Unusual cause of rapidly progressive right-sided heart failure: aortic sinus of Valsalva aneurysm causing ball valve obstruction of the tricuspid valve

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A rare presentation with rapidly progressive right heart failure due to tricuspid inflow obstruction (simulating right-sided valvular heart disease) caused by a non-coronary cusp sinus of Valsalva aneurysm with small perforation is reported. The aneurysm was causing ball valve obstruction at the tricuspid valve, leading to dilated right atrium and back pressure changes which were relieved after successful aneurysctomy.

KEYWORDS
Sinus of Valsalva aneurysm (SAV);
Two-dimensional echocardiography

Introduction
Aneurysm of the sinus of Valsalva is an uncommon disorder, most commonly presenting as a rupture into a cardiac chamber. Small perforations of aneurysms present insidiously; patients may remain asymptomatic for months or years until symptoms of congestive heart failure develop. Most such aneurysms are thought to be congenital in origin, arising because of discontinuity between the aortic tunica media and aortic valve annulus.1 We describe a highly unusual form of a large sinus of Valsalva aneurysm (SVA) with small perforation presenting with rapidly progressive right-sided heart failure due to ball valve obstruction of the tricuspid valve.

Case report
A 30-year-old female was admitted to our institute with a 6 week history of rapidly progressive dyspnoea on exertion, facial puffiness, swelling of the feet, and jaundice. History of orthopnoea, paroxysmal nocturnal dyspnoea, palpitations or syncope was absent. She had no history of fever, cough, or any other constitutional symptoms.

Physical examination revealed a pulse rate of 100 beats/min, blood pressure of 110/80 mmHg, and respiratory rate of 28 bpm. Cardiovascular examination revealed an elevated jugular venous pressure, bilateral pedal oedema with no significant precordial pulsations or heave. Heart sounds were normal. Auscultation revealed a Grade 2/6 systolic murmur with maximum intensity at the mid left sternal border. Lungs were clear. Liver was palpable 3 cm below the right costal margin. Chest roentgenogram revealed no cardiomegaly. Electrocardiogram showed sinus tachycardia.

Two-dimensional echocardiography demonstrated a large cystic mass originating from the base of the aorta, extending to the right ventricular inflow tract, and appearing to straddle the tricuspid valve orifice especially during diastole (Figure 1). Transoesophageal echocardiography clearly showed this to be an aneurysm arising from non-coronary sinus of aorta moving across the tricuspid annulus between dilated right atrium and small right ventricle (Figure 2) (see Supplementary data, Video clip 1).

On cardiac catheterization the mean right atrial pressure was 22 mmHg. Tricuspid valve was crossed carefully with a 0.032 in. Terumo guidewire which was followed by passing a Swan Ganz catheter. The mean pulmonary artery pressure was normal. Ascending aortography demonstrated a large aneurysm with a wide neck originating from the non-coronary sinus with protrusion into right ventricular inflow with a streak of dye flowing from the apex of the aneurysm suggesting its ruptured status which was not clearly noticed on Doppler echocardiography (Figure 3) (see Supplementary data, Video clip 2). There was no aortic regurgitation or ventricular septal defect. Coronary angiogram revealed normal epicardial coronary arteries.

The patient underwent a median sternotomy with cannulation of both venae cavae and the ascending aorta for...
cardiopulmonary bypass. Opening the pericardium revealed a hugely dilated right atrium with disappearance of wrinkling over the appendage, dilated inferior vena cava, and a small right ventricle. A 4 × 3 cm aneurysm was found in right atrium which showed a small perforation, through which cardioplegia was seen flowing. The aneurysm was seen occluding the tricuspid annulus like a ball valve with the posterior wall of the aneurysmal sac adherent to the septal leaflet of the tricuspid valve. Through a transverse aortotomy, the origin of the aneurysm was identified at the base of the non-coronary cusp of the aortic valve. The aneurysm was resected and a pericardial patch closure of the defect was done. The patient was weaned off cardiopulmonary bypass without difficulty with rapid resolution of the symptoms in the post-operative period. Jaundice subsided after about 2 weeks. However, she developed mobitz type II, second-degree atrioventricular block on Day 2 of surgery for which she required a dual-chamber permanent pacemaker. Rest of the post-operative period was uneventful. At 6 month follow-up, the patient was doing well with complete resolution of symptoms.

Discussion

SVAs are rare cardiac anomalies. An aneurysm of the sinus of Valsalva is defined as dilatation of one of the three aortic sinuses between the aortic valve annulus and the sino-tubular junction or supra-aortic ridge. Most SVAs arise from the right and non-coronary sinuses and, rarely, from the left. Aneurysms of right or non-coronary cusp origin commonly involve the right atrium and right ventricular outflow tract. Patients with these lesions have been found to be asymptomatic clinically unless the aneurysm ruptures. Unruptured SVA usually remain asymptomatic and undetected. However, with the advent of echocardiography and other non-invasive imaging modalities, unruptured SVA are diagnosed more frequently. Multiple case reports describe unruptured SVA causing significant anatomic and physiological derangements.2–5

This case highlights an unusual presentation of a large SVA with small perforation (physiologically behaving as an unruptured SAV) simulating primary right-sided valvular heart disease. Isolated unruptured aneurysm of the sinus of Valsalva producing significant haemodynamic compromise is extremely rare.4–8 Though presentation as right-sided heart failure are well described, all of them have been secondary to right ventricular outflow obstruction.9–10 Only one case of congenital unruptured SAV has been previously reported with haemodynamically documented obstruction of the right ventricular inflow.11 Gibbs et al.11 described a 25-year-old Caucasian male with haemodynamic compromise due to dynamic tricuspid stenosis and insufficiency caused by an unruptured noncoronary SVA that protruded into the right atrium and across the tricuspid valve, partially obstructing right ventricular inflow. The unruptured aneurysm was resected successfully. Nisanoglu et al. recently reported a case of large unruptured SAV presenting haemodynamically as right atrial tamponade.12 However, unlike
our case, the authors found it to be an acquired SAV secondary to bacterial endocarditis. Bulkley et al.13 reported five cases of unruptured aneurysm of the sinus of Valsalva simulating right-sided valvular heart disease. In two patients, aneurysms of the non-coronary sinus projected into the right atrium at the level of the tricuspid valve and caused tricuspid incompetence. In three patients, aneurysms of the right sinus of Valsalva projected into the right ventricle immediately below the pulmonic valve and caused pulmonary outflow tract obstruction. In only one of these patients was the aneurysm perforated at necropsy. Although valvular dysfunction was evident clinically, in none was there a clinical suspicion of an SVA.

In the present case, the SVA was found to obstruct the right ventricular inflow like a ball valve causing dilatation of right atrium producing back pressure changes leading to peripheral oedema, raised jugular venous pressure, hepato-megaly, and jaundice. The SVA had a small perforation at its apex (demonstrated on angiogram) which did not produce classic signs like thrill, continuous murmur, or shunt seen in ruptured aneurysm. Unlike the case reported by Gibbs et al.,11 our case did not have associated tricuspid regurgitation and presented with a clinical picture of rapidly progressive right-sided heart failure. Two-dimensional echocardiography permitted non-invasive diagnosis. Traditionally both the ruptured and intact SAV are dealt by open-heart correction under cardiopulmonary bypass. In the recent past, successful efforts have been done to manage the SAV through transcatheter route, but the safety and proficiency of these methods is yet to be established. In the present case, surgical resection of the aneurysm resulted in dramatic clinical improvement barring the complication of post-operative conduction block which necessitated a permanent pacemaker implantation.

Conclusion

This case report highlights the rare presentation of a large aneurysm of sinus of valsalva obstructing the tricuspid valve leading to right heart failure. Echocardiogram is a useful non-invasive investigation to diagnose this condition. The traditional treatment of these aneurysms has been surgical repair. Percutaneous closure, as an alternative in selected cases, has been reported recently. In the present case, as the neck of aneurysm was large, surgery was considered and was successfully performed.

Supplementary data

Supplementary data are available at European Journal of Echocardiography online.

References