

Right Sinus of Valsalva Aneurysm Causing Extrinsic Coronary Compression

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Sinus of Valsalva aneurysm is a rare cardiac disorder. It is more frequently a congenital, but it may also be an acquired condition. Takayasu's disease is an extremely rare cause of this disorder. Most cases of unruptured sinus of Valsalva aneurysm are asymptomatic. Compression of the left coronary artery is an unusual manifestation of the disease that can cause angina, myocardial infarction or death. This report describes a 19-year-old black male with an unruptured right sinus of Valsalva aneurysm caused by Takayasu's disease and manifested by an acute coronary syndrome, successfully treated by surgery.

Introduction

Sinus of Valsalva aneurysm (SVA) is a rare cardiac disorder, occurring five times more often in Asians than in the western population¹. It is more frequently a congenital, but it may also be an acquired condition. Congenital SVA can be caused by the absence of muscular and elastic tissue in the aortic wall behind the sinus of Valsalva or a lack of continuity between the aortic media and the aortic valve annulus². The acquired form can be caused by trauma, infections (bacterial endocarditis, syphilis, tuberculosis), degenerative disease (atherosclerosis), systemic inflammatory diseases (Behçet's disease, ankylosing spondylitis) and connective tissue disorders (systemic lupus erythematosus, Marfan syndrome). Takayasu's disease, an inflammatory arteritis involving large vessels, is an extremely rare cause of this disorder, with only one previously reported case³. Most cases of unruptured sinus of Valsalva aneurysm are asymptomatic and difficult to diagnose. Compression of the left coronary artery is an unusual manifestation of the disease and can cause acute coronary syndrome⁴. This report describes a 19-year-old man with an unruptured right sinus of Valsalva aneurysm caused by Takayasu's arteritis and manifested by ischemic coronary disease successfully treated by surgical repair.

Key Words

Aneurysm; sinus of Valsalva; heart defects, congenital/surgery; Takayasu's disease.

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Case report

A 19-year-old black male was in good health until three years ago when he presented with recurrent headache, fever, fatigue and positive inflammatory markers, bilateral renal artery stenosis and renovascular hypertension. He was submitted to right percutaneous transluminal renal artery angioplasty and to open left renal artery revascularization with success. The diagnosis of Takayasu's arteritis⁵ was confirmed at that time. During clinical follow-up, antihypertensive drugs were withdrawn and he was treated with prednisone.

He was admitted to our institution with a 2-month history of chest pain episodes, suggestive of angina pectoris, dyspnea and palpitations. On physical examination, an ejection systolic murmur over the superior left parasternal area was heard. The laboratory data, including cardiac enzyme levels, showed no abnormalities. The chest x-ray was normal. The ECG showed sinus rhythm and right bundle branch block. Holter ECG revealed bradycardic sinus rhythm alternating with atrial ectopic rhythm. The transthoracic three-dimensional echocardiogram demonstrated an unruptured right sinus of Valsalva aneurysm with a neck of 19mm and normal ventricular function. A multi-slice spiral CT revealed a huge right SVA measuring 70 mm x 40 mm, with a neck of 16 mm, causing compression of the proximal portion of the anterior descending coronary artery (fig. 1A, 1B) and the stress myocardial perfusion MRI test revealed anterior myocardial ischemia. The patient was referred to surgery.

The aortotomy disclosed the orifice of the aneurysm's neck, measuring approximately 20 mm in diameter. It was identified in the right coronary sinus near the right coronary ostium. The aortic valve had a normal appearance. The surgery consisted of the closing of the Valsalva aneurysm neck with a bovine pericardial patch (fig. 2A, 2B) and a coronary artery bypass grafting operation using the internal mammary artery to the left anterior descending coronary artery (LAD). The postoperative was uneventful and the patient is asymptomatic 6 months after surgery. The control CT revealed reduction of the aneurismal sac, decompression of the coronary artery (LAD) and a permeable internal mammary artery (fig. 1C, 1D).

Discussion

This report describes an extremely rare association. An unruptured right SVA manifested by chronic coronary syndrome in a young patient with Takayasu's disease is an association that has never been reported before.

Takayasu's arteritis is a chronic inflammatory disease of unknown etiology that can produce stenosis, occlusion, or aneurismal degeneration of large arteries, predominantly



Figure 1 - Multi-slice spiral CT; A - Pre-operative volume rendering image of a large sinus of Valsalva aneurysm associated with circumferential thrombus (70mmX40mm) causing extrinsic compression of the proximal portion of the LAD; B - Post-operative image of the LAD without the SVA compression. C - Sagittal projection showing the right SVA and the compression of the LAD; D - Post-operative image of the coronary bypass without the SVA.

the aorta and its major branches. It is an extremely rare acquired cause of SVA, with only one previously reported case in the literature³.

SVA is a rare cardiac disorder with a wide clinical spectrum¹. The aneurysm can grow in close relation to the cardiac

chambers and ruptures more commonly into the right ventricle or right atrium, or rarely into the left ventricle or pulmonary artery. Most cases of ruptured SVAs are symptomatic, thereby causing congestive heart failure and arrhythmias. The free rupture into the pericardium is a catastrophic event that causes

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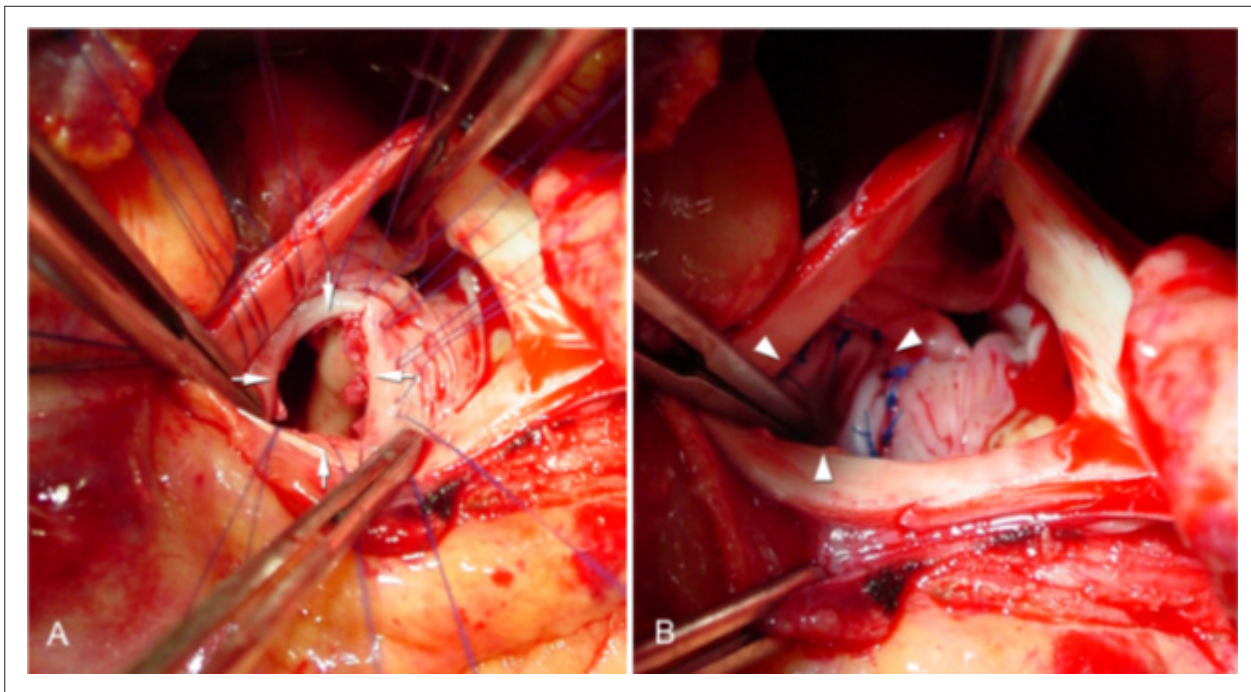


Figure 2 - Intra-operative pictures; A - Closure of the aortic orifice of the right sinus of Valsalva aneurysm (arrows); B - Closure performed with a bovine pericardial patch with separate 5-0 prolene sutures (arrowheads).

sudden death by cardiac tamponade. Unruptured SVAs are in general asymptomatic and difficult to be diagnosed. They can occasionally cause heart block, obstruction of cardiac outflow and even compress the coronary arteries, causing myocardial infarction or angina, which was, in fact, the clinical complaint of this particular patient⁴. In this case, the right SVA was compressing the LAD causing angina. In contrast, in the great majority of the reports, coronary insufficiency is caused by coronary artery compression by a left SVA.

Traditionally, diagnosis has been made with accuracy by echocardiography, either transthoracic or transesophageal, or by cardiac catheterization. More recently, multi-slice spiral CT and MRI have been used with increasing frequency. Besides being non-invasive methods, they give a three-dimensional picture of the aneurysm and allow a more accurate surgical planning. This includes the possibility of evaluating the correlation between the anatomy and patency of the coronary arteries and the aneurysm⁶.

Early surgical repair of SVA is the treatment of choice in symptomatic patients and in ruptured aneurysms. Optimal management is less clear for asymptomatic unruptured aneurysms. Operative risk is low and the long-term outcome is good. Direct closure may cause or worsen aortic regurgitation by deformation of the aortic annulus and may also be responsible for recurrences. Therefore, patch closure is recommended, even when the orifice is small, as was done in this case. Other incisions (atriotomy,

ventriculotomy) may be performed when there is rupture of the aneurysm into other cardiac chambers or when there are associated anomalies. Aortic valve replacement or valvuloplasty may be necessary when there is aortic valve regurgitation⁷. Coronary artery bypass grafting may also be used in cases of SVA associated with coronary compression, even though there have been reports of successful treatment with SVA repair alone [4], which should have been done in this particular case once the control multi-slice CT revealed no coronary compression after the SVA treatment. Nevertheless, the coronary bypass grafting was performed in order to guarantee a distal myocardial perfusion, despite the impossibility of knowing in advance the surgical result of the SVA treatment.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any post-graduation program.

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