

Unruptured multi-compartmental left sinus of Valsalva aneurysm with chronic dissection of inter-atrial septum and left ventricular free wall

Amarnath C¹, Sathyan G², Periakarupan A³, Ganga Devi R⁴, Kanimozhi D⁵

Abstract:

The sinus of Valsalva aneurysm (SVA) is a small dilatation caused by a separation between the aortic media and annulus fibrosus. A 30-year-old female presented with complaint of pain abdomen. She was evaluated with CT abdomen, scanogram showed ventricular wall calcifications. There was history of intermittent palpitation and exertional breathlessness. On further evaluation with echocardiogram and 64 slice CT scanner, it turned out to be unruptured aneurysm arising from the left coronary sinus. It is a unique aneurysm, which bifurcated into superior part giving origin to the left coronary artery and inferior part dissecting as a complex multilobated aneurysm. The aneurysm had three components in the groove between left atrium and aortic root, dissecting aneurysm into the inter-atrial septum with thrombus, and dissecting aneurysm into the anterolateral free wall of the left ventricle, protruding into the lumen.

Keywords: Sinus of Valsalva aneurysm, Coronary sinus, Aortic root, Chronic dissection, Ventricular wall aneurysm.

¹Professor and Head,

²Associate Professor,

⁴Assistant Professor,

⁵Resident, Department of Radio diagnosis, Stanley medical college, Chennai 600001, India

³Consultant Radiologist, Scans world research & education institute, Chennai, amarrd02@yahoo.co.in

Introduction

Sinus of Valsalva is dilatations at the aortic root wall that arise between the aortic valve annulus and the sinotubular ridge. Each sinus is associated with a corresponding right, left or non-coronary aortic valve cusp (1-2). The right and left main coronary arteries arise simultaneously from their respective developing sinuses. Aneurysm of sinus of Valsalva is rare and may be congenital or acquired. They are most common in right coronary sinus followed by non-coronary sinus, left coronary sinus. The rupture of Sinus of Valsalva aneurysm (SVA) causes fatal outcome.

Case History

A 30-year-old female presented to our department with complaints of right loin pain. She was subjected to CT abdomen which showed right renal calculus (4.1 mm), no hydronephrosis, and no ureteric calculus. No significant abnormality noted in other abdominal organs. Scanogram showed cardiac calcification.

On further detailed history, she had intermittent palpitation and exertional breathlessness. She revealed no history of chest pain, orthopnoea, pedal oedema,

abdominal distension or hypertension, trauma, long term medications. General examination was unremarkable, no features suggestive of connective tissue disorders. On systemic examination, no evidence of thrill, heave, systolic / diastolic murmur, added sounds. Abdomen examination showed no hepatomegaly. Electrocardiogram showed sinus tachycardia, otherwise normal.

She was further evaluated with 2D-Transthoracic Echocardiogram which showed normal left and right ventricular function, LVEF – 58%, no regional wall motion abnormality. The left aortic sinus was dilated.

A complex multilobated aneurysm arising from left aortic sinus was noted. As shown in the echo images in Fig. 1, there were three components. One small component without thrombus was seen between the left atrium and the root of aorta. It measured 4.5 cm x 2.5 cm. Another sac measuring 3.5 cm x 3.5cm, with thrombus was seen dissecting into the inter-atrial septum. The largest component measuring 5.5 cm x 4.5 cm with a small luminal thrombus was seen dissecting into the anterolateral free wall of the left ventricle and projecting into the lumen.

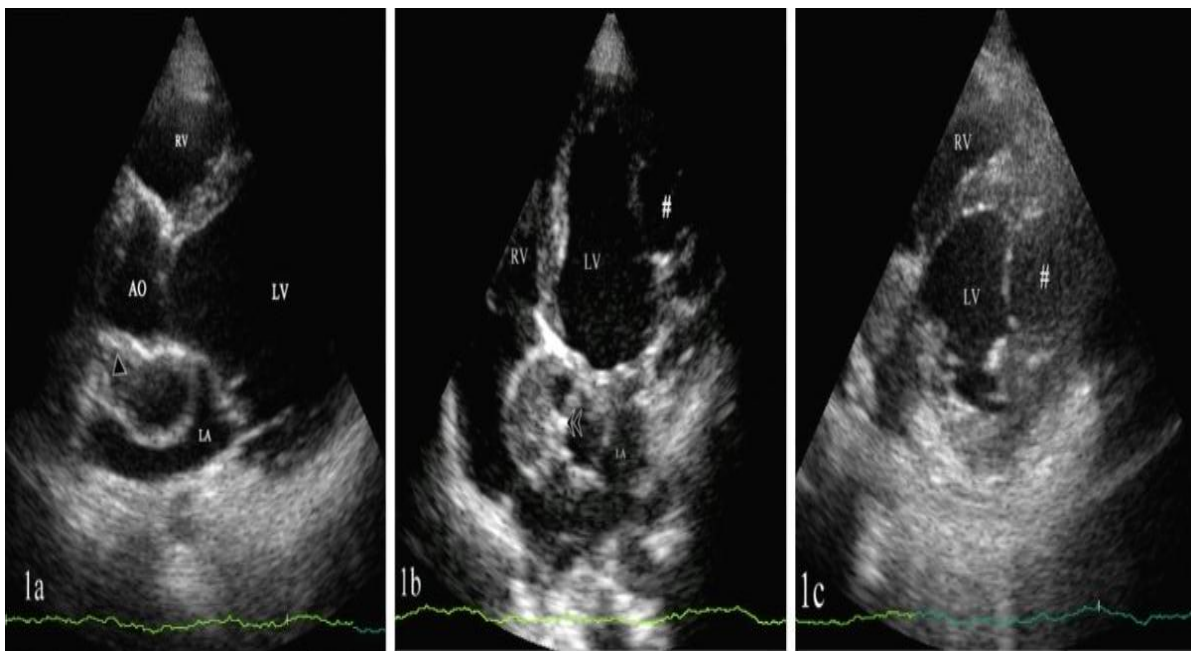


Fig. 1: Trans-thoracic Echocardiogram

a. parasternal long axis view, b. four chamber view, c. two chamber view. ►-aneurysm component in the groove between aorta and left atrium, « - component of aneurysm dissecting into the inter-atrial septum with thrombus, # - aneurysm component dissecting into the left ventricular free wall. LA - left atrium, RA - right atrium, LV - left ventricle, RV - right ventricle, AO – aorta

To further characterise the lesion, to know the size, extent, thrombus, she was further evaluated with a 64 slice CT scanner (Philips Brilliance 64). CT was done with 476 mA, 120kV, 0.3mm slice thickness, 80ml of intravenous iohexol used as contrast material.

The right aortic sinus, non-coronary aortic sinus, right coronary artery, ascending/ descending thoracic aorta, aortic arch, right ventricle were normal. There was abnormal dilatation of left sinus of Valsalva.

The sinus of Valsalva aneurysm bifurcated, superior part giving origin to the left coronary artery and inferior part dissecting as a complex multilobulated unruptured sinus of Valsalva aneurysm. The aneurysm had three components as shown in the Fig. 2.

1. In the groove between left atrium and aortic root,
2. Dissecting into the interatrial septum with thrombus, and
3. Dissecting into the anterolateral free wall of the left ventricle, protruding into the left ventricular lumen.

Fig. 3 demonstrates the aneurysm from the reconstructed MDCT image. Of the above mentioned components, the first and the second components originated from a narrow neck of 4 mm length and 3 mm width. The first multilobulated component measuring 4.8 cm x 1.3 cm x 2.8 cm, was seen tracking posteriorly and superiorly, into the groove between left atrium and aortic root. The second component measuring 3.5 x 2.7 x 3.6 cm with a large thrombus, was seen dissecting into the interatrial septum.

The third component was the largest measuring 5.7 cm x 4.3 cm x 4.5 cm, with a neck of 8.2 mm, was seen dissecting into the anterolateral free wall of the left ventricle protruding into the lumen. It contained a thrombus of in the anterosuperior aspect and calcifications in the wall. No shunt was present.

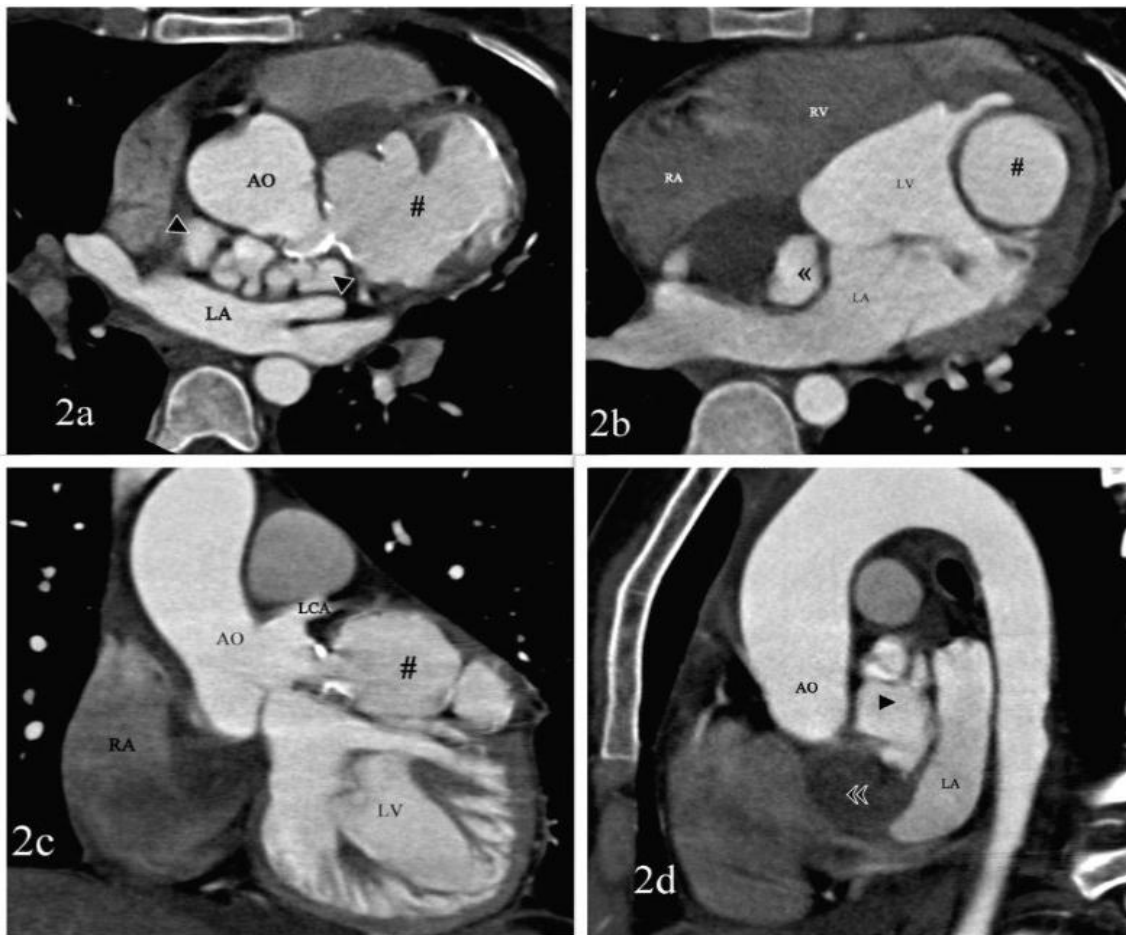


Fig. 2: Computed Tomographic Images

a, b - axial; c, d- multiplanar reconstructed images showing: ▶ - aneurysm component in the groove between aorta and left atrium, « - component of aneurysm dissecting into the interatrial septum with thrombus, # - aneurysm component dissecting into in the left ventricular free wall with calcifications in the wall and thrombus in anterosuperior aspect, LCA - Left coronary artery, LA - left atrium, RA - right atrium, LV - left ventricle, RV- right ventricle, AO – aorta.

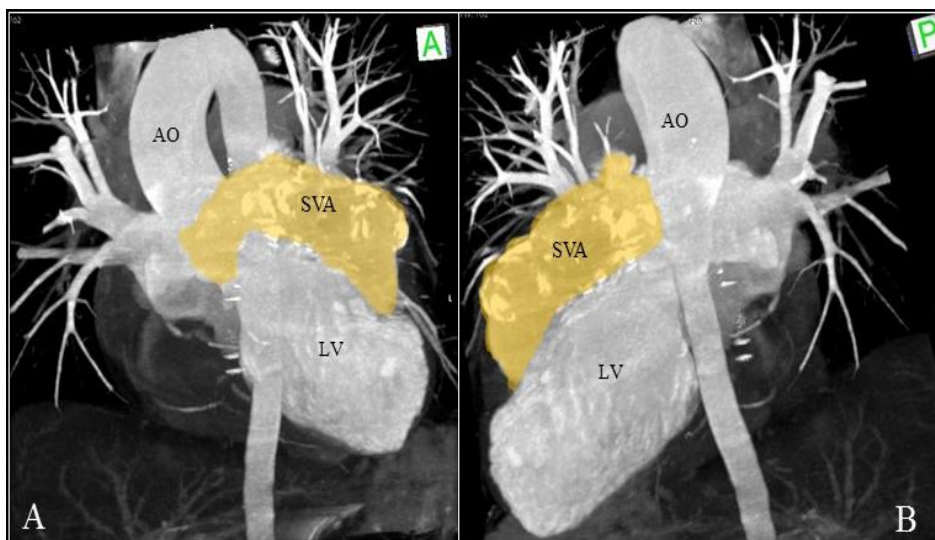


Fig. 3: CT - volume rendered MIP images showing the SVA.LV - left ventricle, AO - aorta

Conventional catheter coronary angiogram was then advised for assessing the flow character of the lesion and presence of any shunt. Patient was subjected to catheter directed selective coronary angiogram which showed lobulated 'wind sock' like lesion originating from the left coronary sinus as shown in Fig. 4. Left coronary artery was originated from a superior out pouching of the left coronary sinus. No significant disease noted in both the coronary arteries.

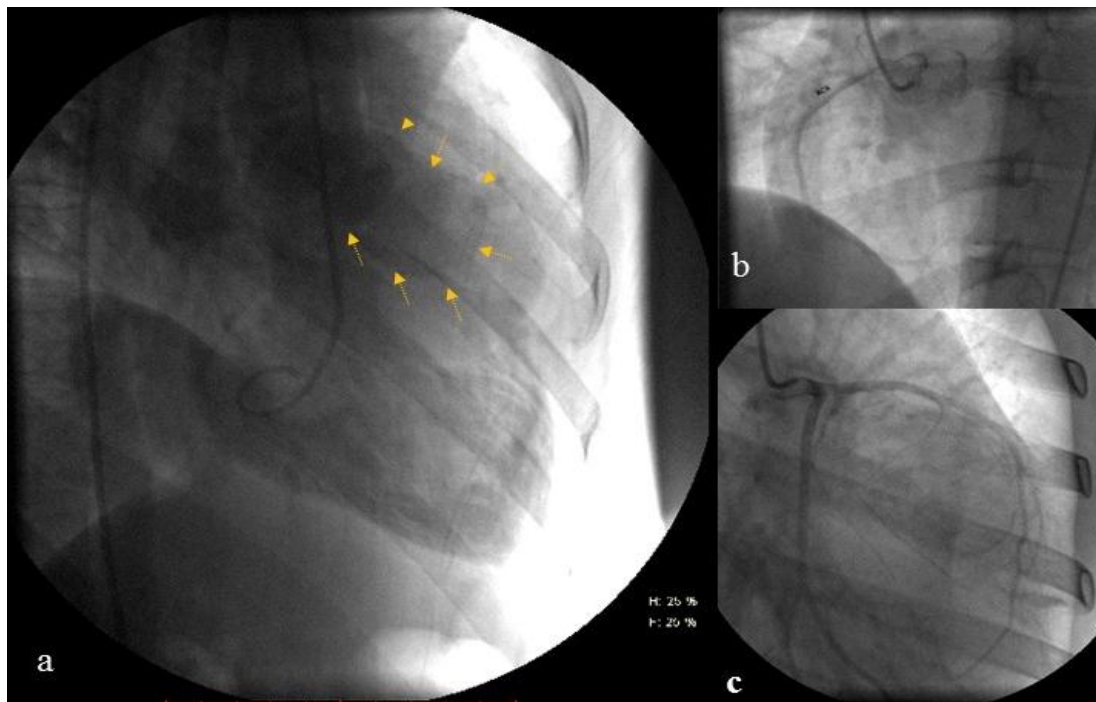


Fig. 4: Conventional catheter coronary angiogram

Spot images: a- windsock like contrast filled structure arising from the root of aorta; b – right coronary artery; c – left coronary artery.

In spite of the need to operate an unruptured aneurysm due to the risk of rupture, sudden cardiac failure and collapse, our patient was not willing for surgery due to the underlying risk. So, she is managed conservatively with Propranolol for tachycardia.

Discussion

Sinus of Valsalva is three subtle dilatations at the aortic root wall that arise between the aortic valve annulus and the Sinotubular ridge. Each sinus is associated with a corresponding right, left or non-coronary aortic valve cusp (1-2). During the 5th week of embryogenesis, left and right truncal swellings develop along the inferior end of truncus, just before septation of the truncus into posterior aortic and anterior pulmonary channels. After septation, cusps of the aortic valve and the pulmonary valve are formed from the three tubercles in ventricular outflow tract. Then, the Valsalva sinuses and leaflets of aortic valve begin to form. The right and left main coronary arteries arise simultaneously from their respective developing sinuses. By the 9th week, formation of the aortic valve leaflets and Valsalva sinuses generally is complete (1-3). Aneurysm of sinus of Valsalva accounts for 0.14 % and very rare (1). Aneurysm are most common in right coronary sinus (72%) followed by non-coronary sinus (22%), left coronary sinus (6%) (1-4).

Aneurysm of the aortic sinus can be either congenital or acquired. Congenital aneurysms are due to localized weakness of the elastic lamina or an underlying deficiency of normal elastic tissue. Common causes of acquired aneurysms included generative conditions (cystic medial necrosis, atherosclerosis), infectious diseases (bacterial endocarditis, syphilis and tuberculosis) and injury (deceleration trauma). They are more common in men (1,5).

Un-ruptured aneurysm may be asymptomatic and incidentally discovered. If symptomatic, it is due to mass effect on adjacent cardiac structures. The most common and dreaded complication of the SVA is rupture. The rupture is most common if the aneurysm is located in the right sinus, followed by non-coronary and left aortic sinus. Rupture may be spontaneous, after trauma, extreme physical exercise or due to endocarditis. Rupture of a SVA occurs principally into the right ventricle. Extra cardiac ruptures are rare, usually fatal(3). Ruptured sinus aneurysm result in an aorto-cardiac shunt and may manifest as insidiously

progressive congestive heart failure, severe acute chest pain with dyspnoea, or, in extreme cases cardiac arrest (6-8).

After treatment, the prognosis is excellent. Thus, prompt and early correct diagnosis is essential (1, 3-4). Most SVA are visualised at echocardiography. ECG gated MR and CT imaging are essential. The former provides valuable functional information, and latter provides excellent depiction of anatomy (2,5).

Medical management involves stabilization of patient clinical condition with medications for heart failure syndrome and peri-operative assessment. Transcatheter closure using Amplatz vascular plug has been performed for ruptured SVA (9). It can also be performed in critically ill patients of ruptured SVA, to avoid sternotomy and cardiopulmonary bypass.

Advanced percutaneous techniques are being performed to correct this condition. But, the treatment of choice is open-heart surgery with or without aortic valve replacement. This procedure carries a mortality of less than 2%. In all patients presenting with rupture, urgent surgical correction should be performed, especially in patients with intra-cardiac shunting. In patients diagnosed to have unruptured SVA, surgical management is recommended to avoid the increased morbidity and mortality (1, 9-10).

We present this case because, aneurysm from the left coronary sinus is rare, presence of multilobulated aneurysm from a single coronary sinus is rare, presence of multi compartmental extension with chronic dissection is hardly reported, coronary sinus aneurysm dissecting into either interatrial septum or left ventricular free wall is hardly reported, and large size of the aneurysms without rupture and without symptoms.

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