

Sinus of Valsalva Rupture or VSD Shunt: Mystery solved by Cardiac CT

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ABSTRACT

An unruptured aneurysm of the Sinus of Valsalva (SOV) is an asymptomatic pathology and diagnosed incidentally. This extremely rare anomaly can be associated with other congenital cardiac anomalies which can make the diagnosis and prognosis even more complex. We are reporting a case of a 12-year-old child with progressive dyspnoea and episodes of syncope. Multimodality imaging confirmed the diagnosis and paved the way for appropriate surgical treatment options.

KEYWORDS: Sinus of Valsalva aneurysm; Ventricular septal defect; Cardiac CT; Surgical plan

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INTRODUCTION

Sinus of Valsalva (SoV) aneurysms is extremely rare cardiac anomalies that may be acquired or congenital. The congenital aneurysm is more common and is most often caused by weakness at the juncture of the aortic media and the annulus fibrosus.^[1] Aneurysms may originate in the right coronary sinus (65%–85%), the noncoronary sinus (10% – 30%), and, rarely, the left coronary sinus (1%–5%).^[2] Although aneurysms usually remain asymptomatic, rupture can complicate the pathophysiology very rapidly. Ruptured sinus of Valsalva aneurysms are frequently associated with perimembranous ventricular septal defects.^[3] We report herein a patient with an unruptured right sinus of Valsalva with the subpulmonic location of a small ventricular septal defect with a curved jet around the aneurysm.

CASE REPORT

A 12-year-old male child with a BMI of 20.6 kg/m² was presented to the outpatient department with symptoms of fatigue, substernal chest pressure, palpitations, and near syncope episodes three times on different days for the past 2

months. He had a history of cardiac murmur since birth and was diagnosed at that time with a small Ventricular Septal Defect (VSD) by Trans Thoracic Echocardiography (TTE) with no further clinical follow-up.

An electrocardiogram showed sinus tachycardia at a rate of 110 bpm and met the voltage criteria for incomplete RBBB. Chest radiography revealed a normal cardiac silhouette with mild congestion of the pulmonary vasculature. TTE showed normal left and right ventricular dimensions; left ventricular systolic function was hyperdynamic. Color-flow doppler echocardiography displayed severe aortic regurgitation with normal biventricular systolic function. Additionally, an abnormal flow curving around the right SoV aneurysm into the right ventricular outflow tract was found (Figure 1 & 2). Interestingly, due to unusually curvilinear thick jet, precise determination of SoV rupture or VSD shunt was not possible on TTE. To determine this complex anatomy, a contrast cardiac computed tomography (CT) scan was performed. On cardiac CT, evidence of contrast-filled outpouching seen arising from right aortic SoV of approximately 16 mm × 16 mm width & 12 mm wide neck suggestive of an aneurysm (Figure 3). The right SoV aneurysm extends to the right ventricle with reflux of contrast into the right ventricle. Additionally, there was evidence of thin track-like communication filled with contrast in the subpulmonic region; confirming the underlying subpulmonic location of VSD (Figure 4).

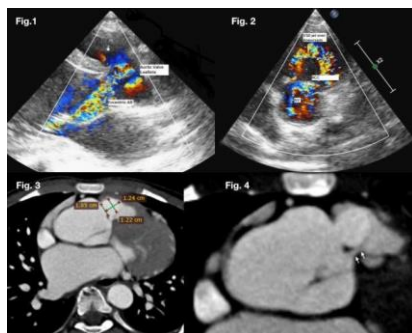


Figure 1: Parasternal long-axis view shows eccentric severe aortic regurgitation jet with dilated aortic root.

Figure 2: Left ventricle to right ventricular shunt with a curved path over an aneurysm.

Figure 3: Cardiac CT showing the RCC aneurysm with contrast reflux extending into RV.

Figure 4: LVOT to RVOT contrast flow at subpulmonic location suggestive of VSD shunt.

DISCUSSION

Sinus of Valsalva aneurysms is very rare, with incidence rates ranging from 0.1% to 3.5% of all congenital heart diseases.^[4] Sinus of Valsalva aneurysms occurs three times more often in males, with the highest incidence in Asian populations.^[5] Most sinus of Valsalva aneurysms arises from the right or the noncoronary sinuses. They commonly rupture into the RV or right atrium. Anatomically, most sinus of Valsalva aneurysms is congenital in origin, caused by the lack of fusion between the media of the aorta and the annulus fibrosus of the aortic valve.^[6] Hemodynamically, the flow-through VSD produces the Venturi effect, a tendency for the related aortic sinus and cusp to pull away from closure. These two mechanisms induce AR, and AR begets AR. Our case confirms the mechanism of this classic effect. Although the first line of investigation in such a case is TTE accuracy is reported approximately 75% in literature.^[7] In our case, TTE confirms the site and size of the aneurysm but fails to determine

abnormal jet as rupture vs. shunt. It was suspected that two different pathologies are difficult to differentiate because of an anatomically adjacent location at the level of the aortic annulus. Cardiac CT identified the exact localization of the VSD which was subpulmonic and the RCC aneurysm was intact with no evidence of contrast spill into the right ventricle.

SURGICAL PLAN

The three-dimensional outline of the complex defect enables a proactive surgical plan and appropriate choice of the surgical method. The importance of advanced imaging in surgical planning is the most critical step in such cases. 3D CT gives accurate anatomy of the region of the aortic root and location of coronary Ostia. In our case, the surgical option includes simple patch closure of VSD with or without aortic valve repair. Another approach is 'Ross' procedure where the diseased aortic valve is replaced by the patient's pulmonary valve and the pulmonary valve is replaced by a homograft. Another surgical approach is the 'Bentall' procedure where composite graft replacement of aortic root, aortic valve, and ascending aorta is performed with reimplantation of coronary Ostia. The choice of surgical approach is determined by the cardiovascular surgical team combining the overall clinical parameters with the anatomy of the aortic root.

CONCLUSION

Cardiac CT with a three-dimensional anatomical outline is the most advanced and preferred imaging method for the appropriate choice of surgical method and risk stratification of surgical outcome. Our case report highlights the importance of comprehensive evaluation of complex congenital heart diseases by clinical parameters and advanced cardiac CT imaging for accurate diagnosis and treatment plans.

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