

# Ruptured Sinus of Valsalva Aneurysm Presenting as Acute Coronary Syndrome with Cardiogenic Shock and aVR ST-Elevation – A Case Report

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## INTRODUCTION

A ruptured sinus of Valsalva aneurysm can present as a clinical emergency and can lead to progressively deteriorating dyspnea. We describe an unusual case of sinus of Valsalva aneurysm (SOVA) presenting with acute chest pain and dyspnea with electrocardiographic ST-segment elevation in the V1 and aVR leads. Diagnostic angiography and cardiac computed tomography angiography showed contrast enhancement from the aorta to the right ventricle and pulmonary artery. The patient was referred to a cardiovascular surgeon for immediate surgical excision and repair. This case highlights the importance of physical examination and echocardiography, especially in the emergency setting, since the disease can manifest in various presentations. To the best of our knowledge, this report may be the first described case to suggest an acute coronary syndrome with aVR ST-elevation occurring from an acute ruptured SOVA.

## CASE

A 53-year-old man with a history of hypertension and chronic renal insufficiency was admitted to the emergency department because of acute onset of chest pain. The symptoms began while he started working at

his office for a few minutes in the morning, and then he developed central chest tightness and shortness of breath while sitting in his chair. At the same time, he also felt weak and diaphoretic. These symptoms did not subside with rest. This sudden onset of substernal, anterior chest discomfort, non-radiating pain persisted until he presented to the emergency department more than 30 minutes later.

On physical examination, he appeared pale-faced, and he had a continuous cardiac murmur detected on cardiac auscultation with clear breath sounds. He had tachycardia (heart rate: 136 beats per minute) and profound shock (blood pressure: 80/46 mmHg) without a significant blood pressure gradient (< 10 mmHg) in his four limbs or engorged jugular vein. His initial 12-lead electrocardiogram revealed > 1.5 mm ST-segment elevation in the V1 and aVR leads with ST depression in leads I, II, and V4 to V6. The ST-segment elevation in the aVR was more prominent than in V1 (Figure 1A). An admission troponin I level of 0.11 ng/ml was reported.

Given the patient's typical electrocardiographic findings of left main coronary artery occlusion, dual antiplatelet therapy with aspirin and ticagrelor were administered. The patient was referred for emergent coronary angiography, which illustrated insignificant coronary artery disease. Left ventriculography with contrast was performed via a 6F pigtail catheter inserted into ascending aorta. Surprisingly, the contrast test revealed a right ventricular image; therefore, we removed the pigtail catheter and performed a manual contrast injection, and the image showed contrast enhancement from the ascending aorta into the right ventricle and pulmonary artery. Thus, we considered a shunt connection from the aorta to the right ventricle (Figure 1B).

Transthoracic echocardiography detected a continuous shunt from the aorta to the right ventricle,

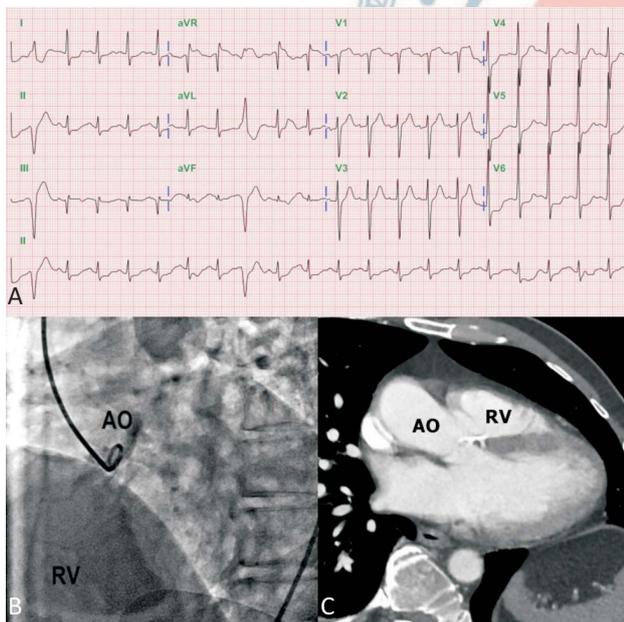
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normal left ventricular systolic function without regional wall motion abnormality or chamber dilatation. These findings suggest a ruptured SOVA on right coronary sinus (Figure 2A, 2B). The patient's previous echocardiographic images were reviewed. They showed moderate aortic regurgitation and an unruptured SOVA on right coronary sinus for at least 5 years (Figure 2C, 2D). Aortic regurgitation had disappeared in the present echocardiography study.

The patient's chest discomfort deteriorated, and profound shock persisted. Cardiac computed tomography angiography (CCTA) confirmed the diagnosis of a ruptured SOVA with a shunt from the aorta to the right ventricle (Figure 1C). The patient underwent emergent excision of the right coronary sinus aneurysm, patch repair, and pericardial effusion drainage. He was discharged without symptoms 7 days later. Follow-up echocardiography after 10 months demonstrated a normal sinus of Valsalva without residual pericardial effusion. He was hemodynamically stable and without chest discomfort 12 months after surgery (Supplementary Figure 1).

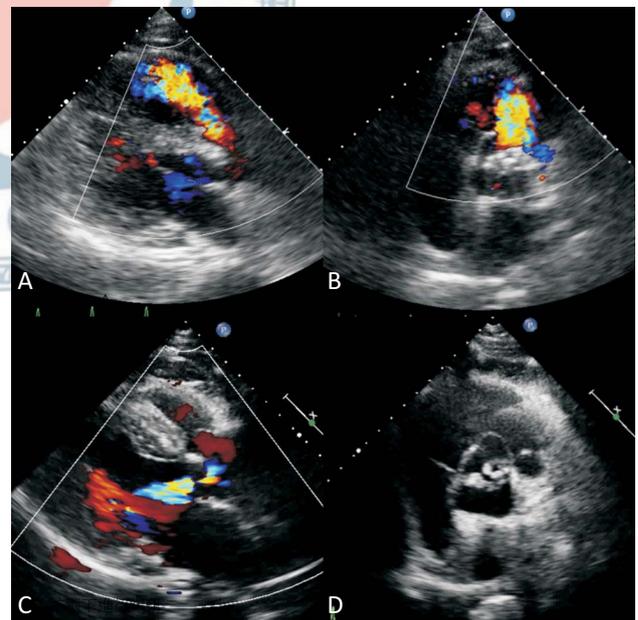


**Figure 1.** (A) Initial 12-lead electrocardiography revealed ST-segment elevation in the V1 and aVR leads with V4 to V6, Lead I and II ST depressions. Note the ST-segment elevation in the aVR lead is more prominent than in the V1 lead. (B) Contrast enhancement from ascending aorta to right ventricle and pulmonary artery, suggestive of a shunt connection from the aorta to the right ventricle. (C) Cardiac computed tomography angiography (CCTA) confirmed the ruptured sinus of Valsalva aneurysm with a shunt from the aorta to the right ventricle.

## DISCUSSION

Ruptured SOVAs can present as a clinical emergency because of formation of aortic-cardiac shunting, primarily toward the right atrium and right ventricle, which can rapidly affect the hemodynamic status. SOVA is rarely considered in the differential diagnosis of acute chest pain with ST-segment elevation on ECG and has been reported in few literature in conjunction with acute ST-elevation myocardial infarction.<sup>1</sup>

Our patient was treated for hypertension and chronic aortic regurgitation according to his previous echocardiography in 2013. The SOVA in our patient was not recognized on his first echocardiogram, and his symptoms stayed stationary until acute chest discomfort developed during this emergency episode. Acute chest pain with lead aVR ST-segment elevation and ST-segment depression in several other leads may predict acute left main coronary artery obstruction.<sup>2,3</sup> The guideline suggests that early initiation of aspirin and P2Y12 inhibitor loading may help to achieve early efficacy. But in cases of ST-segment elevation and myocardial infarction, the diagnosis is not clear yet, delaying P2Y12 inhibitor loading until other etiologies are excluded (e.g.,



**Figure 2.** (A, B) Transthoracic echocardiography detected a continuous shunt connection from the aorta to the right ventricle. No significant aortic regurgitation. (C, D) Previous transthoracic echocardiography detected significant aortic regurgitation and unruptured sinus of Valsalva aneurysm.

aortic dissection) should be considered.<sup>4</sup> Our treatment strategy for the patient was to send him immediately for emergent coronary angiography, which met the percutaneous coronary intervention-mediated reperfusion (wire crossing) time of less than 90 minutes.<sup>5</sup> However, asymmetric radial pulsations (blood pressure) can present in aortic dissection but not in ruptured SOVA, and this phenomenon emphasizes the importance of early differential diagnosis. Early diagnosis and recognition that a patient with possible predictors and risk factors such as previous knowledge of an echocardiogram with an unruptured SOVA, poorly controlled blood pressure and the presence of a new cardiac murmur are critical for early diagnosis. Echocardiography, electrocardiographically gated CCTA, and cardiac magnetic resonance (CMR) imaging can provide an accurate diagnosis. Because the patient presented with persistent, profound cardiogenic shock, we chose CCTA rather than CMR imaging for efficiency and to rush the patient to surgery as soon as possible, even though transesophageal echocardiography is another good method to detect SOVA and CMR imaging can provide excellent anatomic and functional information.<sup>6</sup>

With our patient, after dual antiplatelet therapy with aspirin 300 mg and ticagrelor 180 mg, the cardiac surgeon confronted numerous difficulties with hemostasis during the operation. Fortunately, the patient was discharged smoothly with no residual symptoms.

Reviewing the entire clinical course and management of the patient showed that there are many points to be learned. Firstly, lack of experience with echocardiography to recognize the presence of SOVA on the initial echocardiogram and the patient's lack of awareness of the danger of aneurysm rupture is extremely dangerous. Unruptured SOVA is usually asymptomatic or presents with symptoms of dyspnea, easy fatigability, palpitations, or chest pain.<sup>7</sup> Asymptomatic SOVA is often detected serendipitously by routine 2-dimensional echocardiography. Aortic regurgitation is one of the most common cardiac anomalies that occur in conjunction with SOVA, like in this case. Second, a detailed, symptom-oriented and focused physical examination in the emergency department could reveal a significant continuous murmur in the left lower sternal border that differed from aortic insufficiency.<sup>8</sup> However, both the careful auscultation and bedside echocardiography were

both ignored in order to shorten the door to wire crossing time. When patients presented with an acute myocardial infarction with shock and cardiac murmur, mechanical complication is important differential diagnosis. Eventually a continuous murmur firstly recorded and described in the cardiac care unit in the left lower sternal border, after coronary angiography. The lead aVR is to obtain specific information from the right side of the heart, such as the right ventricle outflow tract and the basal part of the interventricular septum. Rupture of the SOVA could cause an acute interruption of coronary artery blood flow that contributing to the electrocardiographic changes. After the aneurysm has ruptured, the flow into the right ventricle (RV) would be continuous during both systole and diastole according to continuous pressure gradient. The acute left-to-right shunting may cause coronary flow supply-demand mismatch and more severely, cardiogenic shock and then deteriorated coronary insufficiency, contributing to aVR ST elevation and diffuse ST depressions. The mechanism include increased myocardial oxygen consumption because of overactivated sympathetic tone and acute RV injury resulting from acute left-to-right shunting that may induce RV transmural ischemia. Those leads being remote from the RV like lead I and V4 to V6, will record ST depressions.<sup>9</sup> Additionally, all acute cardiovascular conditions should be assessed with echocardiography for differentiation between acute myocardial infarction, aortic dissection, acute pulmonary embolism, and cardiac tamponade.<sup>10</sup> However uncommon, SOVA rupture with shunting is also detected easily. Unnecessary medical prescription and intervention could be avoided by the careful use of these non-invasive examinations.

To the best of our knowledge, this report may be the first described case to suggest an acute aVR ST-elevation myocardial infarction occurring from an acute ruptured SOVA. Early diagnosis by accurate imaging and emergent surgical repair can provide good immediate and 1-year outcomes.

## LEARNING POINTS

- A ruptured SOVA can present as a clinical emergency and can lead to progressively deteriorating dyspnea and acute coronary syndrome. Continuous heart mur-

mur with acute shock hints acute ruptured SOVA.

- Lead aVR ST elevation reflects reciprocal change from ischemia of the left ventricle and ischemia from the RV outflow tract or the right basal part of the interventricular septum region, possible caused not only by left main coronary artery stenosis and severe triple vessel obstructions but also any RV injury.
- All acute cardiovascular conditions should be assessed with echocardiography. However uncommon, SOVA rupture with shunting is also detected easily.
- Early diagnosis by accurate imaging and emergent surgical repair can provide good immediate and 1-year outcomes for ruptured SOVA.

#### ACKNOWLEDGMENT

None.

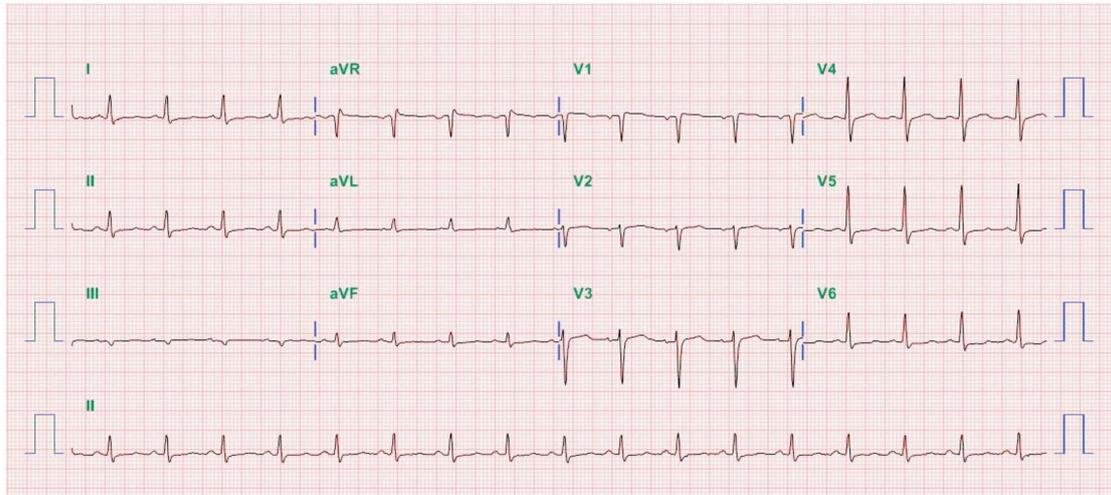
#### CONFLICT OF INTEREST

All the authors declare no conflict of interest.

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SUPPLEMENT



**Supplementary Figure 1.** The post-operation electrocardiography shows those initial ST elevation at lead aVR and other ST depressions all normalized.

