

A Rare Case of Ruptured Sinus of Valsalva Aneurysm in a Young Patient Presenting with Heart Failure

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ABSTRACT

This is a case of Ruptured Sinus of Valsalva aneurysm (RSOV) in a young Asian male patient presenting with symptoms of heart failure. Clinical examination revealed signs of heart failure and the presence of continuous machinery type murmur at the left sternal edge. The diagnosis was made by transthoracic echocardiogram which showed rupture of the non-coronary sinus of Valsalva into the right atrium.

Key words: Ruptured Sinus of Valsalva Aneurysm (RSOV), Oedema and Ascites, TTE.

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DESCRIPTION

An otherwise healthy 18-year-old Nepalese gentleman presented with acute onset symptoms of heart failure preceded by chest pain. The chest pain started suddenly 1 week previously. Two days later, he developed bilateral lower limbs oedema and early morning periorbital oedema. He also noticed new onset cough. His symptoms were exacerbated by physical activity. His functional class of New York Heart Association was III. He had no previous medical history and had no family history of congenital heart diseases. On examination, his blood pressure was 119/61mmHg, pulse rate was 80 beats per minute and the temperature 36.7-degree Celsius. He had bilateral pitting pedal oedema as well as sacral oedema and ascites. Precordial examination revealed grade IV continuous murmur at the left lower sternal edge. Blood investigations showed normal full blood count. The Chest X ray shows cardiomegaly and pulmonary venous congestion (Figure 1).



Figure 1: Chest X ray showing cardiomegaly with pulmonary venous congestion.

Transthoracic Echocardiogram (TTE) showed Ruptured sinus of Valsalva with severe holodiastolic flow reversal in the ascending thoracic aorta. Rupture of non-coronary sinus of Valsalva into right atrium, shunt = 0.877cm, PG ~ 88mmHg (Figure 2).

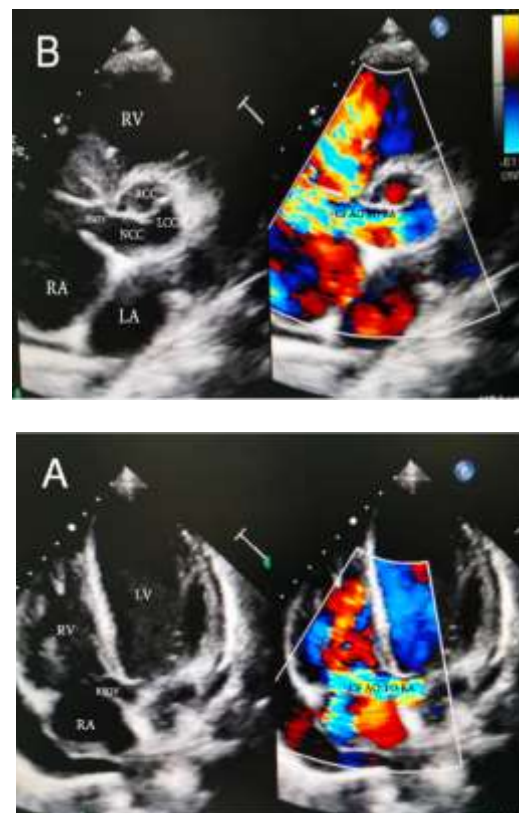


Figure 2: Transthoracic modified 5 chamber view (A) and AV short axis view (B) show colour flow (CF) from aorta (AO) to right atrium (RA) through the ruptured sinus of Valsalva aneurysm. (LV: left ventricle, RV: right ventricle, RA: right atrium, RSOV: ruptured sinus of Valsalva, RCC: right coronary cusp, LCC: left coronary cusp, NCC: noncoronary cusp, CF: colour flow, AO: aorta).

DISCUSSION

Sinus of Valsalva aneurysms (SVA) are rare cardiac conditions. Majority is congenital. However, it can also be acquired. The incidence of congenital SVAs ranges between 0.1% to 3.5% of all congenital heart defects.¹ The incidence among Asian population is 2% according to one study.² Sinus of Valsalva aneurysms are four times more frequently seen in men compared to women.^{1,3} It is associated with a

range of congenital cardiac defects; ventricular septal defect in 30% to 60% of cases, bicuspid aortic valve (10%), and aortic insufficiency.⁴ In one study, the prevalence of aortic insufficiency was reported to be as high as 23.9% in patients with SVA originating from the noncoronary sinus.⁵ Other cardiac defects include; pulmonary stenosis, coarctation of aorta, and subvalvular aneurysm.⁴

There are a number of causes for SVA rupture. Trauma; either blunt or penetrating, Syphilis, endocarditis, Marfan's syndrome and senile-type dilatation of the sinuses are responsible for acquired rupture of sinus of Valsalva aneurysms (RSOV).¹ Moreover, certain infections such as Tuberculosis and fungal infection are said to cause RSOV.¹

Our patient is a young Asian male, who has no features suggestive of Marfan's syndrome. His demographic characteristics are compatible with what has been described in literature. Although Transoesophageal Echocardiogram (TOE) was not done, TTE showed no evidence of infective endocarditis in this patient. History of blunt chest trauma was not reported in this patient.

RSOV can occur into any of the cardiac cavities, and this commonly happens in patients in their thirties and forties.^{1,3,6} The reported mean age of presentation in one study was 26.8 ± 12.1 years.² SVAs affect right, left or noncoronary sinuses.¹ About 65% - 85% of SVAs occur in the right coronary sinus, while 10% to 30% in the noncoronary sinus.³ Only small proportion affects the left coronary sinus (less than 5%).^{3,2} Cheng et al reported that 19.3% of SVAs originate from the noncoronary sinus, and the commonest two sites of SVA rupture were the right ventricle followed by the right atrium (67.9% and 27.4% respectively).² In another study, the site of termination of a ruptured SVA was the right ventricle (60% to 90%), followed by right atrium (10%), then the left atrium (2% to 3%).⁷ Our patient was 18 years old, which is an uncommon age of presentation. Furthermore, the aneurysm was found to have originated from the noncoronary sinus and ruptured into the right atrium. Again, RSOV into the right atrium is an uncommon and so is site of origin; the noncoronary sinus in this case.

The structural defect that leads to formation of sinus aneurysm is lack of unification between the aortic media and the annulus fibrosus of the aortic valve.^{1,8} The high pressure at the root of the aorta will eventually lead to aneurysm formation at this weak point.^{1,8}

Although Sinus of Valsalva aneurysms are usually asymptomatic,^{1,3,7} they may cause compression on the conducting system leading to heart block.³ Furthermore, Valsalva aneurysms may block the right or left ventricular outflow tracts, or cause angina due to disturbance of blood flow through coronary ostia,⁹ or even sudden death.¹ Sudden death can occur if rupture occurs into the pericardium.⁷ In addition, cardioembolic stroke occurs as a result of thrombus formation in an unruptured aneurysm.⁷

The more common symptoms of RSOV, however, are sudden onset of chest pain and dyspnoea, as well as other symptoms of heart failure such as paroxysmal nocturnal dyspnoea and orthopnea.^{1,3} These symptoms were prominent in our patient at presentation along with continuous machinery murmur found on physical

examination. It is worth noting that failure symptoms are due to hemodynamic instability as a result of left to right shunt or left to left shunt,¹ and large proportion of patients will have a continuous machinery murmur heard at the left sternal edge.¹

The chest x-ray findings are non-specific, such as those of enlargement of the cardiac silhouette and pulmonary congestion.¹ Electrocardiography (ECG) may show chamber enlargement which could only suggest the diagnosis.¹

Transthoracic and transoesophageal Echocardiogram are diagnostic.^{7,2} Cheng et al reported that the sensitivity and specificity of TTE for the SVAs and the associated cardiac lesions could be as high as 99.6% and 99.0%, respectively.² "Wind-sock" appearance could be detected on 2-D echocardiogram indicating RSOV.⁷ The diagnosis can later be confirmed by performing colour flow imaging and Doppler studies.¹⁰

RSOV is treated surgically.¹ If left untreated, however, patients die within one year from the onset of symptoms due to heart failure or endocarditis.^{1,7} Our patient was treated for heart failure and referred to a cardiologist and cardiothoracic surgeons for evaluation and repair in Nepal as per patient request.

LEARNING POINTS

- RSOV have been reported to occur in the third or fourth decades of life, but this is a rare presentation of RSOV given the age of the patient.
- Although RSOV is a rare condition, it should still be considered in young individuals presenting with heart failure.

CONFLICT OF INTEREST

NONE

REFERENCES

1. Goldberg, N, Krasnow, N. Sinus of valsalva aneurysms. *Cardiothorac Surg Rev.*, 2012; 836: 463-464.
2. Cheng TO, Yang YL, Xie MX, Wang XF, Dong NG, Su W, Li, L. Echocardiographic diagnosis of sinus of Valsalva aneurysm: A 17-year (1995-2012) experience of 212 surgically treated patients from one single medical center in China. *Int J Cardiol.*, 2014; 173(1): 33-39.
3. Shah AJ, Pocock JM, Belham M, Kydd AC, Watson T. Aneurysm of the sinus of Valsalva. *Cardiol J.*, 2010; 17(3): 312-313.
4. Feldman DN, Roman MJ. Aneurysms of the Sinuses of Valsalva. *Cardiology* 2006; 106(2): 73-81. <http://doi.org/10.1159/000092635>
5. Xin-jin L, Xuan L, Bo P, Hong-wei G, Wei W, Shou-jun L, Sheng-shou H. Modified Sakakibara classification system for ruptured sinus of Valsalva aneurysm. *The Journal of thoracic and cardiovascular surgery.* 2013; 146(4): 874-878. <http://doi.org/10.1016/j.jtcvs.2012.12.059>
6. Smer A, Elsallabi O, Ayan M, Buaisa H, Rayes H, Alshebani Y, Salih, M. Sinus of Valsalva Aneurysm: A

- Rare Cause of Dyspnea. *Case Rep Med.* 2015;3. <http://doi.org/10.1155/2015/467935>
7. Meier JH, Seward JB, Miller FA, Oh JK, Enriquez-Sarano M. Aneurysms in the left ventricular outflow tract: Clinical presentation, causes, and echocardiographic features. *J Am Soc Echocardiogr* 1998; 11(7): 729-745.
 8. Grosz C, Pais R, Poanta L. Rupture sinus of Valsalva in a patient with dextrocardia. Case report. *Med Ultrason* 2011; 13(4): 323-325.
 9. Mirdamadi A, Mirmohammadsadeghi M, Marashinia F, Nourbakhsh M. Sinus of Valsalva aneurysm: An uncommon presentation. *ARYA Atheroscler* 2012; 8(3): 164-166.
 10. Shah RP, Ding ZP, Ng ASH, Quek SSS. A ten-year review of ruptured sinus of valsalva: clinico-pathological and echo-Doppler features. *Singapore medical journal* 2001; 42(10): 473-476.