

CASE REPORT

Valsalva Sinus Aneurysm of the Non-Coronary Cuspid Presenting as Cyclops Sign on Multi-Detector Computed Tomography

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Key Words

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An 8-year-old girl with Williams syndrome was found to have a heart murmur. Cardiac CT demonstrated a Valsalva sinus aneurysm (VSA) of the non-coronary cuspid, with left atrium (LA) indentation, resembling the face of Cyclops on a coronal reformatted image. Williams syndrome is related to some congenital disorders with interruption of the tunica media from the aortic root to the annular fibrous ring or aortic valve. However, it rarely presents at birth. The sinus of Valsalva dilates as time goes by, due to the persistent striking force from the left ventricle (LV). However, it is silent until rupture when cardiac tamponade occurs. The typical imaging appearance of VSA is of a saccular shape and originates above the aortic root, but sometimes involves the entire sinus. Although lethal complications of VSA occur without warning, prophylactic surgical intervention remains controversial. Regular imaging follow-up is advised before complications occur.

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1. Introduction

Valsalva sinus aneurysm (VSA) is a rare structural disease of the heart. It is usually asymptomatic until complications occur. Traditionally, trans-thoracic echocardiography provides a baseline modality for detecting the aneurysm, and cardiac catheterization is used as the definite

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diagnostic tool.¹ Much literature has reported that multi-detector computed tomography (MDCT) with multi-plane reformation (MPR) provides better spatial and temporal resolution in depicting the size and location of a VSA. It also provides the detailed anatomic relationship between the aneurysm and its adjacent structures. We report a case of VSA of the non-coronary cuspid, which has an impressive picture resembling a mythology character (Cyclops) on its coronal reformatted image (CRI) by MDCT.

2. Case Report

An 8-year-old girl with history of Williams syndrome received regular follow-ups in the pediatric cardiology department. She did not have the expected symptoms or signs such as shortness of breath or chest tightness, but instead had progressive grade II–III systolic murmur. Contrast-enhanced cardiac CT with electrocardiography-gated scan and breath-hold technique demonstrated a saccular aneurysm (14.5 mm × 14.2 mm × 12.5 mm) at the Valsalva sinus of the non-coronary cuspid with posteroinferior protrusion. The aneurysm made an indentation into the left atrium (LA), which formed a circular shape on CRI. Combining the left upper pulmonary vein, confluence of right pulmonary veins, lumen of the LA and mitral valve, the image looked like the face of “Cyclops” (Figures 1A and 1B), a famous mythology character. In this study, we also found left ventricular hypertrophy, a small caliber of aorta, and a Kommerell’s diverticulum at the distal aortic arch (Figure 2). Since the patient was asymptomatic and the size of the aneurysm was relatively small, she did not receive further management, but maintained regular follow-up in our outpatient department.

3. Discussion

VSA is an uncommon cardiac disorder and can be congenital or acquired. The age of occurrence ranges from 4 days to 96

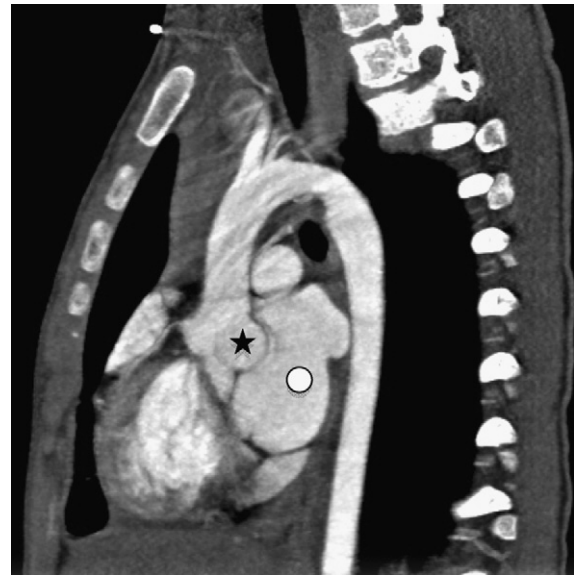


Figure 2 The sagittal reformatted image demonstrates the relationship between the aneurysm (star) and left atrium (LA) (circle).

years old. It occurs more frequently in the oriental male, and the prevalence is about 0.09% in the general population and 0.1–3.5% in patients afflicted with other congenital heart diseases (ventricular septal defect = 30–60%; bicuspid aortic valve = 10%).^{2,3} VSA can arise from any cuspid of the aortic root, in a decreasing order of right coronary cuspid (72%), non-coronary cuspid (22%) and left coronary cuspid (6%).² In acquired aneurysms, it is often associated with the condition that compromises the elastic connective tissue at the junction between the aortic media and annulus, such as infections (syphilis, bacterial endocarditis or tuberculosis), or deceleration trauma. In the congenital situation, it is related to the developmental interruption or weakness between the lamina media of the

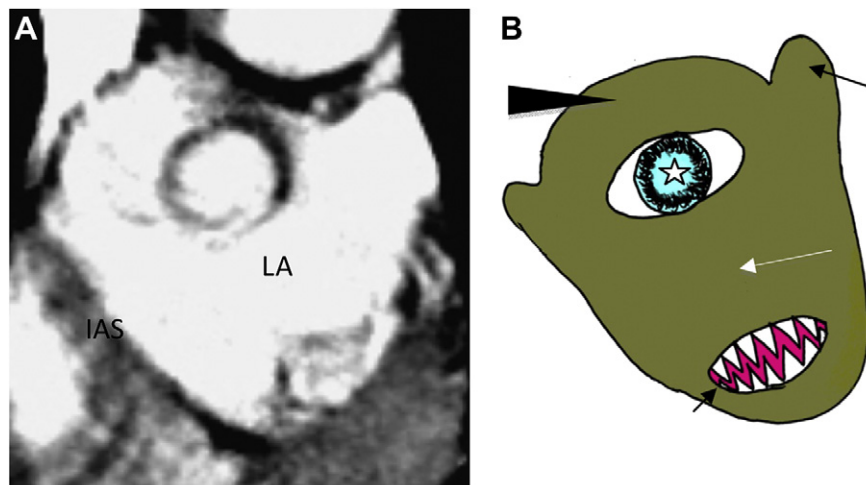


Figure 1 (A) The coronal reformatted image by multi-detector computed tomography demonstrates the Valsalva sinus aneurysm of the non-coronary cuspid; (B) the combination of circular indentation of the Valsalva aneurysm (star), the left upper pulmonary vein (long black arrow), lumen of the LA (white arrow), confluence of the right pulmonary veins (arrowhead) and mitral valve (short black arrow) resemble the face of Cyclops. LA = left atrium; IAS = interatrial septum.



Figure 3 The facial morphology of this patient shows steep mandible angles and a flattened nasal bridge.

aortic root and the annular fibrous ring of the aortic valve, as is commonly found in some connective tissue disorders (Ehlers–Danlos syndrome or Marfan syndrome).⁴ Unruptured VSA is usually silent and found incidentally by image studies when the patient is suspected of having other diseases. The most common clinical sign is cardiac murmur, and some patients may complain of dyspnea or chest tightness. In the case of rupture, the symptoms and signs depend on the size of the VSA, and the rapidity of rupture of the VSA, and the cardiac chamber with which the VSA communicates. Common presentations include severe dyspnea, sudden chest pain or hemodynamic collapse.² Aortic regurgitation is the most common complication of both ruptured and unruptured aneurysms.³ Other complications include arrhythmia, myocardial ischemia, right ventricular outlet obstruction, and interventricular septum

dissection in unruptured aneurysm and aorto-atrial or aorto-ventricular fistula in ruptured aneurysm. Cardiac tamponade is the most lethal complication, but it rarely occurs.^{5,6} Although VSA can occur congenitally, it rarely presents at birth. The sinus of Valsalva progressively dilates as time goes by due to the persistent striking force from the LV. Therefore, the aneurysm during childhood is relatively small and may be difficult to identify. In this case, we found the “Cyclops sign” in a patient with VSA of the non-coronary cuspid, which is formed by the circular indentation of the aneurysm, left upper pulmonary vein, confluence of right pulmonary veins, lumen of the LA and mitral valve.

Although this disease has many complications, including congestive heart failure, aortic regurgitation, and myocardial ischemia, prophylactic surgical intervention of an asymptomatic unruptured sinus valsalva aneurysm remains controversial. Surgery is indicated for complicated unruptured VSA, such as right ventricular outlet obstruction, arrhythmia, and interventricular dissection. Ruptured aneurysm is also an indication for surgical repair owing to congestive heart failure. For small aneurysms, regular follow-up by MR or CT is mandatory (Figure 3).

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