

Sinus of Valsalva Aneurysm Dissecting Into the Interventricular Septum – An Unusual Case

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ABSTRACT

Sinus of Valsalva Aneurysm (SVA) is a cardiac anomaly due to the defective connective tissue at the aortic root. It is usually congenital and sometimes acquired. They can remain asymptomatic and be detected incidentally or can manifest with heart failure and its sequelae when ruptured. We report a case of Sinus of Valsalva aneurysm dissecting into the interventricular septum.

Keywords

Sinus of Valsalva aneurysm, Dissecting aneurysm of the interventricular septum (DAIS), Aortic regurgitation, Heart failure, Left bundle branch block.

Case

A 17-year male, presented with complaints of shortness of breath of 1-month duration which was insidious in onset and gradually progressive. There was no history of cough, wheeze, palpitations or chest pain. There was no history of rheumatic fever. There was a recent history of strenuous physical activity to improve his physique. On examination, he was tall, thinly built, with a high arched palate and flat chest. There was no family history of Marfan's syndrome. The pulse was high volume and collapsing in character. The other peripheral signs of distal run-off, i.e., Corrigan's pulse in carotids and pistol shot sounds over both femoral arteries were present. His blood pressure was 120/50 mm Hg. He had a holodiastolic murmur over the right third intercostal space and along the left parasternal

border and a pansystolic murmur of grade 3/6 at the apex radiating towards the axilla in left lateral position. There were crepitations heard over the base of both lungs. Other systems examination was unremarkable.

The 12-lead electrocardiogram showed sinus tachycardia, left axis deviation with a prolonged PR interval and complete left bundle branch block (Figure 1). The chest radiograph showed cardiomegaly with pulmonary venous hypertension. Echocardiogram revealed an unruptured right SVA dissecting into the proximal one-third of the interventricular septum below the origin of the right coronary artery (Figure 2). There was severe aortic regurgitation (AR). Left ventricular ejection fraction was 34%. There was severe mitral regurgitation secondary to left ventricular annular dilatation. There was severe tricuspid regurgitation with severe Pulmonary hypertension. The right ventricular function was normal. A CT coronary angiography and aortography showed a large SVA dissecting into the interventricular septum (Figure 3a). Coronary

arteries were normal.

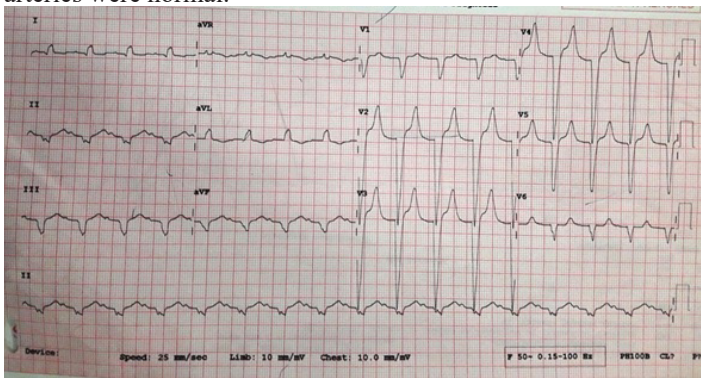


Figure 1: A four-channel 12 lead electrocardiogram showing sinus rhythm, left axis deviation, PR interval of 200msec, poor R wave progression over the precordial leads and complete left bundle branch block with QRS duration of 160 msec.

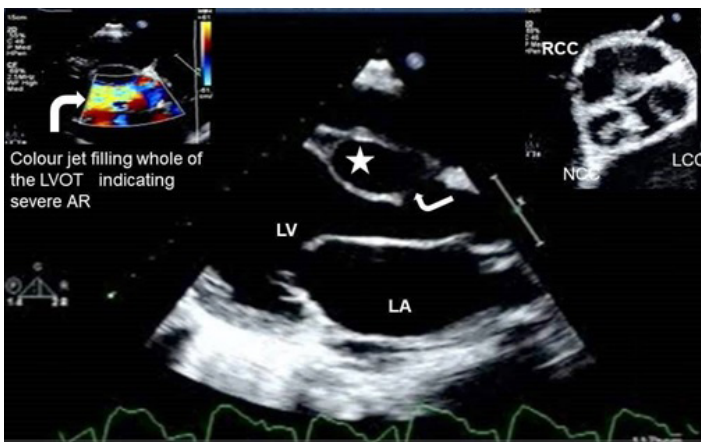


Figure 2: Parasternal Long axis view is showing dilated Left atrium, dilated left ventricle along with an aneurysm burrowing into the interventricular septum (*) with the rent from the aortic sinus shown by the arrow. The upper left image is showing the color Doppler across the left ventricular outflow tract indicating severe aortic regurgitation. The upper right image is showing the three cusps of the aorta in diastole, leading to incomplete coaptation causing aortic regurgitation. Abbreviations: LA – left atrium, LV – left ventricle, RCC- right coronary cusp, LCC – left coronary cusp, NCC- noncoronary cusp, LVOT – left ventricular outflow tract, AR- aortic regurgitation.

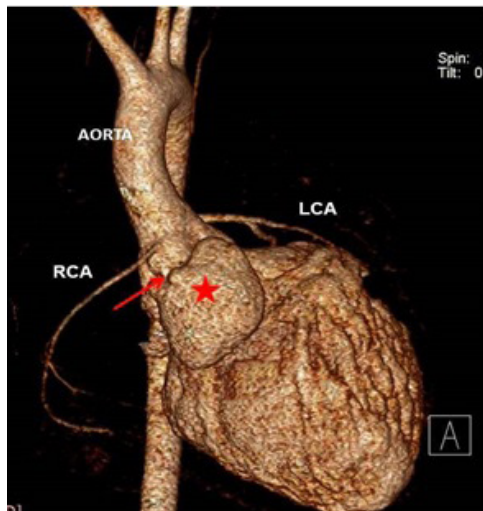


Figure 3a: Three dimensional reconstructed computed tomography image of the left ventricle and the aorta. The sinus of Valsalva aneurysm is seen (*) arising just below the origin of the right coronary artery (indicated by the closed arrow). The coronaries were normal. Abbreviations: RCA – right coronary artery, LCA – left coronary artery.

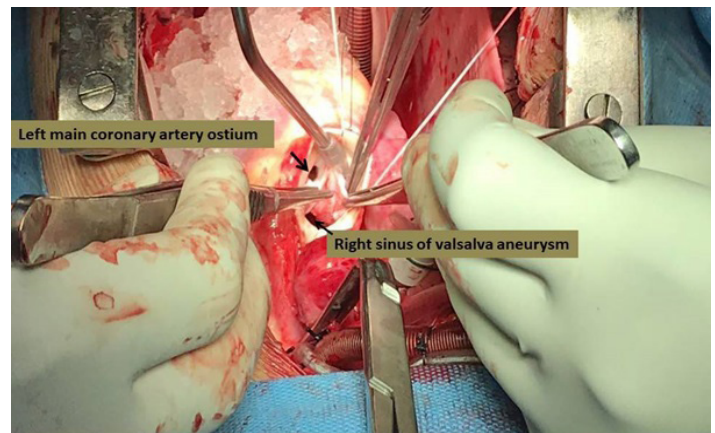


Figure 3b: Intraoperative image (en face view) of the aortic valve showing the left main coronary artery ostium and the opening of the Right sinus of Valsalva aneurysm into the interventricular septum.

Operative findings showed SVA arising from right sinus dissecting into the interventricular septum, distorting the aortic annulus and the valve causing severe AR (Figure 3b). There was dilation of both mitral and tricuspid annulus. A Poly Tetra Fluoro Ethylene (PTFE) patch was used to close the sinus and the aortic valve replaced with 18mm ATS Open Pivot® mechanical valve. A 30 mm Profile 3D® annuloplasty ring (Medtronic) was placed to repair the dilated mitral annulus, whereas for the tricuspid valve bi-cuspidation was done. Postoperatively, he had renal failure requiring hemodialysis. He made a gradual recovery and subsequently discharged after a hospital stay of 40 days.

Discussion

Sinus of Valsalva Aneurysm (SVA), was first described by Hope et al. in 1839 [1]. The congenital SVAs result from failure of fusion of the distal halves of the bulbar septum [2]. The acquired can result from trauma, infective endocarditis, and Behcet disease [3]. Dissecting Aneurysm of the Interventricular septum (DAIS) arises from the right sinus and is a rare entity among the SVAs [4,5].

SVAs usually remain asymptomatic and are detected incidentally. They predominantly rupture into right-sided chambers [6]. Rupture into an intracardiac chamber can be quiescent initially but later can manifest as progressive heart failure due to shunting and AR [6].

The mechanism of AR is the absence of adequate support to the aortic cusp secondary to the extension of aneurysm into the interventricular septum [4]. The increased turbulence and loss of laminar flow will eventually lead to the annular deformity and dilation resulting in further aggravation of the regurgitation (viz. AR begets AR) [4]. The DAIS can produce conduction disturbances varying from the first-degree block to complete heart block requiring permanent pacemaker insertion. The proposed

mechanisms are a mass effect and localized inflammation [4].

Echocardiography is the investigation of choice in diagnosing SVAs [6]. The usual finding is a presence of continuity of the echolucent space with the upper septum and the aortic root at the level of the right aortic sinus [6]. An ECG gated contrast-enhanced multislice CT imaging provides better spatial resolution [7]. The advantages of cardiac MRI include the ability to evaluate the hemodynamics, identify the AR, quantify the shunt or turbulent or fistulous blood flow [8].

The management of SVAs focuses on restoring the function of the aortic valve, the integrity of the ventricular septum, to decrease the size of aneurysm, thereby to improve the cardiac function [5]. Surgery used to be the mainstay of treatment however percutaneous approach is available in select cases. The options are percutaneous catheter closure, direct suture repair, patch closure, endoaneurysmal repair and aortic valve-sparing operation (remodeling) [5,9].

Lilleihei and colleagues have reported the first successful surgical repair of SVA [9]. Unruptured SVAs, though asymptomatic needs early intervention to prevent future complications. Unruptured symptomatic aneurysms require an urgent surgical repair [5]. Conservative treatment is possible if AR is absent or mild and if there is no further increase in the size of aneurysm [5].

DAIS has a progressive course and poor prognosis with the perioperative death of 3.9%. The perioperative complications include obstruction of the ventricular outflow tracts, arrhythmias, complete heart block and communication with the left ventricle resulting in intractable heart failure.

Our patient had complete LBBB secondary to the aneurysm

dissecting into the interventricular septum, severe AR secondary to distortion of the aortic annulus and refractory heart failure.

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