Fatal Sinus of Valsalva Aneurysm and Dissection into the Left Ventricle With Extension to the Interventricular Septum: A Challenging Diagnosis

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INTRODUCTION

Sinus of Valsalva aneurysm (SVA) is a rare condition caused by a weakness of the elastic lamina at the junction of the aortic media and the annulus fibrosus.[1] It can be congenital or acquired secondary to an injury, endocarditis, syphilis, Behçet's disease, Marfan syndrome, and other conditions.[2] There is a high incidence in the Asian population, predominantly in men (4:1).[3]

CASE PRESENTATION

A 42-year-old farmer with no relevant medical history came to the emergency department with dyspnea, chest pain, and altered mental status. His symptoms had limited his daily activities for the past 6 months until they became incapacitating. At admission he had tachycardia (heart rate 116 beats/min), tachypnea (respiratory rate 28 breaths/min), hypotension (blood pressure 70/40 mm Hg), and desaturation (oxygen saturation 87%), as well as marked pallor, capillary filling > 2 sec, distal cyanosis, and generalized coldness. A grade III/VI diastolic murmur on the right parasternal line and a Glasgow Coma Scale score of 7 were observed.

Electrocardiography showed sinus rhythm and a left bundle branch block (Figure 1A). Troponin I level was within the normal range, excluding a myocardial infarction. Chest radiography showed grade III cardiomegaly and a right basal pleural effusion (Figure 1B). Transthoracic echocardiography revealed left ventricular global hypokinesis and dilation of the four chambers (Figure 2) with left ventricular end-diastolic volume of 105 mL/m^2 and end-systolic volume of 61 mL/m^2. There was systolic dysfunction, with a left ventricular ejection fraction of 42% determined using the modified Simpson method and diastolic dysfunction with a restrictive pattern (E/A ratio 1.8 and deceleration time 100 msec; Figure 3). There was also moderate mitral regurgitation (vena contracta 0.53 cm) and severe tricuspid regurgitation (vena contracta 0.71 cm), and elevated systolic pulmonary arterial pressure (75 mm Hg; Figure 4). A dissection of the right coronary sinus of Valsalva into the left ventricle (Figure 5, Videos 1-3) and extending into the left outflow tract and the interventricular septum with severe aortic regurgitation (pressure half-time 170 msec; reverse holodiastolic flow pattern in the abdominal aorta) was also found (Figure 6). No ventricular septal defect (VSD) was detected.

The patient had prerenal acute kidney injury secondary to cardiogenic shock (creatinine level 3.44 mg/dL). Management for cardiogenic shock was established with intravascular volume and vasopressors with a poor outcome, so immediate surgical management was planned. Unfortunately, the patient died before surgical closure.

VIDEO HIGHLIGHTS

Video 1: Parasternal long- and short-axis views. **(A)** Dissection of the right coronary sinus with extension to the perimembranous portion of the interventricular septum, with dilation of the left cavities and mild pericardial effusion. **(B)** Short-axis view at the level of the mitral valve with dissection of the interventricular septum.

Video 2: Three-dimensional short-axis view. Short-axis view at the level of the mitral valve with dissection of the interventricular septum.

Video 3: Two-dimensional and color flow Doppler long-axis view. The right coronary sinus ruptured into an echolucent cavity in the perimembranous portion of the interventricular septum, draining into the left ventricle.

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DISCUSSION

According to the echocardiographic findings, characterized by dilatation of the cardiac cavities, systolic dysfunction with reduced left

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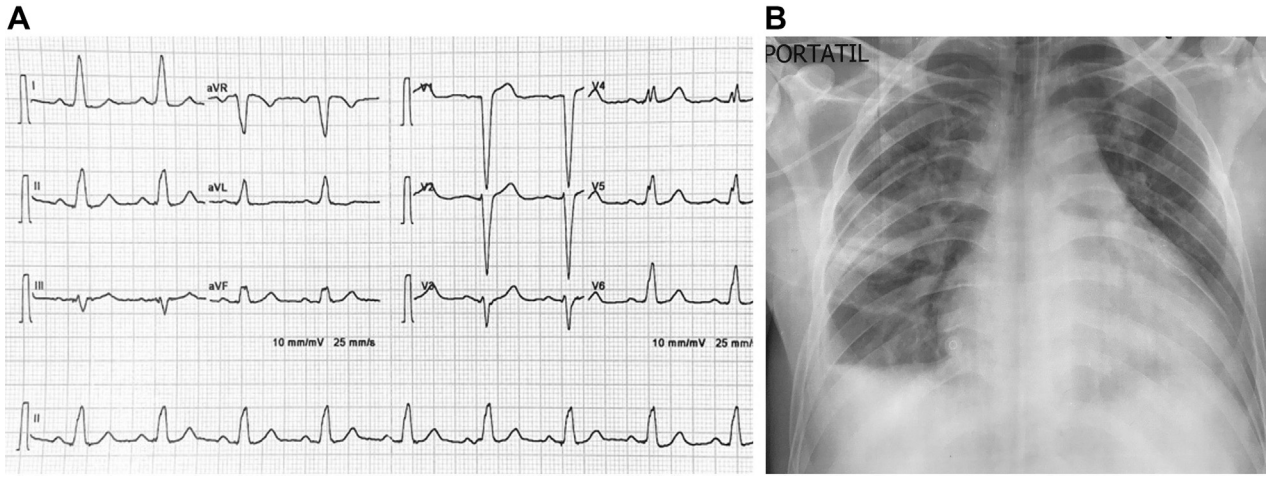

Figure 1 Initial evaluation. A 42-year-old man with progressive dyspnea and exertional chest pain for 6 months. **(A)** Twelve-lead electrocardiography with evidence of a left bundle branch block. **(B)** Posteroanterior chest radiography that shows grade III cardiomegaly (with a cardiothoracic index of 0.62), right basal pleural effusion on the right hemithorax, and indirect signs of pulmonary capillary venous hypertension.

ventricular ejection fraction, and diastolic dysfunction with a restrictive pattern, we can establish that this patient had long-term aortic regurgitation related to severe distortion of the aortic valve annulus secondary to SVA. We believe that the patient's initial symptoms were secondary to hemodynamic decompensation due to the severity of the untreated aortic regurgitation and that the dissection of the aneurysm with rupture toward the left ventricle was what caused the final clinical deterioration with the development of cardiogenic shock that produced a fatal outcome. This result would probably have been different if the patient had sought care while still asymptomatic, but he arrived too late for a successful intervention.

SVA is thought to result from the absence of normal elastic and muscular tissue, which creates thinning of the aortic sinus walls.

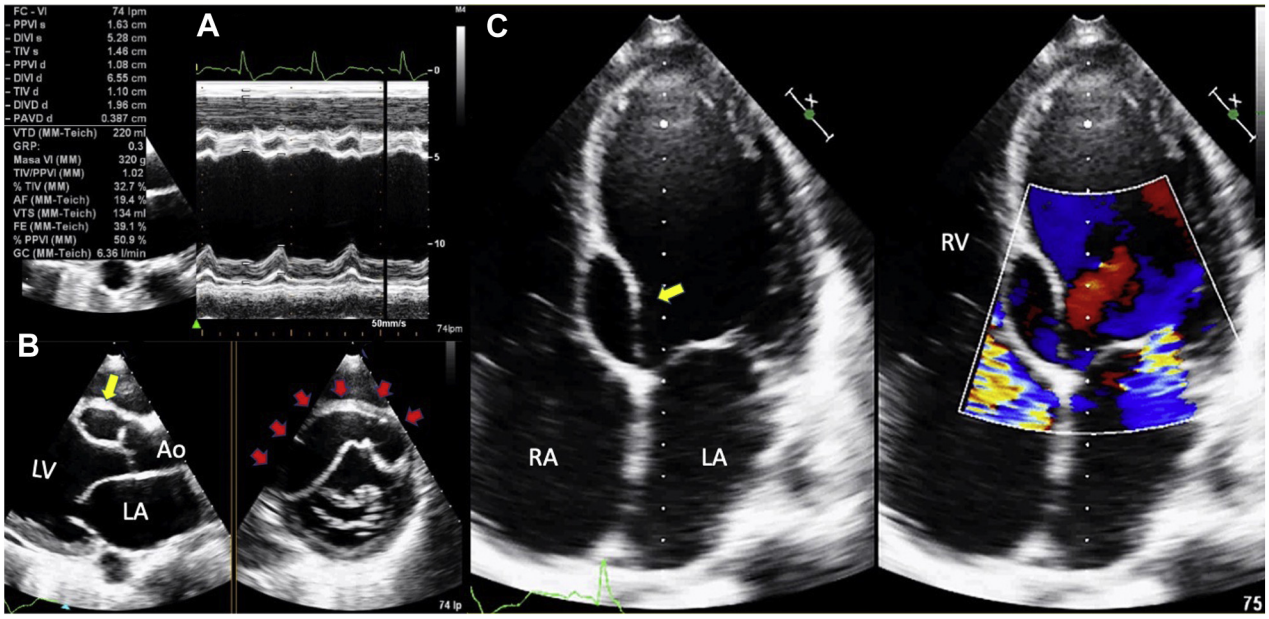

Figure 2 M-mode, two-dimensional, and color flow transthoracic echocardiography. **(A)** M-mode image showing dilation of the left ventricle (LV) and eccentric hypertrophy, with a diastolic diameter of 65 mm, systolic diameter of 52.8 mm, relative wall thickness of 0.3, and left ventricular mass index of 200 g/m^2. These parameters suggest significant dilation of the LV. **(B)** Long-axis parasternal view and short-axis parasternal view at the mitral valve level revealing a dissection *(yellow arrow)* of the right coronary sinus of Valsalva that compromised two thirds of the interventricular septum *(red arrows)*. **(C)** Four-chamber views (two-dimensional and color flow) with dilation of all chambers and generalized hypokinesis of the LV with dissection of the perimembranous portion of the interventricular septum *(yellow arrow)*, moderate mitral regurgitation, and severe tricuspid regurgitation. *Ao*, Aorta; *LA*, left atrium; *RA*, right atrium; *RV*, right ventricle.

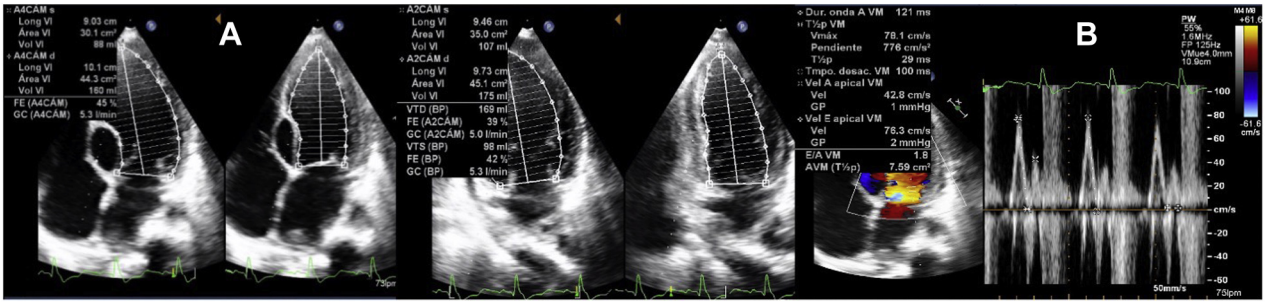

Figure 3 Left ventricular systolic and diastolic function by echocardiography. **(A)** Left ventricular ejection fraction in four- and two-chamber views (calculated using the modified Simpson method) of 42%. **(B)** Four-chamber view with pulsed Doppler of the mitral valve with an E/A ratio of 1.8 and deceleration time of 100 msec, suggesting a restrictive pattern. These studies indicated that the patient had left ventricular systolic and diastolic dysfunction.

SVAs constitute 0.1% to 3.5% of all congenital heart defects.[3] The right coronary sinus is involved in 67% to 85% of reported cases; other locations include 42% in the noncoronary sinus and only 10% in the left coronary sinus.[4] Most aneurysms rupture into the right chambers and rarely into the left chambers and pulmonary arteries.

SVAs are frequently associated with other congenital anomalies, most commonly VSD (30%-60%), bicuspid aortic valve (15%-20%), and aortic regurgitation (44%-50%).[5] In the presence of a coexisting VSD, the large ruptured SVA shunt overlaps the VSD flow, which can be difficult to recognize on two-dimensional echocardiography, but three-dimensional echocardiography identifies the two abnormal flows in most cases.[6] In our patient, the presence of associated VSD was ruled out using the three-dimensional color Doppler technique, which allowed us to demonstrate the absence of a left-to-right shunt.

The differential diagnosis of ventricular cystic masses includes infections secondary to *Echinococcus* that cause hydatid cyst or *Taenia solium* that causes cysticercosis, capillary hemangiomas, cystic thrombi, congenital blood cysts, and intracardiac tumors. The clinical context and characteristics of the masses help establish the most accurate diagnosis.[7]

The clinical presentation ranges from asymptomatic to symptoms such as cough, dyspnea, fatigue, and chest pain.[8] We report a rare case in which the aneurysm extended to the ventricular septum and ruptured into the left ventricle, making it unique and interesting.

The patient's medical history yielded no clues that helped determine the etiology of the aneurysm, so we suspected it to be congenital, as that is the most common cause. Rupture can occur as a spontaneous event, the result of trauma or physical exercise, or

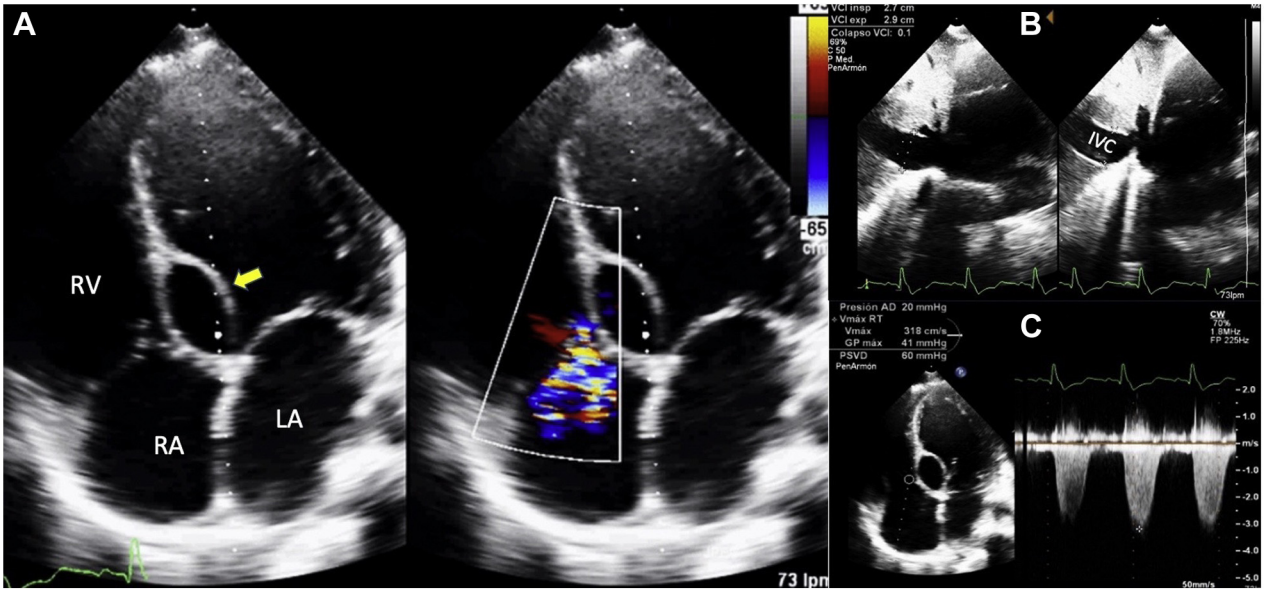

Figure 4 Pulmonary hypertension. **(A)** Two-dimensional and color flow imaging showing severe tricuspid regurgitation with a vena contracta diameter of 7.1 mm. **(B)** Dilation of the inferior vena cava (IVC) in the subcostal view. **(C)** Two-dimensional and continuous-wave Doppler showing peak tricuspid gradient of 60 mm Hg and pulmonary systolic arterial pressure of 75 mm Hg, confirming the echocardiographic diagnosis of pulmonary hypertension. *LA*, Left atrium; *RA*, right atrium; *RV*, right ventricle.

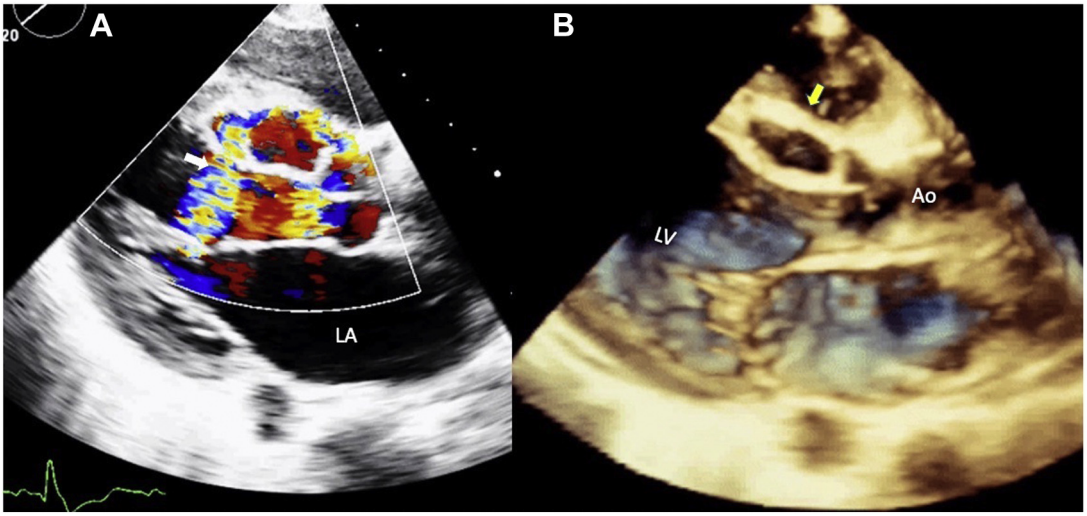

Figure 5 Long-axis view. **(A)** Two-dimensional and color flow Doppler showing an echolucent cavity in the perimembranous portion of the interventricular septum ruptured into the left ventricle (LV; *white arrow*). **(B)** Three-dimensional view with echolucent cavity in the perimembranous portion of the interventricular septum *(yellow arrow)* and enlargement of the left chambers. *Ao*, Aorta; *LA*, left atrium.

infective endocarditis.[9] There were no suggestive data of vegetations in the echocardiographic study, and there was no report of exceptional trauma or unusual strenuous exercise. Thus, the precipitating factor for the rupture remains unclear.

Surgical closure is the definitive treatment for rupture of these aneurysms, with a mortality rate of 3.6%.[1] Transcatheter closure devices have been proposed but are not as effective as surgery. Unfortunately, our patient was unable to undergo surgery because of the instability of his hemodynamic status. On the basis of the present literature review, there are no reports of survivors with cardiogenic shock, with or without surgery. If surgery is performed before hemodynamic decompensation occurs, long-term follow-up is recommended to assess New York Heart Association functional class, with serial echocardiography to evaluate ventricular function and recurrence of SVAs.

CONCLUSION

SVA is an uncommon cause of cardiogenic shock. The clinical features are wide ranging and nonspecific, requiring a high index of suspicion from clinicians. Given the severity of the presentation, once myocardial infarction is excluded, we must consider SVA as an improbable but possible cause for cardiogenic shock, as was the case in our patient.

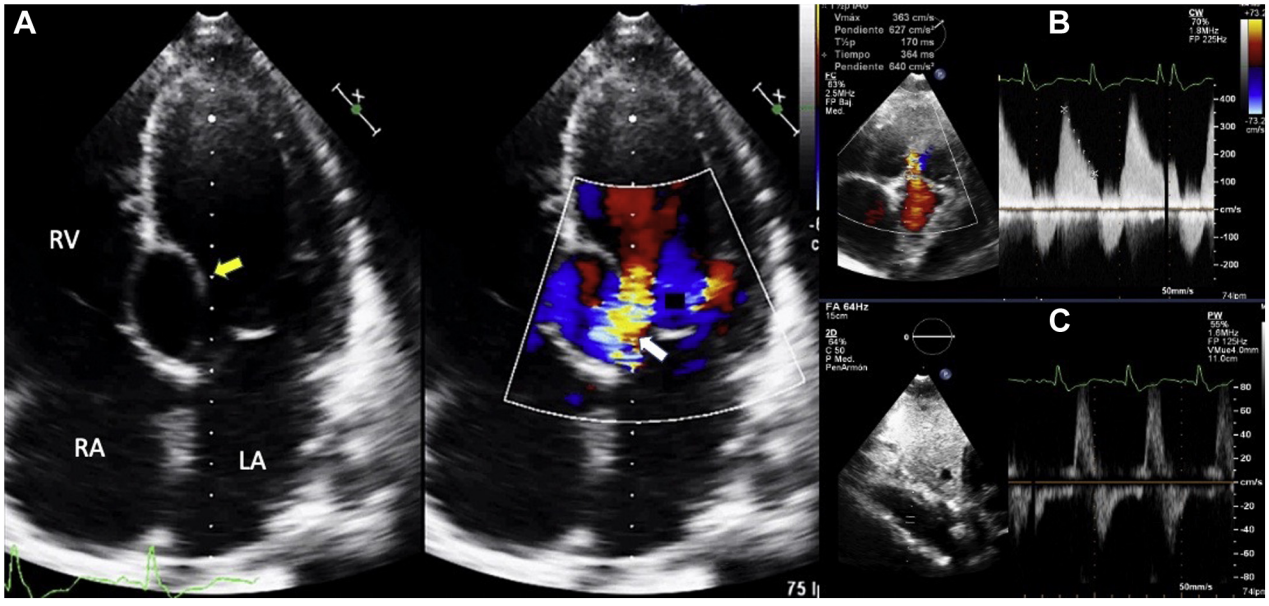

Figure 6 Aortic regurgitation. **(A)** Four-chamber view (two-dimensional and color flow Doppler) showing severe aortic regurgitation *(white arrow)*. **(B)** Pressure half-time 170 msec. **(C)** Subcostal view showing the abdominal aorta with pulsed Doppler showing reverse holodiastolic flow, both **(B, C)** supporting the diagnosis of severe aortic regurgitation. *LA*, Left atrium; *RA*, right atrium; *RV*, right ventricle.

In conclusion, the clinician must have a high index of suspicion, use echocardiography to make a prompt diagnosis, and then offer emergency surgical repair before the anticipated rapid clinical deterioration.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2021.09.011.

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