ORIGINAL ARTICLES

Ruptured Sinus of Valsalva Aneurysm Initially Misdiagnosed as Ventricular Septal Defect by Echocardiography

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ABSTRACT

Background: The sinus of Valsalva aneurysm (SVA) is a small dilatation caused by a separation between the aortic media and annulus fibroses. Its origin may be either acquired or congenital, the right coronary sinus is most frequently affected, with the most common complication being rupture. Clinical cases: We present a case of a 41-year-old male with epigastric pain that deteriorated to acute pulmonary oedema and cardiogenic shock. A grade 4/6 harsh systolic murmur heard at left sterna border, primary clinical diagnosis suspect ventricular septal defect. TTE (Trans-thoracic echocardiography) done revealed left to right shunt suspected rupture sinus of valsalva so TEE (Trans-oesophageal echocardiography) done and confirm the diagnosis. Conclusions: Rupture sinus of Valsalva can be presented by angina pain then deteriorated rapidly into heart failure and cardiogenic shock. Diagnosis was suggested by TTE and confirmed by TEE.

Key words: Sinus of Valsalva, aneurysm rupture

Introduction

Sinus of Valsalva aneurysm is a rare disorder. It is usually congenital, but other origins have been described. The presentation may range from an asymptomatic murmur to acute cardiogenic shock and death. Once ruptured, it often produces hemodynamic instability. Diagnosis should be pursued with transesophageal echocardiography (TEE) or catheterization.

A 41-year-old man was admitted with epigastric pain that deteriorated to acute pulmonary oedema and cardiogenic shock transthoracic (TTE) and transesophageal echocardiography done and revealed ruptured sinus of valsalva at the non-coronary cusp and decided for transfer to tertiary hospital for surgical repair but unfortunately patient deteriorated on the way and unfortunately died.

Case Report:

In October 2011, a 41-year-old man was admitted to our hospital with symptoms suggestive of angina. The patient reported epigastric pain radiated to the back, heaving in nature associated with nausea persistent for three hours with history of twenty days of cough and whitish expectoration.

On physical examination, he was hemodynamically stable with a blood pressure -110/60, heart rate of 100 beats/min, clear chest, auscultation revealed a normal S1 and S2 and a grade 4/6 harsh systolic murmur heard at the left sternal border then the patient deteriorated and went to pulmonary congestion improved with diuretics, blood pressure was 100/60, heart rate 90 beats/min, fine basal crepitation on chest and then the patient had sudden cardiac arrest and ACLS started and came back and mechanically ventilated and started on inotropic support.

Laboratory data revealed high troponin I of 1.6 ug/l then increased to 10.8 ug/l, other lab WBCs 15, Hgb 14.5 mg/dl, platelets 198, total bilirubin 25 umol/l, ALT 1731 U/L, AST 2055 U/L, creatinine 134 umol/l then increased to 433 umol/l, the electrocardiogram showed minimal ST depression 0.5 mm at inferolateral leads.

An echocardiography done at the start and revealed normal systolic function, left to right shunt suspected ventricular septal defect (fig.1,2) then echocardiography repeated and suspected ruptured sinus of valsalva. So TEE done and confirmed the diagnosis of sinus of valsalva aneurysm at non-coronary cusp with rupture (fig.3) with left to right shunt into the right atrium measuring about 1 cm as evidenced by colour turbulent flow passing from the non coronary cusp into the right atrium (RA): (fig.4)
Fig. 1: TTE 2D mode showed an aneurysm with a defect in the non coronary cusp.

Fig. 2: TTE colour mode showed a colour flow from the non coronary cusp to the RA.

Fig. 3: 2D mode showed an aneurysm with a defect in the non coronary cusp.
Fig. 4: TTE colour mode showed a colour flow from the non coronary cusp to the RA just below the tricuspid valve measuring 1 cm.

The patient managed primarily as acute coronary syndrome then with deterioration managed by respiratory support through a mechanical ventilator and inotropic support with noradrenaline infusion then decided to transfer the patient to a tertiary center for urgent surgical repair but unfortunately patient deteriorated and got more hypotension so treated with more inotropic support using dopamine infusion but unfortunately patient not responded and died.

Discussion:

A sinus of Valsalva aneurysm is a rare disorder. It is usually congenital but less commonly is associated with endocarditis, atherosclerosis, trauma, syphilis, or aortic dissection. Males are more affected than females (3:1 ratio) and the prevalence is higher in the Eastern than in the Western population. Patients are usually diagnosed in the 3rd or 4th decade of life, when rupture occurs and symptoms develop. In our case, the patient presented at 41 years of age.

A ruptured aneurysm of an aortic sinus is a major cardiovascular event that demands prompt diagnosis and treatment. Nevertheless, given the relative infrequency of the condition, achieving a definitive diagnosis can be challenging.

Since first reported in the mid-1800s, sinus of Valsalva aneurysm has been described in multiple isolated cases, and the estimated prevalence is less than 1% in patients undergoing open heart surgical procedures. Sinus of Valsalva aneurysms may be asymptomatic, or they may present with angina or with symptoms of valvular insufficiency or outflow obstruction.

Once ruptured, sinus of Valsalva aneurysms may produce serious hemodynamic instability, such as acute heart failure or sudden death. When rupture is suspected, immediate diagnosis should be pursued with transesophageal echocardiography (TEE) or cardiac catheterization.

The right sinus of Valsalva is the most common site of aortic sinus aneurysmal dilatation, followed by the noncoronary sinus; after rupture, a fistulous tract is formed, frequently with the right ventricle in the former instance and with the RA in the latter. Uncommonly, rupture into the pulmonary artery may occur. Aortic regurgitation is present in 18% to 44% of cases and in some patients this is caused solely by a hemodynamic effect (Bernoulli effect) associated with the blood-flow shunt. Our patient had a noncoronary sinus of Valsalva aneurysm and a fistula to the RA.

A diagnosis of aortic sinus aneurysm and fistula can be confirmed by echocardiography, either transthoracic or transesophageal, or by cardiac catheterization (right and left). Transthoracic echocardiography (TTE) can lead to an accurate diagnosis in virtually all of these patients. TEE is useful when TTE is inconclusive. Moreover, TEE may be very helpful for better anatomic definition during preoperative evaluation. Direct visualization of the aneurysmal dilatation and fistulous tract by cardiac catheterization, as well as the measurement of oxygen saturation in different cardiac chambers and the great vessels, can yield a specific diagnosis. Our patient showed an aneurysm at non coronary cusp of sinus of valsalva with a colour turbulent flow from non coronary cusp to the right atrium denoting ruptured sinus of valsalva at the non coronary cusp.

A few patients are misdiagnosed as having isolated ventricular septal defect as in our patient. In this situation, the characteristic Doppler spectrum showing continuous high jet velocity accentuated in diastole as opposed to a high velocity systolic and low velocity diastolic flow which differentiates the two conditions.
The natural history of asymptomatic aneurysm of an aortic sinus is unclear, and variant cases—with rapid clinical deterioration or many years of stabilization—have been described. However, once symptoms develop or rupture occurs, urgent intervention is recommended. Open-heart correction of the aneurysm and fistula, with or without aortic valve replacement, carries a low operative risk and traditionally has been the treatment of choice. More recently, novel percutaneous closure techniques have brought hope of a less invasive method to correct such a condition.

Conclusion:

In summary, we report a case of a sinus of Valsalva aneurysm with rupture into the RA in a patient who presented with anginal symptoms then deteriorated into heart failure and cardiogenic shock. Diagnosis was suggested by transthoracic echocardiography and confirmed by trans-oesophageal echocardiography. But patient died before proceeding to any surgical interference.

References