

**Case report****A Case of Perimembranous Ventricular Septal Defect with worsening clinical condition. Is a relook at the Echocardiography warranted?? – An Interesting Case Report****Dr. Kritika Sharma¹ | Dr Rohit Radhakishan Bunage² | Dr Roly Mishra³ | Dr Maulik Parekh⁴ | Dr Anvay Mulay⁵**

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Introduction:

An aneurysm of Sinus of Valsalva (most commonly right sinus) is a rare finding, which may be congenital or acquired. A ruptured Sinus of Valsalva aneurysm (RSOVA) may lead to serious complications including myocardial ischemia, obstruction of right ventricular outflow tract and cardiac tamponade and warrants urgent repair – either surgical or percutaneous. We present a case which was pre-diagnosed with VSD and referred to our centre where clinical

evaluation revealed continuous murmur and on transthoracic echocardiography a large left to right shunt with continuous flow (predominant diastolic flow) was observed. It was only diagnosed as a RSOVA when Transesophageal echocardiography (TEE) was performed when patient was taken to Cath lab for urgent percutaneous device closure of septal defect. Although rare, a missed diagnosis of RSOVA can be catastrophic as it has serious consequences and warrants urgent repair – either

surgical or percutaneous. The role of TEE cannot be undermined in its diagnosis.

Case report :

A 38yrs old patient with complaints of sudden onset intermittent chest pain, with cough, palpitations and fever from 4 days was diagnosed with aneurysmal ventricular septal defect (VSD) with large left to right shunt and was then referred to our hospital. On evaluation, patient had a loud continuous murmur. Chest X-Ray was suggestive of cardiomegaly; ECG revealed ventricular bigeminy and Transthoracic Echocardiography revealed a large left to right shunt with continuous flow (predominant diastolic flow) which raised suspicion as the flow of VSD has a predominant systolic component and a higher velocity.

He was explained the need for urgent surgical repair or device closure and explained in detail about risks and benefits of each. Subsequently, a combined decision was taken to proceed with trans-catheter device closure which was performed under general anaesthesia with TEE guidance.

After a detailed pre-anaesthesia evaluation, anaesthesia consent was taken. After shifting to cath lab, a wide bore 16g peripheral line was secured. Injection fentanyl 100 µg was given intravenously followed by induction using etomidate 16 mg and atracurium 40 mg IV was used as muscle relaxant. Trachea was intubated with 8.0 mm ID cuffed endotracheal tube and fixed at 22 cm. Anaesthesia was maintained with oxygen, air, and sevoflurane. End-tidal CO₂ monitoring was attached. Transesophageal echocardiography (TEE) probe was inserted gently after lubricating with lignocaine gel avoiding any trauma. Central line was secured in right Internal jugular vein under ultrasonographic guidance. Intraoperatively, multiplane colour doppler, two-dimensional and three-dimensional – TEE examination was performed which revealed RSOVA from right coronary sinus into right ventricle resulting in severe left to right shunt along with a small VSD of 3mm which was not significant. On mid esophageal aortic valve long

axis (ME AV LAX) view (clip 1), RSOVA could be seen arising from right coronary sinus which was further confirmed on applying colour doppler (clip 2). After discussion with cardiologist and with standby surgical team and perfusionist, RSOV Device Closure was done under TEE and fluoroscopy guidance with 14 x 16 ADO1 Duct Occluder in hybrid theater. At the end of procedure, the device was seen sitting well inside the sinus of Valsalva aneurysm on TEE in ME AV LAX view (clip 3) which was confirmed on 3d examination (clip 4) and no impingement of AV leaflets with normal LV function. Optimal position of the device was further confirmed on fluoroscopy in aortogram (clip 5). The procedure was uneventful and patient was extubated at the end of procedure after reversal. He was shifted to ICU after the procedure and his postoperative course was uneventful.

Discussion:

Sinus of Valsalva aneurysm (SVA) is a rare cardiac anomaly which may be congenital or acquired and is more prevalent in men and Asian population. Congenital aneurysms result from defect in the aortic media and is usually seen in patients with Marfans and Ehlers-Danlos syndrome, while acquired aneurysms may result from infections like bacterial endocarditis [1]. Other cardiac anomalies found in association with ruptured sinus of Valsalva (RSOV) includes ventricular septal defects (30-60%), aortic insufficiency (20-30%), bicuspid aortic valve (10%) and coronary anomalies [2]. In our case, the patient had an associated small VSD.

The congenital SVA most commonly arise from right coronary sinus (65-85%) as in our case, followed by the non-coronary sinus and left coronary sinus. [3]

The right coronary sinus ruptures usually into the right ventricle resulting in left to right shunt, non-coronary sinus into the right atrium and left coronary sinuses though rare may rupture into pericardium causing tamponade and lead to serious complications. Based on the site of rupture, ruptured SVA is classified (modified

Sakakibara's classification) into 5 types as follows: :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 III, rupture into the right atrium or right ventricle near or at the tricuspid annulus; type IV, rupture into the right atrium; and type V, other rare conditions, such as rupture into the left atrium, left ventricle, or pulmonary artery. [4]

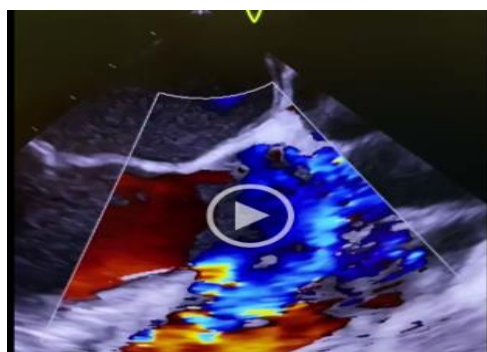
Unruptured aneurysms may be asymptomatic or can cause obstruction to right ventricular outflow tract, complete heart block or myocardial ischemia while ruptured aneurysms may present acutely with dyspnea and chest pain due to severe left to right shunt or may have gradually worsening dyspnea, fatigue, chest pain and peripheral oedema over months to years due to compensatory mechanisms. [5]

The shunting of blood through the aneurysm may lead to a decrease in diastolic coronary perfusion and result in ischemia.

Echocardiography, particularly transesophageal echocardiography (TEE) and magnetic resonance imaging (MRI) are useful for diagnosis and to rule out associated cardiac anomalies.

The treatment options include surgical repair or device closure.

Although, surgical correction has been described as the treatment of choice, lately isolated RSOVs have been successfully closed percutaneously using transcatheter devices.



Clip 1

https://drive.google.com/file/d/1AXqVG_vvALgbrRw9tMJRiw8sOr7w76jt/view



Clip 2

https://drive.google.com/file/d/1WJFjO2AM_t9zeZSCqqdvJDpme1HIGMp/view



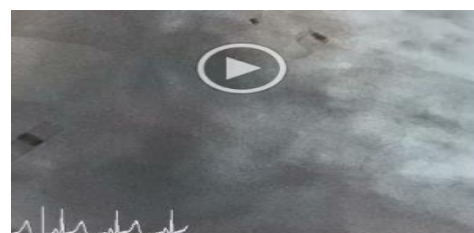
Clip 3

https://drive.google.com/file/d/11aPWh4z8uiq_1wK2GOX9DBrkY2EvPH54/view



Clip 4

<https://drive.google.com/file/d/1GCUQOoGgHSWXjngKCXt6TvVFwta77DLx/view>



Clip 5

<https://drive.google.com/file/d/1GCUQOoGgHSWXjngKCXt6TvVFwta77DLx/view>

On echocardiography, aneurysmal appearance of the involved sinus and the presence of a “windsock,” protruding into the receiving chamber are characteristic of RSOV as noted in Clip 1 and Clip 2. [6] Due to similar location, cases have been reported where RSOV was misdiagnosed as VSD, as in our case. A ruptured SVA may mimic perimembranous VSD if it ruptures in to RV close to tricuspid valve or subpulmonic VSD if close to the pulmonary valve. The ME aortic valve long axis and short axis view and the RV inflow outflow view on TEE can be used to distinguish between VSD and RSOV based on the location. Colour flow and continuous wave Doppler can be used to confirm the diagnosis. While in VSD, the flow occurs predominantly in systole from LV to RV with high velocity; the flow in RSOV has a high velocity throughout cardiac cycle with a prominent diastolic component with a decelerating slope similar to Aortic Regurgitation (AR). A holodiastolic flow reversal may also be seen in descending aorta similar to AR. 3D TEE represents an important adjunctive tool to demonstrate the RSOV with better delineation of its characteristics such as the site of rupture into the cardiac chambers, the size, shape of the defect, associated defects such as ventricular septal defect; however, it has lesser frame rate compared with 2DTEE as seen in Clip 4. [6] After device placement, appropriate position of device is confirmed on TEE and any leak is ruled out as seen in clip 3. This is further confirmed on aortogram (Clip 5).

In our case, the patient was misdiagnosed as an isolated VSD initially, but detailed intraoperative TEE with colour Doppler profile and continuous murmur were suggestive of RSOV which was the main cause of symptoms along with a small, VSD. The size of defect can also be measured on TEE which helps in selection of appropriate device for closure.

Conclusion:

This case highlights the important of Transesophageal echocardiography in the diagnosis of

RSOV (which can be easily misdiagnosed as a VSD) and in proper selection of device for closure.

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