Ruptured sinus of valsalva

George Koshy, MD, DM, Alfred E. Farias, MBBS, MD, Nagi A. Moussa, MBBS, FRCP, Hassan F. Ahmed, MBBS, FRCP.

ABSTRACT

This case report is about a 24-year-old lady, labeled case of ventricular septal defect since birth, who presented with sudden onset of chest pain and palpitation followed by acute pulmonary edema. She was found to have clinical cardiomegaly, biventricular impulse, pansystolic murmur of ventricular septal defect with continuous murmur at the right lower sternal border. Electrocadiogram revealed frequent ventricular premature beat and chest skiagram revealed cardiomegaly with pulmonary venous congestion. Transthoracic color doppler echocardiography revealed ruptured non coronary sinus of valsalva to right ventricle with large left to right shunt, perimembraneous sub aortic ventricular septal defect with left to right shunt and prolapse of right coronary cusp with mild aortic regurgitation and good left ventricular systolic function. She underwent corrective surgery of ruptured sinus of valsalva and ventricular septal defect and aortic valve replacement

Keywords: Ruptured sinus of valsalva, non coronary sinus, perimembraneous sub aortic ventricular septal defect, aortic regurgitation.

Saudi Med J 2002; Vol. 23 (8): 995-998

Congenital aneurysms of sinus of valsalva is an uncommon condition and usually presents after its rupture. In 1839 Hope[1] described "a case of aneurysmal pouch of aorta bursting into the right ventricle" which, was the first published account of a ruptured congenital aneurysm of sinus of valsalva. The most commonly involved sinuses are the right and less commonly noncoronary sinus.[2] The involvement of the left coronary sinus is rare.[3] Aneurysms arising from the right sinus generally communicate with the right ventricle and occasionally to right atrium, however aneurysms originating in the noncoronary sinus almost always ruptures to right atrium.[2] It is to be noted that about 20% of the congenital sinus of valsalva aneurysm are unruptured and were chance findings at autopsy or cardiac sugery[4] but now readily identified by color doppler echocardiography.[5-9] We are presenting a case of rupture of aneurysm of non coronary sinus of valsalva to right ventricle diagnosed by color doppler echocardiography.

Case Report. A 26-year-old unmarried lady labeled to have ventricular septal defect (VSD) since childhood was admitted with history of severe bilateral chest pain for 2 hours followed by palpitations and shortness of breath for 3 days prior to admission. She was asymptomatic prior to this illness and denied history of prolonged fever, undue physical stress or blunt trauma to chest. On admission she was orthopneic, pulse was 110/min collapsing in nature, blood pressure 150/60. Mean pressure of JVP was not elevated. Cardiovascular examination revealed clinical cardiomegaly with biventricular hyperdynamic impulse, prominent systolic thrill along the left sternal border and pulmonary artery pulsations. First and 2nd heart

From the Division of Cardiology and Department of Medicine, (Koshy, Farias, Moussa), Medical Director, (Ahmed), Al Ain Hospital, Al Ain, *United Arab Emirates*.

Received 19th February 2002. Accepted for publication in final form 21st April 2002.

Address correspondence and reprint request to: Dr. George Koshy, Division of Cardiology and Department of Medicine, Al Ain Hospital, PO Box 1066, Al Ain, *United Arab Emirates*. Tel. +971 (3) 7635888. Ext. 755. Fax. +971 (3) 7630040. E-mail: gkcardio@emirates.net.ae

sounds were audible and she had grade 4/6 systolic murmur on the left sternal border, prominent continuous murmur with diastolic augmentation at the right lower sternal border and grade 3/6 ejection systolic murmur at the pulmonary area. She had bibasilar rales upto the infrascapular regions of chest. Electrocardiogram revealed normal PR interval with QRS axis + 60 and frequent ventricular premature beats. Chest skiagram showed cardiomegaly with cardiothoracic ratio 60% with pulmonary venous congestion and increased pulmonary blood flow. Echo colour doppler study revealed left ventricular internal diameter (LVID) diastole 4.6 cm, LVID systole 3.1 cm, left atrium 3.96 cm, Aorta 3.6 cm left ventricular ejection fraction 57%. Echocardiography revealed ruptured aneurysm of non coronary cusp to right ventricle measuring 7 mm **(Figure 1)**, sub aortic perimembranous ventricular septal defect (VSD) 4 mm with trileaflet aortic valve with good opening motion with thickened tips, mild prolapse of right coronary cusp. Mitral valve was normal and so was the pulmonary and tricuspid valves and no vegetations were noticed. Color flow imaging revealed left to right shunt through non coronary sinus and also through the ventricular septal defect **(Figure 2)**. She also had mild aortic regurgitation **(Figure 3)**. Continuous wave doppler interrogation of ruptured sinus revealed continuous flow in right ventricle and aortic regurgitation with pressure half time of 540 milli seconds. There was no evidence of mitral or tricuspid regurgitation. Peak instantaneous gradient across pulmonary valve was 26 mm Hg. Interatrial septum was intact and the systemic and pulmonary venous drainage was normal. Her renal functions were normal and blood cultures did not yield any growth.

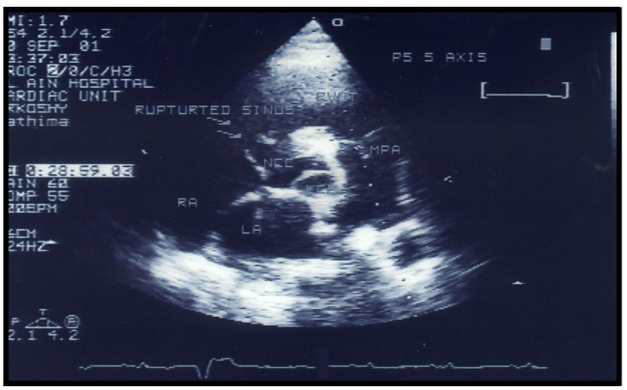

Figure 1 - Two dimensional echocardiography at basal short axis level showing the rupture of non coronary sinus of aorta.

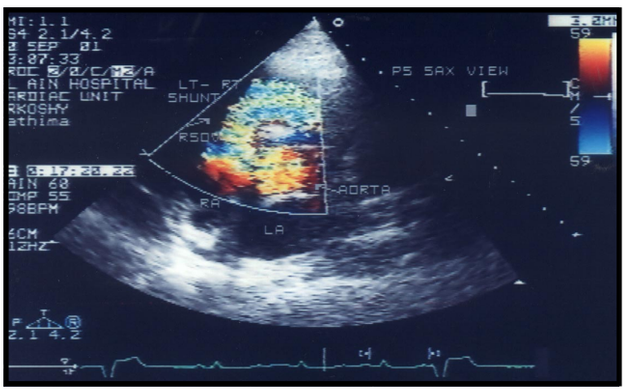

Figure 2 - Color flow evaluation at basal short axis level depicting large right to left shunt through the ruptured sinus of valsalva to right ventricle.

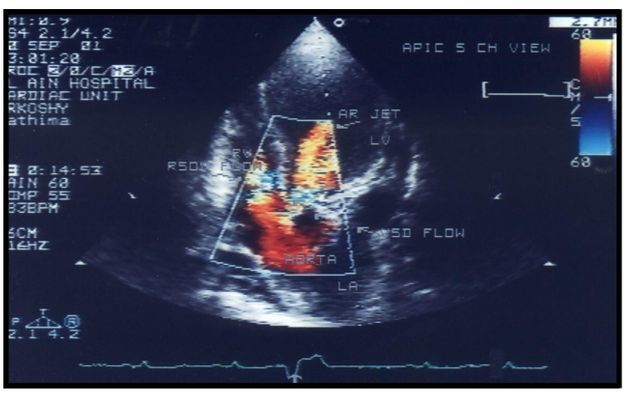

Figure 3 - Color flow evaluation in apical 5 chamber view revealing left to right shunt through the ruptured sinus, flow through the ventricular septal defect, aortic regurgitation.

Discussion. Rupture of sinus of valsalva is an uncommon disorder and results due to aortic medial separation from the anulus fibrosus resulting in aneurysmal dilatation of one or more of the aortic sinus and rupture to form an aortocardiac fistula. Diffuse dilatation of one or more aortic sinuses, with rare presentation of rupture are also reported in Marfans Syndrome.[10] Though rupture of sinus of Valsalva is more commonly observed in adults, cases have also been reported in neonates[11] and in pediatric age group.[12] Depending on the anatomical location of the aneurysmal bulge a number of complications may ensue. These frequently rupture into the right ventricle, right atrium, left ventricle, and pulmonary artery but rarely into pericardium.[9] Dissection into the interventricular septum is a very rare complication of sinus of valsalva aneurysm which originates from the right coronary sinus[9,13,14] Associated congenital abnormalities with ruptured sinus of valsalva included supracristal VSD, Gerbode defect,[15] aortic valve regurgutation, bicuspid aortic

valve,[16,17] atrial septal defect, atrial septal aneurysm,[18] patent ductus arteriosus, coarctation of aorta,[19] single coronary artery,[20] anomalous origin of right coronary artery from pulmonary artery,[21] persistent left superior venacava,[22,23] infundibular pulmonary stenosis,[24] adult polycystic disease of kidneys.[25] Systemic diseases like Syphilis,[26] Bechet's disease[27,28] and infective endocarditis[29-31] have found to be associated with rupture of sinus of valsalva. The usual site of rupture of the aneurysm of the right coronary sinus is to right ventricle and to right atrium,[32] and that of non coronary cusp is to the right atrium.[2] However, this patient had rupture of non coronary cusp to the right ventricle. Unusual sites of rupture and drainage of non coronary cusp to right atrium and right ventricle simultaneously had been reported by Lim et al,[33] rupture of non coronary cusp aneurysm to pericardium,[10] rupture of right coronary cusp aneurysm to left ventricle[34] and rupture of left coronary cusp right atrium,[35] has also been reported by various authors. This patient had acute development of large perforation resulting in sudden onset of dyspnea with large left to right shunt with clinical and radiological evidence of cardiac failure. No atrio ventricular conduction abnormalities were observed. Two dimensional echocardiography plays an important role in diagnosing ruptured or unruptured sinus of valsalva aneurysm,[36,37] and with use of contrast echocardiography,[38] and pulsed doppler,[39] and color doppler imaging help to confirm the defect.[40-43] Use of transesophageal echocardiography,[44] and magnetic resonance imaging[45-47] are complimentary and helps to confirm the anatomical details. Two dimensional color doppler and pulsed doppler echocardiography clearly identified the rupture of non coronary cusp aneurysm of sinus of valsalva to right ventricle with association of perimembraneous sub aortic VSD and mild aortic valvular regurgitation. She improved on decongestive therapy and subsequently she underwent corrective surgery with patch closure of the VSD and ruptured aneurysm with replacement of aortic valve.

In conclusion, congenital ruptured sinus of valsalva is an uncommon disorder but is to be suspected in patients with long history of VSD with sudden onset of cardiac failure with continuous murmur with wide pulse pressure. Of the various aneurysms of sinus of valsalva the occurrence of rupture of non coronary cusp aneurysm to right ventricle is rare. The presence of perimembraneous sub aortic VSD is also not a common association as sub arterial VSD is more often reported with aneurysm of sinus of valsalva. Two dimensional colors doppler and pulsed doppler echocardiography are extremely useful tools for accurate diagnosis and for documenting the communication with various cardiac chambers. These patients require patch closure of the aneurysm and closure of VSD with plication of aortic valve or aortic valve replacement.

References

1. Nowicki GR, Aberden E, Friedman S, Rashkind WJ. Congenital left aortic sinus: Left ventricular fistula and review of aortic cardiac fistula. *Ann Thoracic Surg* 1977; 23: 378-388.
2. Hope J. A Treatise on the Diseases of the Heart and Great Vessels. 3rd ed. London, (UK): J Churchill & Sons; 1839. p. 52-55.
3. Sakakibara S, Kono S. Congenital aneurysms of sinus of Valsalva; anatomy and classification. *Am Heart J* 1962; 63: 405-424.
4. Eliot RS, Wollbrink A, Edwards JE. Congenital aneurysm of left aortic sinus. A rare lesion and a rare cause of coronary insufficiency. *Circulation* 1963; 28: 951-954.
5. Fishbein MC, Obma R, Roberts WC. Unruptured sinus of Valsalva aneurysm. *Am J Cardiology* 1975; 35: 918-922.
6. Kinare SG, Bijur S, Deshpande J. Congenital aneurysms of the sinus of Valsalva with dissection into the interventricular septum. *Indian Heart J* 1989; 41: 330-334.
7. Abad C. Congenital aneurysm of the sinus of Valsalva dissecting into interventricular septum. *Cardiovasc Surg* 1995; 3: 563-564.
8. Rajendra KP, Venkiteswara Rao C, Padmanabhan TNC, Seshagiri Rao, Jaishankar S. Unruptured sinus of Valsalva dissecting into Ventricular septum: Diagnosis by Echocardiography. *Indian Heart J* 1998; 50: 209-211.
9. Munk MD, Gatzoulis MA, King DE, Webb GD. Cardiac tamponade and death from intrapericardial rupture of the sinus of Valsalva. *E J Cardiothoracic Surg* 1999; 15: 100-102.
10. Onat A, Ersanli O, Kanuni A, Aykan TB. Congenital aortic sinus aneurysm with particular reference to dissection of the interventricular septum. *Am Heart J* 1966; 72: 158-164.
11. Malcolm I. Unruptured aneurysm of sinus of Valsalva. *Can J Cardiol* 1996; 12: 783-785.
12. Chen WWC, Tai YT. Dissection of interventricular septum by aneurysm of sinus of Valsalva; A rare complication diagnosed by echocardiography. *Br Heart J* 1983; 50: 293-295.
13. Qibin Xu, Xangping Peng, Ratko PS. Doppler echocardiographic characterstics of sinus of Valsalva aneurysms. *Am Heart J* 1995; 130: 1265-1269.
14. Chasi-Pasha H, Gandhi RG, Logan WF. Ruptured aneurysm of sinus of Valsalva coexisting with single coronary artery. *Eur Heart J* 1989; 10: 180-181.
15. Sunder AS, Fox KA. Anomalous origin right coronary artery from pulmonary artery in association with congenital aneurysm of sinus of Valsalva: Angiographic diagnosis of a rare association. *Br Heart J* 1992; 68: 330-332.
16. Ohkado A, Akiyama K, Hirota J, Shiina Y. A case of ruptured sinus of Valsalva aneurysm with bicuspid aortic valve. *Kyobu Geka* 1996; 49: 567-569.
17. Pannu HS, Shivaprakash K, Bazaz S, Bedi HS, Dhaliwal RS, Singh RK et al. Geographical variation in presentation of ruptured aneurysms of sinuses of Valsalva: Evaluation of surgicalrepair. *J Card Surg* 1995; 10: 316-320.
18. Chu SH, Hung CR, How SS, Chang H, Wang SS, Tsai CH et al. Ruptured sinus of Valsalva in oriental patients. *J Thorac Cardivascular Surg* 1991; 101: 940.
19. Yildirir A, Batur MK, Kabakei G. Ruptured aneurysm of sinus of Valsalva in association with left superior vena cava—a case report. *Angiology* 2000; 51: 167-171.
20. Saito T, Mitshima A, Asano M, Yamamoto S, Sasaki S, Saeki T. Ruptured sinus of a Valsalva aneurysm associated with autosomal dominant polycystic kidney disease in an elderly patient: Report of a case. *Surg Today* 2000; 30: 732-734.
21. Karavidas A, Matsakas E, Foukarakis M, Panou F, Dounis G, Zacha. A Ruptured aneurysm of sinus of Valsalva associated with atrial septal aneurysm. *Echocardiography* 2000; 17: 733-736.

22. Coelho R, Pannu HS, Thakurta SG, Singh RS, Rao SG, Cherian KM. Ruptured aneurysm of sinus of Valsalva and Gerbode defect with severe tricuspid and aortic regurgitation. A case report and surgical correction. *J Cardiovascular Surg* 1997; 38: 531-533.
23. Nojiri C, Endo M, Koyanagi H. Conduction disturbance in Bechet's disease. Association with ruptured aneurysm of sinus of Valsalva into left ventricular cavity. *Chest* 1984; 86: 636-638.
24. Koh KK, Lee KH, Kim SS, Lee SC, Jin SH, Cho SW. Ruptured aneurysm of sinus of Valsalva in a patient with Bechet's disease. *Int J Cardiol* 1994; 47: 177-179.
25. Hanai M, Kurosawa H, Sakamoto Y, Nagahori R, Saito F, Naganum. A case report of aortic valve replacement following rupture of aneurysm of the sinus of Valsalva with bicuspid valve. *Jpn J Thorac Cardiovascular Surg* 1998; 46: 509-512.
26. Yildirir A, Batur MK, Kabakci G. Ruptured aneurysm of the sinus of Valsalva in association with persistent left superior vena cava-a case report. *Angiology* 2000; 51: 167-171.
27. Hurley JM, Govern E. Rupture of sinus of Valsalva aneurysm due to Aspergillus endocarditis. *J Cardiovasc Surg* 1994; 35: 75-77.
28. Anguera I, Quaglio G, Miro JM, Pare C, Azqueta M, Marco F et al. Aortocardiac fistulas complicating infective endocarditis. *Am J Cardiol* 2001; 87: 652-654.
29. Mayer ED, Ruffman K, Saggar W, Butzmann B, Bernhardt-Mayer, Schatton N et al. Ruptured aneurysm of sinus of Valsalva. *Ann Thoracic Surg* 1986; 42: 81-85.
30. Tesler UF, Fiorilli R, Lisanti P. Repair of sinus of Valsalva aneurysm associated with annuloaortic ectasia and coarctation of aorta in a patient with Marfans syndrome. *Tex Heart Inst J* 1997; 24: 134-138.
31. Breviere GM, Vaksmann G, Francart C. Rupture of a sinus of Valsalva in a neonate. *Eur J Pediatr* 1990; 149: 603-604.
32. Oztunic F, Aklin F, Polat B, Bilal MS. Echocardiographic diagnosis of sinus of Valsalva aneurysm rupture in two pediatric patients. *Turk J Pediatr* 1999; 41: 127-132.
33. Lim ST, Lau KW, Ding ZP. An unusual case of ruptured aneurysm of sinus of Valsalva. *Singapore Med J* 1996; 37: 115-116.
34. Kueukoglu S, Ural E, Mutlu H, Ural D, Sonmez B, Uner S. Ruptured aneurysm of the sinus of Valsalva onto the left Ventricle a case report and review of literature. *J Am Soc Echocardiogr* 1997; 10: 862-865.
35. Oberhansli I , Friedli B. Aneurysm of left sinus of Valsalva draining into the right atrium. *Chest* 1979; 76: 322-436.
36. Scaltz R, Schiller NB, Tri TB, Bowen TE, Ports TA, Silverman NH. Two dimensional echocardiographic diagnosis of ruptured right sinus of Valsalva aneurysm. *Chest* 1981; 79: 584-586.
37. Terdjman M, Bourdarias JP, Farcot JC, Gueret P, Dubourg O, Hanania G. Aneurysm of sinus of Valsalva: Two-dimensional echocardiographic diagnosis and recognition of rupture into right heart cavities. *J Am Coll Cardiol* 1984; 3: 1227-1235.
38. Nakamura K, Suzuki S, Satomi G. Detection of ruptured aneurysm of sinus of Valsalva by contrast two-dimensional echocardiography. *Br Heart J* 1981; 45: 219-221.
39. Radhakrishnan S, Bahl VK, Shrivastava S. Non-invasive diagnosis of ruptured sinus of Valsalva aneurysm by pulsed Doppler echocardiography. *Int J Cardiology* 1988; 19: 374-378.
40. Chaing CW, Lin FC, Fang BR, Kuo CT, Lee YS, Chang CH. Doppler and two-dimensional echocardiographic features of sinus of Valsalva aneurysm. *Am Heart J* 1988; 116: 1283-1288
41. Bansal RC, Wangsnes KM, Bailey L. Right aortic sinus of Valsalva-to-right ventricle fistula complicating bacterial endocarditis of membraneous ventricular septal defect; evaluation by two-dimensional, colour flow, and Doppler echocardiography. *J Am Soc Echocardiogr* 1993; 6: 308-311.
42. Dev V, Gowswami KC, Shrivastava S, Bahl VK, Saxena A. Echocardiographic diagnosis of aneurysm of the sinus of Valsalva. *Am Heart J* 1993; 126: 930-936.
43. Xu Q, Peng Z, Rakho PS. Doppler echocardiographic characterstics of sinus of Valsalva aneurysms. *Am Heart J* 1995; 130: 1265-1269.
44. Gurgun C, Ozerkan F, Akin M. Ruptured aneurysm of sinus of Valsalva with ventricular septal defect: The role of transesophageal echocardiography in diagnosis. *Int J Cardiology* 2000; 74: 95-96.
45. Kulan K, Kulan C, Tuncer C, Komsuoglu B, Zengin M. Echocardiography and Magnetic resonance imaging of sinus of Valsalva aneurysm with rupture into the ventricle. *J Cardiovasc Surg* 1996; 37: 639-641.
46. Karaaslan T, Gudinchet F, Payot M, Sekarski N. Congental aneurysm of sinus of Valsalva ruptured into right ventricle diagnosed by magnetic resonance imaging. *Pediatr Cardiol* 1999; 20: 212-214.
47. Erdol C, Gokce M, Baykan M, Celik S, Orem C, Kulan K et al. Rupture of right sinus of Valsalva into right ventricle: Echocardiographic and angiographic imaging. *J Invasive Cardiol* 2000; 12: 435-438.