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CASE REPORT

Rupture of sinus of valsalva aneurysm: earliest presentation in association with ventricular septal defect and aortic regurgitation

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Abstract:
Rupture of sinus of Valsalva aneurysm (SVA) is a rare cardiac lesion, generally occurring in second to fourth decades of life. Ruptured SVAs are frequently associated with other congenital defects, particularly with ventricular septal defect, aortic valve regurgitation, and bicuspid aortic valve. The malformation arises due to lack of continuity between the aortic media and the annulus fibrosus of the aortic valve. The structure becomes aneurysmal and may rupture to form a fistula. They are rare in paediatric patients. We describe here, a four year old female child being managed previously as a case of ventricular septal defect and aortic regurgitation deteriorates progressively. Transthoracic echocardiography revealed rupture of sinus of valsalva aneurysm into the right ventricle along with ventricular septal defect with aortic regurgitation. We report here earliest presentation of ruptured sinus of valsalva aneurysm which had been misdiagnosed as ventricular septal defect.

Keywords: *Sinus of Valsalva aneurysm, Ventricular septal defect, Aortic regurgitation*
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Introduction

Sinus of Valsalva aneurysms are rare cardiac abnormalities that usually originate in the right or noncoronary aortic sinus and may rupture into cardiac chamber. It usually presents in the second to fourth decade of life with variable clinical manifestations and male preponderance [1]. It can be congenital or acquired and is sometimes associated with other cardiac lesions such as ventricular septal defect (VSD), membranous subaortic stenosis, aortic regurgitation (AR) and bicuspid aortic valve [2]. It can be missed clinically in cases associated with to and fro murmur such as VSD and AR. Echocardiography is the main diagnostic tool for identifying ruptured SVA. The present case report describes a four year old female child with VSD diagnosed as ruptured sinus of valsalva aneurysm, found on echocardiography, which is a very early manifestation in a child [3,4].

Case report

A 4-year old female child was admitted to pediatric department with complain of cough, respiratory distress and chest pain. Past history revealed complaints of repeated episodes of respiratory distress since 1 year for which she

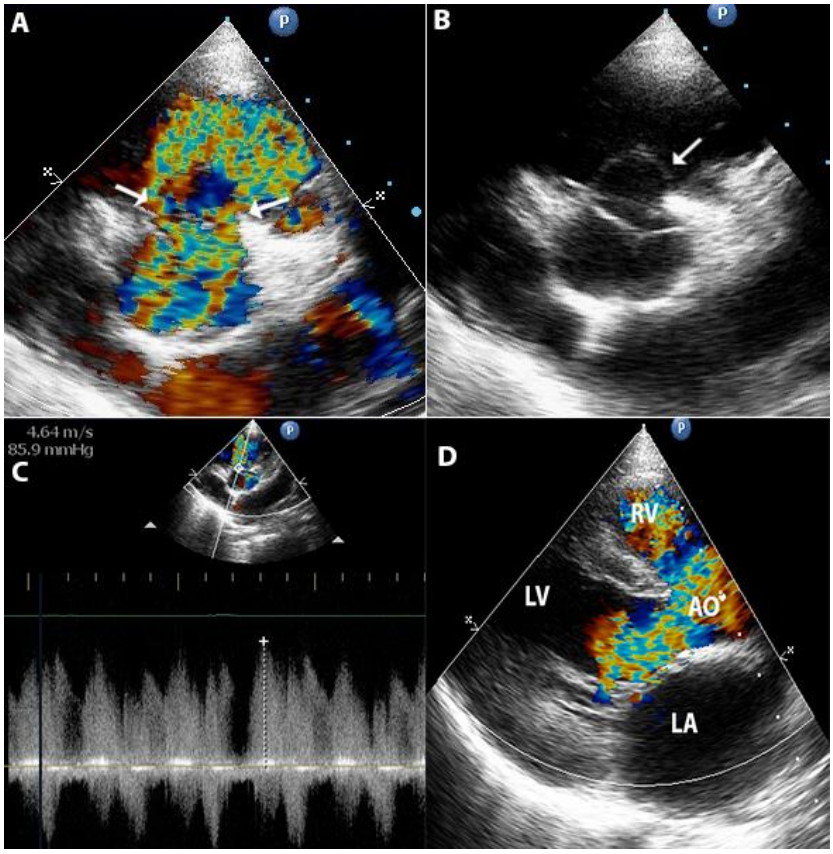

Figure I. 1(A) Short axis view on Colour doppler echocardiography shows ruptured sinus of valsalva aneurysm through right coronary cusp of aortic valve. 1(B)Short axis shows aneurysm of right coronary cusp with characteristic "windsock" deformity. 1(C)Doppler study shows continuous flow through defect with a pressure gradient of 85 mm Hg. 1(D)Long parasternal view shows severe aortic regurgitation and flow through ventricular septal defect.

had been investigated before and initially she was managed as ventricular septal defect with heart failure. However, her condition continued to worsen despite medical management for the past 2 months and was referred to us. She presented with dyspnea while playing (NYHA grade III) with swelling all over her body.

Her weight was 12 kg, height 85 cm with pulse rate of 120 beats per minute. Blood pressure was 94/38 mmHg in both arms and 96/40 in both lower limbs.

Cardiovascular examination revealed continuous murmur at left lower sternal border and grade 2 early diastolic murmur in 3rd left intercostal space. Respiratory examination revealed basal crackles bilaterally, her abdominal examination revealed hepatomegaly and other systemic examinations were normal.

Laboratory investigations showed hemoglobin 9.5 g; total leukocyte count 11,000/mm^3 with 60% neutrophils and 30% lymphocytes. Other biochemical tests including renal and liver function test were within normal limits. Chest radiography revealed a normal cardiac silhouette with mild congestion of the pulmonary vasculature. ECG showed sinus rhythm with left ventricular hypertrophy.

Transthoracic echocardiography (TTE) showed subaortic VSD measuring 5mm with dilated right sinus of Valsalva into right ventricle ('wind-sock'deformity), and enlarged LV with normal systolic function. Colour Doppler

echocardiography confirmed the presence of a ruptured right coronary sinus of Valsalva aneurysm with high-velocity flow (4.6 m/sec) into the right ventricular cavity during both systole and diastole with aortic regurgitation (Figure I).

She was managed as congestive heart failure with diuretics, digitalis and humidified oxygen. Patient underwent surgery after stabilization.

Discussion

Sinus of Valsalva aneurysms (SVAs) are rare with an incidence 0.1 to 3.5% of all congenital heart defects and their incidence is highest in Asian population with marked male preponderance (4:1) [5]. SVA arise from the right coronary sinus (65–85%), noncoronary sinus (10–30%), and rarely from left coronary sinus (5%) [6].

SVA can be congenital or acquired. The congenital aneurysm usually begins as a diverticulum that originates from one of the sinuses of Valsalva as the result of sustained high pressure in the aortic root. Acquired, or noncongenital SVAs are less frequent, caused by conditions affecting the aortic wall, such as infections (syphilis or tuberculosis), degenerative disease (atherosclerosis), connective tissue disorders (Ehlers-Danlos or Marfan syndrome or cystic medial necrosis), or trauma.

20% of SVAs were unruptured [7]. Aneurysms from the right coronary sinus, rupture into the right ventricle mostly (75%), some into the right atrium (19%), left atrium/ ventricle (6%), and least in pulmonary artery (less than 1%) [8]. Most ruptures develop well after puberty and between third to fourth decade of life, rarely seen in infancy or childhood [3]. Associations of ruptured SVA include ventricular septal defects (30–60%), bicuspid aortic valve (10%), aortic insufficiency, pulmonary stenosis, coarctation of the aorta, and subvalvular aneurysms [2]. In patients with VSD, aortic insufficiency can be explained by either lack of supporting tissue near aortic cusps or traction of the related cusp into the VSD during systole or diastole.

The clinical presentation may vary from an asymptomatic continuous murmur to heart failure, cardiogenic shock and death depending upon the onset and size of perforation [9]. The principal VSD associated with a ruptured sinus of Valsalva aneurysm is the subaortic type same as seen in our case. This is the rare case reported so far in medical literature and is unique in being associated with ventricular septal defect and aortic regurgitation with presentation at four years of age.

Transthoracic echocardiography is the initial diagnostic tool for the identification of the ruptured SVA which reveals characteristic ‘wind-sock deformity’ that is finger-like projection of the ruptured aneurysm into the receiving chamber, while cardiac catheterization is used for establishing the diagnosis and evaluating the hemodynamic effects of theruptured SVA. However interventional cardiac catheterization can also be used for therapeutic purposes in these patients. Echocardiography also provides information required for the differential diagnosis of other disorders with continuous murmur, such as patent ductus arteriosus, aortopulmonary window, coronary fistula, as well as co-existing VSD and aortic valve regurgitation. A few patients are misdiagnosed as having isolated ventricular septal defect as in our patient. In this situation, the characteristic Doppler spectrum showing continuous high jet velocity accentuated in diastole as opposed to a high velocity systolic and low velocity diastolic flow which differentiates the two conditions.

The mainstay of treatment for ruptured SVA is surgical intervention to prevent endocarditis and intractable congestive heart failure, else prognosis of patients is very poor. The long-term results after surgical repair are excellent [6]. But nowadays the patients with ruptured SVA can also be treated by transcatheter approach which is a cost-effective modality [10].

In conclusion, ruptured SVA is a rare condition with a varied presentation that can be fatal if not diagnosed early. Echocardiography is an accurate and reliable noninvasive tool to diagnose the lesion and obviates the need for cardiac catheterization in most cases. Clinicians who come across patients with continuous/ pansystolic murmur should always rule out ruptured SVA by echocardiography to avoid complications by early intervention.

References:

1. Feldman DN, Roman MJ. Aneurysms of the Sinuses of Valsalva. Cardiology 2006;106:73–81

2. Yilmaz AT, Demirkilic U, Özal E, Tatar H, Yöztü Ö. Aneurysms of the sinus of Valsalva. J Cardiovasc Surg 1997; 38: 119–124.

3. Perry LW, Martin GR, Galioto FM, Midgley FM. Rupture of Congenital Sinus of Valsalva Aneurysm in a Newborn. Am J Cardiol 1991;68:1255-6.

4. Burakovsky VI, Pcdsolkov VP, Sabirow BN, Nasedkina MA, Alekian BG, Dvinyaninova NB. Ruptured congenital aneurysm of the sinus of Valsalva. J Thorac Cardiovasc Surg 1988;95:836-841.

5.Chu SH, Hung CR, How SS. Ruptured aneurysms of the sinus of Valsalva in oriental patients. J Thorac Cardiovasc Surg 1990; 99: 288–298.

6. Takach TJ, Reul GJ, Duncan MJ, Cooley DA, Livesay JJ, Ott DA, Frazier OH. Sinus of Valsalva aneurysm or fistula: management and outcome. Ann Thorac Surg 1999; 68: 1573–7.

7. Fishbein MC, Obma R, Roberts WC. Unruptured sinus of Valsalva aneurysm. Am J Cardiol 1975; 35: 918.

8. Kar AK, Bhattacharya S, Ray D, Mondal M, Ghosh S, Mazumdar A. Rupture of the sinus of Valsalva into the pulmonary artery. Indian Heart J 2002; 54: 415–417.

9. Jugpal TS, Dixit R, Lohchab S, Garg A. Giant Unruptured Sinus of Valsalva Aneurysm. An Unusual Cause of Right Heart Failure. J Clin Imaging Sci. 2015;30:5:64.

10. Narin N, Özyurt A, Baykan A, Üzüm K. Transcatheter closure of ruptured sinus Valsalva aneurysm with retrograde approach. Arch Turk Soc Cardiol 2014;42:299-301