ESC European Society of Cardiology

European Heart Journal - Case Reports (2019) **3**, 1–4
doi:10.1093/ehjcr/ytz111

CASE REPORT
Congenital heart disease

The value of three-dimensional echocardiography in the diagnosis and management of ruptured sinus of Valsalva aneurysm: a case report

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Received 19 November 2018; first decision 19 December 2018; accepted 27 June 2019; online publish-ahead-of-print 13 July 2019

Background A sinus of Valsalva aneurysm (SVA) is a rare cardiac anomaly. Most SVA's rupture into right heart chambers and can be classified using the modified Sakakibara classification according to the site of rupture. Transoesophageal echocardiography (TOE) is a useful diagnostic tool and aides in treatment planning in patients with congenital anomalies in emergency situations. Three-dimensional TOE (3D-TOE) provides additional value over standard TOE.

Case summary A 38-year-old man with a reported history of ventricular septal defect (VSD) presented to the emergency department complaining of chest pain and epigastric pain lasting several days. Physical examination revealed a continuous heart murmur and signs of acute heart failure. A 3D-TOE revealed an SVA rupture into the right ventricle (Type IIIv) but no evidence of a VSD. Urgent aortic valve replacement with correction of the ruptured SVA was performed. Neither a VSD nor signs of endocarditis were found during surgical exploration. The patient was discharged on post-operative Day 5 in good condition.

Discussion A sinus of Valsalva aneurysm is a rare cardiac condition. Ventricular septal defect, bicuspid aortic valve, or aortic valve regurgitation may coexist with SVA. Xin-Jin *et al*. classified a ruptured SVA into five types according to the site of rupture. Transoesophageal echocardiography is an important tool for diagnosis, anatomical description, and typing of the ruptured SVA. Sinus of Valsalva aneurysm may be misdiagnosed as a VSD, as was the case in our patient, and 3D-TOE can be instrumental for providing both correct diagnosis and critical surgical planning.

Keywords Three-dimensional echocardiography • Congenital cardiac anomaly • Cardiogenic shock • Ruptured sinus of Valsalva aneurysm • Case report

Learning points

- A sinus of Valsalva aneurysm (SVA) is a rare congenital anomaly.
- The clinical signs and symptoms of SVA rupture usually include abrupt dyspnoea and chest pain.
- Transoesophageal echocardiography has a great value in the correct diagnosis, detailed anatomic relations and the treatment in the case of ruptured SVA.

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Handling Editor: Nikos Papageorgiou

Peer-reviewers: Richard Alexander Brown, Georgia Daniel, and Esther Cambronero-Cortinas

Compliance Editor: Amir Aziz

Supplementary Material Editor: Peysh A. Patel

Introduction

A sinus of Valsalva aneurysm (SVA) is a rare cardiac condition. It is most commonly congenital, yet it may also result from endocarditis, trauma, rheumatoid arthritis, or aortitis. The aneurysm predominantly involves the right coronary sinus and is more common in males from Asian descent.[1,2] Ventricular septal defect, bicuspid aortic valve, or aortic valve regurgitation may coexist with SVA.[3] Sinus of Valsalva aneurysms may become symptomatic around 30–45 years of age.[2] Rupture of the aneurysm can lead to acute symptoms such as acute chest pain and dyspnoea. Most aneurysms rupture into the right-sided heart chambers.[3,4] Xin-Jin *et al.* classified the ruptured SVA into five types, known as the Sakakibara classification[5]: Type I—rupture into the right ventricle just beneath the pulmonary valve; Type II—rupture into or just beneath the crista supraventricularis of the right ventricle; Type IIIa—rupture into the right atrium at the tricuspid annulus; Type IIIv—rupture into the right ventricle at the tricuspid annulus; Type IV—rupture into the right atrium; and Type V—rupture into other structures (left ventricle, left atrium, pulmonary artery, etc.).

Timeline

Background	38-year-old smoker, presumed ventricular septal defect (VSD)
3 days before presentation to the hospital	Dyspnoea, chest pain, and epigastric pain
Day of admission (Day 0) 03:00 am	Admission to emergency department: Heart failure symptoms Chest pain Epigastric pain Lactic acidosis Acute renal failure Hand-held transthoracic echo showing presumed VSD with left to right shunt Non-contrast chest and abdominal computed tomography—no contributing findings
Day 0—10:30 am	Admission to intensive cardiac care unit
Day 0—11:30 am	Three-dimensional transoesophageal echocardiography showing ruptured sinus of Valsalva aneurysm into right ventricle
Day 0—12:30	Haemodynamic and respiratory deterioration requiring initiation of mechanical ventilation transfer to tertiary centre for surgery
Day 0—14:00	Mechanical aortic valve replacement and closure of ruptured sinus of Valsalva
Day—5	Discharge home in stable condition

Case presentation

A 38-year-old man with a history of smoking, alcohol use, VSD, and biliary pancreatitis a year earlier, presented to the emergency department complaining of progressive severe chest pain and epigastric pain unrelated to physical effort for three consecutive days. Additionally, the patient complained of dyspnoea and sweating. His vital signs revealed sinus tachycardia of 120 b.p.m., blood pressure 120/70 mmHg, and normal body temperature. The physical examination revealed a loud continuous harsh systolic–diastolic murmur with an intensity of 4/6 and maximal intensity at the lower left sternal border. Jugular venous pressure was elevated. Electrocardiogram showed sinus tachycardia and negative T waves in leads $V_{3–4}$.

Laboratory findings showed acute renal failure, metabolic acidosis with a pH of 7.16 (normal range 7.35–7.45), mildly elevated blood lactate level, leucocytosis, and mild troponin elevation. Chest radiography showed bilateral minimal pleural effusions without cardiomegaly. Bedside hand-held transthoracic echocardiography (TTE) (V scan, GE) performed in the emergency department revealed normal left ventricle function, right ventricle dilatation, left to right ventricular shunt presumed to be secondary to VSD (given the patient's reported history of VSD), and no pericardial effusion. Given the patient's prior history of pancreatitis and ongoing abdominal pain with acute renal failure, he underwent computed tomography (CT) of the chest and abdomen without contrast media. The scan showed bilateral pleural effusions at the lung bases but did not show signs of pancreatitis and there were no blatant signs of aortic dissection or pulmonary emboli in the main pulmonary artery branches, notwithstanding the limitations of a non-contrast tomography. Subsequently, a three-dimensional transoesophageal echocardiography (3D-TOE) (*Figures 1–3*) was performed which revealed an SVA originating from the right coronary cusp which had ruptured into the right ventricle, causing the large left to right ventricle shunt seen on hand-held TTE. No VSD was identified. The edge of the ruptured SVA was 18 mm from the right coronary artery ostium (*Figure 3*). The right ventricle was dilated with preserved function. Immediately following the 3D-TOE, the patient deteriorated haemodynamically and was intubated. The patient was then transferred to a nearby tertiary medical centre for urgent surgical intervention without performing a diagnostic cardiac catheterization given the patient's young age, relatively low-risk profile, and overall critical condition.

Surgery confirmed a fistula from the right coronary cusp aneurysm into the right ventricle (*Figure 4*). A suturing attempt of the fistula was unsuccessful due to prolapse of the right coronary cusp, thus the patient underwent mechanical aortic valve replacement (ON-X 21 mm), along with successful closure of the fistula. In addition, visual exploration confirmed that no VSD was present, nor were there signs of endocarditis.

Echocardiography after the procedure showed no residual shunt and no aortic regurgitation. The patient's condition dramatically improved, including his renal function, and he was discharged home on post-operative Day 5 on anticoagulation.

Figure 1 Right ventricle inflow–outflow view in three-dimensional transoesophageal echocardiogram. Green arrow: fistula from ruptured right coronary cusp sinus of Valsalva aneurysm into right ventricle. A, aortic valve; P, pulmonic valve; RV, right ventricle.

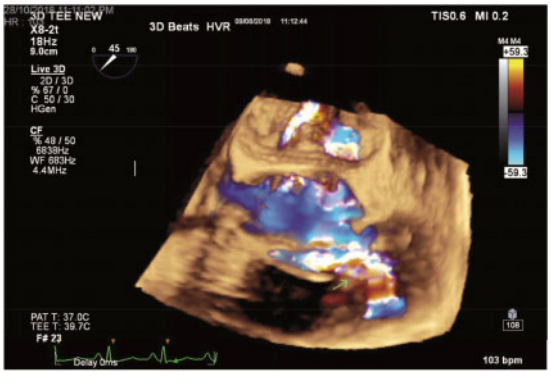

Figure 2 Mid-oesophageal—aortic valve—short axis three-dimensional colour Doppler view. Arrow: flow jet from right sinus of Valsalva aneurysm to right ventricle.

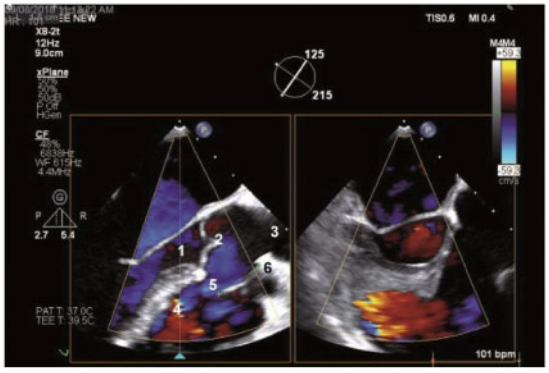

Figure 3 A two-dimensional colour Doppler X-plane transoesophageal echocardiogram shows in the left side, the left ventricular outflow tract (1), the aortic valve (2), the ascending aorta (3), the right ventricular outflow tract (4), the ruptured sinus of Valsalva aneurysm (5), and the right coronary artery origin (6). The distance between the RCA origin and the ruptured sinus of Valsalva aneurysm was measured to be 18 mm.

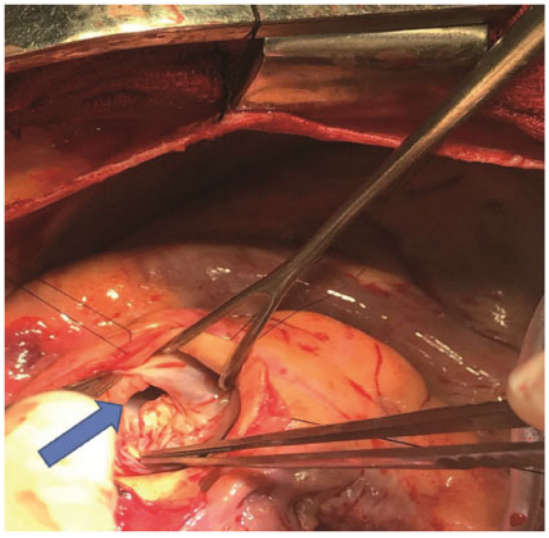

Figure 4 Image from surgery. Mid-sternotomy, open aorta. View through the aortic root. Arrow: inlet of ruptured right sinus of Valsalva aneurysm into the right ventricle.

Discussion

Transoesophageal echocardiography is an important tool for diagnosis, anatomical description, and typing of the ruptured SVA. Magnetic resonance imaging (MRI) may help in the diagnosis of other lesions but is limited to hemodynamically stable patients. Coronary angiography prior to surgery should be considered in older patients or patients with risk factors for coronary artery disease. In two previous case reports, the advantage of 3D-TOE was described.[6,7] In the first case, the 3D-TOE helped to discriminate and administer the appropriate treatment to a patient with both SVA rupture and perimembranous VSD. The second case presented a patient in which the 3D-TOE helped to choose the proper percutaneous equipment for closure of the ruptured SVA.

In our case, the 3D-TOE was instrumental in correctly diagnosing the left to right shunt as being from a ruptured SVA and not from a VSD as initially thought based on a reported history of VSD in the patient's medical record. We believe the anatomical proximity of the previously unruptured SVA to the ventricular septum and likely turbulent flow within the aneurysm projecting into the right ventricle led to the VSD misdiagnosis on prior imaging studies. Our initial hand-held TTE clearly showed the left to right shunting but did not lead us to question the diagnosis of VSD or suggest the presence of an SVA. Rather, the 3D-TOE provided the necessary anatomical

resolution required to make a correct diagnose. It furthermore provided both rapid and accurate information of the anatomical relationship between the main cardiac structures around the SVA in an unstable patient. Other imaging modalities, such as cardiac MRI, CT, or coronary angiography, are not always feasible if not immediately available or if the patient is haemodynamically unstable. In our case, the 3D-TOE provided the information necessary for the thoracic surgeon to proceed with urgent surgical repair.

Conclusion

In the case of an acutely decompensating patient with congenital, acquired or mixed cardiac anomaly, 3D-TOE should be included in the initial evaluation as it is a quick, safe, and reliable bedside imaging technique to achieve an appropriate anatomical diagnosis. Sinus of Valsalva aneurysm may be misdiagnosed as a VSD on TTE alone, leading to delayed diagnosis of SVA rupture. Unappreciated SVA rupture may lead to inappropriate surgical planning.

Lead author biography

Dr. Elias Daud, MD, is a graduate of Münster University School of Medicine in Germany, where he also attended his undergraduate studies in Medical Sciences. Dr. Daud moved to the Western Galilee Medical Center in Israel where he completed his residency in Internal Medicine. Currently, he is completing his clinical fellowship at the Cardiology Department. Dr. Daud holds Israeli and European medical licenses and certifications from the Israeli Board of Internal Medicine. He is also a member of the Israeli Medical Association (IMA), Israeli Society of Internal Medicine and Israeli Heart Society (IHS).

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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