Rupture of Aneurysm of Sinus of Valsalva into the Right Ventricle: Report of One Case

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Abstract: Aneurysm of the sinus of Valsalva starts at the base of one of the 3 sinuses of Valsalva; then it extends towards the cardiac chambers. This is a rare disease which diagnosis is most of the time made at the stage of rupture. This work, report a case of ruptured aneurysm of Valsalva’s sinus into the right ventricle (RV) in a 54-year-old male patient. Our intention is to emphasize the role of Trans Thoracic Echocardiogram (TTE) in the diagnosis of this pathology.

Keywords: Ruptured, Aneurysm of Sinus of Valsalva, Right Ventricle

1. Introduction

The sinus of Valsalva aneurysm (SVA) is defined as a significant dilatation of the aortic wall between the aortic valve and the sinotubular junction [1].

Aneurysm of sinus of Valsalva (ASV) is a rare pathology that represents between 0.15% and 0.96% of surgical cardiac cases [2]. It involves commonly the right coronary sinus (RCS) ~70% followed by noncoronary sinus ~25% and rarely the left coronary sinus (LCS) ~5% [3]. The initial anomaly is a failure to fuse between the aortic tunica media and the other structures of the heart [3]. It can become complicated with rupture or infection. Although all three aortic sinuses can rupture, the right aortic sinus does so most commonly, and the
receiving chamber is right-sided, resulting in a left-to-right shunt. The most common receiving chamber is the right ventricle [4]. The diagnosis is most of the time made at the stage of rupture. Doppler TTE color mode is a key investigation [5]. The rupture is a serious complication, it can occur spontaneously; or induced by trauma, endocarditis or tuberculosis. The treatment was surgical but recently, transcatheter closure of ASV has also recently been proposed. This is a report of a case of ruptured congenital ASV into the right ventricle.

2. The Case

Mr MN was a 54-year-old patient admitted for dyspnea class 3 of the New York Heart Association. His medical history started 3 months earlier with class 2 dyspnea but without orthopnea, chest pain and fever. No chest trauma was recorded. The clinical examination found the following parameters: blood pressure (BP) = 120/60 mmHg; Heart rate (HR) = 85 /min; the weight = 71 kg and the height = 1.65 m. The temperature was normal. The auscultation revealed a continuous murmur that was located at the left sternal border; the grade was 3/6. Other findings were compatible with right sided heart failure. The biology found steady increases in blood creatinine (15.59 mg/L) and uricemia (78.33 mg/L). The other lab tests that included complete blood count, fasting glucose, lipid, ions, inflammation tests and hemostasis tests were normal. The electrocardiogram (ECG) showed sinus rhythm, heart rate 83 bpm, QRS axis +35, right atrium hypertrophy and right ventricle hypertrophy. There was normal repolarization. The chest x rays showed cardiomegaly (cardiothoracic ratio = 65%) with bilateral hilar congestion. The transthoracic echocardiography (TTE) showed no dilated LV; the motion and systolic function were normal. The LA was not dilated. There were moderate mitral regurgitation and moderate aortic insufficiency; there was a no echogenic neocavity that sized 14×11 mm (figure 1).

There was slight dilation of the right ventricle. The diameter of the tear in the right coronary sinus was 4 mm. The sizes of ascending aorta, arch and descending aorta were normal. There was no collected fluid in the pericardium. No vegetation was found. Medical treatment was given to the patient. It consisted of furosemide and nitrate derivatives. The early evolution was satisfying although the persistence of class 2 dyspnea. Currently, the patient is waiting for surgical repair.

Figure 1. No echogenic neocavity of 14 x 11 mm (diameter) located in the right coronary sinus corresponding to an aneurysm of sinus of Valsalva.

It was located at the right coronary sinus. In fact it was a right coronary sinus aneurysm that was ruptured into the right ventricle (figure 2) leading left-to-right shunting from the aneurysmal right coronary sinus to the right ventricle. There were severe pulmonary artery hypertension (pulmonary artery systolic pressure = 67 mmHg) (figure 3).

Figure 2. Aneurysm of sinus of Valsalva.

Figure 3. Shunt from the right coronary sinus to the right ventricle, severe pulmonary artery hypertension (pulmonary artery systolic pressure = 67 mmHg)
3. Discussion

ASV is a rare pathology. It represents from 0.15% to 0.96% of open heart surgery cases [1] and 0.1 to 3.5% of congenital heart diseases [6]. ASV can be asymptomatic until the age of 30 or 40 years old before rupture happens [7]. Sinus of Valsalva aneurysm is more prevalent in Asian countries and in men. OVA ASV can be congenital (Marfan syndrome, Ehlers–Danlos syndrome, or other connective tissue disorders) or acquired (siphils, bacterial endocarditis, tuberculosis, chest trauma, vasculitic diseases, and iatrogenic injury during aortic valve replacement) [8]. Congenital ASV is due to malfusion of aortic media and annulus fibrosis of aortic valve. Most frequently, right coronary sinus is involved in draining into right ventricle or right atrium [9]. No acquired etiology was found in our patient so we guessed that it probably comes from congenital anomaly [2]. Congenital ASV is due abnormal prenatal development in which aortic tunica media fails to fuse with the cardiac structures. Therefore, the absence of elastic layer in the aortic wall makes it flimsy [10, 11]. Like in our patient, the commonest location is the right coronary sinus (70%) whereas location in the left coronary sinus is unusual (5%) [2]. Complications of ASV include aortic regurgitation, compression of coronary arteries, arrhythmia and rupture. The rupture is very rare and happens in male patients during their 30s or 40s [12]. ASV can break into the right ventricle, the right atrium, the pericardium, the pleural cavity or in the left cavities of the heart. The rupture can occur spontaneously; it can also be induced by trauma, endocarditis or tuberculosis. Our case is certainly a spontaneous rupture. TTE is a key investigation for diagnosis of the rupture; it can detect the associated lesions [13]. It has been very helpful for diagnosing ruptured and intact aneurysm and has also proved to be a great guidance tool for intra operative repair. It provides information regarding involved sinuses, protrusion, and associated shunt or coexisting cardiac abnormalities [14]. TEE was thus beneficial in guiding the surgeon about both right coronary sinus aneurysm and LCS aneurysm with rupture of the left coronary sinus aneurysm aneurysm into the LVOT [5]. This emphasizes the role of intraoperative TEE for ruptured sinus of Valsalva [5]. It also guides about the origin, the size of the aneurysm, and associated conditions [5]. Magnetic resonance imaging might be useful in diagnosing the coexisting coronary lesions more precisely. TOE (trans-esophageal echocardiography) is recommended when the results of TTE are unclear. Other investigations such as 3D echogram, contrast tomography and magnetic resonance imaging could help for further analysis of the lesion [15, 16]. Preoperative coronary angiography can identify the associated coronary arteries diseases. Non-symptomatic and intact aneurysms are managed conservatively. Surgical resection is generally recommended in symptomatic cases, when there is compression or distortion of surrounding structures, in cases of rupture, and in the cases in which the SVA is incidentally discovered at the time of surgery for other cardiac reasons [17]. Presence of a continuous murmur along with sudden onset of chest pain and shortness of breath might be secondary to a ruptured sinus of Valsalva. This patient presented a ruptured ASV of the right coronary sinus into the right ventricle. The treatment was surgical but transcatheter closure of ASV was also described in 1994. Since then, there are several case reports. Still the mainstay of treatment for complex ASV with multiple defects or VSD or AR is surgery requiring cardiopulmonary bypass [8].

Early repair prevents from endocarditis and avoids the extension of the rupture. The current surgical procedures provide good outcomes as the surviving rate can reach 97.3% by postoperative year 10 [2, 18]. However increasing numbers of study report some good results with percutaneous closing procedures. These constitute an alternative to open heart surgery [19-20].

4. Conclusion

ASV is a rare and serious aortic disease. Here we report a ruptured aneurysm of sinus of Valsalva into the right ventricle, echogram is the key exam to make the diagnosis it provide exact and comprehensive information about complicated conditions. Although surgical repair is the first-line treatment, percutaneous closing procedures can be an alternative.

Abbreviations

ASV: Aneurysm of sinus of Valsalva
BP: blood pressure
HR: Heart rate
ECG: electrocardiogram
TTE: transthoracic echocardiography
TOE: trans-esophageal echocardiography

References


