

Ruptured sinus of Valsalva aneurysm associated with aortic regurgitation and severe myocardial ischemia

Aort yetersizliği ve ciddi miyokart iskemisine yol açan yırtılmış Valsalva sinüsü anevrizması

Abdulkadir Yıldız, M.D., Aytun Çanga, M.D., Nihat Şen, M.D.

Department of Cardiology, Türkiye Yüksek İhtisas Heart-Education and Research Hospital, Ankara

Sinus of Valsalva aneurysm (SVA) is a rare cardiac anomaly either presenting as a congenital heart disease or occurring secondary to cardiac surgical interventions. A 19-year-old male patient presented with chest pain and shortness of breath. On auscultation, a grade 4/6 early diastolic murmur was heard over the left lower sternal border and Erb's area with a thrill. Crepitating rales were heard over bilateral basal lung fields. The electrocardiogram showed right bundle branch block and ST depression. Troponin and CK-MB levels were increased. Shortly after admission, he developed ventricular fibrillation and was defibrillated three times. After restoration of hemodynamic stabilization, transthoracic echocardiography was performed, which showed grade 4 aortic regurgitation, patent foramen ovale, and an aneurysm of the sinus of Valsalva arising from the right coronary sinus, with rupture into the right ventricle. The patient underwent surgery under cardiopulmonary bypass, for repair of the ruptured SVA and patent foramen ovale and aortic valve replacement. He was discharged on the fifth postoperative day following an uneventful operation and postoperative course.

Key words: Aortic aneurysm/complications; aortic rupture/surgery; aortic valve insufficiency sinus of Valsalva/pathology.

Sinus of Valsalva aneurysm (SVA) is a rare cardiac anomaly accounting for 1.2% of all congenital heart diseases and occurring in 0.2% to 1.5% of all cardiac surgical interventions.^[1-3] It may be asymptomatic, or it may present as angina or with symptoms of valvular insufficiency or outflow obstruction. The most common complication is rupture with an incidence range of 0.23% to 1.5%, often producing hemodynamic instability.^[1,2]

CASE REPORT

A 19-year-old male patient presented to our emergency department with severe chest pain and shortness of

Valsalva sinüsü anevrizması, doğuştan kalp hastalığı olarak ya da kardiyak cerrahi girişimler sonucu ortaya çıkan nadir bir kardiyak sorundur. On dokuz yaşında erkek hasta göğüs ağrısı ve nefes darlığı yakınmalarıyla başvurdu. Oskültasyonda sol alt sternal sınırdaki ve Erb noktasındaki titreşim ile birlikte 4/6 dereceli erken diastolik üfürüm duyuldu. İki taraflı bazal akciğer alanlarında krepitasyonlu raller vardı. Elektrokardiyogramda sağ dal bloku ve ST çökmesi görüldü. Troponin ve CK-MB düzeyleri yüksek bulundu. Yatıştan kısa süre sonra hastada ventrikül fibrilasyonu gelişti ve üç kez defibrilasyon uygulandı. Hemodinamik stabilizasyon sağlandıktan sonra yapılan transtorasik ekokardiyografide derece 4 aort yetersizliği, foramen ovale açıklığı ve sağ koroner sinüsten köken alan ve yırtılma sonucu sağ ventriküle giren Valsalva sinüsü anevrizması görüldü. Hastaya kardiyopulmoner baypass altında yırtılmış anevrizmanın ve foramen ovale açıklığının onarımı ve aort kapağı değişimi uygulandı. Ameliyatla ilgili ya da ameliyat sonrası takipte herhangi bir sorun yaşamayan hasta cerrahiden beş gün sonra taburcu edildi.

Anahtar sözcükler: Aort anevrizması/komplikasyon; aort yırtığı/cerrahi; aort kapağı yetersizliği; Valsalva sinüsü/patoloji.

breath. His heart rate was 103 beats per minute, and blood pressure was 100/60 mmHg. On auscultation, a grade 4/6 early diastolic murmur was heard over the left lower sternal border and Erb's area with a thrill. Crepitating rales were heard over bilateral basal lung fields. The electrocardiogram showed right bundle branch block and ST depression (Fig. 1). Among laboratory findings, increased troponin and CK-MB levels were noted. At the fifth minute of admission, the patient developed ventricular fibrillation and was defibrillated three times. After restoration of hemodynamic stabilization, transthoracic echocardiography

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Correspondence: Dr. Abdulkadir Yıldız, Türkiye Yüksek İhtisas Eğitim ve Araştırma Hastanesi, Kardiyoloji Kliniği, 06100 Sıhhiye, Ankara, Turkey. Tel: +90 312 - 327 71 90 e-mail: gadirr@hotmail.com

was performed, which showed grade 4 aortic regurgitation, patent foramen ovale, and an aneurysm of the sinus of Valsalva arising from the right coronary sinus, with rupture into the right ventricle (Fig. 2). Because of the presence of aortic regurgitation, percutaneous closure of the ruptured SVA with an Amplatzer occluder device was not appropriate, so an invasive diagnostic procedure was not considered.

The patient underwent surgical intervention through a median sternotomy and cardiopulmonary bypass was instituted. Retrograde continuous cold blood cardioplegia was used during cardiac arrest. A right atriotomy and right ventricular outflow tractotomy were performed. There was dilatation of the right sinus of Valsalva and rupture into the right ventricle outflow tract. The ruptured SVA and patent foramen ovale were repaired with fiber patches and the aortic valve was replaced with a 23-mm aortic valve prosthesis (St. Jude Medical). The patient was discharged on the fifth postoperative day following an uneventful surgery and postoperative course.

DISCUSSION

Sinus of Valsalva aneurysms are defined as enlargement of one of the aortic sinuses between the valve annulus and sinotubular ridge. Loss of lamellar elasticity results in focal weakening of the aortic wall, leading to aneurysmal dilatation and ultimately to rupture. These aneurysms are four times more common in males.^[4] They are usually congenital, but less commonly may be associated with endocarditis, atherosclerosis, trauma, syphilis, or aortic dissections. Unruptured SVAs may be medically followed-up,^[5] uncomplicated rup-

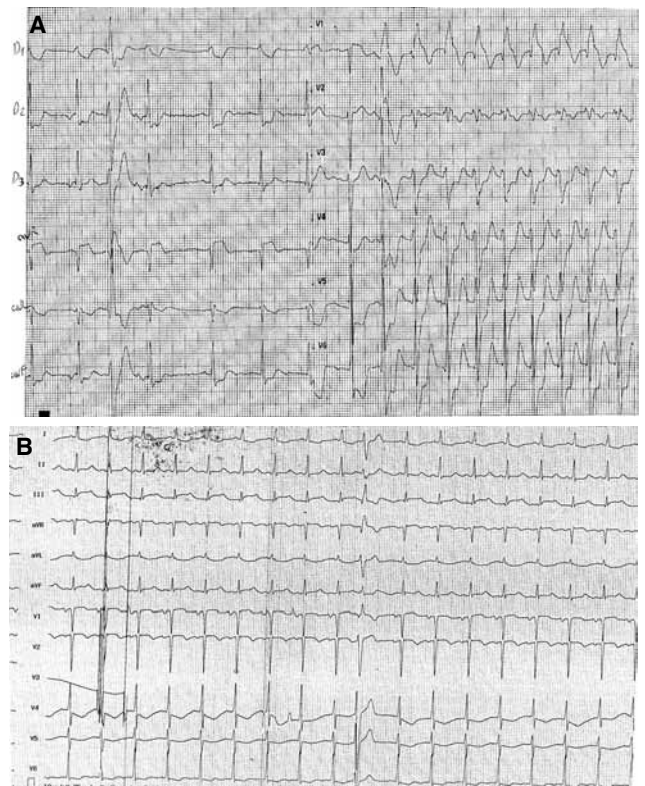


Figure 1. (A) The electrocardiogram of the patient showing right bundle branch block and ST depression as indicators of myocardial ischemia. (B) Postoperative electrocardiogram.

tured SVAs may be closed with the Amplatzer occluder device, but the majority of patients with a ruptured SVA undergo surgery between 20 to 40 years of age.^[6]

Ruptured SVAs cause acute symptoms in over 40% of the patients, the most common being dyspnea, cough, chest pain, and palpitations. In our pa-

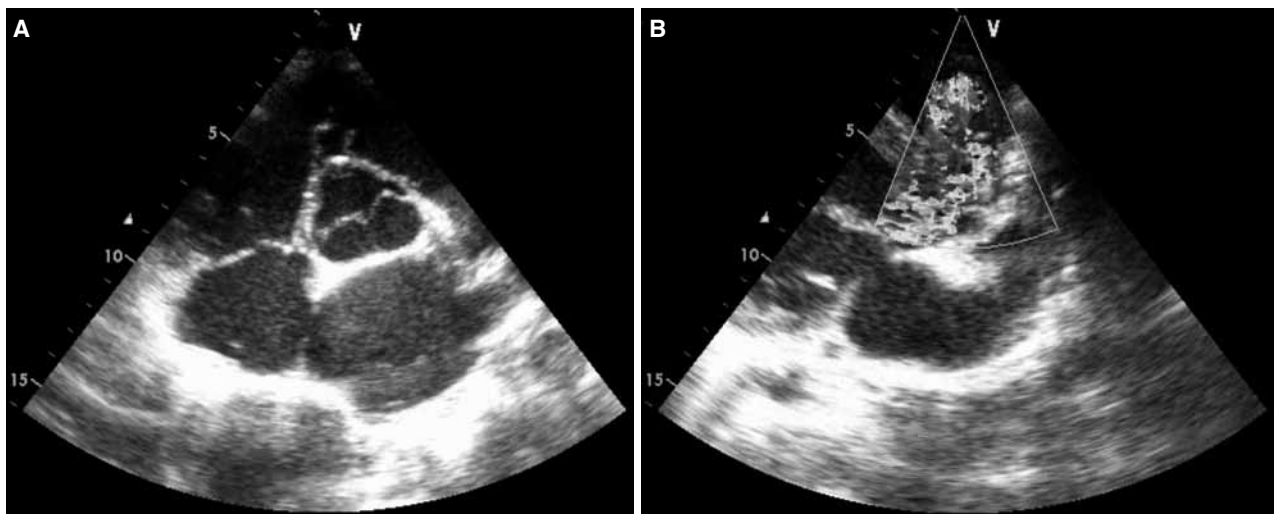


Figure 2. (A) Echocardiogram showing the windssock appearance. (B) Doppler-flow demonstration of rupture of the sinus of Valsalva aneurysm into the right ventricle.

tient, the main symptoms were angina and dyspnea. The incidence of ischemia after rupture of SVAs is not well known. Because of aortic regurgitation and left-to-right shunt from the aorta to the right ventricle, severe coronary oxygen supply/demand mismatch occurs, causing myocardial ischemia and anginal chest pain.^[7] Our patient developed ventricular fibrillation due to myocardial ischemia, which was converted to sinus rhythm with defibrillation.

Sinus of Valsalva aneurysms may be associated with ventricular septal defect, aortic regurgitation, bicuspid aortic valve, or connective tissue disorders such as Marfan and Ehler-Danlos syndromes. Aortic valve abnormalities and incompetence are common in patients with a ruptured SVA, sometimes having an adverse influence on prognosis. The incidence of aortic regurgitation has been reported to be 25% to 45%.^[1,6,8]

In conclusion, a ruptured SVA should be borne in mind in the differential diagnosis of young patients presenting to the emergency department with typical chest pain. Clinical recognition and early echocardiographic diagnosis followed by urgent surgical repair were lifesaving in our patient having a ruptured SVA accompanied by aortic regurgitation, myocardial ischemia, and patent foramen ovale.

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