Case Report
A rare cause of sudden dyspnea and unexpected death in adolescence: fistula from aortic sinus of Valsalva to right atrium

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Abstract: Aneurysm of the sinus of Valsalva is a rare cardiac abnormality. Unruptured sinus of Valsalva aneurysm is usually asymptomatic, however, a ruptured aneurysm typically leads to an aortocardiatic shunt and progressively worsening heart failure. We report a case of a 17-year-old male who suffered an aneurysm of the sinus of Valsalva rupture into the right atrium who manifested sudden dyspnea and acute heart failure.

Keywords: Aneurysm of the sinus of Valsalva, heart failure, adolescence

Background

Sinus of Valsalva aneurysm is a rare cardiac anomaly, occurring in between 0.09% and 0.15% of cases, and accounts for up to 3.5% of all congenital cardiac anomalies [1, 2]. This type of aneurysm is typically congenital and may be associated with heart defects. It is concerned with the lack of continuity between the aortic media and the aortic annulus leading to subsequent weakening, avulsion and aneurismal formation. It is sometimes associated with Marfan syndrome or Loeys-Dietz syndrome but may also result from Ehlers-Danlos syndrome, atherosclerosis, syphilis, cystic medial necrosis, chest injury, or infective endocarditis. There is a male to female predominance of 4:1, with the highest incidence in the Asian population [3].

Unruptured sinus of Valsalva aneurysm is usually asymptomatic, therefore they are more frequently diagnosed with the noninvasive imaging modalities such as echocardiography and MRI for other reasons. We present a case of ruptured congenital aortic sinus aneurysm, which manifested sudden dyspnea and acute heart failure, and was diagnosed using two-dimensional echocardiography.

Case report

A 17-year-old male was brought to the Emergency Department with acute onset dyspnea and hypotension. He was a student and reported nothing remarkable about his health until he suddenly collapsed after playing basketball 2 days ago. He had nausea, vomiting, hypoxia, tachypnea, and tachycardia accompanied by acute orthopnea with palpitation. On examination, his blood pressure was 80/50 mmHg, heart rate was 110-150 bpm, and respiratory rate was 25/min. Pulse oximeter measured an oxygen (O2) saturation of 85-90%. Electrocardiography showed sinus tachycardia with normal axis. Chest CT showed congestion of both lung fields, with normal heart size. Arterial blood gas analysis revealed oxygen desaturation with respiratory alkalosis. The creatine kinase, creatine kinase MB, cardiac troponin T and pro-BNP all increased. And the white blood cell was high. So, a provisional diagnosis of acute viral myocarditis was made, which lead to acute heart failure. However, a “machine-type” continuous murmur of grade III was audible along the left and right paraesternal border. Further, the transthoracic echocardiogram revealed the presence of a continuity solution between the right coronary sinus and the right atrium, com-
patible with a fistula between a small right coronary sinus aneurysms and the right atrium (Figure 1). In modified views, this ring-like image was found to be a protrusion of the SV to the RA. In color Doppler, there was a high velocity jet directly to the atrial side of the tricuspid through a 3 mm orifice. Because of poor socioeconomic conditions, the urgent surgical procedures weren’t carried out. The patient’s condition got worse, he suddenly lost consciousness during preparing the surgical correction of the fistula at same day. Cardiopulmonary resuscitation was immediately performed in hospital, but he failed to recover and died.

Comment

Aneurysms of sinus of Valsalva are thought to result from the absence of normal elastic and muscular tissue, which leads to thinning of the wall of the aortic sinus. Unruptured sinus of Valsalva aneurysms are usually asymptomatic. However, if rupture, the presentation is acute with dyspnea or heart failure. Sudden shunting of blood from aorta to the right heart chambers leads to an abrupt increase in venous pressure with decrease in aortic diastolic pressure, which leads to acute symptoms and ultimately culminates into biventricular volume over-load due to systemic-pulmonary shunting and congestive heart failure. The severity of symptoms depends on the location of rupture and the size of defect. It is always confused with acute heart failure with other factors. In young and middle-aged patients with symptoms of acute heart failure and new heart murmurs, lack of medical history of fever, ruptured Aneurysms of sinus of Valsalva should be considered in the differential diagnosis. Two or three-dimensional echocardiography is an important tool to confirm the diagnosis of ruptured sinus of Valsalva. Additional imaging tools include cardiac catheterization, multidetector computed tomography, and cardiovascular magnetic resonance.

The present case was made a definite diagnosis by transthoracic echocardiography. But with large size of defect, multiple organ dysfunction emerged in short term, which may be the main reason of dead. Of course, cardiac catheterization or autopsy wasn’t performed, so we can’t exclude the other factor to cause sudden cardiac death [4].

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Disclosure conflict of interest

None.

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