CASE REPORT

Atrioventricular and Interventricular Groove and Septal Extension of Right Sinus of Valsalva Aneurysm: A Rare Cause of Complete Heart Block

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Abstract

A 26 years old male presented with vertigo and history of fall. The electrocardiogram revealed 2:1 second-degree heart block and later progression to complete heart block. Transthoracic echocardiography revealed aneurysm at the site of ascending aorta and computed tomographic scan showed an aneurysm of right sinsus of Valsalva extending into right atrioventricular and interventricular groove and causing complete heart block by compression on the conduction system. He also suffered from lymph node tuberculosis. This case report is unique because of rare presentation as complete heart block.

Key Words: *Sinus of Valsalva. Aneurysm. Complete heart block.*

INTRODUCTION

Sinus of Valsalva aneurysm (SVA) was first described by John Thurnam in 1840. Also described by Hope in 1939. It is a rare congenital anomaly. A congenital SVA is usually clinically silent but may present in a variety of ways.[1] Extension of aneurysm into interventricular septum is also a rare complication.[2] Rarely, it can also present as right heart failure and complete heart block.[3] Approximately 65 – 85% of cases originate from the right sinus of Valsalva, while others originating from non-coronary (10 – 30%) and left sinuses (< 5%) are exceedingly rare.[4,5]

Pathologically, there is separation of aortic media and the annulus fibrosus leading to dilation of sinus of Valsalva. A decrease of elastic tissue in the aortic wall and mal-development of bulbus cordis have been associated with the development of aneurysm.[3] Two-dimensional echocardiography is likely to determine a higher incidence of SVA. SVA is more prevalent in Asian surgical series (0.46 – 3.5%) and correlates with more supracristal ventricular septal defects (60.%).[6] The true natural history of SVA is unclear. Clinical complications from SVA are often the initial presentation of SVA. Rupture of SVA is the main cause of death and rarely occurs before the age of 2 years in congenital SVA.[6]

Race differences in SVA are unclear, although a higher frequency was observed in the Asian surgical series. Male-to-female ratio is 4:1, including frequencies of both ruptured and unruptured SVA. Most ruptured SVAs occur from puberty to age 30 years and are often diagnosed or presented clinically at this age.[7]

This report describes this rare anomaly in a young man with an atypical presentation.

CASE REPORT

A 26 years old male, previously of good health presented in March 2009 with a history of vertigo and fall. On admission, he was clinically in 2:1 second degree heart block and developed tachy-brady rhythm after giving atropine. He developed complete heart block during the process of investigation (Figure 1). At the time of presentation, he was vitally stable with a blood pressure of 110/60 and a pulse of 75 beats per minute. Auscultation was unremarkable for any positive finding. A temporary pacemaker was implanted at the time of admission. Chest radiograph revealed widening of lower right heart border. Transthoracic echo revealed aneurysm at the site of ascending aorta. It also revealed moderate left ventricular dysfunction and mild aortic regurgitation and tricuspid regurgitation. Computed tomographic chest scan revealed evidence of aneurysm arising from the right aortic cusp. The aneurysm showed widening and bilobed expansion beyond the neck and extending into the interventricular and right atrio-ventricular groove and interventricular septum (Figure 2) the expanded portion of the aneurysm measured 4.8 into 4.0 centimeters and the neck portion of the aneurysm measured 1.0 centimeter. The right coronary artery was arising from the neck portion of the aneurysm. The left coronary was not involved. The aneurysm was also compressing the aortic valve causing its narrowing. Left heart catheterization was also performed and revealed huge aneurysmally dilated right coronary cusp

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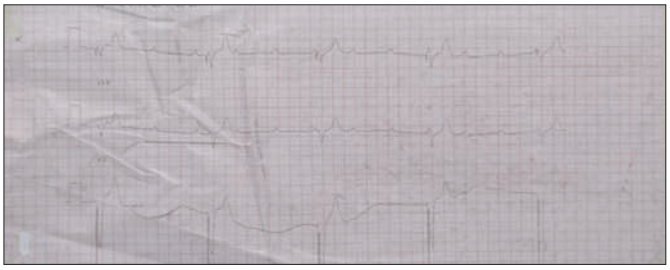

Figure 1: Showing complete heart block.

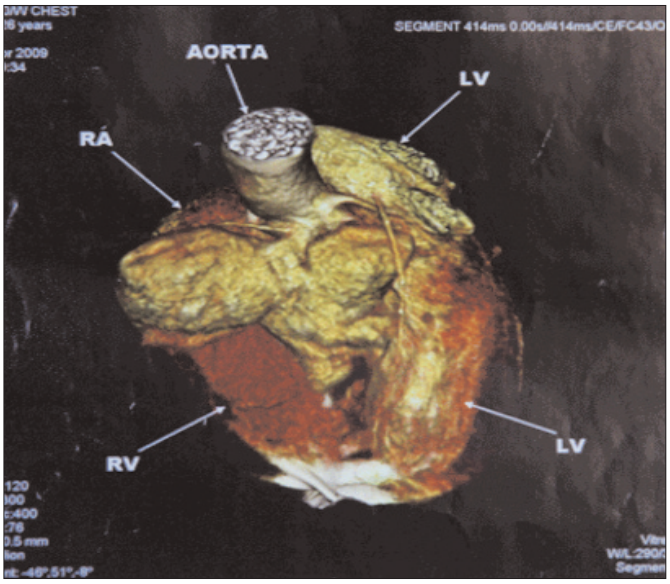

Figure 2: Showing CT angiogram with extensions of abnormal sinus of Valsalva aneurysm.

with evidence of clot in it compromising filling of right coronary artery.

Surgery was contemplated. Peroperatively, aneurysmal dilatation at the sinus of right coronary cusp extending upto the interventricular and atrioventricular groove was seen, causing deviation of the interventricular septum towards the right ventricle. Aneurysmectomy and patch closure with Dacron was performed. Surgery was uneventful.

DISCUSSION

Causes of SVA include a congenital anomaly associated with other congenital defects such as ventricular septal defect and aortic insufficiency. Secondary causes include atherosclerosis, syphilis, cystic medial necrosis or Marfan syndrome, blunt or penetrating chest injury and infective Endocarditis. Rare association with polycystic kidney, Behcet's disease, and tuberculosis has also been reported.[7,10]

Diagnosis is established by Cine magnetic resonance imaging (MRI) and multiplane transesophageal echocardiography (TEE) and two-dimensional transthoracis echocardiography (TEE) may detect as many as 75% of all patients with SVA. Usually TEE or MRI is needed to confirm the diagnosis and for perioperative assessment. Electrocardiogram usually presents with sinus tachycardia, although conduction defects may occur.[8]

Medical management usually involves stabilization (e.g., optimizing medications for heart failure syndrome) and perioperative assessment and management.[7] Aggressive surgical correction of unruptured SVA is often recommended because of its association with increased morbidity and mortality.[6] A review of the literature revealed very rare case reports of SVAs arising from the right sinus of Valsalva and extending into interventricular and atrioventricular groove and interventricular septum.[2,9] The most common clinical manifestation of SVA is rupture into the right ventricle, although rupture into the right atrium, left ventricle, interventricular septum, pleural space, pulmonary artery, and pericardium can also occur.[10] The pattern of presentation does, however, vary widely, and unruptured aneurysms are often 'silent' or present only with vague non-specific symptoms. Cardiac conduction disturbance due to SVA can occur at several levels including sinoatrial conduction disruption and various levels of his bundle block. Transient atrioventricular block and persistent complete atrioventricular block are also reported.[8] In this patient, the unruptured right SVA had extended into the interventricular septum and through transient compression, had compromised normal atrioventricular (A-V) node/his bundle function resulting in complete heart block. This presentation is seen rarely and there are hardly few cases reported in literature in the past.

We conclude from this case and a review of the literature that an unruptured sinus of Valsalva aneurysm presenting as complete heart block and showing possible association with tuberculosis is very rare and merits reporting. This patient also had multiple lymph node enlargement in right axilla and the biopsy revealed chronic granulomatous inflammation consistent of tuberculosis that could have caused this problem although not proved unequivocally. We were not able to prove association as no tissue was preserved for histopathological examination at the time of surgery.

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