Ruptured Left Sinus of Valsalva Aneurysm in a Female Nigerian Septuagenarian

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Abstract

Sinus of Valsalva Aneurysm (SVA) is a rare cardiac condition that is commonly congenital. Acquired SVA is very rare, resulting from infections affecting the aortic wall, degenerative diseases or traumatic injury to the chest. Left SVAs are usually acquired, especially when associated with rupture into the left heart. We present a case of ruptured left SVA in a female Nigerian Septuagenarian who presented with congestive cardiac failure.

Keywords: Sinus of valsalva; Aneurysm; Heart failure; Septuagenarian

Introduction

Sinus of Valsalva Aneurysm (SVA) is an uncommon cardiac anomaly that may be congenital or acquired. Congenital SVA is more common, and results from weakness of the aortic media at its junction with the annulus fibrosus [1]. The right coronary sinus is the most common site involved as observed in 75% to 90% of cases, followed by the non-coronary sinus in 10% to 25%. Congenital lesion of the left coronary artery is very rare, probably because it does not originate from the bulb septum [2]. Acquired SVA can result from trauma, endocarditis, syphilis, tuberculosis and connective tissue diseases [3-5].

The reported incidence of SVA ranges from 0.1-3.0% of all congenital heart defects, accounting for 0.14% of all open heart surgical procedures, while autopsy series suggests a prevalence of 0.09% in the general population [6,7].

Most unruptured SVA remain asymptomatic and incidentally discovered during echocardiography. However, the presentation in ruptured SVA is variable, depending among other things the receiving chamber, and whether the rupture is acute or chronic. Ruptured SVA commonly present with continuous murmur, often suspected to be due to patent ductus arteriosus or coronary fistula, or congestive cardiac failure [1]. We present a case of a ruptured SVA in a 70-year old lady presenting with heart failure and mitral incompetence.

Case Presentation

A 70-year old lady was referred to our unit with a four-week history of progressive shortness of breath, paroxysmal nocturnal dyspnea, orthopnea, cough, abdominal and leg swelling. There was a history of palpitation but no syncope or chest pain. She developed fever a week prior to presentation. She was not known to be hypertensive or diabetic, and had not suffered myocardial infarction, trauma to the chest, stroke or transient ischemic attack.

Physical examination revealed a frail elderly lady comfortably lying in cardiac position with bilateral pitting edema extending to the knees. There were no Oslerian features of subacute endocarditis. The pulse was regular at 118 bpm, with thickened arterial wall. All peripheral pulses were present and normal. Blood pressure was 142/90 mm Hg from the right arm in cardiac position. Jugular venous pressure was 10 cm of H\textsubscript{2}O with normal wave forms. A heaving point of maximum apical impulse was felt in the fifth left intercostal space 3 cm lateral to the mid-clavicular line. There was an S\textsubscript{3} gallop of left ventricular origin with 3/6 murmur of mitral regurgitation. There was also a murmur of aortic regurgitation. She had features of right sided pleural effusion. The liver was palpable 2 cm below the right costal margin with a smooth, sharp and tender edge. A diagnosis of biventricular heart failure was made.

Chest radiograph revealed a cardiothoracic ratio of 53% with a curvilinear opacity overlying a prominent aortic knuckle and pleural effusion involving the right lower zone. The mediastinum was not dilated. A 12 lead ECG showed a wandering pacemaker with an irregular ventricular rate of 90 cycles per minute, a unifocal PVC of LV origin, QRS axis of +90 degrees and left ventricular hypertrophy by Sokolow's criteria. Transthoracic echocardiogram showed left ventricular end-diastolic diameter of 56 mm and interventricular septum/posterior wall thickness of 12/12 mm. The estimated left ventricular ejection fraction was 40-45%. A SVA involving the non-coronary cusp was visualised in parasternal long axis and short axis views. An apical four chamber view showed the SVA simulating an interatrial septal cyst measuring 38 mm × 40 mm (Figure 1). The lesion was observed to have an expansile systolic movement, and echocardiography discontinuity was noted on the edge of the aneurysm, resulting in a communication with the left atrium (Figure 2). Doppler flow imaging confirmed a communication between the SVA and LA with the latter serving as the receiving chamber. Mitral annular calcification was observed with a posterior jet of moderate mitral incompetence. Aortic valve had a degenerative morphology with mild aortic regurgitation. There was no aortic stenosis. There was a trivial tricuspid regurgitation. No ventricular or atrial septal defect was detected. Transesophageal echocardiography, cardiac CT/MRI and cardiac catheterization were not available in our centre. Complete blood count revealed a WCC of 5.5 × 10\textsuperscript{11}/L with a normal differential leucocytes count. The blood film

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revealed microcytosis and anisocytosis with packed cell volume was 34%. The erythrocyte sedimentation rate was 33 mm/h.

She was placed on furosemide, lisinopril, spironolactone and made remarkable improvement. Carvedilol was initiated at low dose following the clearance of peripheral edema and pulmonary congestion. She improved to NYHA class II and discharged after two weeks of hospitalization. There was no onsite cardiothoracic surgical service, and referral to a centre with such capability was out rightly rejected by the patient. She remained stable on medical treatment and was followed-up for four months after which the relative reported her sudden unexpected death.

Discussion

Unruptured SVA is commonly asymptomatic, diagnosed incidentally by echocardiography in adolescents and young adults. However, ruptured SVA presents with murmurs and/or congestive cardiac failure. Few cases have been reported as incidental findings in elderly patients. Our patient presented in her seventies with symptoms of heart failure considered to be due to hypertensive heart disease. Sinus of Valsalva aneurysm was not considered.

Whether our patient had acute heart failure resulting from the ruptured aneurysm or she has had a smouldering background chronic heart failure that did not classically manifest because of age-related limitation of physical activity could not be ascertained. Interestingly, the communication resulting from the rupture did not generate a continuous murmur as expected. This may probably be due to the small nature of the rupture or the fact that it is communicating with the left ventricle since continuous murmur is commonly encountered in ruptures communicating with the right side of the heart.

Common anomalies coexisting with SVA include aortic regurgitation and ventricular septal defects. Although our patient had aortic regurgitation, the aortic valve had morphologic appearance consistent with degenerative valve disease on echocardiography, a common finding in hypertensive septuagenarians.

Unruptured SVA are commonly asymptomatic [1]. Though previously considered an uncommon entity, the advent of echocardiography has led to a more frequent diagnosis of SVA. The presentation in ruptured SVA is quite variable, depending on the size of the rupture, the rapidity with which it develops and the receiving chamber [8]. Two clinical patterns are largely recognised: acute rupture (usually of a large SVA) and the insidiously evolving small perforations [1,8]. Abrupt onset of distressing retrosternal chest discomfort, upper abdominal pain, and severe breathlessness with haemodynamic instability characterizes acute rupture of a large SVA. Presentation is subtler in small perforations, remaining asymptomatic for many months, with insidious onset of symptomatic congestive cardiac failure in months to years. Our case highlights the coexistence of ruptured SVA, presumably of the insidious type, with congestive cardiac failure.

Differentiating acquired aneurysm from the congenital forms could be difficult. However, acquired aneurysm from senility involves all the sinuses because of degenerative dilatation, resulting in multiple SVA. In a criterion proposed by Jones and Langely [9], the following findings favour acquired SVA: involvement of an additional SV, extracardiac location, often extending superiorly, and presence of a coexisting cardiac disease (other than congenital heart disease). Some workers alluded that involvement of the left coronary sinus is most consistent with acquired SVA, especially when associated with rupture into the left heart [5,10].
Transthoracic echocardiography is the commonly employed method for the initial evaluation of SVA. Transesophageal echocardiography is highly sensitive for the identification localisation of ruptured SVA as well as assessment of associated abnormalities, especially VSD. 3D transesophageal echocardiography is increasingly being utilized for assessment of SVA, but more importantly, during repair [11]. Other imaging modalities include cardiac catheterization, contrast-enhanced CT scan and cardiac MRI. Our patient only had transthoracic echocardiography.

Surgical intervention remains the treatment of choice for ruptured and symptomatic unruptured SVA [12], with prompt intervention recommended in cases of ruptured SVA to prevent endocarditis or enlargement of the rupture [6]. Although the operative risk associated with repair is generally low, the outcome will greatly depend on patient factors especially in the elderly. Our patient was not subjected to repair because of unavailability of onsite cardiothoracic services, and she declined referral to other centres. She responded to medical treatment of heart failure and was discharged.

The patient was reported to have suffered sudden death after four months of uneventful follow up. Although it was difficult to decipher the cause of her sudden death, especially when no autopsy was done, it may not be closely related to SVA since the rupture in question is deemed insidious. Sudden cardiac death in SVA results from tamponade, myocardial ischemia related to left anterior descending artery ostial occlusion, conduction system defects and arrhythmia [13,14].

This case illustrates the difficulties associated with differentiating between congenital and acquired Sinus of Valsalva aneurysm, an uncommon cardiac problem presenting in the elderly. Although the diagnosis of ruptured SVA aneurysm was made using transthoracic echocardiography, we did not have the benefit of doing additional investigations using transesophageal echocardiography, 3D echocardiography, cardiac catheterization, cardiac CT or cardiac MRI. This as well as lack of onsite cardiothoracic services posed major limitations to the management of this case, a prevailing scenario in most of tertiary hospitals of sub-Saharan African countries.

References