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 was diagnosed as having giant unruptured SVA arising from the right coronary sinus (RCS) with extracardiac protrusion by echocardiography. Contrast-enhanced computed tomography (CT) scan revealed a huge calcified aneurysm with mural thrombi originating from the aortic root, and right coronary artery (RCA) was involved as about 80% stenosis at the initial segment. 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

• Characteristics of echocardiogram 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 upper portion of right ventricular outflow tract\(^1\). Extracardiac unruptured SVA was 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 causing myocardial ischemic is one of the most common complications of unruptured SVAs\(^2\)\(^-\)\(^3\). With a literature review of some cases of giant extracardiac unruptured SVAs, the pathological manifestations included absence of medial elastic fibers, mucoid degeneration, myxomatous degeneration and medial infiltration of the eosinophils\(^4\)\(^-\)\(^7\). Few of case revealed a SVA complicated with atherothrombosis only located on the aneurismal wall.

In this report, we present an extremely rare case of a giant extracardiac unruptured SVA arising from the right 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 170/110 mmHg, a regular pulse rate of 70 beats/min, and normal temperature. There was no murmur in auscultation of the heart and the lungs. Both electrocardiogram and chest X-ray were non-specific. 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,
computed tomography (CT) scan and renal angiography. Moreover, standard serological test for syphilis showed 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 $52 \times 37$ mm in size. On the parasternal long-axis and short axis views, a flap-like appearance can be visualized in the aneurysm (Figure 1). Multiple views indicated this giant SVA extracardiac protruded outward without right atrium and right ventricle compression. There were no other intracardiac anomalies including aortic regurgitation and ventricular septal defect in color doppler imaging (Figure 1). Cardiac multislice CT demonstrated a giant unruptured extracardiac aneurysm arising from the RCS, complicated with calcification and mural thrombi in it. In addition, CT coronary angiography revealed right coronary artery (RCA) originated from this SVA and there was 70% to 80% stenosis at the initial segment of the RCA (Figure 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 done via median sternotomy with cardiopulmonary bypass. The intraoperative examination demonstrated a large aneurysm of the RCS (Figure 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 atheromatous necrosis materials (Figure 3). On the inner side of the aneurysm, a small localized intima tearing and mural thrombosis were detected. The orifice of the aneurysm was located in the RCS and demonstrated an oval measuring $35 \times 35$ mm (Figure 3). The ostium of the RCA was seen 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 right-size prosthetic vascular patch (Figure 3). After this, the ostium of the RCA was closed. And right great saphenous vein graft was used to connect the ascending aorta to the proximal RCA with bridge connection. Finally, the incision of the aneurismal wall was folded closed and the patient was easily weaned from cardiopulmonary bypass. The pathological examination with hematoxylin/eosin staining demonstrated significant foam cells, inflammatory cells and thrombosis in the aortic wall (Figure 4).

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

Discussion

SVA is dilatation of one of the three aortic sinuses between the sinotubular junctions and the aortic valve annuluses supra-aortic ridge, which commonly involving the right or noncoronary sinus. As a kind of rare cardiac anomalies, SVA is mostly detected in the situation of complication of a rupture. Patients with ruptured SVAs have symptoms such as dyspnea, chest pain, cough, or peripheral edema. Physical examination could reveal continuous, mechanical-sounding murmur. In contrast, an unruptured SVA usually remains asymptomatic and undetected unless expanding SVA affecting the adjacent tissues. In patients without any obvious complications, SVAs may only be accidentally detected by examinations.

Currently, echocardiography is the initial imaging choice to detect SVA in suspected patients. On echocardiograms, SVA commonly had a thin-walled saccular lesion arising from the aortic root in continuation with the aortic annulus. According to the echocardiographic features including origin, protruding position, and whether ruptured or not, SVAs could be classified into different patterns. In previously reported patients, a few unruptured SVA arising from RCS with extracardiac protrusion was detected. In fact, it was easily confused with dilation of
RCA for inexperienced doctors since echocardiography was not always satisfactory in
imprecisely delineating the anatomic relations of the aneurysm and its associated lesions.
Further CT coronary angiography and 3D reconstruction was helpful in confirming the RCA
arising from the remote of the aneurysm. Different from common simple SVAs, prominent
mural thrombi and calcification were found in the aneurysm. In addition, this young patient
was complicated with severe RCA stenosis. Compressing coronary arteries to cause a
myocardial ischemic event is severe complication of unruptured SVA, which can even result
in myocardial infarction or angina⁵⁻⁶. In fact, the mechanism of coronary flow obstacle is
different between the left and right sinus aneurysms⁸. Multiple case reports described
unruptured left SVA compressing the LCA to give rise to LCA stenosis, and a few right SVA
could also cause the proximal part of RCA compression. However, coronary flow obstruction
due to right SVA is mainly because of involvement of RCA ostium. More specifically, RCA
may be occluded by a thrombus in the aneurysm or surrounded by a haematoma, leading to a
stenosis in the RCA ostium⁸. Polat et al. reported a right SVA causing acute myocardial
infarction, They presented a thrombotic right SVA associated with RCA occluded at ostium
with the thrombus, which led to acute myocardial infarction and ischemic stroke⁹. 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 except a localized intima
tearing in the aneurysm. Although there was a flap-like appearance in the aneurysm on
echocardiographic views, we couldn’t decide whether it indicated the intima tearing or not.
This patient report demonstrated imaging modalities sometimes could not 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. And laboratory examination results could rule out the possibility of infective disease such as Kawasaki or Lues. Although in the absence of atherosclerosis anywhere else, the aneurismal wall presented as marked foam cells and inflammatory cells infiltration. Most of previous SVA cases were complicated with coronary arterial atherosclerosis which caused coronary arteries obstruction. In our case, unruptured extracardiac SVA with atherothrombosis involving the SVA wall was observed. Moreover, the reason for almost obliteratorive RCA was secondary to localized atherosclerosis of the aneurysm not compression of SVA. To our knowledge, this is the first case report of such a condition.

In present reported case, 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 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 exactly diagnosed by echocardiography, CT coronary angiography and even surgical exploration and pathological examination. Surgery repair is main treatment for SVA.

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Declaration of interest
The authors declare there is no conflict of interest that could be perceived as prejudicing the impartiality of the research reported.
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.

References


Legends to figures

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 52×37mm in size. A weak-echogenic flap-like appearance can be seen in the aneurysm (Arrow).

C & D On parasternal long axis and short axis views, color doppler imaging showing no other intracardiac anomalies including aortic regurgitation and ventricular septal defect.

SV A= 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.

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 SV A complicated with mural thrombi.

B. CT coronary angiography confirming RCA originated from this SV A with 70% to 80% stenosis (Yellow arrow) at the initial segment.

C. Three dimensional reconstruction showing the SV A with LCA and RCA.
SVA = sinus of Valsalva aneurysm; LCA = left coronary artery; RCA = right coronary artery.

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 aneurysmal 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.

Figure 4 – Hematoxylin/eosin staining of the aneurysm wall.
A. Extensive foam cells and inflammatory cells infiltration in the aortic wall with deposition of calcium salt. Hematoxylin/eosin staining, ×4.
B. Mural thrombosis in this aneurysm. Hematoxylin/eosin staining, ×4.
C. At high magnification, network of platelets and fibrin containing red blood cells. Hematoxylin/eosin staining, ×40.

Figure 5 – Echocardiography and CT coronary angiography images after surgery.
A. Postoperative echocardiography: RCS is remodeled with patch repair (Yellow arrow).
B. Postoperative CT three dimensional reconstruction: RCS was remolded with patch repair.
C. Postoperative CT coronary angiography: the GSV graft connecting the ascending aorta and the proximal RCA.

LA = left atrium; RA = right atrium; R = right coronary sinus; L = left coronary sinus; N = noncoronary sinus; GSV = great saphenous vein.
Figure 1 – Images of echocardiography before surgery.
86x86mm (600 x 600 DPI)
Figure 2 – Contrast-enhanced CT and three dimensional reconstruction demonstrating a giant unruptured aneurysm arising from the RCS with extracardiac protrusion.

172x369mm (600 x 600 DPI)
Figure 3 – Images of intraoperative examination.
59x59mm (600 x 600 DPI)
Figure 4 – Hematoxylin/eosin staining of the aneurysm wall.
43x27mm (600 x 600 DPI)
Figure 5 – Echocardiography and CT coronary angiography images after surgery.

159x317mm (600 x 600 DPI)