

CASE REPORT

An extracardiac unruptured right sinus of valsalva aneurysm complicated with atherothrombosis

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Summary

We present quite a rare case of extracardiac unruptured right sinus of valsalva aneurysm (SVA) complicated with atherothrombosis in a young adult man. A 35-year-old male with a giant unruptured SVA arising from the right coronary sinus (RCS) with extracardiac protrusion was diagnosed by echocardiography. Contrast-enhanced computed tomography (CT) revealed a huge calcified aneurysm with mural thrombi originating from the aortic root, and about 80% stenosis at the initial segment of the right coronary artery (RCA). Intraoperative exploration demonstrated a giant unruptured aneurysm arising from the RCS. Different from other SVAs reported before, this aneurismal wall appeared thick and atheromatous-like. In this aneurysm, there was a small localized intima tearing and mural thrombosis, and the orifice of the RCA was almost blocked. This patient underwent surgical patch repair to prevent aneurysm rupture and coronary artery bypass grafting for RCA revascularization. In conclusion, the pathological examination demonstrated marked foam cells, inflammatory cells, and thrombosis in the aneurismal wall.

Learning points:

- Echocardiographic characteristics of sinus of valsalva aneurysm (SVA).
- Diagnostic evaluation of extracardiac unruptured SVA.
- Pathology of rare SVA.

Background

Sinus of valsalva aneurysm (SVA) is an unusual cardiovascular anomaly, most commonly rupturing or protruding into the upper portion of right ventricular outflow tract (1). Extracardiac unruptured SVA are rarely reported. Since it is usually asymptomatic, SVA is often incidentally revealed by echocardiography or other imaging modalities for other indications. SVA is either congenital or secondary to infectious diseases, aortic trauma, and degenerative diseases, such as connective tissue disorders, or cystic medial necrosis, giving rise to coronary flow obstruction that causes

myocardial ischemia, which is one of the most common complications of unruptured SVAs (2, 3). A literature review of some cases of giant extracardiac unruptured SVAs revealed that pathological manifestations include absence of medial elastic fibers, mucoid degeneration, myxomatous degeneration, and medial infiltration of eosinophils (4, 5, 6, 7). Few cases reported an SVA complicated with atherothrombosis localized only on the aneurismal wall.

In this report, we present an extremely rare case of a giant extracardiac unruptured SVA arising from the right



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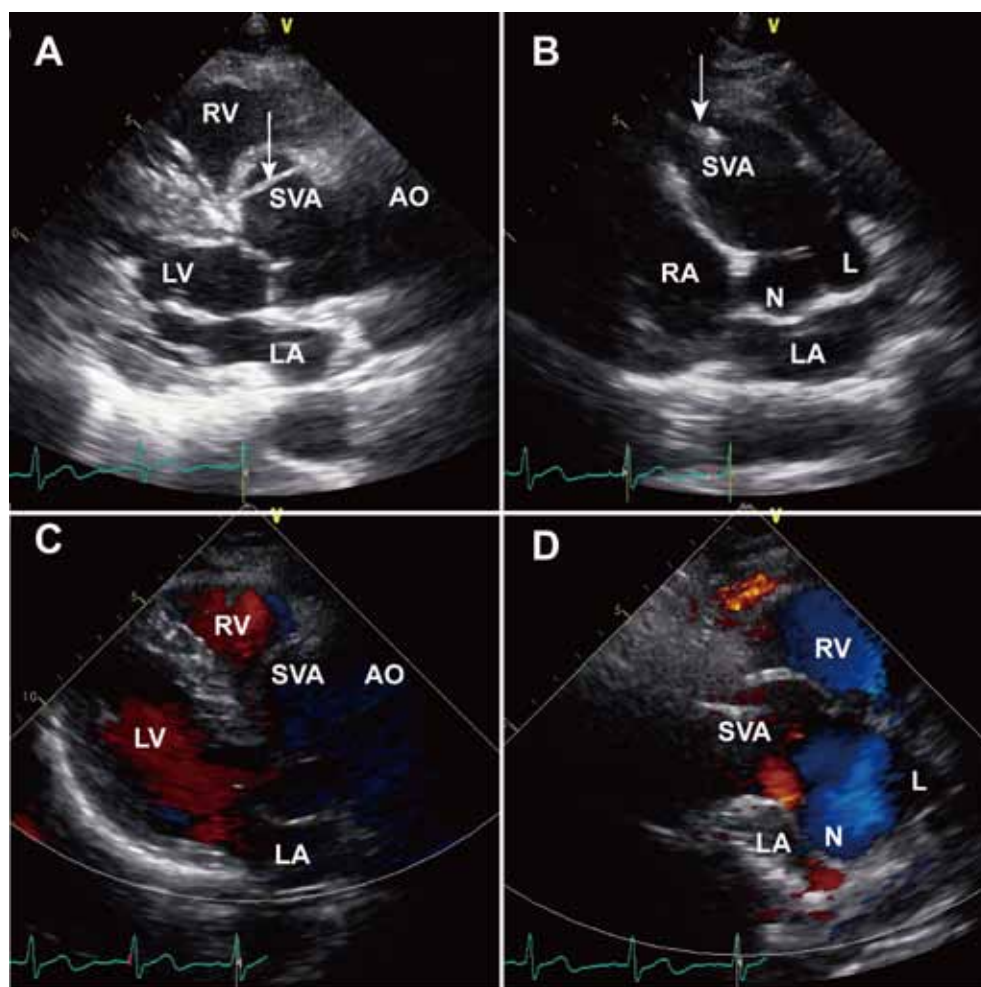


Figure 1

Images of echocardiography before surgery. (A) In transthoracic echocardiography, the parasternal long-axis view showing a cystic mass located in the right coronary sinus with a flap-like appearance in the aneurysm (arrow). (B) On parasternal short-axis view, a giant saccular aneurysm located in the right coronary sinus, approximately 52×37 mm in size. A weak-echogenic flap-like appearance can be seen in the aneurysm (arrow). (C and D) On parasternal long-axis and short-axis views, color Doppler imaging showing no other intracardiac anomalies including aortic regurgitation and ventricular septal defect. SVA, sinus of Valsalva aneurysm; LV, left ventricle; LA, left atrium; RV, right ventricle; RA, right atrium; AO, aorta; L, left coronary sinus; N, noncoronary sinus.

coronary sinus (RCS) with localized atherothrombosis in a young adult man.

Case presentation

A 35-year-old male was referred to our hospital with chest distress. At admission, physical examination revealed a blood pressure of 170/110mmHg, a regular pulse rate of 70 beats/min, and normal temperature. There was no murmur on auscultation of the heart and the lungs. Both electrocardiogram and chest X-ray were nonspecific. There were no abnormalities in whole blood lipid analysis

and renal function examination. Considering the high blood pressure, secondary hypertension was further ruled out after laboratory investigations, CT, and renal angiography. Moreover, standard serological test for syphilis was *Treponema pallidum* negative and CRP, ESR, platelet, and other indicators of serum fluid were normal.

Investigations

Two-dimensional echocardiography on admission incidentally revealed a giant aneurysm located in RCS, approximately 52×37 mm in size. On the parasternal long-axis

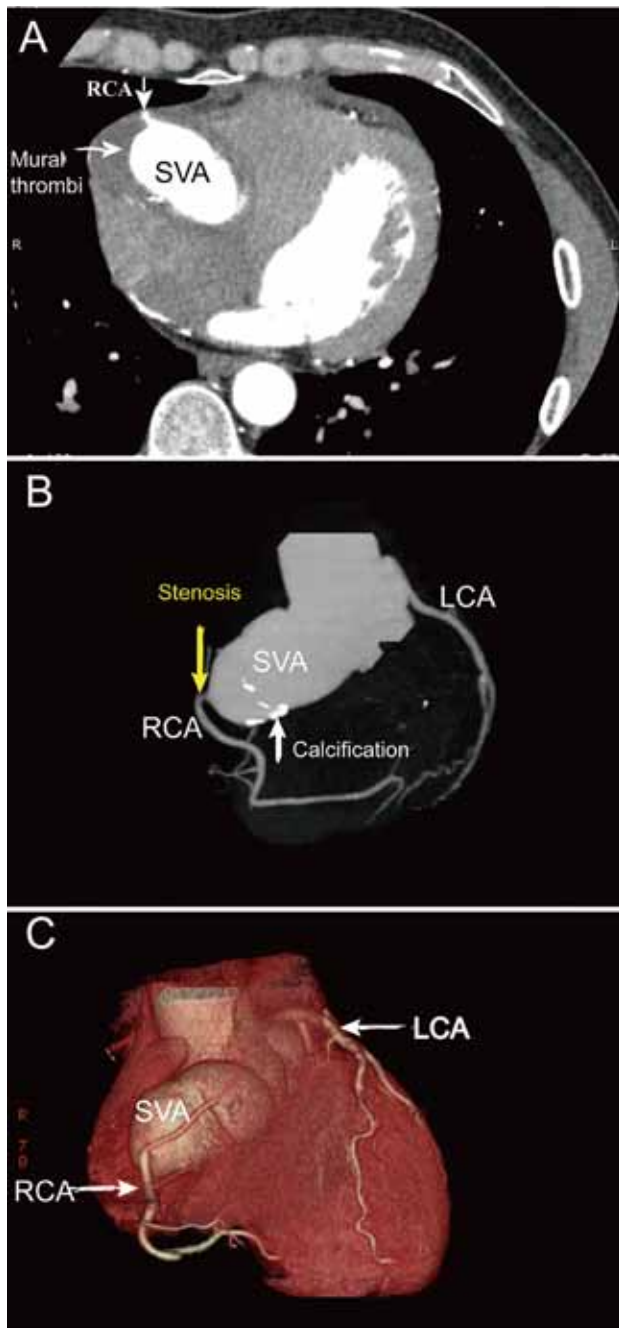


Figure 2

Contrast-enhanced CT and three-dimensional reconstruction demonstrating a giant unruptured aneurysm arising from the RCS with extracardiac protrusion. (A) A view showing the SVA complicated with mural thrombi. (B) CT coronary angiography confirming that the RCA originated from this SVA with 70–80% stenosis (yellow arrow) at the initial segment. (C) Three-dimensional reconstruction showing the SVA with LCA and RCA. SVA, sinus of valsalva aneurysm; LCA, left coronary artery; RCA, right coronary artery.

and short-axis views, a flap-like appearance was visualized in the aneurysm (Fig. 1). Multiple views indicated a giant SVA extracardiac protruded outward without compressing the right atrium and right ventricle. No other intracardiac anomalies such as aortic regurgitation or ventricular septal defect were observed in color Doppler imaging (Fig. 1). Cardiac multislice CT demonstrated a giant unruptured extracardiac aneurysm arising from the RCS, complicated with calcification and mural thrombi. In addition, CT coronary angiography revealed that the right coronary artery (RCA) originated from this SVA and there was 70–80% stenosis at the initial segment of the RCA (Fig. 2).

Treatment and outcomes

Although there were no indications of SVA rupture, immediate SVA patch repair, combined with RCA bypass grafting was performed to prevent potentially life-threatening complications. The procedure was performed via median sternotomy with cardiopulmonary bypass. The intraoperative examination demonstrated a large aneurysm of the RCS (Fig. 3). After dissecting the epicardial fat around the aneurysm and aortic clamping, a longitudinal incision was made on the aneurysm. The aneurismal wall appeared very thick and filled with yellow necrotic atheromatous material (Fig. 3). On the inner side of the aneurysm, a small localized intima tearing and mural thrombosis was detected. The orifice of the aneurysm was located in the RCS and was oval in shape measuring 35×35 mm (Fig. 3). The ostium of the RCA was observed to be close to the orifice and almost totally blocked. The aortic valve was intact, and the other sinuses were normal. Patch closure of the orifice of this SVA was performed using the correct size of prosthetic vascular patch (Fig. 3). After this, the ostium of the RCA was closed, and a right great saphenous vein graft was used to connect the ascending aorta to the proximal RCA as a bridge connection. Finally, the incision of the aneurismal wall was folded and closed, and the patient was easily weaned from cardiopulmonary bypass. Pathological examination with hematoxylin/eosin staining demonstrated significant foam cell formation, infiltration of inflammatory cells, and thrombosis in the aortic wall (Fig. 4).

Postoperative echocardiography and CT demonstrated successful reconstruction of the RCS and RCA revascularization (Fig. 5). Three months after surgery, the patient had recovered well and follow-up echocardiography demonstrated that there was no aortic regurgitation.

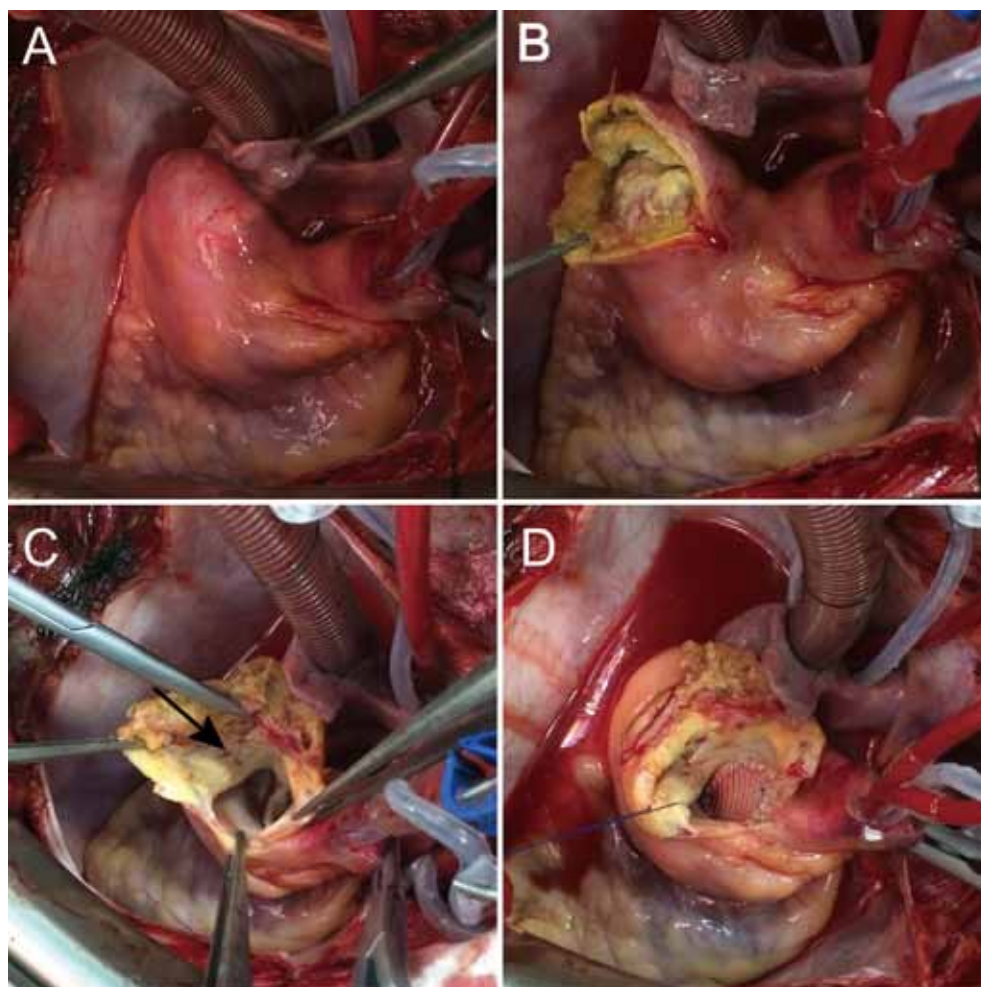


Figure 3

Images of intraoperative examination. (A) Intraoperative examination demonstrating a large SVA with extracardiac protrusion. (B) After a longitudinal incision on the aneurysm, the aneurismal wall appearing very thick and filled with yellow atheromatous necrosis materials. (C) Intraoperative view of the orifice of the aneurysm of the right sinus of Valsalva. The ostium of the right coronary artery close to the orifice and explored almost totally blocked (arrow). (D) Patch closure of the aneurysm was performed.

Discussion

SVA is dilatation of one of the three aortic sinuses between the sinotubular junctions and the aortic valve annulus supra-aortic ridge, which commonly involve the right or noncoronary sinus. As it is a rare type of cardiac anomaly, SVA is mostly detected in the event of a rupture. Patients with ruptured SVAs have symptoms such as dyspnea, chest pain, cough, or peripheral edema. Physical examination could reveal a continuous, mechanical-sounding murmur. In contrast, an unruptured SVA usually remains asymptomatic and undetected unless they expand and affect adjacent tissues. In patients without any obvious

complications, SVAs may be only accidentally detected during examinations (2, 3). Currently, echocardiography is the initial imaging choice to detect SVA in patients. On echocardiograms, SVA commonly demonstrate a thin-walled saccular lesion arising from the aortic root in continuation with the aortic annulus. Based on echocardiographic features which include origin, protruding position, and whether ruptured or not, SVAs could be classified into different patterns (1). In previously reported patients, a few unruptured SVAs arising from the RCS with extracardiac protrusion were detected. In fact, inexperienced doctors can easily be confused when RCA dilation is observed, because echocardiography is not always satisfactory in

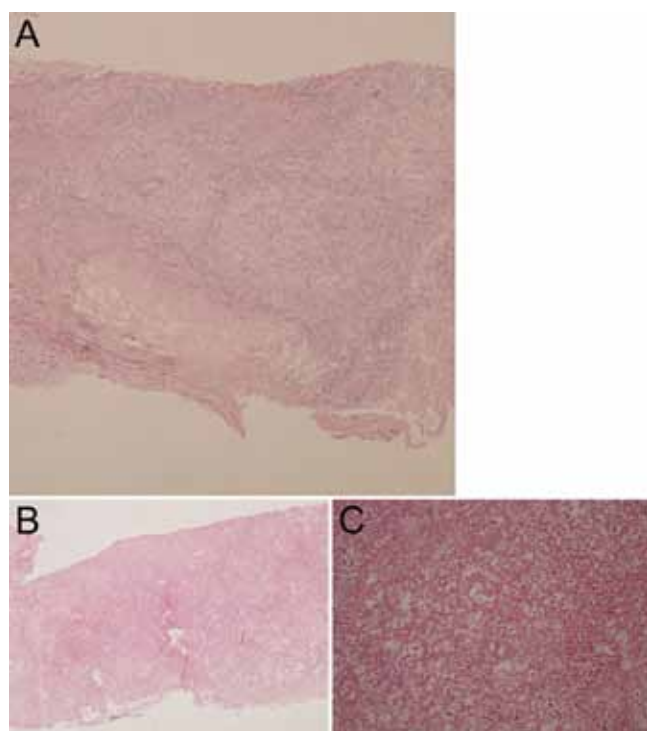


Figure 4

Hematoxylin/eosin staining of the aneurysm wall. (A) Extensive foam cell formation and inflammatory cell infiltration into the aortic wall with deposition of calcium salt. Hematoxylin/eosin staining, x4. (B) Mural thrombosis in this aneurysm. Hematoxylin/eosin staining, x4. (C) At high magnification, network of platelets and fibrin containing red blood cells. Hematoxylin/eosin staining, x40.

precisely delineating the anatomic relationships of the aneurysm with its associated lesions. Further CT coronary angiography and 3D reconstruction were helpful in confirming that the RCA arose from the distal portion of the aneurysm. In contrast to the more common simple SVAs, prominent mural thrombi and calcification were found in the aneurysm described here. In addition, this case was complicated by severe RCA stenosis. Compression of coronary arteries can cause a myocardial ischemic event, which is a severe complication of unruptured SVA, and may even result in myocardial infarction or angina (2, 3). In fact, the mechanism behind coronary flow obstruction is different in left and right sinus aneurysms (8). Multiple case reports describe how unruptured left SVA compress the LCA to give rise to LCA stenosis, and in a few cases right SVA can also cause compression of the proximal part of the RCA. However, coronary flow obstruction due to right SVA mainly involves the RCA ostium. More specifically, RCA may be occluded by a thrombus in the aneurysm or surrounded by a hematoma, leading to a stenosis

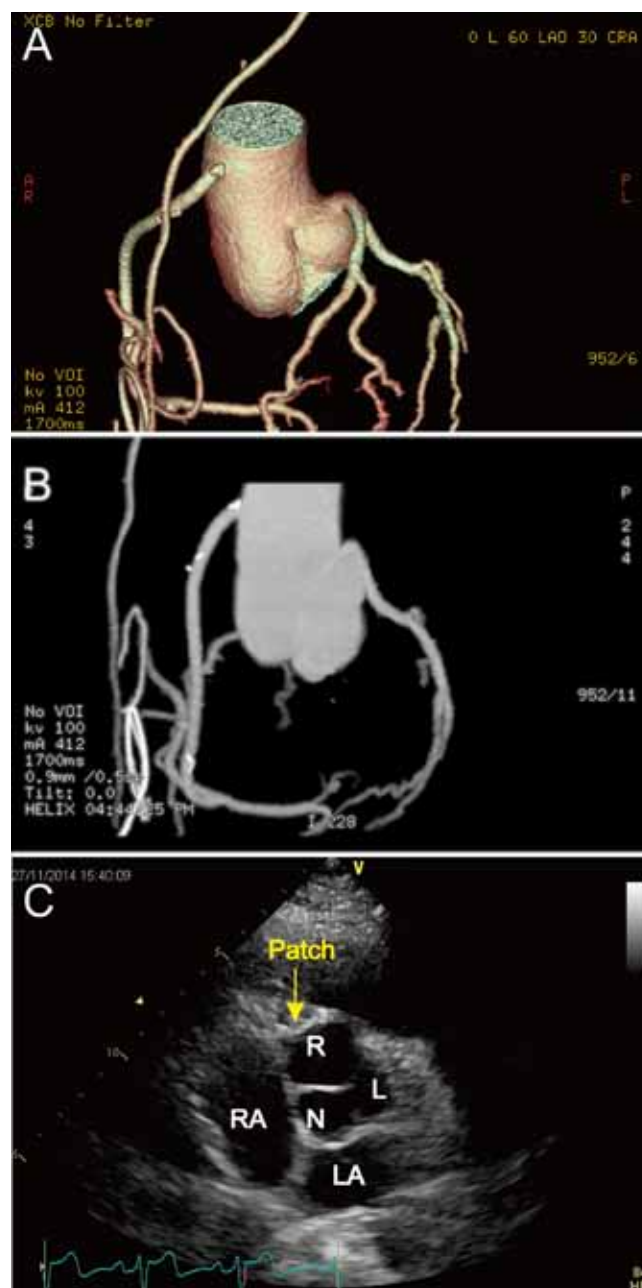


Figure 5

CT coronary angiography and echocardiography images after surgery. (A) Postoperative CT three-dimensional reconstruction: RCS was remodeled with patch repair. (B) Postoperative CT coronary angiography: the GSV graft connecting the ascending aorta and the proximal RCA. (C) Postoperative echocardiography: RCS is remodeled with patch repair (yellow arrow). LA, left atrium; RA, right atrium; R, right coronary sinus; L, left coronary sinus; N, noncoronary sinus; GSV, great saphenous vein.

in the RCA ostium (8). Polat and coworkers reported a right SVA which caused acute myocardial infarction (9). Their case presented with a thrombotic right SVA

associated with RCA occlusion at ostium with thrombus, which led to acute myocardial infarction and ischemic stroke (9). In our case, the localized mural thrombi and calcification that obstructed the ostium of RCA may be the cause of the clinical complaint of this particular patient.

These imaging results coincided with the final surgical exploration, apart from a localized intima tearing in the aneurysm. Although echocardiographic view showed a flap-like appearance in the aneurysm, we could not decide whether it indicated the intima tearing or not. This case report demonstrates that imaging modalities sometimes cannot make a comprehensive diagnosis, especially in the SVA with rare complications. Therefore, surgical exploration was quite important in confirming the final diagnosis.

SVA can be congenital in origin or may be acquired through infection, degenerative diseases, trauma, or atherosclerosis. In our case, the pathological finding was atherosclerotic change. The patient denied any history of trauma and infections. Laboratory examination results could rule out the possibility of infective disease such as Kawasaki or Lues. Even in the absence of signs of atherosclerosis elsewhere in this patient, the aneurismal wall presented with marked foam cell formation and inflammatory cell infiltration. Most of the previous SVA cases were complicated with coronary arterial atherosclerosis that caused coronary obstruction of arteries. In our case, an unruptured extracardiac SVA with atherothrombosis involving the SVA wall was observed. Moreover, almost obliterative RCA was secondary to localized atherosclerosis of the aneurysm rather than from compression of the SVA. To our knowledge, this is the first case report of such a condition.

In this case, a conventional surgical patch repair was performed to prevent aneurysm rupture and thromboembolic events. Meanwhile, coronary artery bypass grafting was also performed for RCA revascularization. Three months after surgery, the patient had recovered well and imaging examinations showed successful repair.

In summary, we present an extracardiac unruptured SVA complicated with localized atherothrombosis and obstruction of RCA ostium. SVA should be diagnosed by echocardiography, CT coronary angiography, surgical exploration, and pathological examination. Surgery is the main treatment option for SVA.

Declaration of interest

The authors declare there is no conflict of interest that could be perceived as prejudicing the impartiality of this case report.

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Patient consent

Written consent was obtained.

Author contribution statement

Jun Zhang collected images, wrote the manuscript and completed a literature review. Yani Liu and Youbin Deng reviewed the manuscript prior to submission and assisted with review of the literature and included images. Ligang Liu provided the clinical information of the patient and performed the operation.

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