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Sinus of valsalva aneurysm with fistula to the right atrium presented as acute heart failure in a young man

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Male, 23
Final Diagnosis: Sinus of valsalva aneurysm (SVA)
Symptoms: Chest pain • low O2 saturation • short of breath
Medication: —
Clinical Procedure: —
Specialty: Cardiology

Objective: Rare disease

Background: Sinus of valsalva aneurysm (SVA) is a rare cardiac anomaly. It may be congenital or acquired; a coexisting cardiac lesion might be present. Rupture of the aneurysm, where it usually occurs in the right atrium, can cause acute symptoms of heart failure. Echocardiography (particularly TEE) can provide all necessary diagnostic data for safe surgical treatment. Treatment of choice is surgery.

Case Report: A 23-year-old male, previously healthy, presented to the emergency room (ER) with shortness of breath for the last 10 hours after lifting a heavy object. The patient had central chest pain. His O2 sat was 88%. ECG showed ischemic changes. Diagnosis of AMI was made, but auscultation revealed a murmur followed by a TTE and TEE, which revealed a ruptured sinus of valsalva aneurysm.

Conclusions: This case report highlights the superiority of TEE over TTE in diagnosis and in planning adequate surgical treatment for patients with ruptured SVA, as well as the importance of ultrasonographer experience in the diagnosis. The optimal management for a ruptured SVA is surgical repair, with an acceptably low operative risk and good long-term outcome.

Key words: sinus of valsalva • aneurysm • transesophageal echocardiogram

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Background

Sinus of valsalva aneurysm (SVA) is a rare cardiac anomaly, occurring in between 0.09% and 0.15% of cases, and accounts for up to 3.5% of all congenital cardiac anomalies [1,2]. It may be congenital or acquired; a coexisting cardiac lesion might be present [1]. Rupture of the aneurysm, where it usually occurs in the right ventricle, can cause acute symptoms of heart failure [3]. Echocardiography (transthoracic, TTE and, more accurately, transesophageal TEE) can provide all necessary diagnostic data for safe surgical treatment [4]. However, preoperative catheterization and routine aortic angiography may still be necessary to confirm the diagnosis and identify associated conditions [5]. Treatment of choice is surgery, with low operative risk and good long-term survival [3].

Case Report

A 23-year-old male, previously healthy, presented to the emergency room (ER) with shortness of breath for the last 10 hours after lifting a heavy object. The patient had central chest pain, palpitation, nausea, and vomiting. On examination, his blood pressure (Bp) was 115/55 mmHg, heart rate (HR) was 125–150 bpm, and respiratory rate was 25/min. Pulse oximeter measured an oxygen (O2) saturation of 88%. Chest X-ray showed congestion of both lung fields (Figure 1), with normal heart size. Electrocardiogram (ECG) (Figure 2) showed left ventricular (LV) strain and ST depression in lateral leads. Serum troponin was positive. A provisional diagnosis of acute myocardial infarction (MI) was made. However, a diastolic murmur was heard with difficulty because of the rapid heart rate. TTE was not conclusive, but it showed grade I mitral valve regurgitation (MR), grade II aortic valve regurgitation (AR), and a pulmonary artery pressure (PAP) of 70 mmHg. TEE showed acute AR grade III–IV, possible a ruptured non-coronary cusp with fistula formation to the right atrium (Figure 3) and possible type A aortic dissection starting at the non-coronary sinus. These findings were confirmed in the operating room. Surgery included a composite mechanical graft and repair of the right atrium.

The patient's father died, unexpectedly, at the age of 43 years, which suggested ischemic heart disease (no autopsy).

Discussion

Sinus of valsalva Aneurysm (SVA) is an exceedingly rare congenital or acquired degeneration of connective tissue, and is due to either atherosclerosis, infection, or trauma [6,7] resulting in detachment of the media of the aortic wall at the aortic annulus and subsequent dilation of 1 of the 3 aortic sinuses: most commonly the right sinus in 94% of the cases

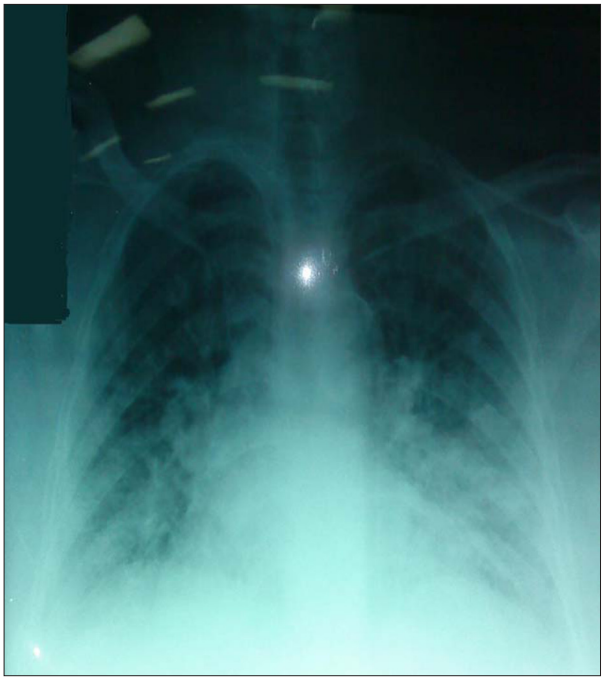

Figure 1. CXR showing bilateral lung congestion.

and rarely in the other 2 sinuses (non-coronary in 5%, and left in 1%) [8,9].

Unruptured SVAs are usually clinically silent and found incidentally during routine TTE [10]. About half are associated with AR (10). Occasionally, the physical presence of SVA can precipitate arrhythmias, obstruct a coronary artery resulting in myocardial ischemia, or disrupt normal hemodynamics [11].

Symptoms occur in 80% of patients, most commonly between 30 and 45 years of age [11]. In 35% of cases, undiagnosed SVA ruptures lead to acute symptoms in one fourth of the patients [12]. Symptoms associated with rupture are shortness of breath, chest pain, and fatigue [13,14]. The size of the shunt, the presence of other lesions, and age at presentation determine the severity of symptoms [9].

In order of frequency, the aneurysm usually ruptures into the right ventricle (60%), the right atrium (29%), and, less frequently, either into the left atrium (6%) or left ventricle (4%). Rupture into the pericardial cavity is exceptional (1%) [15].

Initially, invasive angiography was considered the gold standard for diagnosing this disease. TTE and /or TEE have emerged as preferred modalities because they provide a quick and noninvasive method that can delineate the size and location of aneurysmal dilatations, the presence of fistulous tract, the presence or absence of cardiac chamber involvement, the degree of aortic valve insufficiency, and can identify any associated anomaly or complication.

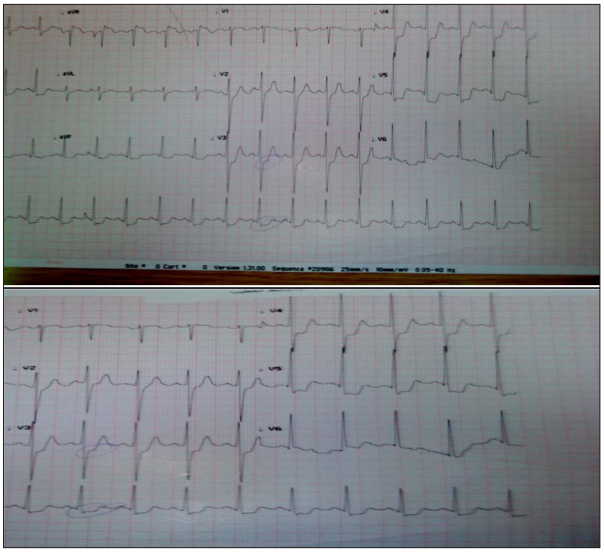

Figure 2. ECG showing LV strain, ST depression in lateral leads.

The mean survival in patients with untreated ruptured SVA is about 4 years [16]. The most used surgical technique is the 'dual exposure technique', wherein both the aorta and the chamber of termination are explored. The aneurysmal sac is excised and the resultant defect is repaired either by direct suturing or patch closure. Coexisting lesions are repaired within the same surgical procedure. Today, the 10-year survival rate after surgical repair of a ruptured SVA is 90% [17]. However, aortic regurgitation might occur after surgical repair [2,16].

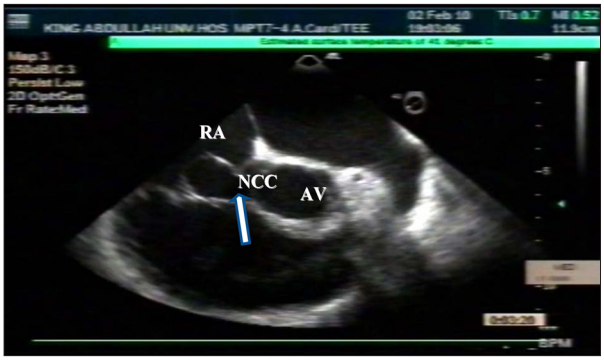

Figure 3. TEE showing the non-coronary sinus with aneurysmal dilatation and rupture (fistulization) to the right atrium (arrow): RA – Right Atrium, AA – Aortic Valve area, NCC – Non-Coronary Cusp of the aortic valve.

Conclusions

This case report highlights the superiority of TEE over TTE in diagnosis and planing adequate surgical treatment for patients with ruptured SVA, as well as the importance of ultrasonographer experience in the diagnosis. The optimal management for a ruptured SVA is surgical repair, with an acceptably low operative risk and good long-term outcome. However, the best therapy for an asymptomatic and unruptured SVA is currently not clear. The report also highlights the importance of proper physical examination for patients who present with acute chest pains.

References:

1. Post MC, Braam RL, Groenemeijer BE et al: Rupture of right coronary sinus of valsalva aneurysm into right ventricle. Neth Heart J, 2010; 8: 9–11
2. Jung SH, Yun TJ, Im YM et al: Ruptured sinus of valsalva aneurysm: transaortic repair may cause sinus of valsalva distortion and aortic regurgitation. J Thorac Cardiovasc Surg, 2008; 135: 1153–58
3. van Son JA, Danielson GK, Schaff HV et al: Long-term outcome of surgical repair of ruptured sinus of valsalva aneurysm. Circulation, 1994; 90 (5 Pt 2): II20–29
4. Tuncer M, Eryonucu B, M.D., Güntekin Ü et al: Acute rupture of a congenital sinus of valsalva aneurysm after severe exercise. Arch Turk Soc Cardiol, 2009; 37(3): 190–92
5. Przybojewski JZ, Blake RS, de Wet Lubbe JJ et al: Rupture of sinus of valsalva aneurysm into both right atrium and right ventricle. A case report. S Afr Med J, 1983; 63: 616–25
6. Freedom RM, Yoo SJ: Sinus of valsalva aneurysm. In: Freedom RM, Yoo SJ, Mikailian H, William WG (eds.), The natural and modified history of congenital heart disease. 1st ed. New York: Blackwell Publishing, 2004; 183–85
7. Vereckei A, Vándor L, Halász J et al: Infective endocarditis resulting in rupture of sinus of valsalva with a rupture site communicating with both the right atrium and right ventricle. J Am Soc Echocardiogr, 2004; 17: 995–97
8. Ott DA: Aneurysm of the sinus of valsalva. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu, 2006; 1: 165–76
9. Guo DW, Cheng TO, Lin ML, Gu ZQ: Aneurysm of the sinus of valsalva: A roentgenologic study of 105 Chinese patients. Am Heart J, 1987; 114: 1169–77
10. Shah AJ, Pocock JM, Belham M et al: Aneurysm of the sinus of valsalva. Cardiol J, 2010; 17(3): 312–13
11. Moustafa S, Mookadam F, Cooper L et al: Sinus of valsalva aneurysms – 47 years of a single center experience and systematic overview of published reports. Am J Cardiol. 2007; 99: 1159–64
12. Yan F, Huo Q, Qiao J et al: Surgery for sinus of valsalva aneurysm: 27-year experience with 100 patients. Asian Cardiovasc Thorac Ann, 2008; 16: 361–65
13. Reichert CL: Ruptured sinus valsalva aneurysm, a rare cause of heart failure. Neth Heart J, 2008; 16: 60–61
14. Weijerse A, van der Schoot MJ, Maat LP et al: Cardiac tamponade due to a ruptured aneurysm of the sinus of valsalva. J Card Surg, 2008; 23: 256–58
15. Gaitán-Román D, López-Salguero R, Álvarez-Rey I, Mejía-Vianaa SJ: Rupture of a Left Sinus of valsalva Aneurysm Into the Pericardial Cavity. Rev Esp Cardiol, 2010; 63(6): 740–50
16. Yan F, Huo Q, Qiao J et al: Surgery for sinus of valsalva aneurysm: 27-year experience with 100 patients. Asian Cardiovasc Thorac Ann, 2008; 16: 361–65
17. Wang ZJ, Zou CW, Li DC et al: Surgical repair of sinus of valsalva aneurysm in Asian patients. Ann Thorac Surg, 2007; 84: 156–60