Rupture of the right sinus of Valsalva aneurysm and formation of ventricular septal dissection and third-degree atrioventricular block: A case report

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Abstract
Sinus of Valsalva aneurysm is a rare congenital or acquired cardiac structural abnormality. The sinus of Valsalva aneurysm refers to the local development defect of the aortic sinus wall. Under aortic hypertension, the sinus wall becomes thinner and expands outward. Its clinical manifestations range from asymptomatic to severe life-threatening complications caused by compression or rupture of important surrounding structures. A few cases of aortic aneurysm sinus with arrhythmias have been reported, including complete heart block. Here, we present the case of a 50-year-old male patient with recurrent syncope due to a third-degree atrioventricular block. We found a right sinus of Valsalva aneurysm that ruptured into the basal segment of the interventricular septum and formed a basal septal dissection on echocardiography, which could have caused a third-degree atrioventricular block. This case report highlights the importance of finding secondary etiologies in the sudden or transient onset of third-degree atrioventricular block and using echocardiography to evaluate patients with heart block.

Keywords
Sinus of Valsalva aneurysm, ventricular septal dissection, third-degree atrioventricular block, syncope, echocardiography

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Introduction
Sinus of Valsalva aneurysm (SVA) is a rare congenital or acquired cardiac structural abnormality. SVA refers to the local development defect of the aortic sinus wall. Under aortic hypertension, the sinus wall becomes thinner and expands outward. About 65%–85% of SVAs originate from the right coronary sinus, 10%–30% from the non-coronary sinus, and <5% from the left coronary sinus.1 It is usually more common in men (4:1) and has a higher incidence rate in the Asian population.1 SVA results from the weak elastic layer at the junction of the aortic middle layer and annulus fibrosus.2 Since SVA is usually asymptomatic, it is rarely diagnosed. With the development and widespread use of echocardiography and the emergence of other noninvasive imaging methods, SVA can be found early and accurately. Unruptured SVA can cause compression of the coronary artery and exertional dyspnea, palpitation, and chest pain. Compression of the cardiac conduction system can also cause serious arrhythmias, such as ventricular tachycardia, atrial fibrillation, and atrioventricular block.

Case report
On 15 September 2021, a 50-year-old man appeared in the emergency room with dizziness for more than a month and three syncope episodes occurred within an hour. He had no history of chest tightness, chest pain, headache, fever, recent use of drugs, alcoholism, and heavy physical activity. He was in good health and had no family history of heart disease. At admission, the blood pressure was 133/74 mmHg, heart rate was 61 beats per minute (bpm), and the breathing rate was 16/min. There was no murmur on cardiac auscultation. Physical examinations showed no abnormalities. The
emergency room electrocardiogram (ECG) showed sinus rhythm with ventricular escape rhythm, ventricular escape segment rate of 48 bpm, third-degree atrioventricular block, and complete right bundle branch block (Figure 1(a)). The patient was hospitalized and implanted with a temporary pacemaker. His emergency head computed tomography (CT) scan showed no abnormality. Laboratory tests showed the concentration of high-sensitivity troponin T to be 0.040 ng/mL (normal range: 0–0.14 ng/mL) and N-terminal pro-B-type natriuretic peptide to be 1225.00 pg/mL (normal range: 0–300 pg/mL). His blood routine, renal function, liver function, thyroid function, and electrolyte tests were within normal ranges.

Transthoracic echocardiography showed that the right coronary sinus was dilated (approximately 36 × 27 mm in size), causing the ventricular septum to become torn, the sinus wall to float with the blood flow, expanding into the left ventricular outflow tract in diastole, and sinking in systole. The myocardium of the left ventricular apex became thinner and the motion amplitude of the middle left inferior
ventricular wall and the apical segment decreased (Figure 2). To further confirm this diagnosis, we performed computed tomography angiography (CTA) of the heart and large vessels. The CTA showed that the right sinus of Valsalva was dilated and expanded into the left ventricular outflow tract, with a size of approximately $38 \times 25$ mm, resulting in stenosis of the left ventricular outflow tract. We also detected the formation of an interventricular septal dissection above the bottom of the interventricular septum, with a range of about $10 \times 9$ mm (Figure 3).

The patient was then transferred for cardiovascular surgery. The patient underwent aortic valve replacement, aortic sinus aneurysm resection, and repair under general anesthesia. Preoperative transesophageal echocardiography showed that the right sinus of Valsalva was dilated (about $37 \times 25$ mm in size), protruding into the left ventricular outflow tract during diastole, close to the anterior mitral valve, and retracted during systole. There was a tear of approximately $3$ mm on the side of the left ventricular outflow tract and an anechoic area of approximately $8 \times 7$ mm in the interventricular septum communicating with the right sinus of Valsalva. Thoracotomy revealed that the right coronary sinus was significantly enlarged and the wall of the sinus aneurysm protruded from the left ventricular outflow tract. The sinus aneurysm had a $5$-mm diameter laceration, and a $10 \times 20$ mm cavity was visible between the sinus wall and the ventricular septum.

The patient’s ECG showed 1:1 atrioventricular conduction on the first day after operation (Figure 1(b)). After repeated operation, the 24-hour ambulatory ECG showed 1:1 atrioventricular conduction. The patient recovered well and left the hospital on the 19th day after the operation. After a year of telephone follow-ups, the patient had no symptoms of dizziness, headache, chest tightness, chest pain, and other discomfort, and the ECG showed 1:1 atrioventricular conduction.

Figure 2. Parasternal long-axis view of the left ventricle: (a) the right coronary sinus was dilated and the sinus wall expanded into the left ventricular outflow tract during diastole; (b) the right coronary sinus wall was sunken in systole; (c) the right coronary sinus was dilated and blood flow signals can be seen at the bottom of the ventricular septal base and the right coronary sinus region; (d) parasternal short axis: the diastolic tumor can be seen expanding into the left ventricular outflow tract and close to the anterior mitral leaflet.

RV: right ventricle; LV: left ventricle; RA: right atrium; LA: left atrium; AML: anterior mitral leaflet; MVO: mitral valve orifice; PML: posterior mitral leaflet; IVS: interventricular septum.
Discussion

SVA is a rare cardiac abnormality, with an incidence rate of 0.09% in the general population and 0.1%–3.5% in all congenital heart diseases.\(^1\)\(^,\)\(^3\) Congenital SVA is often associated with Marfan syndrome, Ehlers-Danlos syndrome, or other connective tissue diseases. The acquired aortic sinus aneurysm is associated with trauma, atherosclerosis, infective endocarditis, iatrogenic injury during aortic valve replacement, syphilis, and collagen vascular diseases.\(^1\)\(^,\)\(^2\)\(^,\)\(^4\) Our patient did not have a supportive etiology, so we believe he had a congenital SVA.

Acute SVA rupture often occurs after weightbearing or strenuous exercise. The clinical manifestation and severity of the rupture depend on the speed of the rupture, the size of the ruptured hole, and the chamber of rupture. Most patients have chest pain, dyspnea, palpitation, fatigue, and syncope. A continuous and loud machine-like murmur with tremors can be heard between the third and fourth ribs at the left edge of the sternum. Our patient’s right coronary sinus tumor ruptured in the ventricular septum, so no cardiac murmur was detected. The natural history of unruptured asymptomatic SVA is unknown, and the best treatment is unclear.\(^4\) SVA rupture requires early surgical treatment because the median survival without treatment is 3.9 years.\(^5\) Treatments usually include primary closure, patch repair, or aortic root replacement, with or without valve replacement. Perioperative mortality ranges from 1.9% to 3.9%.\(^6\)\(^,\)\(^7\) After repair, the life expectancy of patients is close to that of healthy people. The prognosis is good after surgical repair, and the 15-year event-free survival rate is about 90%.\(^6\)\(^,\)\(^8\)

In this case, we reported a rare third-degree atrioventricular block due to the right coronary sinus aneurysm that invades the interventricular septum and forms an interventricular septal dissection. Pier Paolo Bocchino et al.\(^9\) reported a similar case of complete atrial ventricular block after right sinus rupture in Valsalva. We believe that the patient’s atrioventricular block was caused by rupture of the right coronary sinus aneurysm into the ventricular septum and impact and compression of blood flow on the conduction bundle, resulting in change of conduction function or compression of the conduction bundle blood supply system, resulting in various degrees of ischemia. This situation demonstrated the importance of searching for reversible causes when a complete atrioventricular block occurs suddenly or transiently. In these cases, echocardiography is a valuable diagnostic tool that can rule out the need for permanent pacemaker implantation or search for reversible causes. Echocardiography of the patient showed myocardial ischemia, including thinning of the left ventricular apical myocardium, interruption of the left ventricular inferior wall, and weakening of the apical motion, which could be the manifestation of transient myocardial ischemia caused by compression of a coronary artery due to the right coronary sinus aneurysm. In our current case, we believe that syncope is the initial clue that leads to the diagnosis of the right coronary sinus aneurysm of the aorta. This can be explained by an expansion of the right coronary sinus aneurysm, extending to the ventricular septum, causing a partial tear of the upper part of the ventricular septum, damaging the normal atrioventricular node or bundle function, and resulting in complete atrioventricular block and syncope. On the first day after the operation, the patient’s
ECG showed 1:1 atrioventricular conduction, which corroborated our hypothesis.

**Conclusion**

Finding a reversible cause when a third-degree atrioventricular block occurs suddenly or transiently is crucial. An aortic sinus aneurysm might be asymptomatic before acute rupture. Echocardiography is the preferred initial diagnostic tool. Finally, the prognosis is poor once the rupture occurs and surgical repair is required.

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**Author contributions**

C.Z., J.F., and W.S.Z. participated in patient treatment and collection of relevant data, S.Y.Y. and Z.Z. reviewed the literature and participated in the drafting of the manuscript; Y.Y. was responsible for the revision of the manuscript for important intellectual content; all authors issued final approval of the version to be submitted.

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**Informed consent**

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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**References**

1. Feldman DN and Roman MJ. Aneurysms of the sinuses of Valsalva. *Cardiology* 2006; 106: 73–78.
2. Weinreich M, Yu PJ and Trost B. Sinus of Valsalva aneurysms: review of the literature and an update on management. *Clin Cardiol* 2015; 38(3): 185–189.
3. Hope J (ed.). *A treatise on the disease of the heart and great vessels*. 3rd ed. Philadelphia, PA: Lea and Blanchard, 1839, pp. 466–471.
4. Takach TJ, Reul GJ, Duncan JM, et al. Sinus of Valsalva aneurysm or fistula: management and outcome. *Ann Thorac* 1999; 68: 1573–1577.
5. Sawyers JL, Adams JE and Scott HW Jr. Surgical treatment for aneurysms of the aortic sinuses with aorticoatrial fistula. *Surgery* 1957; 41: 26–42.
6. Vural KM, Şener E, Taşdemir O, et al. Approach to sinus of Valsalva aneurysms: a review of 53 cases. *Eur J Cardiothorac Surg* 2001; 20(1): 71–76.
7. Moustafa S, Mookadam F, Cooper L, 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.
8. Sarikaya S, Adademir T, Elibol A, et al. Surgery for ruptured sinus of Valsalva aneurysm: 25-year experience with 55 patients. *Eur J Cardiothorac Surg* 2013; 43(3): 591–596.
9. Bocchino PP, Fasano R, Fortuni F, et al. Transient complete atrioventricular block due to rupture of the right sinus of Valsalva. *Circ Cardiovasc Imaging* 2021; 14(8): e012708.