Curriculum in Cardiology

Case Discussion

Ruptured Sinus of Valsalva Aneurysm: Clinical Case Presentation and Management

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Abstract

A 27-year-old female presented with generalized body swelling and progressive shortness of breath for the past 4 months. On examination, she had a loud continuous murmur, elevated jugular venous pressure, and gross ascites. The examination findings, electrocardiogram, chest radiography, and echocardiogram are discussed in a step-wise manner to arrive at a diagnosis and plan of management of a patient with ruptured sinus of Valsalva aneurysm is discussed along with review of relevant literature.

Keywords: Continuous murmur, raised jugular venous pressure, sinus of Valsalva aneurysm

Clinical Presentation

A 27-year-old female, resident of Uttar Pradesh, and a homemaker by occupation presented with the chief complaints of

- Episode of sudden-onset epigastric pain 4 months back
- Progressive shortness of breath × 4 months
- Pedal edema and abdominal distension × 4 months
- Exertional palpitations × 3 months.

The onset of symptoms was 4 months back when the patient developed sudden-onset epigastric pain with no accompanying gastrointestinal symptoms. While pain abdomen settled spontaneously within a day, the patient developed progressive shortness of breath increasing to the New York Heart Association class II over 20 days. Two days after the epigastric pain, she noticed swelling over both lower limbs which progressed to involve whole body over 1 month. The patient also complained of exertional palpitations for the duration of illness. There was no history of trauma, fever, cough, decreased urine output, or altered mentation. There was no history of sore throat, migratory joint pain, abnormal body movements, or subcutaneous nodules. She did not complain of repeated chest infections or cyanotic spells.

Summary

A 27-year-old female developed sudden-onset epigastric pain associated with shortness of breath 4 months back. Two days after this event, she noticed swelling over lower limbs along with abdominal distension which gradually progressed in 4 months.

Discussion

What is first differential diagnosis on the basis of your history?

The first differential diagnosis is ruptured sinus of Valsalva (RSOV) aneurysm. The points in favor are sudden-onset epigastric pain associated with sudden-onset dyspnea which was followed by symptoms of progressive right-sided heart failure.

What are the other differential diagnosis to consider in such a case?

Other possibilities are

1. Pulmonary embolism: sudden-onset shortness of breath followed by right heart failure can occur in pulmonary embolism. However, the patient did not have any predisposing factors for pulmonary embolism. There was no history of pleuritic chest pain or hemoptysis

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2. Aortic dissection: It can present with sudden-onset epigastric pain. Shortness of breath can be secondary to tamponade or acute aortic regurgitation. However, cardiac tamponade or acute aortic regurgitation is unlikely to present so late or survive without intervention
3. Acute valvular regurgitation with infective endocarditis: sudden onset of symptoms can be seen with acute aortic regurgitation or mitral regurgitation caused by infective endocarditis. However, infective endocarditis rarely presents without fever in otherwise healthy adults
4. Constrictive pericarditis and restrictive cardiomyopathy: both can present with right heart failure and onset is generally insidious
5. Chest trauma: patients with chest trauma can present with right heart failure secondary to traumatic tricuspid regurgitation. However, such a history was absent in the given patient.

Examination

The patient was a lean, cachectic young female. Blood pressure was 98/36 mmHg and 112/44 mmHg in the right upper limb and right lower limb, respectively. Pulse was 90/min, regular, bounding in character, no radio-radial or radio-femoral delay, and all peripheral pulses are palpable. The patient was afebrile to touch. Jugular venous pressure (JVP) was elevated, 7 cm above sternal angle with prominent A wave. There was the presence of bilateral pitting pedal edema up to the knees. There is no pallor, icterus, cyanosis, clubbing, or lymphadenopathy. Her arm span to height ratio was <1 and other signs of Marfan's syndrome were absent.

Cardiovascular Examination

On inspection, the precordium was hyperkinetic, and apex beat was visible in the 6th intercostal space two centimeters lateral to midclavicular line.

On palpation, apex was diffuse and hyperdynamic. Thrill with both systolic and diastolic components was present at the left parasternal border in the 5th intercostal space. Prominent ill-sustained parasternal pulsations were also present.

On percussion, bilateral 2nd intercostal spaces were resonant, right cardiac border was behind the sternum, and left cardiac border was coincident with the apex.

On auscultation, S1 was normal and S2 was normally split with loud P2 component. There was a high-pitched, grade V continuous murmur heard over the entire precordium but best heard along the left parasternal area in 5th intercostal space, in sitting forward position, and increasing in intensity with clenching of fists.

Systemic Examination

Respiratory system examination revealed bilateral diffuse crepitations.

Abdomen was distended, liver was enlarged 3 cm below right costal margin with smooth surface, round margin and was nontender. Shifting dullness was present.

Discussion

Summary of clinical findings

The patient had a bounding pulse with wide pulse pressure, elevated JVP, and pedal edema. The apex was hyperdynamic, shifted down and out, and ill-sustained systolic left parasternal pulsations were present with a prominent thrill having both systolic and diastolic components. There was a grade V continuous murmur heard in left lower parasternal area. There were bilateral chest crepitations, ascites, and hepatomegaly.

What are the causes of continuous murmur?

A continuous murmur starts in systole and extends uninterrupted up to the diastole, not necessarily occupying the entire duration of either. It arises from continuous blood flow from a high pressure to a low-pressure chamber and reflects a persistent pressure gradient between the two during systole and diastole.

Important causes of continuous murmur are [Table 1]:

Patent ductus arteriosus

The murmur of a patent ductus arteriosus (PDA), also known as Gibson's murmur, is a common cause of continuous murmur. The murmur has two components, crescendo systolic and decrescendo diastolic and the intensity of murmur peaks around second heart sound (S2). It is often associated with the presence of Eddy sounds or clicks at the end of systole and at the beginning of diastole. The murmur is best heard at the second left intercostal space or the left infraclavicular area. The duration of murmur reflects the size of shunt. The murmur may only be systolic in the setting of a large PDA and the length of murmur progressively shortens as pulmonary artery hypertension develops. The presence of differential cyanosis further aids in recognition of these patients (cyanosis of lower

Table 1: Important causes of continuous murmur

Causes of continuous murmur
Patent ductus arteriosus
Coronary arteriovenous fistula
Ruptured sinus of Valsalva aneurysm
Aortic-pulmonary window
Anomalous left coronary artery origin from pulmonary artery
Coarctation of aorta
Pulmonary artery atresia
Truncus arteriosus
Pulmonary artery stenosis
Severe arterial stenosis
Systemic arteriovenous fistula
Pulmonary arteriovenous fistula
Mammary souffle
Venous hum
Cruveilhier-Baumgarten syndrome

limbs and left upper limb sparing the face, lips and upper right limb). Differential clubbing may also be present.

Coronary cavernous fistula

There is abnormal communication between the coronary artery and any heart chamber or a cardiac vein. Most commonly the right coronary artery (RCA) is involved. When the communication is into right-sided chambers, coronary sinus, or pulmonary artery, it produces continuous murmur because of unrestricted flow throughout the cardiac cycle. The murmur is louder during diastole and site of maximum intensity depends on artery of origin and draining cavity. The RCA draining into right atrium or ventricle (RA or RV) produces murmur located along parasternal area, whereas circumflex artery and coronary sinus communication produces a murmur in left axilla.

Ruptured sinus of Valsalva aneurysm

Sinus of Valsalva aneurysm most commonly ruptures into the RA or RV which are low-pressure chambers producing a continuous murmur. Murmur is classically louder in diastole and this feature can be used to differentiate from the murmur of PDA or arteriovenous (AV) fistula. It is generally located along the left parasternal border with the site of loudest murmur and thrill suggesting the possible location of rupture. Systolic suppression of murmur may occur because of mechanical narrowing of fistulous tract during systole, and the murmur may be only diastolic in case of rupture into the left ventricle (LV).

Aortic-pulmonary window

This rare congenital disorder entails a communication between aorta and pulmonary artery above semilunar valves. The continuous murmur of aortic-pulmonary window is difficult to distinguish from that of PDA. It is usually present along left parasternal border, much lower than the expected site of PDA murmur. Like PDA, the length of diastolic component of the murmur depends upon pulmonary artery pressure and pulmonary vascular resistance.

Anomalous left coronary artery origin from pulmonary artery

This disorder generally presents in the infancy with LV dysfunction, but presentation can be rarely delated till adulthood. Retrograde flow through the anomalous artery results in a left-to-right shunt from aorta into pulmonary artery often producing a continuous murmur.

Coarctation of aorta

Coarctation of aorta is typically associated with an ejection systolic murmur, but continuous murmurs can be heard in severe coarctation when collateral circulation is insufficient producing a continuous systolic and diastolic gradient. The murmur is heard posteriorly at interscapular area with maximum intensity in systole. Some patients with coarctation can have a continuous murmur from dilated intercostal collateral arteries.

Pulmonary artery atresia

In pulmonary artery atresia, pulmonary blood flow occurs through dilated bronchial collaterals. Bronchial collaterals originate in the aorta (systemic circulation) and communicate with pulmonary circulation producing continuous murmur which is widely distributed on the surface of chest with characteristics similar to that of ductus arteriosus.

Truncus arteriosus

It can sometimes lead to continuous murmur due to low pulmonary resistance and high pulmonary blood flow.

Pulmonary artery stenosis

Pulmonary artery stenosis usually causes systolic murmur but when severe stenosis is present, the murmur extends into diastole producing continuous murmur. It is heard over interscapular, infraclavicular, or axillary area and rarely it may be precordial. This is often associated with William's syndrome or rubella syndrome.

Severe arterial stenosis

Arterial stenosis >80% can cause continuous murmur at the site of stenosis in carotid, subclavian, femoral, or renal arteries with louder systolic and soft diastolic component. With formation of collaterals, continuous murmur of stenosis may disappear completely.

Systemic arteriovenous fistulas

Systemic AV fistulas may be congenital, arise after trauma, iatrogenic injury, or spontaneously. Continuous murmur is audible at the site of fistula, is rustling in character and is associated with a bruit. Compression of the artery proximal to AV fistula results in reflex bradycardia because of increase in peripheral resistance and afterload leading to reflex bradycardia. This is known as Nicoladoni–Branham sign.

Pulmonary arteriovenous fistula

Pulmonary AV fistulas are frequently associated with hereditary telangiectasia and can cause cyanosis. The murmur is continuous which is superficially heard over the location of pulmonary AV malformations in chest frequently in lower lobes. The phenomenon of orthodeoxia is characteristic.

Mammary soufflé

Mammary soufflé is an innocent murmur associated with increased mammary blood flow in the late pregnancy or postpartum period and disappears at the end of lactation. Murmur is best heard directly over the breast and may be continuous or only systolic. This innocent murmur is usually of higher frequency and louder in systole.

The venous hum

It is a physiological continuous murmur frequently heard in young and healthy children. It is a low-pitch murmur which is louder in diastole, has a harsh and noisy character and is best heard in sitting position with the bell of the stethoscope. Venous hum is heard in the supraclavicular fossa near the sternocleidomastoid muscle and is more likely to be heard on the right side because the right jugular vein is larger and drains about 2/3rd of intracranial venous blood. The murmur disappears after compression of jugular vein or changing position from sitting to supine, while increases with rotation

of the head to the opposite side. Anemia, thyrotoxicosis and other conditions that increase cardiac output may exacerbate cervical venous hum.

Cruveilhier–Baumgarten syndrome

It is a continuous murmur may be heard in epigastric or paraumbilical area in portal venous hypertension due to dilated paraumbilical venous collaterals.

What is the diagnosis and how to justify the clinical findings?

The first possibility is RSOV aneurysm. The possible site of rupture is into RA.

RSOV causes wide pulse pressure with bounding pulse because of diastolic reversal of flow in aorta through ruptured sinus. When the aneurysm ruptures into RA or RV, it leads to raised JVP. Increased blood flow into right chambers of heart leads to pedal edema, abdominal distension, hepatomegaly, and raised JVP. The left-to-right shunt results in increasing pulmonary blood flow. Volume overload of the left side results in left heart failure presenting as cardiomegaly and pulmonary edema. Ill-sustained left parasternal pulsations can be explained by the volume overload of RV. RSOV produces continuous murmur because of continuous flow from the aorta into the receiving chamber.

Describe the electrocardiogram and X-ray of patient

Electrocardiogram (ECG) [Figure 1] shows sinus rhythm with a heart rate of 94 beats/min. The QRS axis is -30°. PR interval, QRS duration, and QT intervals are normal. There is no evidence of chamber enlargement. There are no significant ST-T wave changes.

Chest X-ray [Figure 2] shows cardiomegaly, left ventricular enlargement, prominent pulmonary artery segment, pulmonary plethora, and pulmonary venous hypertension. RA does not seem much enlarged. Costophrenic angles are clear.

Describe the echocardiographic views and findings in this patient

Modified parasternal short-axis, apical-five chamber, suprasternal long-axis, and apical four-chamber views are shown here [Figures 3-6]. Aneurysm of sinus of Valsalva arising from the noncoronary cusp and protruding into RA is seen in the form of wind-sock. RA is dilated along with the presence of moderate tricuspid regurgitation.

What is sinus of Valsalva aneurysm?

There are three-specialized dilatations, known as sinuses of Valsalva in aortic lumen near the aortic valve which are named in relation to their valve cusps. From the right and left coronary sinuses give rise to the right and left coronary arteries, respectively. However, posterior sinus does not have any coronary ostium, hence also called as noncoronary sinus.

Sinus of Valsalva aneurysm was first described by John Thurman.[1] Sinus of Valsalva aneurysm is a rare condition

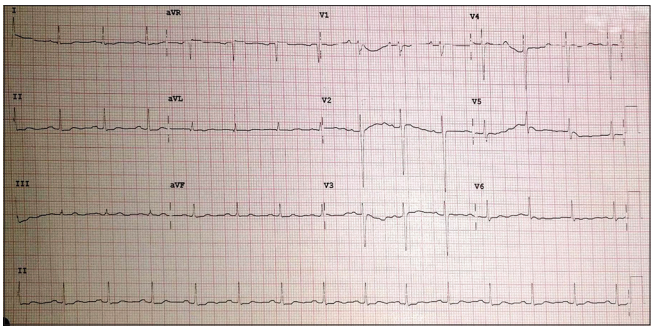

Figure 1: Electrocardiogram shows normal sinus rhythm with QRS axis of -30°. Electrocardiogram is not suggestive of any cardiac chamber enlargement. There is no heart block or evidence of myocardial ischemia in Electrocardiogram.

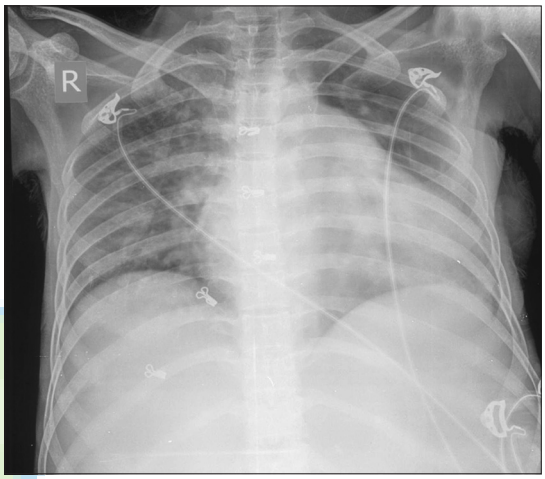

Figure 2: Chest radiograph showing cardiomegaly, right atrial enlargement, dilated pulmonary artery segment, pulmonary plethora, and pulmonary venous hypertension. Combination of these findings is strongly suggestive of ruptured sinus of Valsalva.

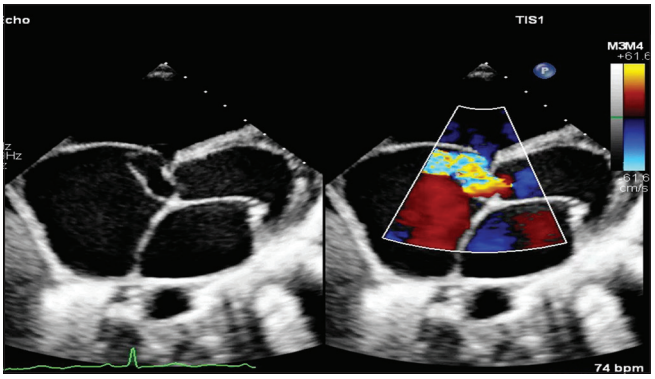

Figure 3: Modified parasternal short-axis echocardiographic image showing aneurysm of sinus of Valsalva arising from the noncoronary cusp and protruding into right atrium in the form of a wind-sock (arrow). Rupture at the tip is appreciated (arrowhead) leading to continuous turbulent flow into right atrium.

which results from separation between aortic media and annulus fibrosis.[2] It can be associated with Marfan's syndrome, syphilis, and infective endocarditis of aortic valve.

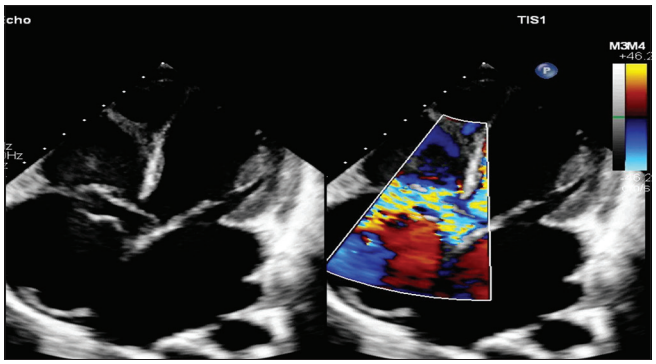

Figure 4: Modified apical-five chamber echocardiographic image showing sinus of Valsalva aneurysm (arrow) with rupture into right atrium just above the tricuspid valve.

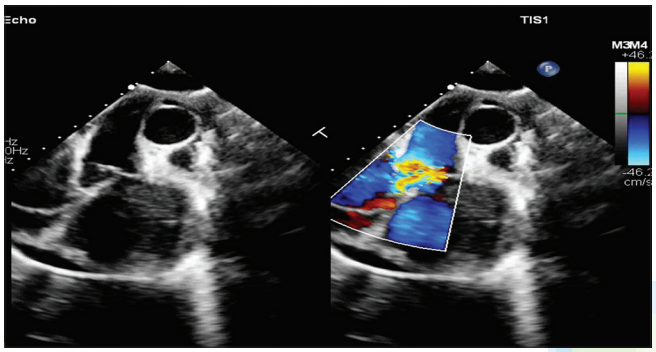

Figure 5: Suprasternal long-axis echocardiographic image showing sinus of Valsalva aneurysm with rupture into the right atrium.

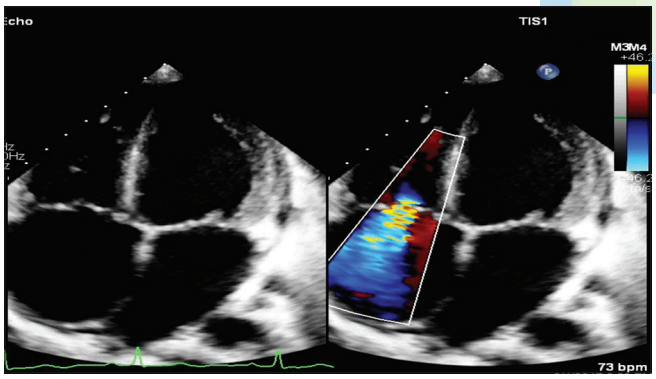

Figure 6: Apical four-chamber echocardiographic view showing prominent dilation of right atrium with bulging of inter-atrial septum to the left and moderate tricuspid regurgitation.

Acquired sinus of Valsalva aneurysm can result from surgery to aortic root or valve and coronary intervention.[3]

Sinus of Valsalva aneurysms originate most commonly from the right coronary sinus (70%–90%), followed by noncoronary sinus (10%–20%) and rarely from left sinus (<5%).[4] Aneurysm from right coronary sinus usually ruptures into RV and from noncoronary sinus into RA. Rupture of left coronary sinus is rare and may rupture into pericardial cavity. Modified Sakakibara classification system for RSOV is based on the chamber into which rupture occurs, and divides patients into 5 types.[5] RSOVs account for 1% of congenital anomalies of the heart and most of them involve single sinus, rarely >1 sinus is involved.

What is the natural history of sinus of Valsalva aneurysm?

Unruptured aneurysms are usually asymptomatic, often incidentally detected on echocardiography unless very large and compress mediastinal structures. Rupture typically occurs in young males after puberty but before the age of 30 years with male-to-female ratio of 4:1.[1] RSOV is five times more common in Asians as compared to Western population.[6] An aneurysm that ruptures acutely may present with acute-onset chest pain that resolves after some time but is soon followed by right-sided heart failure symptoms which progress relentlessly. Sometimes there may be a small rupture that enlarges over years to produce symptoms related to volume overload. Only one-third of the patients present with chest pain or severe dyspnea of acute onset, and the rest usually develop breathlessness gradually which keeps on worsening with time. Death from heart failure typically occurs within a year after rupture and so all the RSOV aneurysms should be repaired either percutaneously or surgically. However, small slowly progressive perforation can be associated with long survival. Sometimes sudden death can occur following perforation into pericardium causing tamponade or by rupture into base of interventricular septum, where AV node is located, causing complete heart block. Large aneurysms can compress coronary artery causing angina such as symptoms, most commonly RCA is compromised.

What are the finding you expect on physical examination of these patients?

The patients have wide pulse pressure and bounding pulses because of run off from the aorta to low-pressure chambers of heart. The rupture usually occurs into RV or RA, so JVP is elevated and signs of volume overload appear in the form of pedal edema, ascites, hepatomegaly, and later on crepitations in the chest because of pulmonary edema. Since the rupture from aortic sinus usually occurs into low-pressure chambers, a loud continuous murmur is audible at the precordium. Tetrad of features including continuous murmur, elevated pulsating JVP, bounding pulse along with a history of sudden-onset chest or epigastric pain is highly suggestive of RSOV aneurysm. We call this as "tetrad of RSOV."

What cardiac defects can be seen in association with sinus of Valsalva aneurysm?

Approximately, 50% of individuals have associated ventricular septal defect (VSD) which is the most common associated defect in patients of RSOV.[6] Supracristal type of VSD is more common in Asians, whereas perimembranous VSD is more frequently seen in Western population.[6] Aortic regurgitation can be seen in 25% of patients of RSOV.[6] Bicuspid aortic valve, coarctation of aorta, pulmonary stenosis, and atrial septal defect are less commonly associated. Approximately, 10% of Marfan's syndrome patients might have sinus of Valsalva aneurysm.

How will you evaluate and confirm your diagnosis in a suspected case of ruptured sinus of Valsalva aneurysm?

Diagnosis of RSOV is often established by the history and clinical examination. ECG may show evidence of chamber enlargement and features of volume overload of heart. Chest X-ray may show cardiomegaly with increased pulmonary vascularity. Blood cultures should be done for the detection of infective endocarditis.

Transthoracic two-dimensional echo shows the typical "windsock deformity" which can be better detected with transesophageal echocardiography (TEE) allowing identification of structural anomalies and shunt locations. Definitive diagnosis can be made with thoracic aortography with cardiac catheterization and ascending aortic angiography. Noninvasive tools such as cardiac computed tomography and magnetic resonance imaging may also be used.

What are the indications for intervention in ruptured sinus of Valsalva patients and what are the available treatment options?

All RSOV need definitive therapy. Patients often need initial stabilization and control of heart failure symptoms with diuretics, vasodilators, and inotropes. Treatment options include surgical repair and percutaneous device closure. The most important factors determining the choice of treatment are anatomic location and associated defects. All RSOV should be closed. Initially, the symptoms of heart failure may be controlled with medications, but eventually, symptoms worsen and may lead to death, often within 1 year from the rupture.

Surgical repair

Surgical repair is the gold standard of care. Acute and long-term results after surgery are excellent with hospital mortality of <5%. Mayo clinic experience reports a 95% survival after 20 years of surgical repair.[7] For isolated ROSV, catheter closure is the current practice. Surgery carries very good immediate and long-term results, but nowadays, it is restricted to the following situations: patients who have an associated VSD, especially when juxta-arterial, significant AR where an aortic valve resuspension or aortic valve replacement is needed, failure of catheter closure due to a large perforation, or prolonged/severe hemolysis after device placement.[8] Other indications include large RSOV with aortic end >12 mm, RSOV with multiple rupture sites, or those with any suspicion or evidence of infective endocarditis.

Catheter closure

Percutaneous closure of RSOV was first attempted by Cullen *et al*. in 1994 using Rashkind umbrella.[9] Catheter closure of RSOV has now become safe and feasible and it is the treatment of choice when there are no associated lesions that require surgery. About 70% of patients with RSOV are potentially eligible for catheter closure.[10] The common approach is to access both femoral vessels to create an AV loop. Most commonly used occlusion device is Amplatzer duct occluder (ADO), a nitinol-based plug, but other devices such as Amplatzer septal occluder and muscular VSD closure device have also been used successfully. Sizing of the RSOV and of the device is done with a combination of angiography and TEE. If the ADO is used for closure, the size of the device should be 2–4 mm larger than the landing zone. If a muscular VSD device is used, the size should be equal to the landing zone, as these devices have a bigger rim than the ADO. Complications of catheter closure include device migration, hemolysis, encroachment of aortic valve leaflets, AV conduction disturbances, and coronary compromise. Other generic complications include infection, thromboembolic events, and internal bleeding.

Our patient was managed with diuretics and vasodilators initially followed by surgical repair of RSOV aneurysm because of insufficient margins required for the percutaneous closure.

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.

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Conflicts of interest

There are no conflicts of interest.

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