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Case report

Atypical presentation of ruptured right sinus of Valsalva aneurysm with severe aortic insufficiency

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ABSTRACT

The sinus of Valsalva aneurysm (SVA) is a rare abnormality that affects less than 0.1% of the general population. We present the case of a 37-year-old woman with a clinical picture characterized by dyspnea, palpitations, and syncope of 6 years of evolution. The echocardiographic study revealed a right SVA with a subpulmonary interventricular perforation of 8mm, resulting in a regurgitant jet into the right ventricle. This led to the dilatation of the right ventricular outflow tract, pulmonary artery, and severe aortic insufficiency (Carpentier's ID). The patient underwent a successful repair of the defect and interventricular perforation, and valve replacement was not required. Timely echocardiographic diagnosis and early surgery are the main predictors that make the difference between an excellent outcome and certain death.

Keywords: Valsalva Sinus; Ventricular Septal Defect; Aortic Regurgitation (source: MeSH-NLM).

Introduction

The sinus of Valsalva aneurysm (SVA) is a rare abnormality, affecting less than 0.1% of the general population, with a prevalence of 1.2-1.8% in the Chinese population and 0.14-0.96% in the Western population with congenital heart disease [1,2]. The age of presentation varies widely, ranging from 2-74 years, with a mean age of 39 years and a male predominance [2]. They are classified as congenital and acquired. Congenital cases are more common, accounting for 3.5% of all congenital heart diseases. They are often associated with other congenital disorders such as ventricular septal defect (mostly supracristal), aortic insufficiency (41.9%), pulmonary stenosis (9.7%), aortic stenosis (6.5%), aortic coarctation (6.5%), patent ductus arteriosus (3.2%), and tricuspid insufficiency (3.2%) [3-5]. SVA are frequently right-sided (75-90%), followed by non-coronary (10-25%), and the remaining in the left coronary sinus [2]. They usually occur when there is an abnormality in the fusion between the middle layer of the aortic wall and the fibrous ring of the aortic valve [5-7]. Acquired defects share similarities in their location with congenital ones and have been associated with conditions such as endocarditis, syphilis, Behcet's syndrome, and Marfan syndrome [6]. The most common complication of this defect is rupture (35.6%), which can occur spontaneously or after trauma, extreme physical exertion, or endocarditis. However, endocarditis can also occur as a complication of a ruptured SVA (RSVA) [8]. Ruptured sinus of Valsalva aneurysm (RSVA) mostly occurs towards the right heart

chambers, with 60% of cases involving the right ventricle and 29% involving the right atrium. It rarely occurs to the left side or pericardium, accounting for approximately 10% of cases [5,9]. Extracardiac rupture rarely occurs; however, if it does happen, it is usually fatal. It occurs into the pleural space and is frequently associated with acquired RSVA [9].

Case report

We present the case of a 37-year-old woman, with no relevant medical history, who was admitted to our institution due to clinical symptoms of 6 years of evolution, characterized by dyspnea (progressing from functional class I to III), palpitations and syncope. Physical examination revealed a systolic murmur in the aortic focus (IV/VI); echocardiographic study showed severe aortic insufficiency and right sinus of Valsalva aneurysm with an 8 mm continuity solution. The latter causes a regurgitant jet into the right ventricle (RV), and the RV outflow tract (RVOT) and the pulmonary artery (PA) dilatation (Figure 1). The severe aortic insufficiency is classified as Carpentier's ID, with a vena contracta of 5 mm, regurgitant volume of 70 ml, regurgitant fraction of 60%, aortic annulus of 18 (indexed at 11.3), sinus of Valsalva of 31 mm (indexed at 19.4), sinotubular junction of 33 mm (indexed at 20.7), ascending aorta of 34 (indexed at 21.3), and preserved left ventricular systolic function ([left ventricular ejection fraction] LVEF) of 65%. Due to this structural cardiac condition causing right-sided chamber overload and uncompensated heart failure, surgical repair was proposed promptly.

During the surgical procedure, a RSVA to RV, perforation of the right aortic valve leaflet plus calcification and fibrosis, retracted right leaflet cusp, right atrioventricular shunt (sinus of Valsalva to RVOT), as well as a subpulmonary ventricular septal defect of approximately 8 mm were identified. The repair was performed under extracorporeal circulation and consisted of closure of the RSVA to RV with "u" stitches with pledgets, closure of the perforation of the right coronary leaflet with continuous 4/0 polypropylene sutures; in addition, plication of the free edge of the right coronary leaflet, subcommissural annuloplasty, and primary closure of the ventricular septal defect without patch were performed (Figure 2). The surgical procedure was performed without any perioperative complications and the patient was admitted to the Intensive Care Unit (ICU) for postoperative recovery.

The postoperative period had a favorable course, initially with the support of intravenous antihypertensive medications and low-dose inotropic agents. The follow-up echocardiogram reported a LVEF of 51% associated with mild aortic insufficiency and no residual shunts. After 72 hours in the ICU, the patient was extubated, and after a 7-day hospital stay, a subsequent echocardiographic examination showed an improved LVEF of 62%. As a result, the patient was discharged with instructions for outpatient follow-up in an external clinic.

Discussion

SVA is a rare condition that predominantly affects males and has a higher prevalence in the Asian population, accounting for 32% of cases [3,5]. Its presentation in our region is infrequent, and including the present case, there have been five published reports on this condition so far [6-8]. Its main etiology is congenital, resulting from thinning of the aortic tunica media and incomplete fusion of the

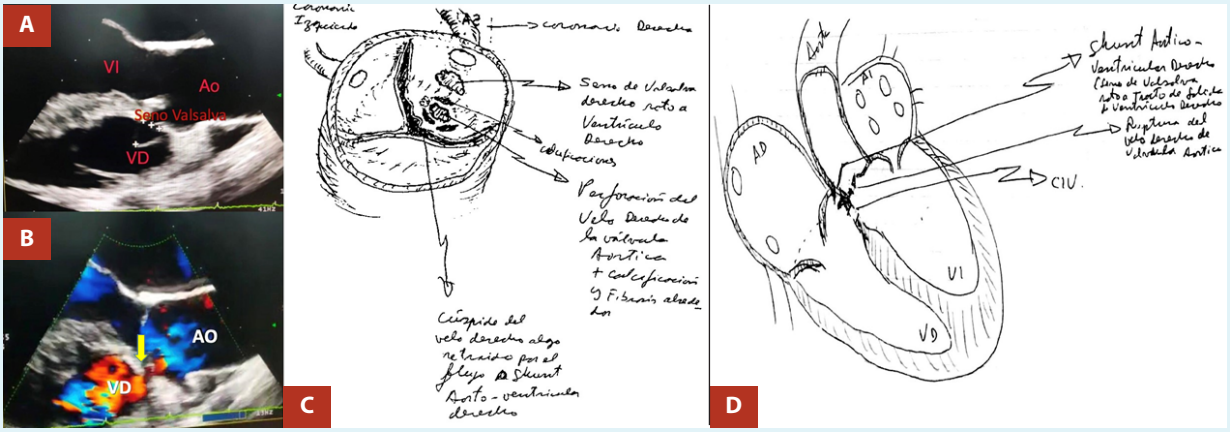

Figure 1. A-B. Preoperative echocardiographic study showing the 8mm RSVA with fistulous connection to the right-sided chambers and regurgitant jet (Arrow). **C-D.** Transverse and coronal sectional views detailing the findings identified at the level of the right coronary sinus and interventricular septum. AO=aorta, LV=left ventricle, RV=right ventricle.

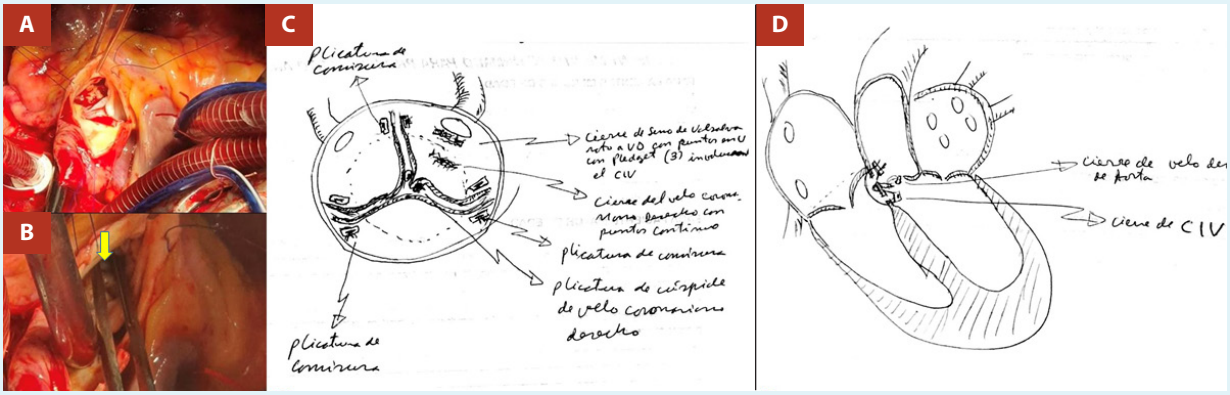

Figure 2. A-B. Intraoperative images confirming the fistulous connection (sinus of Valsalva to right ventricle) with a surgical clamp (Arrow). **C-D.** Graph showing RSVA repair, VSD closure, commissural and right coronary leaflet cusp plication.

distal bulbar septum, which separates the pulmonary artery from the aorta and connects to the fibrous ring of the valve [2, 4]. Since the right sinus and two-thirds of the coronary sinus originate from this septum embryologically, the defect is primarily located in these two regions [1,3]. However, involvement of the left sinus of Valsalva is not uncommon and an acquired origin should be suspected, with infective endocarditis being the most frequent cause in this subgroup [5].

Pathophysiologically, this defect leads to wall weakness, which, under high-pressure conditions in the aorta, can cause dilation and eventually rupture [2]. The disease often progresses asymptomatically, and when symptoms occur, they can manifest in two ways. The first, in a compressive manner, due to its growing size to compress other structures and generate ischemia or thrombosis at the coronary level and the second, generally of acute onset, due to rupture, resulting in a variable clinical symptom depending on the site of fistulization (chest pain, cardiac tamponade, heart failure, etc.) [1-3]. RSVA with fistulization into lower-pressure chambers, particularly the right atrium and ventricle, is likely the main form of presentation for this condition [3,5]. When the fistulous connection is established, progressive and rapidly evolving heart failure symptoms can appear, being possible to auscultate a continuous murmur in machinery, of high intensity and mesocardial predominance [4].

Regarding diagnosis, the definitive identification is made through echocardiographic studies, which show the aneurysmal area and continuous flow through the fistula, this can be complemented by a transesophageal view [1]. However, due to the low frequency of SVAs, the correct diagnosis of complications arising from their rupture requires a high index of suspicion and careful echocardiographic examination, as it was in our case. This examination should be focused on the differential diagnosis with other more common conditions that can also present with aortic wall rupture, such as complications derived from infective endocarditis and aortic dissection [9]. Additional diagnostic tests include computed tomography or magnetic resonance imaging, which are reserved for evaluating the entire aorta and better characterization of the defect, especially in the context of definitive treatment. However, in this case, it was not necessary since the diagnosis was successfully identified with echocardiography study.

The literature has not reached a consensus on specific treatment indications, but current guidelines describe surgical options for aneurysmal dilations greater than 55 mm in the general population, 50 mm for patients with Marfan syndrome, 45 mm in the presence of risk factors, and 55 mm or more for patients with a bicuspid valve [3,9]. There are several reports describing the unusual presentation of SVA, its complications, and various surgical repair possibilities; however, many of these reports conclude that the repair of a RSVA should be performed systematically at the time of diagnosis [1,5]. Early surgery is indicated even in asymptomatic patients because aorto-atrial fistulas, if left untreated, have a poor prognosis, and severely compromise the patient's life expectancy to less than 30% within 2 years [1-4]. The prognosis is influenced by the progression to heart failure and the risk of serious complications such as endocarditis, which occurs in 21% of cases [3,5,10]. Only in asymptomatic and very small defects, close quarterly surveillance with echocardiographic studies could be considered, assuming that a high percentage of these defects (43.4%) will require intervention within the first two years of follow-up [3,11,12].

This clinical case represents the paradigm of complicated SVA with rupture, which is a complex and very rare condition. In addition to a high clinical suspicion, a thorough and detailed

echocardiographic study is crucial, along with timely surgical intervention to ensure good patient survival. This particular case is unusual due to the coexistence of three simultaneous lesions: ruptured right sinus of Valsalva to the right ventricle, perforation of the right leaflet of the aortic valve and a subpulmonary ventricular septal defect, making our case a real challenge for diagnosis and management.

Author contributions: MÁSS: conceptualization, methodology, writing. ASE: conceptualization, methodology, writing. WSC: conceptualization, methodology, writing.

References

1. Arcario MJ, Lou S, Taylor P, Gregory SH. Sinus of Valsalva Aneurysms: A Review with Perioperative Considerations. J Cardiothorac Vasc Anesth. 2021;35(11):3340-3349. doi: 10.1053/j.jvca.2020.12.016.
2. Nguyen Q, Vervoort D, Phan K, Luc JGY. Surgical management for unruptured sinus of Valsalva aneurysms a narrative review of the literature. J Thorac Dis. 2021 Mar;13(3):1833-1850. doi: 10.21037/jtd-20-2682.
3. Joshi AS, Aironi BD, Kotkar NS, Kulkarni DV, Jadhav UE, Nagare KM. Ruptured sinus of Valsalva aneurysms - our surgical experience of 7 years. Kardiochir Torakochirurgia Pol. 2021;18(2):100-104. doi: 10.5114/kitp.2021.107471.
4. Xu B, Kocyigit D, Godoy-Rivas C, Betancor J, Rodríguez LL, Menon V, et al. Outcomes of contemporary imaging-guided management of sinus of Valsalva aneurysms. Cardiovasc Diagn Ther. 2021;11(3):770-780. doi: 10.21037/cdt-20-630.
5. Wang Y, Wu B, Li J, Shu X. Sinus of Valsalva Aneurysm in Females. Int Heart J. 2022;63(6):1201-1204. doi: 10.1536/ihj.22-156.
6. Carrión-Arcela JP, Custodio - Sánchez P, Rodríguez Urteaga Z, Ríos Vásquez C. Aneurisma del seno de Valsalva roto hacia aurícula derecha en el adulto: a propósito de un caso. Arch Peru Cardiol Cir Cardiovasc. 2021;2(1):63-7. doi: 10.47487/apcyccv.v2i1.106.
7. Paredes-Muñoz Martín Abel, Montoya-Rojas Héctor Alberto, Pino-Zavaleta Francis Robert, Ponce-Mendoza Víctor Manuel, Reyes-Chávez Jhunior Darwin Heber, Camacho-Saavedra Luis Arturo. Rotura del seno de Valsalva: Reporte de caso. Rev Med Hered. 2020;31(2):116-118.
8. Paredes-Muñoz MA, Montoya-Rojas HA, Pino-Zavaleta FR, Ponce-Mendoza VM, Reyes-Chávez JDH, Camacho-Saavedra LA. Rotura del seno de Valsalva: Reporte de caso. Revista Médica Herediana. 2020;31(2):116-8. doi: 10.20453/rmh.v31i2.3773.
9. Regeer MV, van der Zwaan HB, Schoof PH, van Aarnhem EEHL. Surgical Repair of a Sinus of Valsalva Aneurysm. World J Pediatr Congenit Heart Surg. 2022;13(6):807-809. doi: 10.1177/21501351221109193.
10. Qiu J, Xie E, Wang Y, Wang W, Yu C, Luo X. Surgical Interventions Are Effective for Treating Unruptured Sinus of Valsalva Aneurysms. Front Cardiovasc Med. 2021;8:707147. doi: 10.3389/fcvm.2021.707147.
11. Nakamura Y, Burkhart HM. Sinus of Valsalva aneurysm: Defining the optimal approach. J Card Surg. 2022;37(12):4456-4458. doi: 10.1111/jocs.17060.
12. Chaganti YS, Husain SM, Iyer VR, Desai N. Sinus of Valsalva aneurysm: A single institutional experience with 216 patients over 30 years. J Card Surg. 2022;37(12):4448-4455. doi: 10.1111/jocs.17021.