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CASE REPORT
Cardiac imaging

Large non-coronary sinus of Valsalva aneurysm: a case report of an unusual cause of angina pectoris

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Background Sinus of Valsalva aneurysm (SVA) is a rare cardiac abnormality occurring in 0.09% of the general population, but few reports have examined its recurrence. Unruptured SVAs are usually asymptomatic.

Case summary A 50-year-old woman presented with chest pain and a history of surgery for a ruptured right coronary SVA 32 years prior. Echocardiography showed the recurrence of an unruptured SVA of the non-coronary sinus with moderate aortic regurgitation, severe mitral regurgitation, and severe tricuspid regurgitation. Cardiac computed tomography (CT) revealed compression of the right coronary artery (RCA) between the SVA and sternum. Adenosine triphosphate stress myocardial perfusion imaging (MPI) identified reversible ischaemia of the inferior wall. The patient underwent patch closure of the SVA, aortic valve replacement, mitral valvuloplasty, and tricuspid annuloplasty. Post-operative MPI showed no residual ischaemia, and CT confirmed both successful repair of the SVA and intact RCA.

Discussion This case provides two noteworthy findings. First, the SVA recurred after 32 years. Second, a non-coronary SVA causing myocardial ischaemia is extremely rare given the long anatomical distance between the non-coronary sinus and coronary arteries. In our patient, the non-coronary SVA grew large enough within the anterior mediastinum to cause RCA compression.

Keywords Sinus of Valsalva aneurysm • Angina pectoris • Cardiac computed tomography • Case report

Learning points

- Acquired sinus of Valsalva aneurysms (SVAs) are known to be associated with connective tissue disease, infections, aortitis, and chest trauma.
- The patient developed a recurrent sinus of Valsalva aneurysm 32 years after the first surgery without any findings of underlying disease.
- Non-coronary SVA causing extrinsic compression of coronary artery is extremely rare because of the long anatomical distance between non-coronary sinus and coronary arteries; however, it can occur when the non-coronary SVA develop large enough to fill in the anterior mediastinum.

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Introduction

Sinus of Valsalva aneurysm (SVA) is a rare cardiac abnormality, with a prevalence in the general population of 0.09%, as estimated from an autopsy series of 8138 individuals.[1] The recurrence of SVA is rare, with only a few reported cases.[2,3] Ruptured SVAs often cause symptoms, including substernal chest pain and mild to severe dyspnoea, whereas most unruptured SVAs are asymptomatic.[4] Here, we report a case of an unruptured SVA that caused angina pectoris.

Timeline

32 years prior to admission	History of surgery for a ruptured right sinus of Valsalva aneurysm (SVA)
1 month prior to admission	Chest pain on exertion
Day of admission (Day 0)	Chest X-ray showing bulging of the right cardiac border
	Echocardiography revealing the recurrence of SVA of the non-coronary sinus
	Contrast-enhanced computed tomography (CT) imaging revealing severe narrowing of the right coronary artery (RCA) due to extrinsic compression by SVA
Day 10	Adenosine triphosphate stress myocardial perfusion imaging (MPI) identifying reversible ischaemia in the inferior segment
Day 34	Patch closure of SVA, aortic valve replacement, mitral annuloplasty, and tricuspid annuloplasty
	Admission to intensive care unit
Day 37	Chest drainage and transfusion due to right haemothorax and anaemia
Day 47	Discharge from intensive care unit
Day 93	Post-operative CT imaging demonstrating successfully repaired SVA and intact RCA
Day 98	Post-operative MPI showing no residual ischaemia
Day 100	Discharge home in stable condition
Outpatient clinic (3 years)	Asymptomatic and uneventful

Case presentation

A 50-year-old woman with a history of patch closure and resection of a ruptured right SVA 32 years earlier was referred to our hospital due to chest pain on exertion of 1 month duration. In the years after SVA surgery, she had been followed up without recurrence until the age of 30, after which her follow-up hospital visits had been discontinued. She had no history of atherosclerotic disease, connective tissue disease, or chest trauma and no family history of connective tissue disease or aortic aneurysm. On physical examination, a systolic murmur with an intensity of III/VI at the apex and an incision scar following a median sternotomy, but no physical features of connective tissue disease, were seen. Electrocardiography showed a sinus rhythm and slight ST depression in the inferior leads. Chest X-ray revealed bulging of the right cardiac border, which was not obvious on the images obtained 3 years earlier (*Figure 1*). Echocardiography revealed an unruptured SVA of the non-coronary sinus with moderate aortic regurgitation (AR), severe mitral regurgitation (MR) due to floppy mitral valve, and severe tricuspid regurgitation (TR) presumably due to compression by the SVA (*Figure 2A*, Supplementary material online, *Video S1*). Three-dimensional reconstruction of the cardiac computed tomography (CT) images showed a large non-coronary SVA and severe narrowing of the right coronary artery (RCA). In addition, the transverse and sagittal images revealed that the RCA was compressed between the SVA and sternum (*Figure 3A*, Supplementary material online, *Video S2*). Adenosine triphosphate stress myocardial perfusion imaging (MPI) identified reversible ischaemia in the inferior segment (*Figure 4A*).

The patient underwent surgical intervention for the recurrent SVA. After patch closure of the SVA orifice, water testing revealed residual TR due to an enlarged tricuspid annulus. Tricuspid valve annuloplasty was performed (Medtronic, Contour 3D). Because the MR was thought to be due to a functional prolapse, mitral valve annuloplasty was performed using a Memo 3D ring. Subsequent water testing demonstrated no residual MR. After the aorta had been unclamped, transoesophageal echocardiography revealed severe AR, and aortic valve replacement was performed using an 18 mm ATS-Advanced Performance. Intraoperatively, no marked atherosclerotic changes in the RCA were observed. The pathology report described a loss of medial elastic fibres and fibrous thickening but no inflammation or myxomatous changes in the aneurysmal wall. Post-operative echocardiography showed successfully repaired SVA and AR, trivial MR, and mild to moderate TR (*Figure 2B*, Supplementary material online, *Video S3*). Post-operative CT imaging demonstrated the thrombosed SVA and intact RCA (*Figure 3B*), but no residual ischaemia was seen on the MPI study (*Figure 4B*). The patient was discharged home and has been asymptomatic during the follow-up period. At the 3 years after surgery, echocardiography and CT imaging demonstrated no recurrence of SVA.

Discussion

This case provides two noteworthy findings. First, the SVA recurred after 32 years. Secondly, a non-coronary SVA causing myocardial ischaemia is extremely rare.

To our knowledge, this is the third case of SVA recurrence in another sinus of Valsalva.[2,3] Although acquired SVAs are associated with connective tissue disease, infections, aortitis, and chest trauma, none of these additional pathologies were evident from the clinical history and physical examination of our patient. A mutation in the FBN1 or MFAP5 gene, either of which may cause of thoracic aortic aneurysms complicated by mitral valve prolapse,[5] may have accounted for the recurrent SVA in our patient given that it was

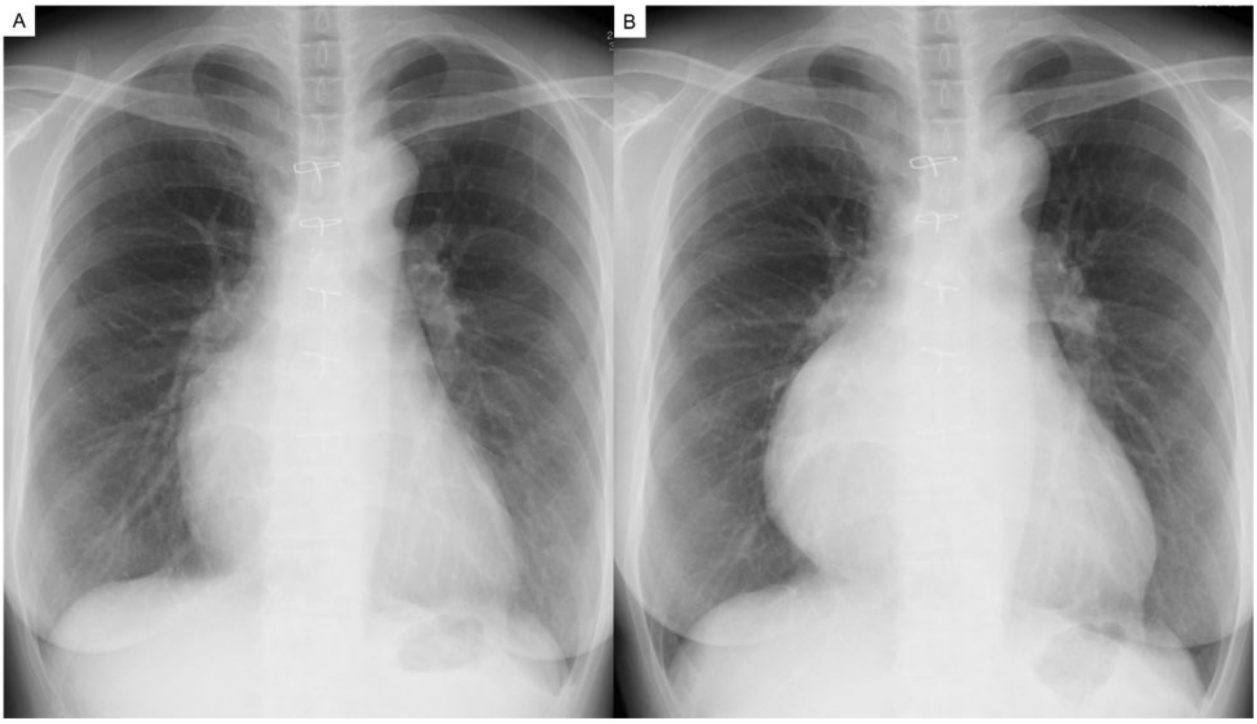

Figure 1 Chest X-rays 3 years prior (*A*) and on admission (*B*).

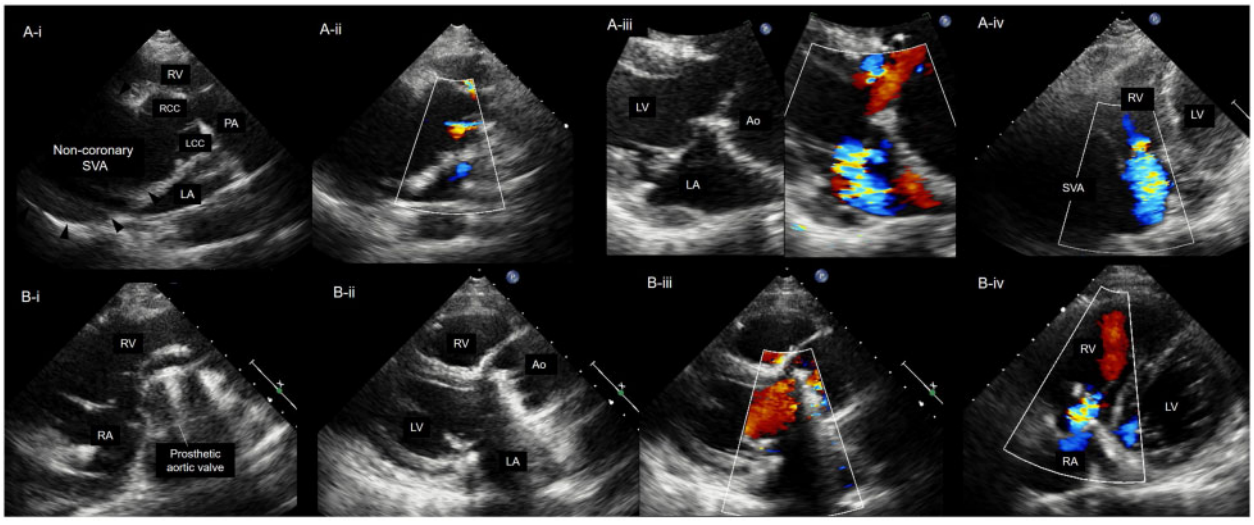

Figure 2 Transthoracic echocardiographic images. (*A*) Pre-operative and (*B*) post-operative results. (*A-i*) Parasternal short-axis view demonstrating the non-coronary SVA (arrowheads). (*A-ii*) Colour flow Doppler shows moderate aortic regurgitation. (*A-iii*) Severe mitral regurgitation due to floppy mitral valve and (*A-iv*) severe tricuspid regurgitation. (*B-i*) Parasternal short-axis view shows a prosthetic aortic valve but not the sinus of Valsalva aneurysm. (*B-ii, iii*) Parasternal long-axis view shows trivial mitral regurgitation without mitral valve prolapse. (*B-iv*) Apical four-chamber view shows mild to moderate tricuspid regurgitation. Ao, ascending aorta; LA, left atrium; LCC, left coronary cusp; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RCC, right coronary cusp; RV, right ventricle; SVA, sinus of Valsalva aneurysm.

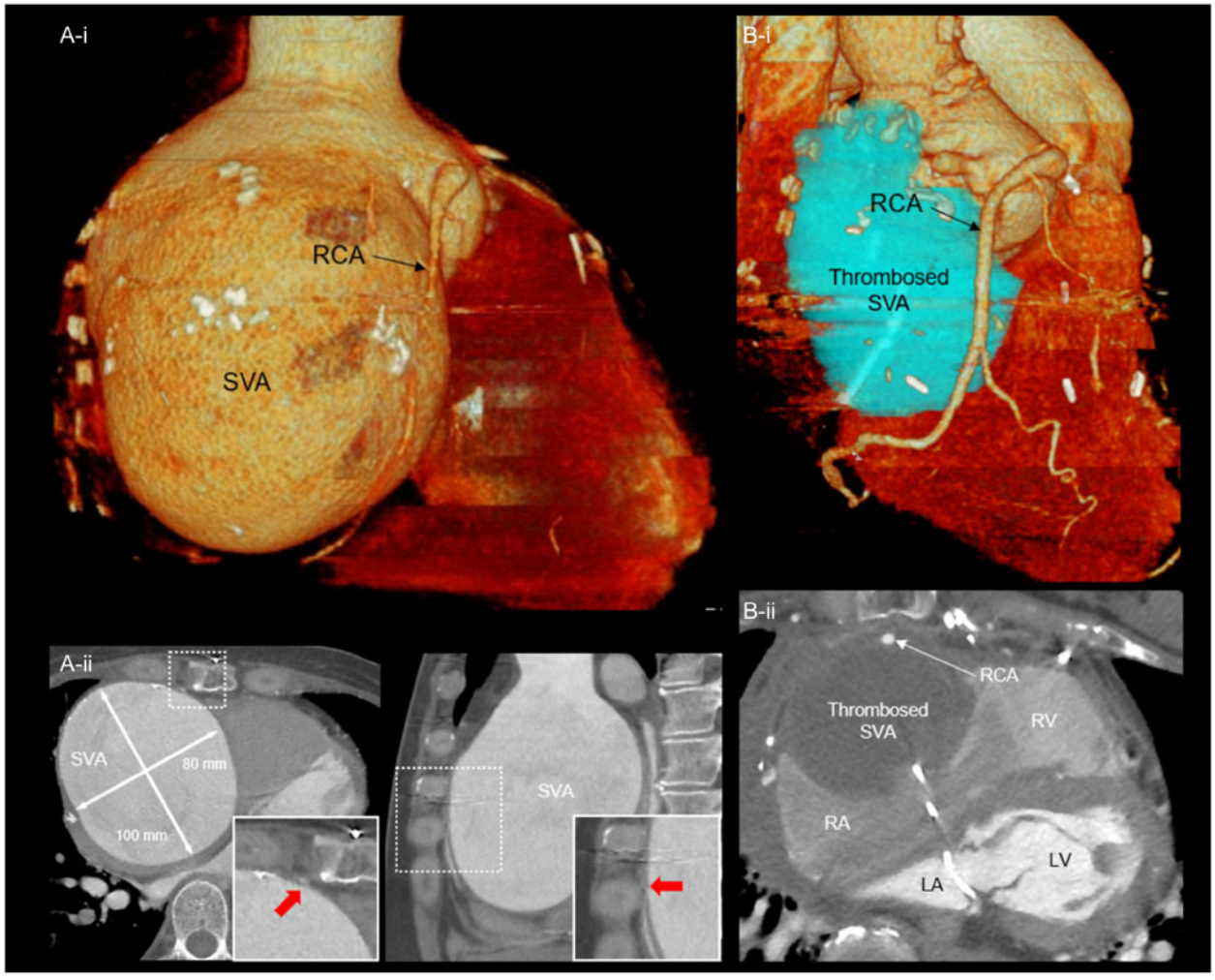

Figure 3 Pre-operative (*A*) and post-operative (*B*) computed tomography imaging. (*A-i*) Three-dimensional reconstruction image shows a large sinus of Valsalva aneurysm and severe narrowing of the right coronary artery. (*A-ii*) Transverse and sagittal images reveal compression of the right coronary artery (red arrows) between the sinus of Valsalva aneurysm and sternum. (*B-i, ii*) Three-dimensional reconstruction and axial image demonstrating the thrombosed sinus of Valsalva aneurysm (light blue mass) owing to a patch repair and an intact right coronary artery. Ao, ascending aorta; LA, left atrium; LCC, left coronary cusp; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RCC, right coronary cusp; RV, right ventricle; SVA, sinus of Valsalva aneurysm.

accompanied by a floppy mitral valve. However, the patient and her family refused genetic testing such that neither FBN1 or MFAP5 gene mutation nor connective tissue disease could be ruled out. This case demonstrates that a patient with a surgically repaired SVA should undergo long-term follow-up by CT imaging even in the absence of evidence of connective tissue disease.

Unruptured SVAs are generally asymptomatic, whereas SVAs that rupture into the internal or external cavity of the heart result in acute heart failure or cardiac tamponade, respectively.[4] However, SVAs rarely cause chest pain due to an external coronary artery compression.[6,7] In those cases, the left or right coronary artery is usually compressed by an aneurysm originating from the coronary sinuses. A non-coronary SVA causing myocardial ischaemia is extremely rare[7], given the long anatomical distance between the non-coronary sinus and coronary arteries. In our patient, the non-coronary SVA grew large enough within the anterior mediastinum to cause RCA compression. Coronary artery bypass grafting (CABG) is considered in patients with SVA causing coronary artery insufficiency,[6–8] with only a few reports of successful repair by primary closure of the aneurysmal orifice without CABG.[9–11] In our patient, the RCA narrowing was likely caused by compression between the SVA and sternum and not by atherosclerosis, for which she had no risk factors nor were marked atherosclerotic changes in the RCA seen intraoperatively. Therefore, The SVA was repaired by patch closure, which was expected to resolve the coronary insufficiency by reducing the

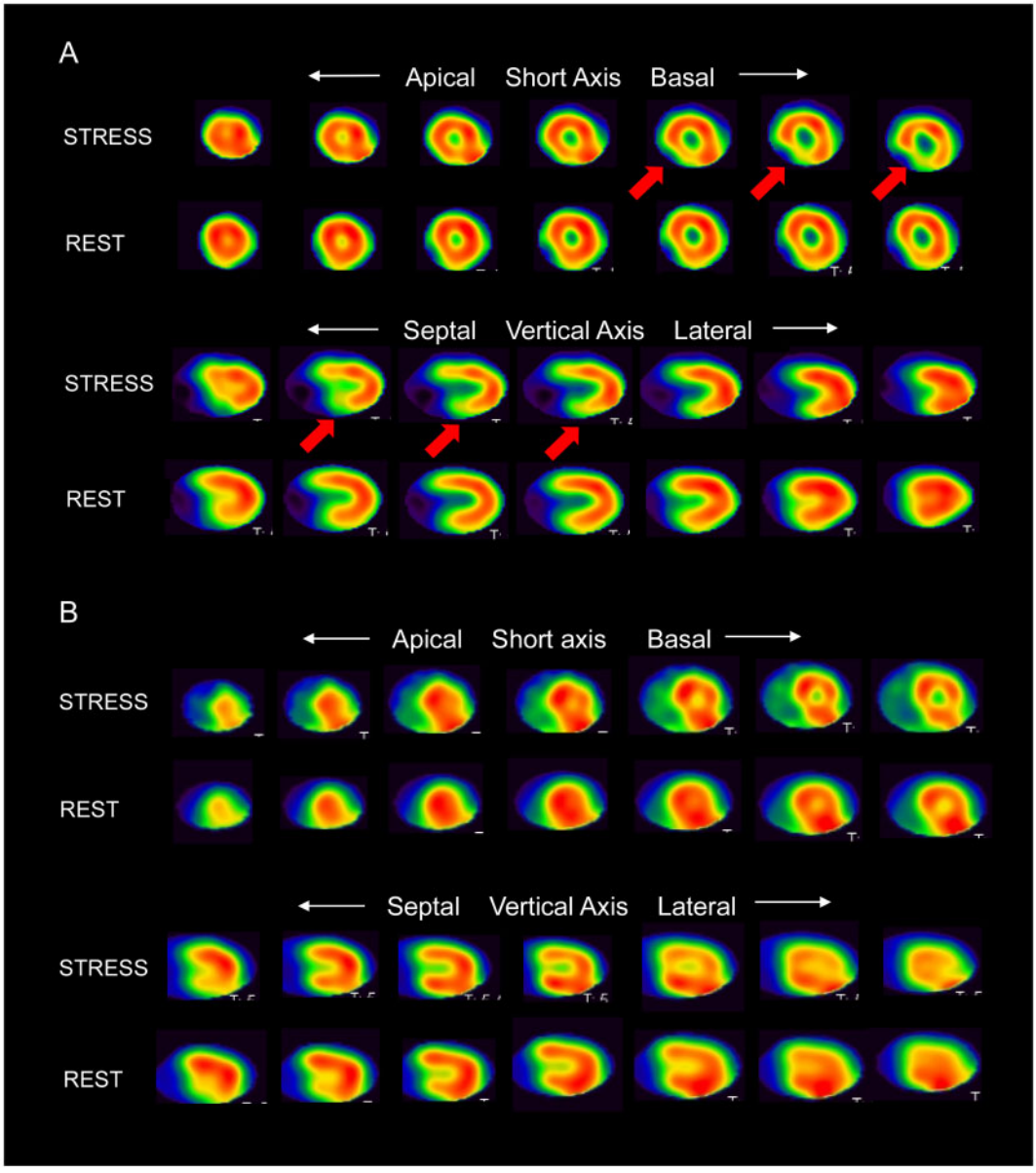

Figure 4 Pre-operative (*A*) and post-operative (*B*) myocardial perfusion imaging. (*A*) Red arrows indicate a reversible perfusion defect in the inferior segment.

volume of the SVA. As expected, residual stenosis and ischaemia were not detected on the post-operative cardiac CT and MPI images.

Conclusion

We reported the case of non-coronary SVA that caused angina pectoris. Severe narrowing of the RCA was resulted from extrinsic compression between the large SVA and sternum, which was precisely diagnosed using multimodal imaging.

Lead author biography

Dr Hidehiro Iwakawa, MD, PhD, graduated from Akita University School of Medicine in 2011. He obtained his doctor's degree in medicine from Akita University Graduate School of Medicine in 2019. Currently, he is a cardiologist in Akita University Hospital, and his clinical and research interests are electrophysiology and cardiac implantable electronic devices.

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 guidelines.

Conflict of interest: none declared.

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