necessitating aortic valve replacement alongside operative repair of the SVA. Additionally, large SVAs may act as a thrombus nidus leading to external compression on the coronary arteries and subsequently resulting in myocardial infarction (MI).

Only a few cases of SVA accompanied by myocardial ischaemia have been reported in the international literature.

We present a case of an unruptured and partially thrombosed left SVA, presenting as anterior MI and congestive heart failure.

Summary figure



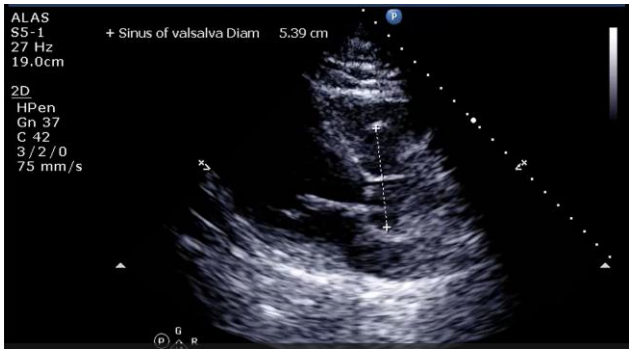


Figure 1 Initial bedside transthoracic echocardiogram with unruptured sinus of Valsalva aneurysm in parasternal long axis (PLAX) view.

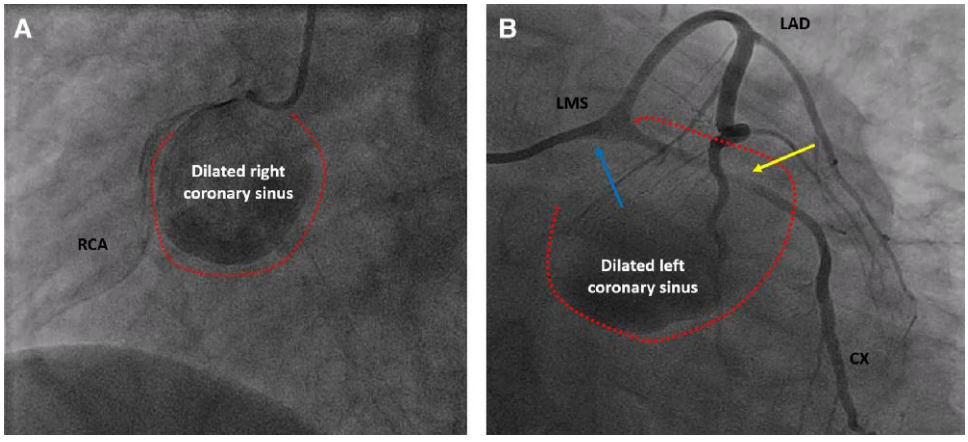


Figure 2 Angiogram showed hugely dilated right (*A*) and left (*B*) coronary sinuses, with a hazy lesion in circumflex (yellow arrow) possibly indicating thrombus. The dilated left coronary sinus causes extra-luminal compression of the left main stem ostium (blue arrow) leading to immediate pressure damping on guide engagement. CX, circumflex; LAD, left anterior descending artery; LMS, left main stem; RCA, right coronary artery.

Case presentation

A 55-year-old gentleman was admitted to the emergency department with worsening shortness of breath and retrosternal chest pain radiating to the left arm and jaw. His medical history consists of hypercholesterolaemia, traumatic neck injury that necessitated spinal surgery, and a previous COVID-19 infection a few months ago. He was an ex-smoker.

During examination, he was found to be tachycardiac, hypoxic, and tachypneic with an oxygen saturation of 80% on room air. His heart rate was 120 b.p.m., and his blood pressure was 110/70 mmHg. Chest auscultation revealed normal heart sounds, no audible murmurs, and bilateral crepitation.

The preliminary diagnosis was a late presenting ST-elevation MI complicated by congestive heart failure. We also considered mechanical complications of MI, such as acute mitral regurgitation due to papillary muscle rupture and ventricular septal defect.

His presenting electrocardiogram showed sinus rhythm with anterior upsloping ST segment elevation, Q waves, and poor R wave progression (see Supplementary material online, *Figure S1*). His high sensitivity Troponin I level exceeded 50 000 ng/L (>50 SI unit) and his creatinine kinase level was 795 U/L. A bedside transthoracic echocardiogram (TTE) showed an unruptured aneurysm of the sinus of Valsalva with mild aortic regurgitation. There was moderate functional mitral regurgitation. The left ventricle was normal size with severely impaired systolic function with regional wall motion abnormalities and an ejection fraction of 20% (*Figure 1*; see Supplementary material online, *Video S1*). His chest X-ray (see Supplementary material online, *Figure S2*) showed cephalization and indistinctness of the pulmonary vasculature compatible with pulmonary oedema.

The patient received a 60 mg intravenous bolus of furosemide and was commenced on a continuous infusion at a rate of 10 mg/hour. Following improvement from pulmonary oedema 2 days later, a coronary angiogram was performed. The procedure utilized a 6F sheath with a right radial artery approach. The right coronary artery was unobstructed and dominant, while the left main stem (LMS) was challenging to locate, requiring several diagnostic and guide catheters. Eventually, a Voda Left 4 guide (Boston Scientific) successfully engaged the LMS, but immediate pressure damping was noted upon engagement. The angiogram revealed a tapering LMS with no flowback and a moderate to severe hazy lesion with TIMI 3 flow in the circumflex indicating a possible thrombus. The left anterior descending artery (LAD) was found to be unobstructed, and a prominent left sinus of Valsalva was noted on the angiogram (*Figure 2*; see Supplementary material online, *Video S2*). It was

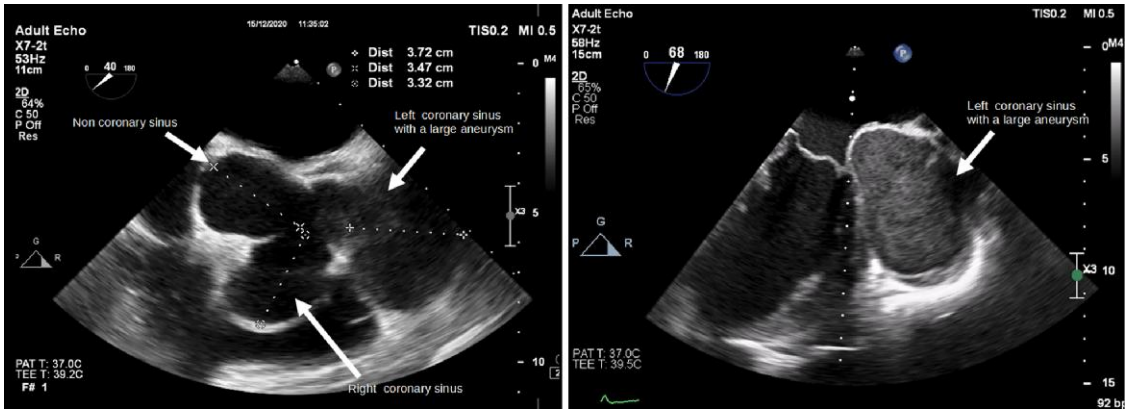


Figure 3 Transoesophageal echocardiogram showing hugely dilated left coronary sinuses with spontaneous echo contrast and increased echogenicity at bottom suggestive of thrombus.

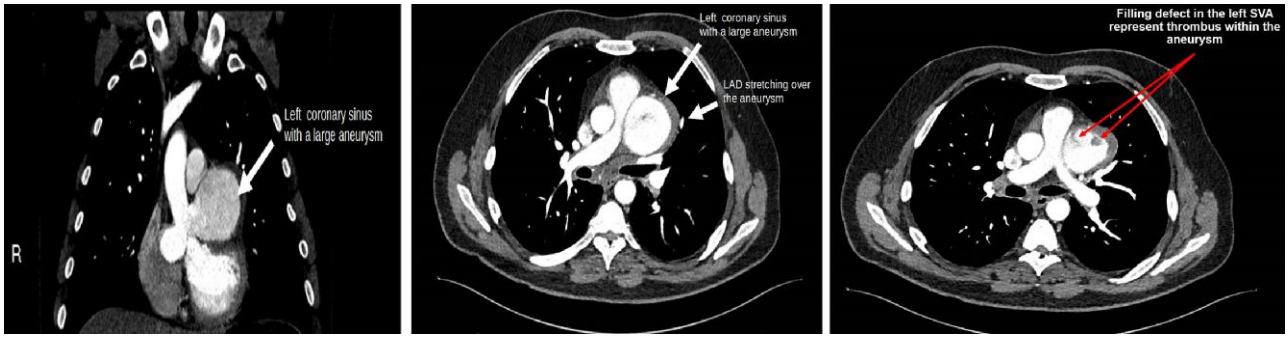


Figure 4 Contrast computed tomography showing hugely dilated left coronary sinuses in sagittal and horizontal plan, in addition to small thrombus at the left superior aspect of the aneurysm (red arrow).

evident that the patient had significant stenosis in both his LMS and LAD. This is likely a result of external compression, potentially from the enlarged left sinus of Valsalva.

A transoesophageal echocardiogram (TOE) (*Figure 3*; see Supplementary material online, *Video S3*) was performed, revealing a large SVA involving the left coronary cusp measured 9.9 cm from cusp to commissure, with spontaneous contrast and laminar thrombus noted within the anterior portion. The study confirmed the TTE finding of severe impairment of left ventricular systolic function secondary to regional wall motion abnormalities in LAD territory. There was evidence of mild aortic regurgitation and mild functional mitral regurgitation.

Further examination with a computed tomography (CT) angiogram of the aorta confirmed aneurysmal dilatation of the three coronary sinuses (*Figure 4*), with the left coronary sinus being the hugely dilated, measuring 63 × 59 mm and extending to the left and cranially, with a small amount of laminar thrombus at the left superior aspect of the aneurysm. On the 3D reconstruction of the CT images, the LAD was observed to be small calibre and stretching over the aneurysm (*Figure 5*). The right coronary sinus was dilated to approximately 31 × 22 mm, while the non-coronary sinus was dilated to 33 × 32 mm. The remainder of the aorta, including the main arch vessels, appeared normal and satisfactory, with no dilatation.

After a full auto-immune and infectious screen was negative, a cardiac magnetic resonance (CMR) imaging was performed to evaluate function and myocardial viability. It showed that only one segment (the apical anterior) was non-viable, while the remaining LAD territories (5/17) were found to be viable with <50% sub-endocardial infarct (see Supplementary material online, *Figure S3* and *Video S4*). Following the discussion at multi-disciplinary team meeting, it was decided that the patient would undergo SVA repair. His heart failure medications (2.5 mg of bisoprolol, 25 mg of eplerenone, 2.5 mg of ramipril, and 40 mg of furosemide once daily) remained on therapeutic dose while awaiting SVA repair surgery. The surgical intervention occurred 30 days subsequent to the initial presentation, as the patient initially declined the procedure. The patient underwent aortic valve and root replacement. Unfortunately, the patient passed away 28 days after surgery, following a protracted postoperative course marked by haemodynamic instability from his severely impaired left ventricular function. During this period, the patient required post-bypass extra-corporeal membrane oxygenation and later transitioned to a biventricular assist device (BiVAD). The case underwent comprehensive discussion within the transplant multi-disciplinary team, resulting in the determination that the patient was not a suitable candidate for transplantation. Towards the end of his stay in the intensive care unit, the patient developed disseminated intravascular coagulation, sepsis, and renal failure. Despite inotropic support and assistance from the BiVAD, the patient remained hypotensive, eventually progressing to multi-organ failure. Regrettably, he passed away on the 28th day post-operatively.

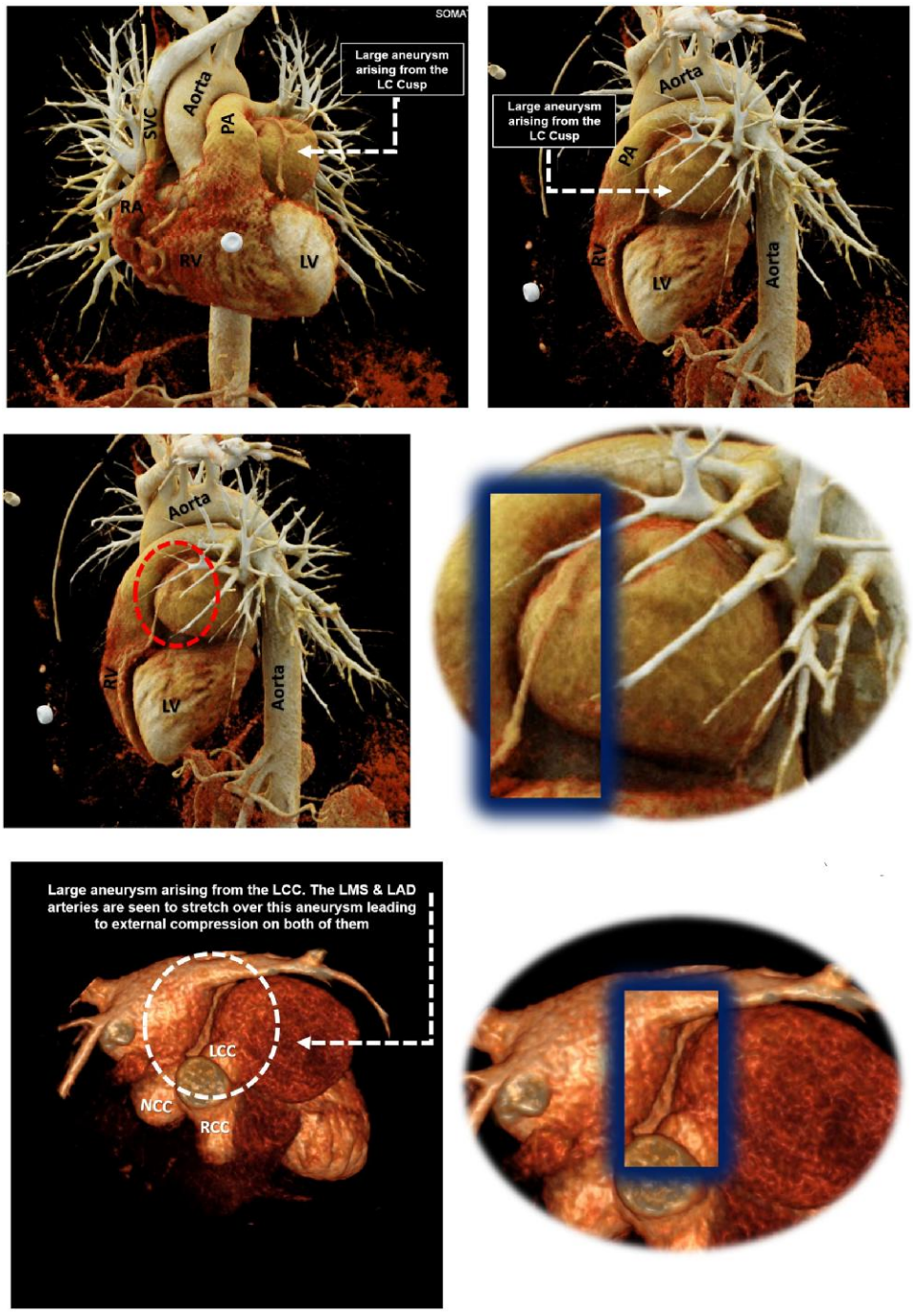


Figure 5 A 3D reconstruction computed tomography scan showed dilatation in all coronary cusps with a huge aneurysm arising from the left coronary cusp. The LMS and LAD are seen to stretch over this aneurysm causing external compression on both the left main stem and left anterior descending artery. The remainder of the aorta including the main arch vessels appear satisfactory and normal with no dilatation and no calcified atheroma. LAD, left anterior descending artery; LCC, left coronary cusp; LMS, left main stem; LV, left ventricle; NCC, non-coronary cusp; PA, pulmonary artery; RA, right atrium; RCC, right coronary cusp; RV, right ventricle.

Discussion

Sinus of Valsalva aneurysm is a rare but potentially life-threatening condition that has been reported in medical literature since the early 19th century. While SVAs can be acquired through infectious processes, degenerative changes, or traumatic injury, most cases are congenital in nature.[2] In these cases, the aneurysm can affect any of the sinuses of Valsalva, the ascending aorta, and even project into the pericardium.

One of the most frequent and dangerous complications of SVA is rupture, which can result in an opening into the right atrium or ventricle or the pleural or pericardial cavity. The diagnosis of a ruptured SVA is typically easier to establish because of the severe clinical picture that accompanies

the condition.[3] However, diagnosing an unruptured SVA is difficult as it is usually silent, showing no clinical manifestations until it becomes infected, produces embolism, or compresses surrounding structures.

While only a few cases of SVA accompanied by myocardial ischaemia have been reported in the international literature, the most common complications of SVAs include obstruction of the right ventricular outflow tract,[4] aortic regurgitation, rhythm disorders,[5] and, in rare cases, myocardial ischaemia due to compression of the coronary arteries.[6,7] While the latter is uncommon, it can be potentially lethal because aneurysmal expansion may be very rapid, requiring emergency surgery. Sinuses of Valsalva aneurysm rarely affect the left coronary sinus (<1%), predominantly affecting the right coronary sinus.[8] Only a handful of case reports have reported on myocardial ischaemia as a complication of left SVAs.[9] There is a higher risk of myocardial ischaemia in left than right SVAs, possibly due to the central part of the left atrial coronary sinus being directly exposed to the pericardium.[10]

Traditionally, surgical management is required for ruptured SVAs, although endovascular closure devices have been used with good outcomes.[11,12] Non-ruptured SVAs should be surgically repaired if there are associated significant symptoms or rapid enlargement.[13] In our case, SVA remained silent until a late anterior MI occurred.

Conclusion

Left SVAs are uncommon, often remain silent, and are discovered incidentally. Due to the close proximity of the left coronary system, they can present with myocardial ischaemia due to extrinsic compression of the coronary system. We were able to perform a comprehensive multi-modality assessment of left SVA, which helped establish this unusual diagnosis and guide management. Transthoracic echocardiogram and TOE helped assess the SVA and demonstrated the thrombus *in situ*, aortic valve insufficiency, and cardiac function. The CT aided in accurately defining the extent of the aneurysm and the extent of compression of the left coronary system and CMR was able to demonstrate viability in LAD and circumflex territory.

Lead author biography

Dr Ghaith M. Maqableh, MBBS, MD, MSc, FRCP, is a board-certified physician from Jordan. He is a graduate of Jordan University of Science & Technology, where he earned his MD degree. Following this, he embarked on a residency programme at King Abdullah University Hospital. Dr Maqableh has pursued a fellowship in Clinical and Interventional Cardiology at Queen Elizabeth Hospital Birmingham (QEHB). He is currently in the final year of his Interventional Cardiology Fellowship training at QEHB. Notably, in 2023, he achieved the prestigious FRCP certificate in Cardiology from the Royal College of Physicians-London, attesting to his excellent expertise in the field.

Supplementary material

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

Consent: The authors confirm that verbal and written informed consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

Funding: No fund was needed to write this case report.

Data availability

All data are incorporated into the article and its online supplementary material.

References

1. Weinreich M, Yu P-J, Trost B. Sinus of Valsalva aneurysms: review of the literature and an update on management. *Clin Cardiol* 2015;**38**:185–189.
2. Ott DA. Aneurysm of the sinus of Valsalva. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2006;**9**:165–176.
3. Moustafa S, Mookadam F, Cooper L, Adam G, Zehr K, Stulak J, et al. Sinus of Valsalva aneurysms—47 years of a single center experience and systematic overview of published reports. *Am J Cardiol* 2007;**99**:1159–1164.
4. John SH. A rare case of unruptured sinus of Valsalva aneurysm obstructing the right ventricular outflow tract. *J Cardiovasc Ultrasound* 2010;**18**:55–57.
5. Agarwal P, Jain A, Singh P, Singh H, Geelani MA, Mehta V. Unruptured right sinus of Valsalva aneurysm dissecting into interventricular septum causing complete heart block: can early surgical correction revert rhythm disturbances? *World J Cardiovasc Dis* 2018;**8**:353–359.
6. Mohammed R, Dandekar U. Large sinus of Valsalva aneurysm presenting as acute coronary syndrome. *Asian Cardiovasc Thorac Ann* 2021;**29**:411–413.
7. Shin JK, Jung JP, Park CR, Lee SG, Shin ES, Kim SJ, et al. Acute myocardial infarction due to unruptured aneurysm of left sinus of Valsalva with aortic valve regurgitation. *J Card Surg* 2005;**20**:545–548.
8. Guo DW, Cheng TO, Lin Lin M, Gu ZQ. Aneurysm of the sinus of Valsalva: a roentgenologic study of 105 Chinese patients. *Am Heart J* 1987;**114**:1169–1177.
9. Gurbuz A, Iscan S, Gokalp O, Karaagac E, Girgin S, Aksun M. Surgical management of left sinus of Valsalva aneurysm presenting with exertional chest pain. *Braz J Cardiovasc Surg* 2020;**35**:573–576.
10. Pedroza AJ, Brewer ZE, Lee AM. Emergency valve-sparing aortic root replacement and coronary artery bypass grafting for giant left sinus of Valsalva aneurysm presenting as acute coronary syndrome. *J Thorac Cardiovasc Surg* 2018;**156**:e81–e84.
11. Cullen S, Somerville J, Redington A. Transcatheter closure of a ruptured aneurysm of the sinus of Valsalva. *Br Heart J* 1994;**71**:479–480.
12. Olearchyk AS, Grasso MA, Danielewski GL. Endoaneurysmal repair of a congenital right coronary sinus aneurysm. *J Card Surg* 1997;**12**:81–85.
13. Vural KM, Şener E, Taşdemir O, Bayazıt K. Approach to sinus of Valsalva aneurysms: a review of 53 cases. *Eur J Cardiothorac Surg* 2001;**20**:71–76.