Sinus of Valsalva Aneurysm with Rupturing into the Right Atrium — A Case Report and Review of the Literature

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Sinus of Valsalva aneurysm accounts for only 1% of congenital cardiac anomalies. Sinus of Valsalva aneurysm can cause aortic insufficiency, coronary artery flow compromise, cardiac arrhythmia, or aneurysm rupture. A 44-year-old male presented with a 3-month history of palpitation and exertional dyspnea. Cardiac auscultation revealed a grade III/VI “to and fro” murmur along the left sternal border. Electrocardiography showed ventricular bigeminy. Transthoracic and transesophageal echocardiography revealed an aneurysm of the right sinus of Valsalva rupturing into the right atrium. Aortography also confirmed the presence of an aneurysm of the right coronary sinus, extending into the right atrium. The patient underwent open heart surgery to close the ostium of the aneurysm and to suture the right atrial fistula directly. The patient was well upon follow-up after the operation.

Key Words: Sinus of Valsalva aneurysm • Right atrium • Transthoracic echocardiography • Transesophageal echocardiography

INTRODUCTION

Sinus of Valsalva aneurysm, also known as coronary sinus fistula, is a rare cardiac abnormality. Aneurysms, either congenital or acquired, arise from a discontinuity between aortic tunica media and aortic valve annulus. Sinus of Valsalva aneurysms can coexist with other congenital disease and can rupture into the cardiac chambers, causing acute clinical symptoms. The right coronary sinus is the commonest site of aneurysm formation. The majority of right coronary cusp aneurysms rupture into the right ventricle. Rupture of sinus of Valsalva aneurysm can be easily diagnosed by echocardiography. Early diagnosis and surgical treatment can save lives for most of the patients.

We present a rare case of right sinus of Valsalva aneurysm that ruptured into the right atrium. The usefulness of various diagnostic methods, as well as the surgical options, are also discussed.

CASE PRESENTATION

A 44-year-old man with hypertension was admitted with chief complaints of sudden-onset palpitations, light headedness, and breathlessness. On examination, he was aptyrexial, with blood pressure of 166/70 mm Hg and pulse rate of 110 beats per minute. He had grade III/VI “to and fro” murmur along the left sternal border. The electrocardiogram showed sinus rhythm with ventricular bigeminy, and chest X-ray showed cardiomegaly. Full blood count and cardiac enzymes were normal. The patient had a transthoracic echocardiography (TTE) a day later, which showed dilatation of four chambers with good systolic function, and no vegetations or valvular abnormalities. An aneurysmal dilation of the right sinus
of Valsalva protruded into the right atrium with a turbulent-color Doppler flow. Additionally, a systolic-diastolic flow was presented in the continuous-wave Doppler (Figure 1); ruptured sinus of Valsalva aneurysm was highly suspected. Transesophageal echocardiography (TEE) confirmed the presence of a ruptured right sinus Valsalva aneurysm (Figure 2). Urgent cardiac catheterization revealed elevated right heart pressures and a step-up in oxygen saturations at the right atrial level. Ascending aortic pressure was 170/60 mm Hg. The aortogram confirmed that the right sinus of Valsalva aneurysm had ruptured into the right atrium (Figure 3). The patient was referred for surgical repair of the ruptured sinus. The patient recovered completely and has been well for three years till the present time.

DISCUSSION

The three sinuses of Valsalva are located in the most proximal portion of the aorta, just above the cusps of the aortic valve; the sinuses end in the areas of the sino-
tubular junction. Causes of sinus of Valsalva aneurysm are classified into congenital or acquired. Congenital type occurs where a lack of fusion exists between the aortic media and the annulus fibrosis of the aortic valve. Acquired sinus of Valsalva aneurysms can exist in patients with endocarditis, syphilis, injury, Bechet’s
disease and Marfan’s syndrome. Deficiency of the aortic media at the attachments on the aortic annulus portion may progressively lead to dilation of aortic sinus, usually over many years. Therefore, sinus of Valsalva aneurysm can remain subclinical for several years until it eventually ruptures between the 3rd and 4th decades of life. The commonest sites of aneurysm ruptures are cardiac chambers, interventricular septum, or pericardial space. Dilated sinus of Valsalva aneurysm also can lead to distortion of aortic valve and aortic regurgitation.

Un-ruptured sinus of Valsalva aneurysms usually do not cause any symptom and are often found during echocardiogram examination and cardiac catheterization. However, un-ruptured sinus of Valsalva aneurysm may cause right ventricular outflow tract obstruction and myocardial ischemia due to the mass compression effect of aneurysms. Symptomatic aortic regurgitation (AR) can occur in cases with progressive dilation of sinus of Valsalva aneurysm.

The ruptured sinus of Valsalva aneurysms most often involve with the right coronary sinus (76.8 to 83.3%) or with other non-coronary sinus, while an aneurysm of the left sinus is rare. Most right sinus of Valsalva aneurysms rupture into the right ventricle (86.7%), or into the right atrium. Patients may remain asymptomatic for several years even if the sinus of Valsalva aneurysms has ruptured. However, dyspnea and exercise intolerance may develop because of the increased shunting and volume overloading. Sudden massive rupture may result in acute chest pain and dyspnea. Patients with a smaller insidious rupture may be asymptomatic, but also may be associated with progressive symptoms of exertional dyspnea and heart failure. In general, the in-and-out blood flow from a ruptured sinus of Valsalva aneurysm can produces a continuous murmur. A diastolic murmur of aortic regurgitation may be heard if aortic cusp dilation occurs. If physical findings are suggestive of a sinus of Valsalva aneurysm, patients may be evaluated using a combination of TTE, TEE, magnetic resonance imaging and cardiac catheterization. Traditionally, the gold standard for the diagnosis of ruptured sinus of Valsalva aneurysm is cardiac catheterization. The cardiac catheterization is used to detect the ruptured site and to establish the patency of coronary artery. With the advancement of newer-generation echocardiographic machines, TTE and TEE have played an important role in the diagnostic confirmation of Valsalva aneurysm sinus.

TTE and TEE can investigate the proximal aorta, sinuses, aortic valve, and surrounding structures. Doppler findings may identify the exact shunt location. The “windsock” dilatation of sinus of Valsalva aneurysm is the feature of sinus of Valsalva aneurysm. Doppler features include a continuous high velocity flow from the aorta to the heart chamber. The diagnosis usually is confirmed by color Doppler flow mapping. The color Doppler flow mapping may reveal a continuous mosaic flow jet from the aneurysm to the heart chamber. Continuous rotation of a multiplane TEE may be particularly helpful to identify the exact point of rupture. Echocardiography is also useful for the differential diagnosis of other disorders, such as patent ductus arteriosus, coronary fistula, or aortopulmonary window that cause similar continuous murmur. Additionally, echocardiography can provide information for coexisting AR.

No specific medical therapy exists for sinus of Valsalva aneurysm. Treatment of congestive heart failure may be required if rupture of the aneurysm does occur. Additionally, concurrent cardiac arrhythmias and infectious endocarditis should be treated. Surgical repair of the ruptured sinus of Valsalva aneurysm is the definitive treatment. In 2001, Vural et al. reported the therapeutic categories for sinus of Valsalva aneurysm. Surgical repair was required for both ruptured sinus of Valsalva aneurysm and for symptomatic unruptured aneurysm. If the size of the unruptured aneurysm is greater than 50% of the other two normal coronary sinuses or is increased in the consecutive echocardiographic examinations, surgical intervention also should be considered. Surgery is also required for the unruptured aneurysm compressing to the adjacent tissues.

Surgical treatments for sinus of Valsalva aneurysm include direct suturing of the aneurysm or suturing with a patch. Attempts should be made to preserve the aortic valve, if possible. However, aortic valve replacement may be necessary when AR is significant. Patients receive surgical repair for sinus of Valsalva aneurysm usually have low operative risk and high long-term survival rates. Therefore, an early surgical intervention should be encouraged.
REFERENCES


右主動脈竇動脈瘤破裂至右心房
—病例報告及文獻回顧

張釗監1、秦志輝1、陳孟麟1、陳瑞雄2、羅鴻舜1
台北市國泰綜合醫院 心臟內科1 心臟外科2

主動脈竇動脈瘤是少見的，大都破裂後才被發現。一位44歲男性，有3個月心悸及運動時呼吸困難病史，心臟聽診在胸骨左緣聽到“去返性”心雜音，心電圖顯示心室性期外收縮，胸前及經食道超音波發現右主動脈竇動脈瘤破裂至右心房，主動脈攝影確定右主動脈竇動脈瘤破裂至右心房，冠狀動脈攝影是正常的。患者於開心手術中接受動脈瘤開口縫合，患者術後狀況良好。

關鍵詞：主動脈竇動脈瘤、右心房、胸前心臟超音波、經食道心臟超音波。