Ruptured sinus of Valsalva aneurysm associated with noncompaction of the ventricular myocardium

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Rupture of a sinus of Valsalva aneurysm is a rare, but life-threatening cardiac abnormality that requires surgical correction when diagnosed, and is frequently associated with other congenital defects, particularly with ventricular septal defect, aortic valve regurgitation, and bicuspid aortic valve. We present the case of a 21-year-old man who had a ruptured aneurysm of the noncoronary sinus into the right atrium, a ventricular septal defect, a persistent left superior vena cava and a noncompaction of the ventricular myocardium diagnosed by two-dimensional echocardiography. Surgical repair was carried out and the patient made an uneventful recovery.

KEYWORDS
Ruptured sinus of Valsalva aneurysm; Noncompaction of the ventricular myocardium

Case report
A 20-year-old man, under evaluation by the internal medicine clinic for anemia, was referred to echocardiography because of his continuous murmur. He was well until 1 year earlier when, exertional dyspnea and generalized fatigue developed after a fall from a height. On examination, the patient was thin and appeared pallor. His blood pressure was 110/65 mmHg, and his pulse rate was 84 bpm. A loud, harsh, continuous grade 4/6 murmur was audible near the right sternal border. On transthoracic echocardiography, the left and right ventricular dimensions and functions were normal. A ruptured sinus of Valsalva aneurysm (SVA) into the right atrium with a typical 'windsock' appearance was detected at the parasternal short axis (Figure 1A, arrow) and modified apical four-chamber views (Figure 2B, arrow). Doppler echocardiography showed an abnormal flow consistent with rupture of a noncoronary SVA into the right atrium and detected a 100-mmHg peak gradient between the aorta and the right atrium. The ventricular septal defect (VSD) was also seen at the modified parasternal short axis (Figure 1B, immediately below the aortic valve) and the apical four-chamber views (Figure 2A, arrow). The coronary sinus was dilated (Figures 2B and 3A, black arrow). In order to reveal the cause of coronary sinus dilatation an agitated saline injection was given into the left antecubital vein. The contrast entered first into the coronary sinus and subsequently appeared in the right atrium. The typical prominent trabeculations and intertrabecular recesses, which are pathognomonic for ventricular noncompaction were also seen on echocardiogram (Figure 3A,B). In view of the aforementioned echocardiographic findings, the diagnoses of the ruptured SVA, the VSD, the persistent left superior vena cava and the noncompaction of the ventricular myocardium were made, and the patient was referred to cardiovascular surgery for corrective surgery. The thin-walled aneurysmal portion was resected, and communication between the aorta and the right atrium was closed by a Dacron patch. The VSD was also closed with a patch. The patient made an uneventful recovery and was discharged from the hospital 5 days after surgery.

Discussion
Ruptured SVA is a rare, but life-threatening cardiac abnormality that requires surgical correction when diagnosed, and is frequently associated with other congenital defects, particularly with VSD, aortic valve regurgitation, bicuspid aortic valve and less commonly pulmonary stenosis, coarctation, left superior vena cava and atrial septal defect. The clinical presentations vary from asymptomatic to progressive heart failure following rupture of the aneurysm into an adjacent cardiac chamber depending mainly upon the rapidity of the rupture, the size of the ruptured aneurysm and the chamber into which it ruptures.

Noncompaction of the ventricular myocardium is a rare congenital cardiac disorder characterised by an excessively...
prominent trabeculations in ventricular wall segments and deep intratrabecular recesses.\(^3\) Currently, transthoracic echocardiography is the most beneficial noninvasive diagnostic modality of choice for both ruptured SVA and noncompaction of the ventricular myocardium. The aetiology of SVA is usually a congenital defect in aortic media tissue and abnormal development of bulbous cordis.\(^4\) Noncompaction of the ventricular myocardium is also thought to be caused by arrest of normal embryogenesis of the myocardium.\(^3\) In presented case, their coexistence may be coincidental,
but, it may reflect the common congenital pathway in origin.

References