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Case Report

Transcatheter closure of ruptured sinus of Valsalva aneurysm in a pregnant woman

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ABSTRACT

Sinus of Valsalva aneurysm (SVA) is a rare cardiac anomaly that is usually congenital, but may be acquired. They are usually asymptomatic unless they compress adjacent structures, develop thrombosis, or rupture. A ruptured SVA (RSVA) can lead to rapid hemodynamic deterioration and often needs to be addressed emergently. Surgical correction has traditionally been the treatment of choice for RSVA; however, lately they have been successfully closed percutaneously using various transcatheter devices. Few cases of RSVA during pregnancy have been reported which were conservatively or surgically managed. There is no documented case of transcatheter closure of RSVA during pregnancy. We report the first case of successful percutaneous device closure of RSVA using an Amplatzer duct occluder in a pregnant woman presenting with heart failure due to RSVA at 26 weeks of gestation.

<Learning objective: Ruptured sinus of Valsalva aneurysm (RSVA) is traditionally repaired by surgery but more recently amenable to percutaneous intervention. Management of RSVA during pregnancy is complex and has been managed by surgery in the past incurring significant risk to fetus due to effects of cardiopulmonary bypass. We report a case of RSVA in pregnancy that was closed by transcatheter closure for the first time, thereby significantly reducing maternal and fetal risks. While risks are present during pregnancy, emergently indicated life-saving invasive cardiac procedures should not be denied solely on the pregnant state.>

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Introduction

A sinus of Valsalva aneurysm (SVA) is a rare cardiac anomaly, usually congenital in origin, rarely acquired, most often arising from right coronary sinus (80.7%), less often from non-coronary sinus (15.8%), and rarely from left (3.5%) sinus [1]. SVA are asymptomatic unless they compress adjacent structures, form a thrombus, or rupture causing left to right shunting or aortic valve insufficiency and congestive heart failure that often requires urgent surgical resolution. There are few documented cases of ruptured SVA (RSVA) in pregnant patients some of whom were managed by surgical repair [2] and for others intervention was postponed until after delivery [3]. We present for the first time, a case of RSVA that was closed by transcatheter closure during pregnancy.

Case report

A 25-year-old, 26-week pregnant primigravida weighing 58 kg with a past history of small perimembranous ventricular septal defect (VSD) was transferred to our center from an obstetric hospital for evaluation and management of acute heart failure. On examination, she was mildly dyspneic at rest (respiratory rate: 22/min), tachycardic (heart rate: 120/min), and normotensive (BP: 110/60 mmHg). Her jugular venous pressure was elevated; precordium was hyperdynamic with a prominent systolic and diastolic thrill. S1 and S2 were normal; a diffuse grade 4/6 continuous murmur was heard. She had 3-cm hepatomegaly and bilateral mild pitting pedal edema. She did not have any symptoms or signs of connective tissue disorder. Electrocardiogram showed normal sinus rhythm with infrequent premature ventricular contractions. A transthoracic echocardiogram (TTE) revealed mildly dilated left ventricle with normal function, a 7-mm perimembranous ventricular septal defect (VSD), SVA of right coronary sinus that had ruptured into the right ventricular outflow tract (RVOT) through a 7-mm defect (Fig. 1, and see supplementary data online, Video 1). Color Doppler revealed continuous flow from

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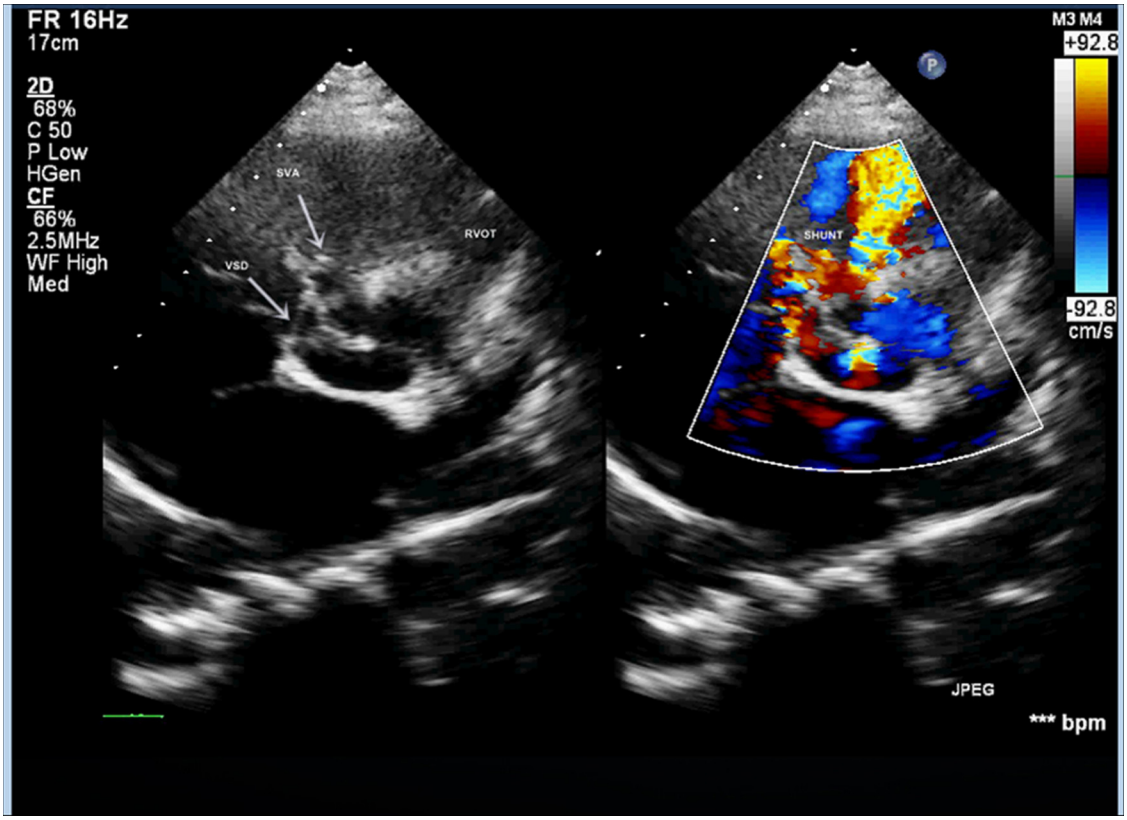

Fig. 1. Transthoracic echocardiographic images in parasternal short-axis view demonstrating small perimembranous ventricular septal defect and ruptured aneurysm of right coronary sinus of Valsalva with left to right shunt into the right ventricular outflow tract.

the aortic root into RVOT, mild aortic regurgitation and mild pulmonary hypertension. There was no pericardial effusion. Simultaneous ultrasound and fetal echocardiogram confirmed fetal well-being and absence of fetal cardiac defects. Her symptoms of acute heart failure progressed and her hemodynamic condition worsened overnight despite medical therapy, necessitating intervention. After some consideration of surgical vs. catheter closure, we decided to proceed with the percutaneous route, because the latter procedure was safer for the mother and also carried less fetal risk.

General anesthesia was avoided; the procedure was performed under local anesthesia with TTE guidance after her abdomen was properly shielded with lead. 7F short sheaths were secured in right femoral vein and bilateral femoral arteries and heparin administered. Aortic root angiogram was done which showed SVA from right coronary sinus measuring 8 mm at its origin, with contrast spilling into RVOT from the ruptured site (Fig. 2 and see supplementary data online, Video 2). The coronary arteries appeared normal. Antegrade approach was employed. Arteriovenous loop was created by passing through the defect from the aortic side using an angled tip 0.035″ glide wire (Terumo Inc., Tokyo, Japan) in 6F Judkins right coronary catheter. Glide wire was exchanged with a 260 cm, 0.035″ Amplatz support stiff guide wire (Cook Medical Inc., Bloomington, IN, USA) over which a 7F Amplatzer long sheath (St. Jude Medical, Inc. St. Paul, MN, USA) was tracked from the venous side and parked in ascending aorta. We chose to use an Amplatzer Duct Occluder (ADO1; St. Jude Medical, Inc.) because we thought it suited best for the "wind-sock"-like defect this patient had, with a broader aortic end. We chose to oversize it to 4 mm greater (ADO 12/14) than the defect size (8 mm) because the margins of this wind-sock-like defect appeared flimsy. The device was loaded over the cable, passed through the long sheath and deployed across the defect. Angiogram performed via pigtail catheter introduced through the second femoral arterial sheath confirmed optimum position and absence of residual shunt. After TTE revealed no impingement of device on aortic leaflets and no change in degree of aortic insufficiency, the device was released (Fig. 3 and see supplementary data online, Video 3). All pregnancy precautions, such as keeping flouroscopy time short (22 min), reducing number of cineangiograms (6 at 15 fps), radiation dose (dose area product-23,796 mGy cm^2), and contrast volume were taken. Soon after the procedure, the patient reported improvement in her symptoms; clinically diastolic component of murmur disappeared (holosystolic murmur of VSD persisted). TTE after the procedure revealed the device to be in a good position without any residual shunt (Fig. 4). Fetal echocardiography confirmed an active fetus with normal fetal heart rate and rhythm. We decided to follow the small VSD medically as the shunt was not hemodynamically significant enough to close. Aspirin at 150 mg once daily was commenced, which was continued throughout pregnancy. Mother and fetus were regularly followed and elective cesarean section was performed at near term to deliver a healthy 3-kg baby.

Discussion

Incidence of SVA ranges from 0.23% to 1.5%, being higher in Asians [1]. Pathogenesis likely involves incomplete fusion of distal bulbar septum and truncal ridges, leading to weakness between aortic media and annulus fibrosus resulting in aneurysmal enlargement of sinus. SVA ruptures in 35% of cases, leading to acute symptoms of dyspnea, palpitation, chest pain, and fatigue in one fourth of the patients.

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