Annals of Clinical Case Reports

Research Article
Published: 22 Jul, 2022

Rupture of Sinus of Valsalva Aneurysm: Classical Windsock in Bicuspid Aortic Valve

Poorzand H[1], Afshar S[2] and Teshnizi MA[3]*

[1]Department of Cardiology, Vascular Surgery Research Center, Mashhad University of Medical Sciences, Iran

[2]Department of Cardiology, Imam Reza Hospital, School of Medicine, Mashhad University of Medical Sciences, Iran

[3]Department of Cardiothoracic Surgery, Imam Reza Hospital, School of Medicine, Mashhad University of Medical Sciences, Iran

Abstract

Background: Sinus of Valsalva Aneurysm is a rare cardiac anomaly that could be congenital or acquired after trauma or specific medical conditions, including endocarditis.

Methods: Here, we present successful management of a case of Ruptured Sinus of Valsalva Aneurysm (RSVA) with a classic windsock appearance in a Bicuspid aortic valve in a young female.

Results: A 17-year-old female patient was admitted to the cardiac emergency unit because of palpitations, lower-extremity edema, and sudden-onset dyspnea. There was moderate cardiomegaly and bilateral pulmonary congestion on the chest X-ray. Transthoracic echocardiography shows a medial aneurysmal sinus with rupture into the right atrium, appearing as a prolonged hypermobile classic windsock deformity. The patient's parents did not agree to catheterization and candidate for cardiac surgery. The damaged tissue of the involved cusp was excised during the surgery, and a pericardial patch was inserted in the hole entering the right atrium. A bioprosthetic aortic valve replaced the aortic valve.

Conclusion: Surgical interventions can successfully treat RSVA, and windsock deformity is a diagnostic clue in RSVAs opening to the RA.

Keywords: Transthoracic echocardiography; Transesophageal echocardiogram; Sinus of Valsalva aneurysm; Rupture of Sinus of Valsalva aneurysm; Classical windsock

OPEN ACCESS

Correspondence:
Sara Afshar, Department of Cardiology, Imam Reza Sqr, Imam Reza Hospital, School of Medicine, Mashhad University of Medical Sciences, Mashhad, Khorasan Razavi Province, Iran, Tel: +98-513-8583854; Fax: +98-513-8583854;
E-mail: afshars@mums.ac.ir

Received Date: *26 May 2022*
Accepted Date: *18 Jul 2022*
Published Date: *22 Jul 2022*

Citation:
Poorzand H, Afshar S, Teshnizi MA. Rupture of Sinus of Valsalva Aneurysm: Classical Windsock in Bicuspid Aortic Valve. Ann Clin Case Rep. 2022; 7: 2260.

ISSN: 2474-1655.

Introduction

Ruptured Sinus of Valsalva Aneurysm (RSVA) is a rare cardiac abnormality presenting with typical symptoms, including dyspnea, palpitation, and chest pain [1]. While most of the ruptures occur in cardiac cavities and predominantly the right ventricle; however, extracardiac ruptures are rarely reported [1]. Transthoracic (TTE) and Transesophageal Echocardiography (TEE) are the primary diagnostic modalities in RSVA [2]. While delayed management of RSVA could be lethal, performing cardiac surgery is considered as the primary treatment as soon as the diagnosis is made [3]. Here, we present a case of RSVA with a classic windsock appearance in a Bicuspid aortic valve undergoing successful surgical repair.

Case Presentation

A 17-year-old female patient was admitted to the cardiac emergency unit because of palpitations, lower-extremity edema, and sudden-onset dyspnea. At admission, the patient complained of shortness of breath (NYHA class III) and fatigue. Physical examination revealed a systolic/diastolic blood pressure of 90/40 mmHg, heart rate of 110 beats/min, an axillary temperature of 36.5°C, jugular vein distension, and bilateral leg edema. A continuous murmur (intensity IV/VI) was heard on the left sternal border and an ejection-type systolic murmur (III/VI) over the right second inters costal space. A 12-lead electrocardiogram demonstrated a Left Ventricular (LV) strain pattern, possibly due to left ventricular hypertrophy. The chest roentgenogram revealed moderate cardiomegaly and bilateral pulmonary congestion.

A TTE was ordered for the patient. The TTE revealed normal LV size and systolic function with mild hypertrophy. D-Shape configuration of LV was noted. The aortic valve was Bicuspid with a small raphe between the Right Coronary Sinus (RCC) and None Coronary Cusp (NCC), with

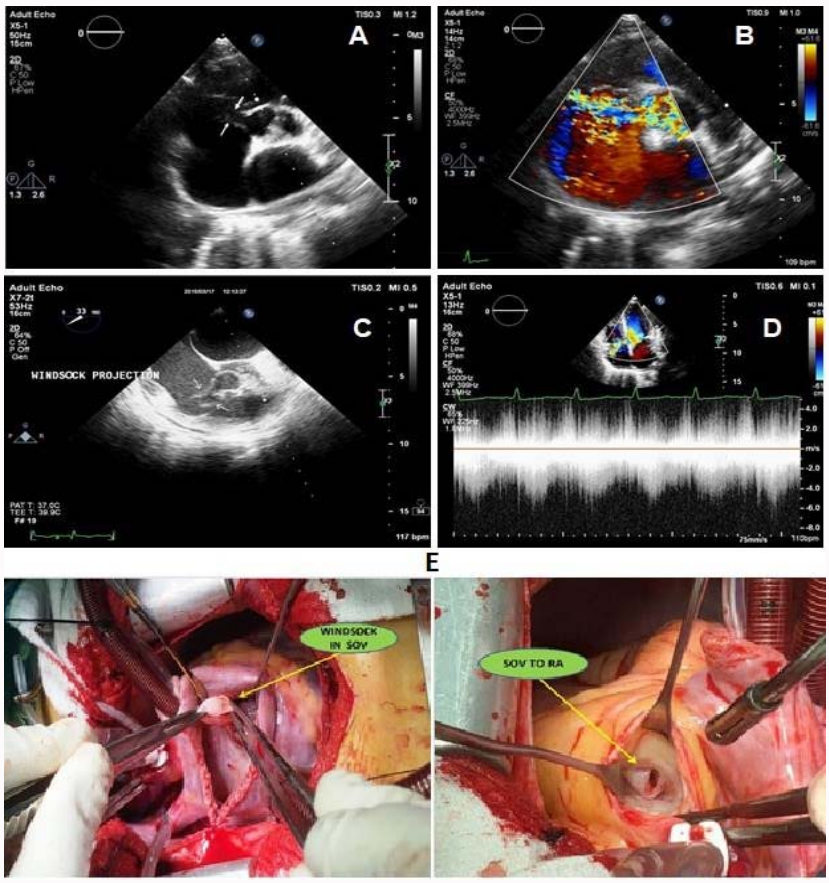

Figure 1: Windsock shape the aneurysmal medial sinus. Turbulent continues flow jet between the Right Atrium (RA) and medial aneurysmal sinus circulating in the RA. A: Short-axis view of transthoracic echocardiography showing the windsock; B: The color of A; C: Mid-esophageal transesophageal echocardiography (short-axis view of the aorta-windsock); D: Continues flow of ruptured aneurysm in transthoracic echocardiography (four-chamber view). E: Large aneurysm of the right coronary sinus with windsock formation.

thickened leaflets, moderate aortic regurgitation, and stenosis.

The medial sinus was aneurysmal with rupture into the right atrium, appearing as a prolonged hypermobile classic windsock deformity, with continuous turbulent flow directed into the Right Atrium (RA) encircling the cavity (systolic peak gradient: 100 mmHg). The rupture point was observed over the anterior and septal leaflets of the Tricuspid Valve (TV). Windsock shape aneurysmal medial sinus was hypermobile with encountering TV leaflets in most beats, but the TV annulus was normal in size, and only trace TR was observed (Figure 1). The Trans Esophageal Echocardiogram (TEE) confirmed the TTE findings, and the size of the ostium at the ruptured site was 7 mm to 8 mm. There were no other associated anomalies, including defects in the ventricular septum.

While the patient's parents did not agree to catheterization, the patient candidate for cardiac surgery and the cardiac surgeon was advised to examine the TV leaflets during surgery. The cardiopulmonary bypass was set after a median sternotomy and coronary ostial cardioplegia infusion used with moderate hypothermia (32°C). After aortotomy and right atriotomy, a large aneurysm of the right coronary sinus with windsock projection observed. The ostium of the rupture was at the point of the commissures, between anterior and septal TV leaflets, which destroyed the base of leaflets by the high-velocity jet (Figure 1).

The damaged tissue of the involved cusp was excised, and a pericardial patch was inserted in the hole entering the right atrium. A bioprosthetic aortic valve replaced the aortic valve. The patient had an uneventful in-hospital course, without residual shunt in follow-up TTE, and complete recovery.

Discussion

The present report demonstrated a case of RSVA's into RA in a young patient successfully managed by surgical approach. RSVA is an uncommon cardiac anomaly occurring due to congenital or acquired defects accounting for about 3.5% of all congenital cardiac anomalies [4]. The Valsalva sinus aneurism could be associated with genetic syndromes involving connective tissue, Ehlers Danlos syndrome, and Marfan syndrome [5]. To the best of our knowledge, neither our patient nor his family had a previous history of any genetic syndrome or connective tissue disorder. Moreover, there was no history of events related to acquired forms of aneurism, including trauma, infective endocarditis, or other causes, including atherosclerosis [5]. While most of the congenital cases of aneurysms are associated with other cardiac anomalies [5] and our patient had a bicuspid aortic valve, we may conclude that she has a congenital form of the disorder, although the possible confirmatory genetic tests not ordered due to the financial concerns. The association of the bicuspid aortic valve and Valsalva sinus aneurysm is previously reported in the literature. The Carità et al. [6] study reported a case of a congenital bicuspid aortic valve and a ruptured aneurysm of Valsalva sinus in a 61 old male patient. They hypothesized that the abnormal blood flow against

the arterial wall resulting from the abnormal aortic valve might be the reason for the aneurysm development.

Moreover, many other studies, including Arcos et al. [7] reported 27 years old male with a bicuspid aortic valve and RSVA [7-12]. The coexistence of these two cardiac anomalies strengthens the hypothesis that the abnormal flow against the arterial wall may be responsible for aneurysm development. However, in contrast to other reports indicating these two anomalies in adult patients, our patient was younger than 20, and therefore the effect of bicuspid aortic valve on the development of aneurysm may not be favorable [8-12]. Therefore, a congenital sequence rather than a genetic syndrome could be the leading cause of cardiac abnormalities. The aneurysm usually occurs following the aortic root's dilatation due to separating the aortic media from the annulus fibrous. The common complications of RSV may present as a compressive effect on near structures, including coronary arteries and adjacent conduction systems resulting in the development of ischemia or arrhythmia [4]. However, the patients mostly remain asymptomatic or rarely become symptomatic when a rupture occurs [4]. Although RSVA can be diagnosed at any age; however, the rupture usually happens in the third decade of life [1]. The RSVA could be from a congenital defect of the aortic media or resultant bacterial endocarditis or trauma [4]. The gradual development of symptoms after a rupture could be tolerated and primarily seen in patients with a small and progressive perforation.

The rapidity of the symptoms' development depends on the defect size and the chamber that the rupture opens into [3]. Although a wide range of clinical manifestations has been reported, similar to our patient, the symptomatic patients usually develop dyspnea and palpitation [13]. The rupture usually occurs in RV following RA, LA, and LV but rarely extracardiac ruptures reported into the pericardium or other extracardiac spaces [13]. In our patient, the ruptured Valsalva sinus aneurysm opening to RA was diagnosed by TTE at first. Both TTE and TEE have considerable diagnostic accuracy for the detection of RSVA and allow discrimination of the involved sinus and the size of the rupture and other associated abnormalities, including orientation of the surgical procedure [2]. Classical windsock deformity is an uncommon echocardiographic finding developed in our patient. The windsock deformity is mainly seen during a TEE study [14,15].

Cardiac surgery is the treatment of choice for RSVA [3]. The surgical repair should be considered as soon as it was diagnosed, but some articles report percutaneous intravascular repair as a therapeutic strategy [3]. While other cardiac anomalies usually accompany the congenital forms of the RSVA, the management could be complex in some patients, and the outcomes mainly depend on the type of repair and underlying cardiac abnormalities [5]. The most common surgical approach for RSVA is the 'dual exposure technique' in which both the aorta and involved chamber are explored. During the repair procedure, the aneurysmal sac excised, and the defects repaired by direct suturing or patch closure. Simultaneously coexisting lesions will be repaired. The survival rate after successful surgery is about 90% in 10 years, and aortic regurgitation is the most common side effect after this surgery [13].

Conclusion

RSVA is a rare cardiac anomaly that surgical interventions could manage. The present report demonstrated a case of RSVA opening to the RA presenting with classical windsock deformity on TTE.

References

1. Moustafa S, Mookadam F, Cooper L, Adam G, Zehr K, Stulak J, et al. Sinus of Valsalva aneurysms--47 years of a single center experience and systematic overview of published reports. Am J Cardiol. 2007;99(8):1159-64.
2. Dhawan I, Malik V, Sharma KP, Makhija N, Pangasa N, et al. Transthoracic echocardiography versus transesophageal echocardiography for rupture sinus of Valsalva aneurysm. Ann Card Anaesth. 2017;20(2):245-6.
3. Sarıkaya S, Adademir T, Elibol A, Büyükbayrak F, Onk A, Kirali K. Surgery for ruptured sinus of Valsalva aneurysm: 25-year experience with 55 patients. Eur J Cardiothorac Surg. 2013;43(3):591-6.
4. Weinreich M, Yu P-J, Trost B. Sinus of Valsalva aneurysms: Review of the literature and an update on management. Clin Cardiol. 2015;38(3):185-9.
5. Doost A, Craig JA, Soh SY. Acute rupture of a sinus of Valsalva aneurysm into the right atrium: A case report and a narrative review. BMC Cardiovasc Disord. 2020;20(1):84.
6. Carità P, Dendramis G, Novo G, Bertolino EC, Neglia L, Mignano A, et al. Bicuspid aortic valve and unruptured sinus of Valsalva aneurysm, a rare association. Int J Cardiol. 2016;202:103-5.
7. Arcos LC, Medina HM, Sandoval N. A case of sinus of Valsalva aneurysm rupture in a patient with bicuspid aortic valve. CASE (Phila). 2020;4(1):47-52.
8. Murase T, Tamura S, Ohzeki Y, Ebine K. [Ruptured aneurysm of the sinus of Valsalva accompanied with a bicuspid aortic valve inan elderly man; report of a case]. Kyobu Geka. 2017;70(10):851-4.
9. Ameta D, Kharwar RB, Aga P, Sethi R, Chandra S, Dwivedi SK, et al. Bicuspid aortic valve with severe stenosis and ruptured sinus of Valsalva aneurysm to left ventricle. Indian Heart J. 2016;68(Suppl 2):S114-7.
10. Ertaş G, Cetinkaya AS, Mehmetoğlu ME. Ruptured sinus of Valsalva aneurysm association with bicuspid aortic valve. Neth Heart J. 2013;21(1):48-9.
11. Mizia-Stec K, Haberka M, Mielczarek M, Szymański L, Skiba W, Szkutnik M, et al. [Recurrent rupture of a sinus of Valsalvaaneurysm in a patient with a bicuspid aortic valve treated with percutaneous implantation of an Amplatzer occluder]. Kardiol Pol. 2008;66(3):344-7.
12. Aggarwal SK, Iyer VR. Ruptured sinus of Valsalva with bicuspid aortic valve. Clin Cardiol. 2009;32(6):E97-8.
13. Murashita T, Kubota T, Kamikubo Y, Shiiya N, Yasuda K. Long-term results of aortic valve regurgitation after repair of ruptured sinus of Valsalva aneurysm. Ann Thorac Surg. 2002;73(5):1466-71.
14. Hirapur I, Veeranna RM, Agrawal N. Classical windsock deformity of ruptured sinus of Valsalva: An unusual appearance on transthoracic echocardiography. BMJ Case Rep. 2014;26:bcr2014204493.
15. Xu B, Kocyigit D, Betancor J, Tan C, Rodriguez ER, Schoenhagen P, et al. Sinus of Valsalva aneurysms: A state-of-the-art imaging review. J Am Soci Echocardiogr. 2020;33(3):295-312.