

From clues to clarity: multimodality imaging uncovers ruptured sinus of Valsalva aneurysm masquerading as tricuspid valve endocarditis

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Abstract

Ruptured sinus of Valsalva aneurysm (RSOVA) is a rare but life-threatening condition that may be misdiagnosed due to nonspecific symptoms. We report the case of a 24-year-old postpartum female who presented with acute right heart failure and was initially misdiagnosed with tricuspid valve infective endocarditis. Detailed evaluation revealed RSOVA originating from the right coronary sinus with left-to-right shunting into the right ventricle. This case highlights the importance of clinical suspicion and multimodal imaging for accurate diagnosis in postpartum females presenting with dyspnea, chest pain, and a continuous murmur. Early diagnosis and timely intervention are crucial to improving outcomes.

Key words: postpartum cardiovascular complications, multimodality imaging, right heart failure.

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Introduction

The sinus of Valsalva is an anatomical dilation of the ascending aorta located just above the aortic valve. There are three sinuses: the right coronary sinus, the left coronary sinus, and the non-coronary sinus, each located above the corresponding cusps of the aortic valve. A sinus of Valsalva aneurysm (SOVA) is a rare congenital or acquired anomaly characterized by abnormal dilation of one or more of these sinuses. Rupture of this aneurysm can occur spontaneously or as a result of infection, trauma, atherosclerosis, or, rarely, during pregnancy. Most ruptures occur between the second and fourth decades of life, with the most common site being the right coronary sinus aneurysm rupturing into the right ventricle (RV) [1]. Here, we present the case of a 24-year-old postpartum female initially misdiagnosed with tricuspid valve infective endocarditis, later confirmed to have ruptured SOVA (RSOVA).

Case Report

The approval to write this case report was previously obtained from the Institutional Ethics Committee of SMS Medical College, Jaipur, Rajasthan, India, vide no. 966/MC/EC/2024 dated 2/11/2024. The informed consent was signed by the patient on 06/11/2024.

A 24-year-old postpartum female with a history of normal vaginal delivery 3 weeks back was referred from a peripheral hospital to our center with a diagnosis of tricuspid valve infec-

tive endocarditis with acute right heart failure. She received treatment with intravenous antibiotics and was referred as her symptoms of dyspnea worsened. A detailed history revealed acute onset of dyspnea from the day after delivery, which progressively worsened and was associated with chest pain. There was no history of fever during or prior to the illness. Physical examination demonstrated a raised jugular venous pulse, pedal edema, ascites, bilateral chest crepitations, hepatomegaly, and a harsh, continuous murmur was heard in the aortic region. The patient was afebrile, with a blood pressure of 130/88 mmHg. An electrocardiogram showed normal sinus rhythm, with sinus tachycardia. A chest X-ray revealed mild cardiomegaly and bilateral pulmonary congestion. Laboratory reports showed no significant abnormality with normal leukocyte counts and negative C-reactive protein and serum procalcitonin levels.

Given the clinical findings, we strongly suspected RSOVA. An initial transthoracic echocardiography (TTE) was inconclusive, revealing a mobile mass of size 25×19 mm likely attached to the tricuspid valve leaflets, with moderate tricuspid regurgitation, mild eccentric aortic regurgitation (AR), borderline enlargement of the right atrium (RA) and RV, and mild pericardial effusion. Incidentally, a continuous wave (CW) Doppler done across the tricuspid valve showed a continuous waveform, suggesting shunting of blood through an RSOVA (Figure 1, Video 1). A transesophageal echocardiography (TEE) was done, which demonstrated an RSOVA with a left-to-right shunt and no involvement of the tricuspid valve. The neck of the aneurysm, however, could not be clearly visualized (Figure 2). Hence, to



confirm the diagnosis, a computed tomography (CT) angiography of the aorta was performed, which demonstrated an RSOVA originating from the right coronary sinus, with a neck measuring 9.2 mm and communicating with the RV outflow tract (Figure 3). Furthermore, to completely exclude infective endocarditis, blood cultures and a rapid BioFire FilmArray assay were done, both of which returned negative.

To manage her acute right heart failure, she was initially treated medically. This included fluid restriction, intravenous furosemide 20 mg twice daily, and oral spironolactone 25 mg once daily. For heart rate control, oral ivabradine 5 mg twice

daily was started initially, which was later switched to oral metoprolol succinate 25 mg once daily. A percutaneous device closure of the RSOVA was not considered due to the large size of the aneurysmal sac, which may limit the stability of the device. She was referred to the cardio-thoracic surgery unit, and she underwent open surgical repair after 3 days. The aneurysmal sac was excised, and the rupture of right coronary sinus was repaired using a pericardial patch. The surgery was uneventful, and she remained stable during the postoperative period. A post-operative TTE revealed no residual defect or shunt and she was discharged in a stable condition.

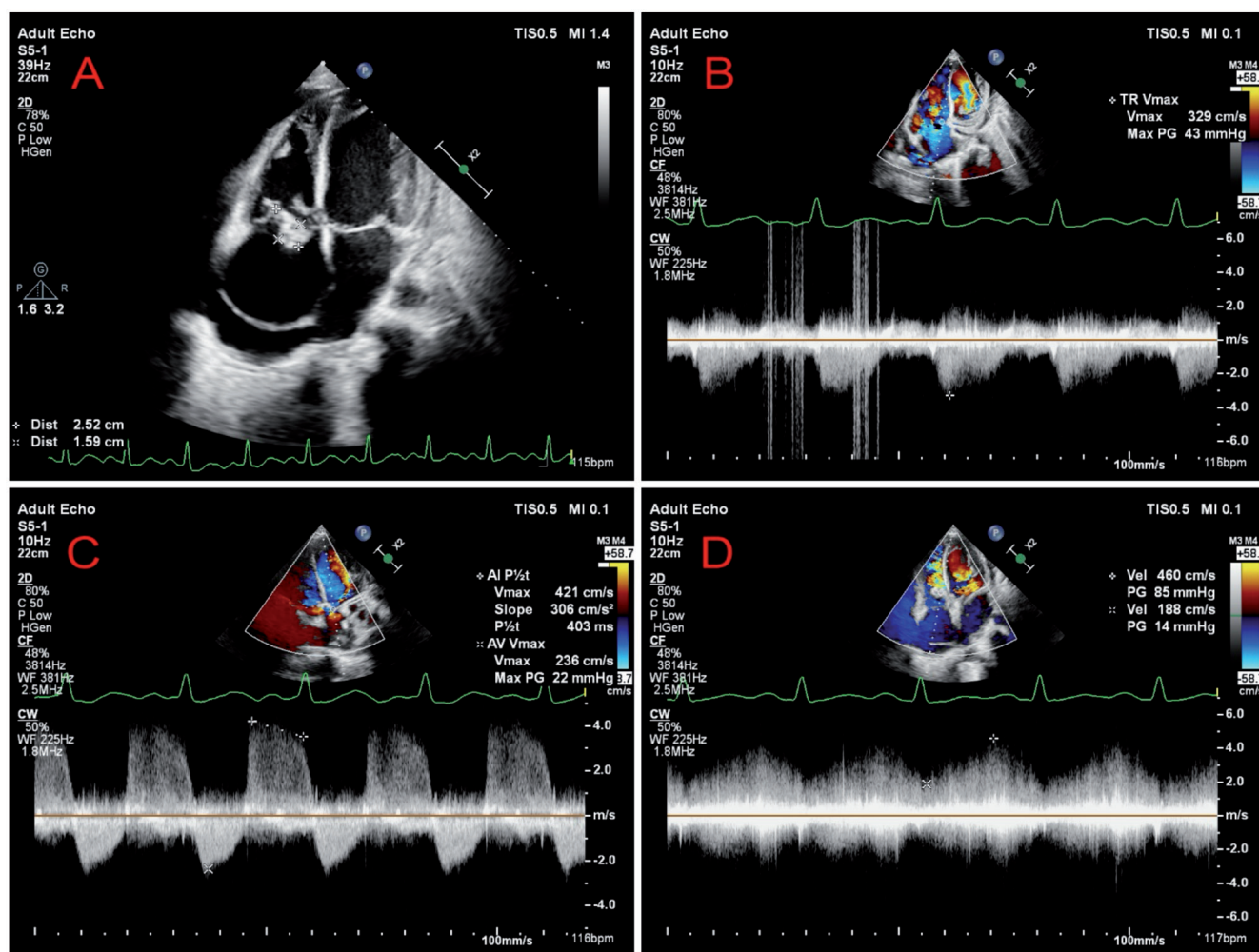


Figure 1. Transthoracic echocardiography images. A) Apical four chamber (A4C) view demonstrating mass of size 25×16 mm around tricuspid valve with borderline enlargement of right atrium and right ventricle, and mild pericardial effusion; B) continuous wave (CW) doppler across tricuspid valve demonstrating moderate tricuspid regurgitation with maximum pressure gradient of 43 mmHg; C) CW doppler across aortic valve demonstrating mild eccentric aortic regurgitation with pressure half time (PHT) of 403 ms; D) CW doppler across tricuspid valve demonstrating continuous flow with a maximum and minimum gradient of 85 mmHg and 14 mmHg respectively.

Discussion

RSOVA present with history of sudden onset chest pain and progressively worsening shortness of breath. Clinical signs typically indicate acute decompensated right-sided heart failure, often accompanied by a continuous murmur caused by an acutely developed left-to-right shunt. Other cardiovascular complications associated with RSOVA include infective endocarditis, AR, ventricular arrhythmias, cardiac tamponade, and sudden cardiac death [1].

There is limited data reporting RSOVA during pregnancy. Most reports have focused on antepartum patients, and the presence of RSOVA in the postpartum period remains anecdotal [2]. It is likely that hyperdynamic circulation during pregnancy may increase wall shear stress, leading to the rupture of an existing SOVA. Theoretically, in the immediate postpartum period, uterine contraction increases inferior vena cava drainage and preload to the heart, leading to increased forward flow and a higher risk of rupture [3].

The diagnosis of RSOVA is primarily reliant on imaging modalities, which include echocardiography, CT angiography, and multiplanar cardiac magnetic resonance imaging (MRI). TTE is the pre-

ferred initial screening modality due to its high sensitivity, ease of access, and simplicity of the procedure. Existing data reports a sensitivity of up to 90% in detecting RSOVA. The main limitation is its ability to detect the site of rupture and delineate the neck of the aneurysm [4]. In certain cases, as in ours, doppler assessment provides the first clue to RSOVA by demonstrating continuous flow across the site of rupture. Therefore, TEE, or alternatively 3D-TEE, may be used in such cases, as it provides a better acoustic window, resulting in higher resolution and improved characterization of RSOVA. Additionally, 3D TEE may provide real-time imaging, which can be used to guide the placement of the closure device, ensuring accurate positioning and minimizing complications [5].

Contrast-enhanced ECG-gated multidetector cardiac CT angiography is a noninvasive modality well-suited for the evaluation of RSOVA, cardiac chambers, and their relationships with the coronary arteries. It offers high spatial and temporal resolution, thus facilitating localization of the RSOVA neck, with contrast extending into the adjacent cardiac chambers. However, in cases with large neck RSOVA, a high-volume left-to-right shunt can lead to pan-cardiac enhancement, thus obscuring contrast passage and visibility of annular structures [6]. In such rare instances, cardiac MRI may be indicat-

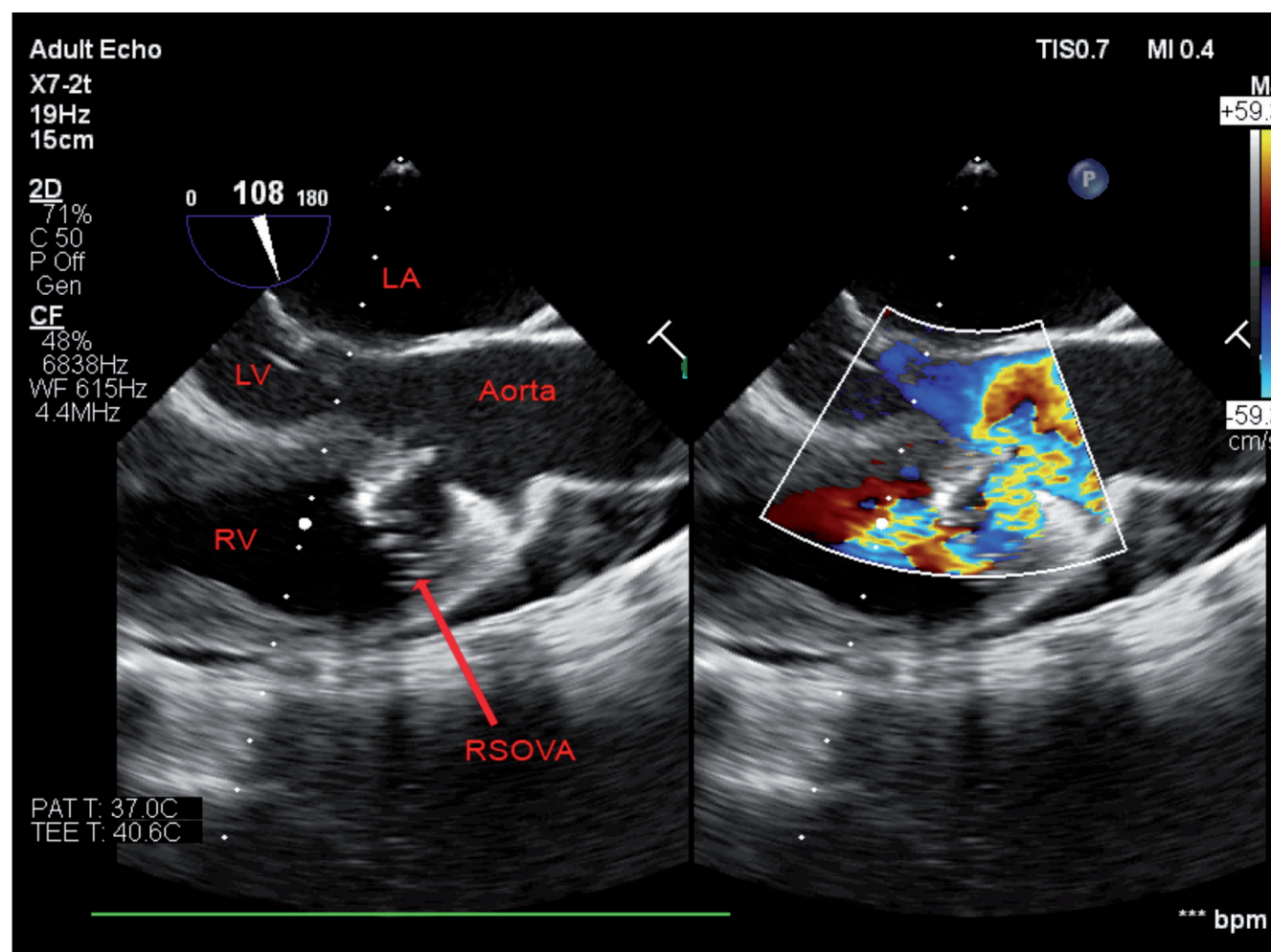


Figure 2. Mid-esophageal long axis view with color doppler demonstrating communication between the aorta and right ventricle with significant left to right shunting of blood. LA, left atrium; LV, left ventricle; RV, right ventricle; RSOVA, ruptured sinus of Valsalva aneurysm.

ed for adequately assessing the RSOVA origin, course, and the large shunt across it.

RSOVA is often misdiagnosed as a ventricular septal defect, infective endocarditis, or tricuspid valve disease. The misdiagnosis typically occurs because the rupture site, the shunt, or the aneurysm may not be clearly identified during the initial echocardiographic evaluation [7]. Therefore, a strong clinical

suspicion, based on the patient's history of acute-onset dyspnea, chest pain, and the presence of a continuous murmur, as in our case, is essential for accurate diagnosis. The average life expectancy of an unrepaired RSOVA is 1 year, highlighting the need for early diagnosis and definitive management, either through surgical open repair or transcatheter device closure [1,6,7].

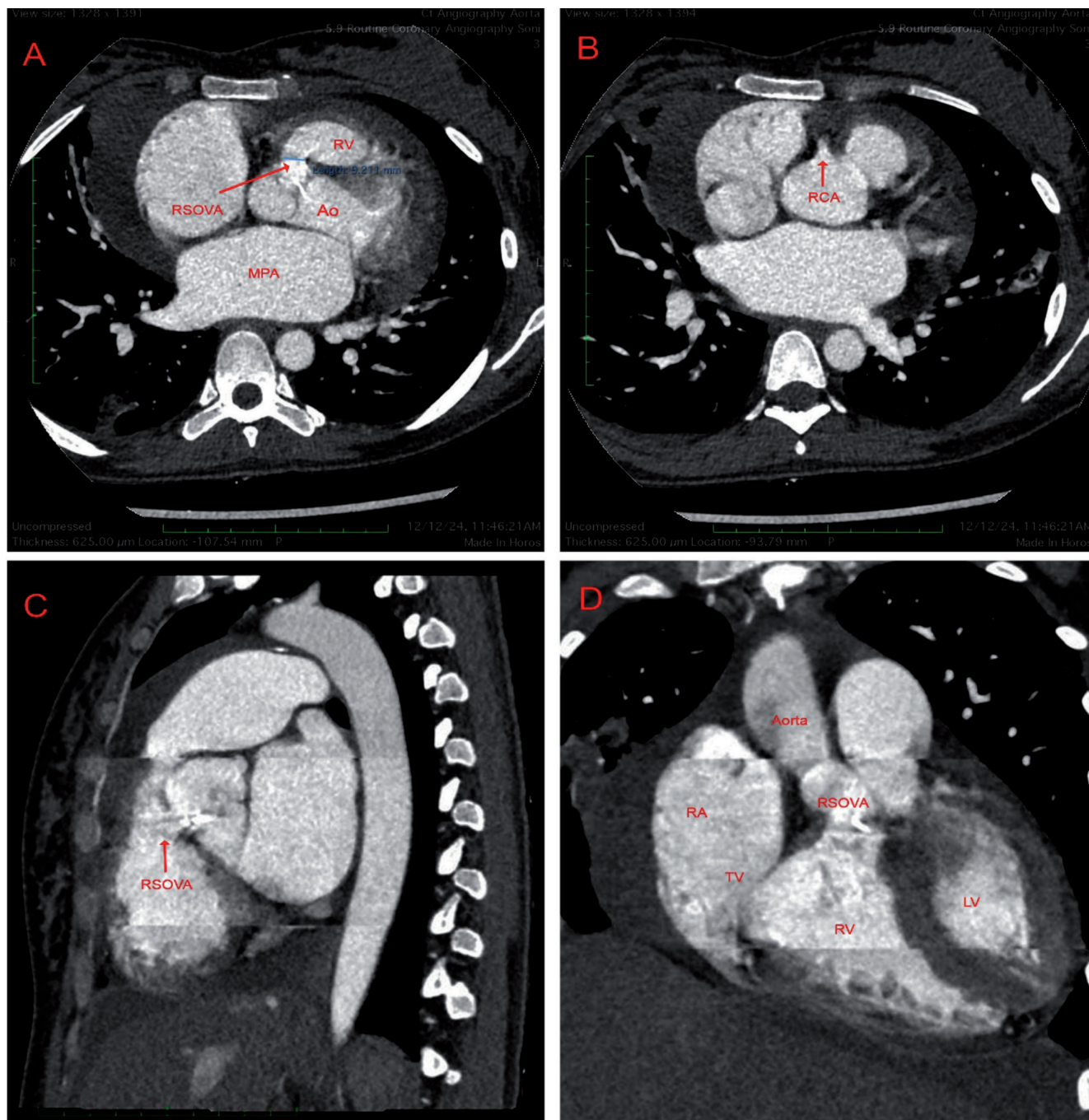


Figure 3. Computed tomography angiogram images. A) Axial section showing ruptured sinus of Valsalva aneurysm (RSOVA), measuring 9.2 mm arising from right coronary cusp communicating with the right ventricle (RV); B) main pulmonary artery (MPA) is mildly dilated; C) axial section showing origin of right coronary artery (RCA), confirming the rupture in right coronary cusp; D) sagittal and coronal sections delineating the site of rupture. Mild pericardial effusion was also noted. RA, right atrium; LV, left ventricle; Ao, aorta; TV, tricuspid valve.

Conclusions

RSOVA is a rare but serious condition that may be missed or misdiagnosed during routine imaging and clinical examination. This case highlights the importance of maintaining a high level of clinical suspicion in patients, including postpartum females presenting with sudden-onset dyspnea, chest pain, and a continuous murmur. Early diagnosis, facilitated by appropriate imaging modalities such as echocardiography, CT angiography, and, in some cases, MRI, is crucial for accurate identification, prompt intervention, and improving patient outcomes.

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Online supplementary material:

Video 1. TTE cine loop of A4C and parasternal short axis view (PSAX). In A4c view, a mobile mass around tricuspid valve is seen with borderline enlargement of RA and RV, and mild pericardial effusion. In PSAX view the right coronary cusp is obscured.

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