



Thrombosed Aneurysm of the Left Sinus of Valsalva Presenting as an Intramyocardial Mass

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Aneurysms of the sinuses of Valsalva are uncommon in clinical practice. Most are congenital, but secondary causes are also recognized. Congenital aneurysms of the left sinus of Valsalva are particularly rare. The authors report a fatal case in which a nonruptured aneurysm of the left sinus of Valsalva dissected into the interventricular septum and presented as heart failure. The concurrent presence of dilated cardiomyopathy and the mechanisms that may have led to it are discussed on the basis of the anatomic and histologic features found at autopsy. (*J Am Soc Echocardiogr* 2010;23:1223.e1-1223.e3.)

Keywords: Valsalva sinus aneurysm, Dilated cardiomyopathy, Heart failure

CASE PRESENTATION

A 37-year-old black man, native of Angola, was referred to our hospital for the evaluation and management of an intramyocardial mass and dilated cardiomyopathy. His medical history started 7 years earlier, when he developed progressive exertional dyspnea, orthopnea, and peripheral edema. There was recent worsening of the heart failure symptoms to New York Heart Association class IV, and he had been hospitalized in Angola for the past 3 months before being transferred to our hospital. He was on standard heart failure therapy, including an angiotensin-converting enzyme inhibitor, digoxin, a diuretic, and an aldosterone antagonist.

There was no prior diagnosis of congenital heart disease, chest trauma, or family history of premature cardiovascular death. He denied having had a prolonged febrile illness or invasive vascular procedures in the past, including coronary catheterization.

As part of the initial diagnostic workup, a transthoracic echocardiographic examination was performed (Figure 1), which showed a dilated left ventricle with global hypokinesis and severely impaired systolic function (ejection fraction of 17%). A nonexpandable heterogeneous cystic-like mass with calcified contour was in close proximity to the left sinus of Valsalva and extended through the left ventricular outflow tract to the anterolateral portion of the mitral annulus and into the interventricular septum (Figures 1A-1C, Video 1).

Contrast echocardiographic imaging (SonoVue; Bracco Diagnostics, Milan, Italy) was used to assess the perfusion of the mass. There was clear delimitation of the mass from the cardiac chambers, and there was no significant mass perfusion (Video 2).

Transesophageal echocardiography (Video 3) confirmed that the mass was closely associated with the left sinus of Valsalva. In the interventricular septum, there were small echo-free spaces, but color and pulsed-wave Doppler interrogation revealed no flow between the cystic mass, the aortic root, and the cardiac chambers. There was a central jet of mild aortic regurgitation.

The mass was also evaluated by computed tomography (Figure 2A, Video 4), which revealed a calcified cystic mass with hypodense content in the interventricular septum localized next to the aortic root. There was no density enhancement by intravenous contrast agent.

Coronary angiography was performed to exclude concomitant coronary artery disease. It showed angiographically normal coronaries. Aortography (Figure 2B) showed the passage of contrast from the left sinus of Valsalva into the interventricular septum, below the left coronary artery origin, confirming the diagnosis of aneurysm of the left sinus of Valsalva (ALSV). There was negative contrast inside the aneurysm sac, suggesting the presence of a thrombus inside the aneurysm.

To exclude an infectious etiology for the cardiac mass, ova and parasite testing as well as serologic tests for syphilis, toxoplasmosis, leishmaniasis, brucellosis, hydatid cyst, Q fever, cytomegalovirus, human immunodeficiency virus, and hepatitis viruses were done, and all results were negative. Antinuclear antibodies and an autoimmunity panel were also negative.

During the hospital stay, the heart failure progressively worsened, and the patient died of refractory pump failure in the cardiac critical care unit.

An autopsy study was performed to reach a final diagnosis. The macroscopic pathology examination of the heart showed dilatation of the cardiac chambers and increased heart weight (1020 g). The aortic valve was tricuspid, with thickened cusps but otherwise normal morphology. In the left sinus of Valsalva, there was a round opening measuring 16 mm in diameter with rounded, regular margins, which led to a large intramyocardial cavity (36 × 135 mm) extending to the left ventricular outflow tract, interventricular septum, and mitral annulus. This cavity was almost completely filled with an organized, laminated, and calcified thrombus (Figures 3A and 3B). Microscopic examination showed that it was best described as a pseudoaneurysm, as there was no true aneurysm wall, just fibrotic tissue. Myocardial

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Abbreviation

ALSV = Aneurysm of the left sinus of Valsalva

histology showed myocyte hypertrophy and interstitial fibrosis consistent with unspecified dilated cardiomyopathy. The coronary arteries were normal.

Congenital aneurysm of the sinus of Valsalva is thought to be due to a defect in the aortic media,⁴ which arises from defective development of the distal bulbar septum, the structure that separates the systemic and pulmonary halves of the primitive exit tube of the heart.⁵ The left aortic cusp does not arise from the bulbar septum, thus explaining the rarity of congenital ALSV.

The combination of ALSV with intraluminal thrombus is also extremely rare, although there have been anecdotal reports of patients presenting with embolic events and syncope.^{6,7} In our patient, the rare combination of ALSV, intramyocardial thrombus, and dilated cardiomyopathy made noninvasive diagnosis difficult, emphasizing the importance of a multimodality imaging approach for rare and complex lesions.

DISCUSSION

Aneurysm of the sinus of Valsalva is a rare entity, reported in only 0.09% of 8138 autopsy cases,¹ but it has been shown to carry a poor prognosis.² In our patient, secondary causes were excluded,

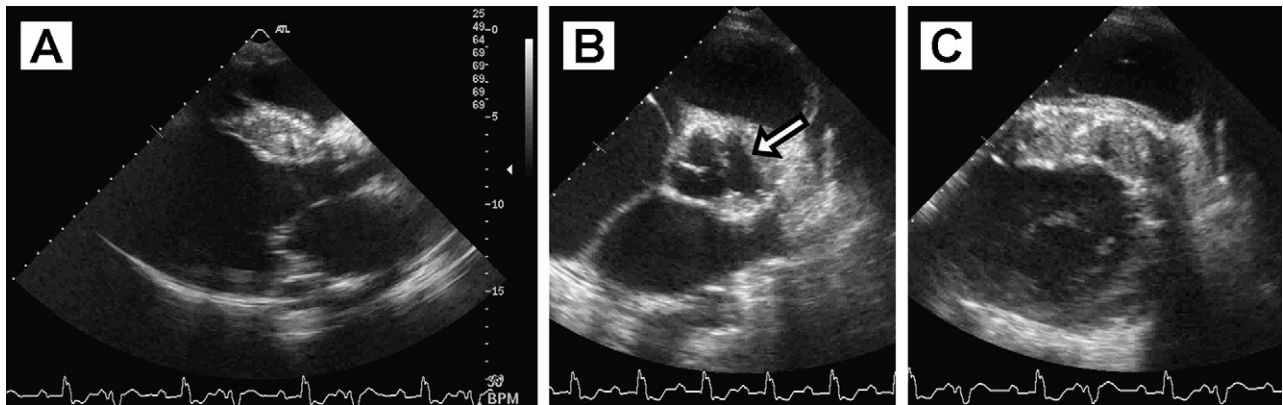


Figure 1 Transthoracic echocardiogram showing a complex cystic mass in the interventricular septum (A). The mass was in close relation with the left sinus of Valsalva (B) and extended to the anterolateral mitral annulus (C).

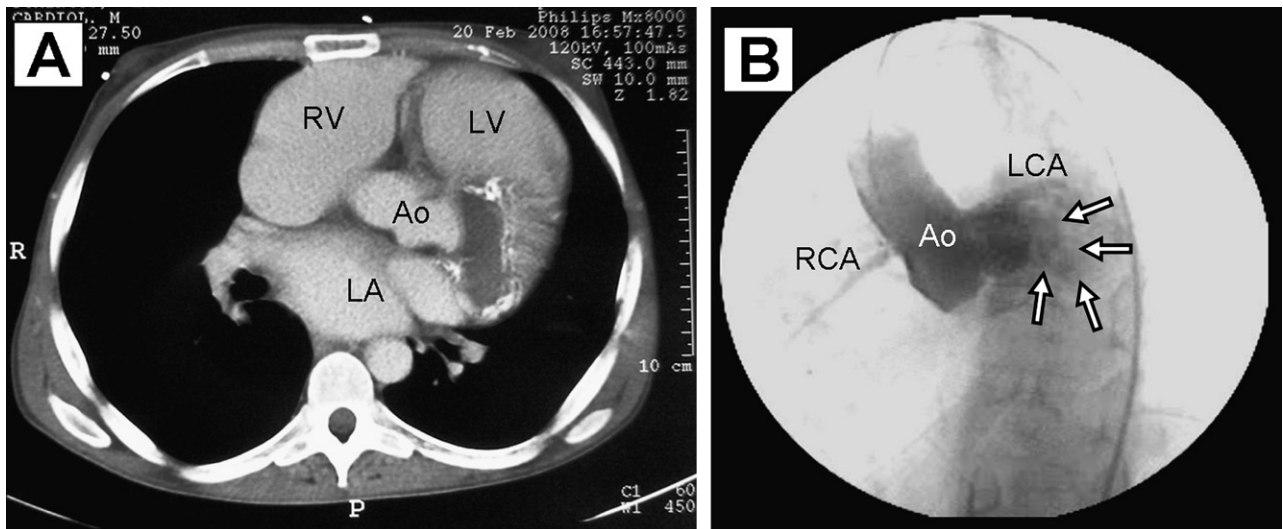


Figure 2 (A) Computed tomography of the heart showing a calcified mass with hypodense content. (B) Aortic root angiogram, showing early phase filling of aorta with faint opacification of the ALSV (arrows). Ao, Aorta; LA, left atrium; LCA, left coronary artery; LV, left ventricle; RCA, right coronary artery; RV, right ventricle.

and the features were consistent with a congenital ALSV. This finding is extremely rare. In a Mayo Clinic report,² there were 4 cases of left origin among 86 Valsalva sinus aneurysms, and in a report from China,³ there was 1 ALSV among 94 ruptured aneurysm cases.

The echocardiographic features we found in this case are similar to those seen after surgical closure of the aneurysm mouth,⁸ which leads to aneurysm thrombosis. The thrombus may have evolved from slow blood velocities inside the aneurysmal sac and progressive clotting, as

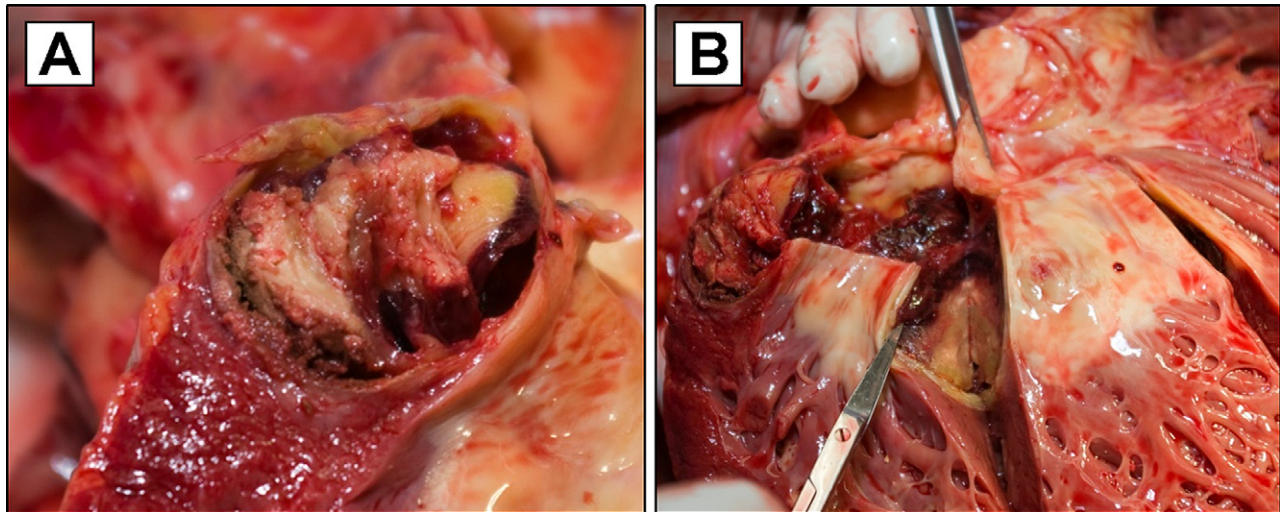


Figure 3 ALSV dissecting into left ventricle outflow tract, interventricular septum, and mitral annulus with transmural involvement.

suggested by the lamellar appearance of the thrombus upon microscopic examination. Low-output dilated cardiomyopathy may have contributed to stasis inside the aneurysmal sac and to thrombus formation.

We report a case of ALSV that presented with left ventricular dilatation and impaired systolic function. Mechanisms implicated as causes of dilated cardiomyopathy in patients with nonruptured aneurysms of the sinus of Valsalva include aortic regurgitation,⁹ coronary artery obstruction,¹⁰ and left ventricular tract obstruction.¹¹ Our patient had mild aortic regurgitation and normal coronary arteries on angiography, and there was no flow obstruction on echocardiography. Although we cannot unequivocally rule out that any of these features might have been present in the past, they are improbable as causes of dilated cardiomyopathy in our patient.

Other common causes of dilated cardiomyopathy have been considered. Atherosclerotic coronary artery disease was excluded by normal results on coronary angiography and the absence of infarcted areas at autopsy. Dallas criteria for acute myocarditis were not met, and a panel of serologic tests for common infectious diseases all had negative results. There was no significant alcohol consumption in the past to justify alcoholic dilated cardiomyopathy.

Insight into explanation for the occurrence of dilated cardiomyopathy in our patient comes from a previous report¹² in which septal destruction was pointed out as a cause for persisting heart failure after surgical repair of aneurysms of the sinuses of Valsalva that dissected into the interventricular septum. Arguing against that are the global myocardial hypokinesis and the diffuse histologic findings. Idiopathic dilated cardiomyopathy remained the final diagnosis because other causes were considered and excluded.

This case report illustrates a rare case of nonruptured ALSV and underlines the importance of multimodality imaging in the diagnosis of complex cardiac masses.

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