Echocardiographic Evaluation of Aorta to Right Atrial Fistula Secondary to Ruptured Sinus of Valsalva Aneurysm

Abstract

We present the case of a 37-year-old male who presented with new onset dyspnea, tachycardia, palpitations, and chest tightness. His initial workup demonstrated a dilated pulmonary artery with reflux of contrast dye into the inferior vena cava. Transthoracic echocardiogram identified a “windsock” appearance indicating Sinus of Valsalva aneurysm (SVA) and severe aortic regurgitation. As a result, the patient was taken for emergent surgery where the windsock was surgically repaired with bovine pericardial patch. This case illuminates the uncommon occurrence of SVA and the ability to recognize these findings on multiple imaging modalities including transthoracic, transesophageal two and three dimensional echocardiography as well as direct surgical field visualization.

Keywords: Coronary to atrial fistula, emergency cardiac surgery, three dimensional echocardiography, transesophageal echocardiography

Introduction

Sinus of Valsalva aneurysm (SVA) is an uncommon cardiac anomaly that may be congenital or acquired. SVA occurs when the aortic media separates from the annulus fibrosis, with aneurysmal rupture representing the most common complication of this cardiac anomaly. The aneurysm predominantly originates from the right coronary sinus and may rupture up to 35% of the time, most commonly to the right cardiac chambers. Diagnosis of a ruptured sinus of Valsalva can be made by the classic “windsock” appearance on echocardiography. Emergent surgical intervention should be the treatment of choice for ruptured SVA due to the possibility of rapid cardiac deterioration.

Case Report

A 37-year-old Caucasian male with no previous medical history presented to the emergency department with sudden onset of tachycardia, palpitations, and chest tightness for 6 days. On examination, the patient had sinus tachycardia with a heart rate of 130 and new holosystolic murmur. A computed tomography angiogram was performed in the emergency department for suspicion of pulmonary embolism which subsequently demonstrated a dilated pulmonary artery with reflux of contrast dye into the inferior vena cava. The patient was admitted for further evaluation. At that time, a transthoracic echo identified a 1.2 cm × 1.2 cm cavity associated with the right sinus of valsalva and severe aortic regurgitation with ejection fraction 64%. Transesophageal echocardiogram (TEE) confirmed the presence of the right coronary SVA with fistulization into the right atrium and deformation of the right coronary leaflet. The patient was taken for emergent repair of the aorta to right atrial fistula. Cardiopulmonary bypass was initiated. On TEE examination, the aortoatrial fistula was visualized just above the tricuspid annulus, visualized in two, and three dimensional views [Figures 1 and 2]. The classic “windsock” appearance was noted, and color flow Doppler identified continuous flow from the right coronary cusp (RCC) to the right atrium [Figure 1]. Three dimensional imaging was used to acquire images of the aortic valve and better visualize the fistula tract to the RCC [Figure 2]. On opening the right atrium, tissue fistulization was identified from the RCC into the right atrium [Figure 3]. Intraoperative TEE also identified a patent foramen ovale (PFO), likely due to a high-pressure right to left
shunt caused by the Sinus of Valsalva fistula. The aorta to right atrial fistula was closed with bovine pericardial patch and PFO surgically repaired. The patient tolerated the procedure well, and he was taken to the Intensive Care Unit for immediate postoperative recovery. The remainder of his care was uneventful with extubation on postoperative day 1 and discharge to home on postoperative day 5.

Discussion

SVA incidence is uncommon and occurs in <0.1% of the general population.[4] Trauma, infection, and inflammatory disease can cause these aneurysms; however, the most common etiology is a congenital defect in the aortic media.[4,5] The most common location for an aortoatrial fistula after SVA rupture is into the right atrium, right ventricle, or both. In contrast, a rupture in the left atrium from a noncoronary or left SVA is rare.[4] A ruptured SVA should be remembered as etiology of sudden, symptomatic aortic regurgitation. Early identification and repair can prevent complications such as arrhythmias, cardiac tamponade, and myocardial ischemia.[3] Intraoperative TEE offers two and three dimensional evaluation to assist with real-time surgical decisions and management. TEE is instrumental for identification of unruptured and ruptured SVA, concomitant cardiac pathology, and optimization of perioperative management in cardiac surgical patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Afshar AH, Kolesnikov S, Pourafkari L, Nader ND. Right valsalva sinus aneurysm protruding into the right ventricle: A Case report. J Cardiovasc Thorac Res 2015;7:126-8.
2. Post MC, Braam RL, Groenemeijer BE, Nicastia D, Rensing BJ, Schepens MA, et al. Rupture of right coronary sinus of valsalva aneurysm into right ventricle. Neth Heart J 2010;18:209-11.
3. Togashi K, Hermsen JL, Joffe DC. Sinus of valsalva aneurysm rupture misdiagnosed as a ventricular septal defect. Anesth Analg 2017;124:1437-9.
4. Moreo A, De Chiara B, Bruschi G, Tarelli G, Parolini M, Mauri F, et al. An unusual cause of pulmonary edema: Acute rupture of noncoronary sinus of valsalva aneurysm into the left atrium. J Am Soc Echocardiogr 2006;19:938.e9-11.
5. Narumi T, Watanabe T, Iwayama T, Ichikawa K, Wanezaki M, Nishiyama S, et al. Ruptured right sinus of valsalva aneurysm caused by suspected takayasu’s arteritis. Intern Med 2016;55:1305-8.