Role of Transesophageal Echocardiography in a Case of Unruptured Sinus of Valsalva Aneurysm Causing Complete Heart Block

Rajan Anand1, Debadas Bagchi2, Nishita Palraj3, Sai S Dharmapuri4

ABSTRACT

Sinus of Valsalva aneurysm is an uncommon disease. Rarely, the aneurysm presents with evidence of obstruction of the right ventricular outflow tract, aortic insufficiency, coronary artery compression, or conduction abnormalities in the absence of rupture. We report a case with an unruptured aneurysm of the right sinus of Valsalva extending into the interventricular septum causing complete heart block, which was managed successfully by surgery. We emphasize the rarity of this pathology and highlighting the importance of multipane transesophageal echocardiography in its assessment, providing a complete anatomic functional characterization, allowing precise identification of structural anomalies, valve abnormalities, and cardiac function, thereby guiding appropriate surgical management.

Keywords: Complete heart block, Sinus of Valsalva aneurysm, Transesophageal echocardiography.

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BACKGROUND

Sinus of Valsalva aneurysm (SVA) is an uncommon disease with an incidence ranging from 0.1 to 3.5%. It is more frequently congenital, but it may also be an acquired condition. Aneurysmal dilatation and progressive enlargement are one of the sinuses of Valsalva, most often the right coronary sinus normally goes undetected during life until rupture occurs.1 Rarely, the aneurysm presents with evidence of obstruction of the right ventricular outflow tract, aortic insufficiency, conduction abnormalities, or coronary artery compression in the absence of rupture.2 We report a case with an unruptured aneurysm of the right sinus of Valsalva extending into the interventricular septum causing complete heart block, highlighting the role of transesophageal echocardiography (TEE) in perioperative management.

CASE DESCRIPTION

A 27-year-old man presented with a short history of giddiness, presyncope, and worsening dyspnea, was in NYHA class IV. There was no history of recent chest pain or injury. Auscultation revealed a grade IV diastolic murmur in the aortic area. Chest X-ray demonstrated moderate cardiomegaly prominent aortic knuckle and prominent bronchovascular marking. Electrocardiogram revealed complete heart block with an atrial rate of 150/minute and ventricular rate of 45/minute. Transthoracic two-dimensional echocardiography (TTE) and TEE demonstrated an unruptured aneurysm of the right coronary sinus, burrowing into basal septum with mild aortic regurgitation (AR) but competent aortic valve, mild mitral regurgitation (MR), and trivial tricuspid regurgitation (TR). Cardiac magnetic resonance (CMR) imaging confirmed the presence of a giant unruptured aneurysm arising from the right sinus of Valsalva measuring 9.5 × 6 cm with opening measures of about 29 mm. The aneurysm was involving the basal and mid interventricular septum causing compression of the left ventricle (LV) inlet, LV outlet, and right ventricle (RV) inlet leading to mechanical mitral and tricuspid stenosis. The left ventricle was normal in size and has moderately reduced systolic function. The right ventricle was normal in size and shape and has normal function. The patient was stabilized in the coronary care unit with the help of inotropic infusion (dobutamine 5 μg/kg/minute), elective intubation, temporary pacemaker implantation, and intravenous furosemide and then referred for cardiac surgery.

After induction of anesthesia, TEE examination was performed using a 6 VT ultrasound probe and GE vivid E9 ultrasound machine. Transesophageal echocardiography confirmed the preoperative findings and revealed the unruptured aneurysm of the right coronary sinus with extension into the interventricular septum with characteristic systolic “emptying” and diastolic expansion into the left ventricular outflow tract (Figs 1 to 3).

At operation, a large SVA was confirmed. It was originating from the right coronary sinus, involving the entire right coronary sinus, entering into the interventricular septum, and bulging into the LV and right atrium with a closed pouch. The walls of the sac were intact. The aortic valve was tricuspid and normal except that the right coronary cusp was noticeably redundant and was sagging into the left ventricular cavity and right atrium. The opening of the aneurysm was closed with a Gor-Tex patch and 5-0 prolene suture.

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Any distortion of the aortic valve was avoided and there was no intraoperative evidence of significant aortic incompetence. Two atrial and two ventricular pacing wires were inserted. The patient was weaned off from cardiopulmonary on dobutamine and isoprenaline infusion with AV sequential pacing on. Intraoperative TEE demonstrated no leak from the patch. Partially thrombosed residual sinus of Valsalva pouch protruding into the LV cavity and partially into the right atrium was seen with no significant RV inflow and left ventricle outflow tract (LVOT) obstruction. Aortic and mitral valves were normal, without any evidence of stenosis or regurgitation (Figs 4 to 7). The left ventricle was globally hypokinetic with an ejection fraction of 15–20%. The postoperative course in ICU was prolonged on account of severe LV dysfunction requiring high inotropic support in the form of adrenaline infusion. A permanent pacemaker was implanted through right subclavian access on day 18 following surgery, in view of the persistent features of complete heart block. Post pacemaker implantation recovery was uneventful. Serial TTE was done, which revealed a substantial reduction in the size of partially thrombosed residual sinus of Valsalva pouch protruding into the LV cavity and partially into the right atrium without any flow and no significant RV inflow and LVOT obstruction with normal aortic and mitral valves and improvement in left ventricular function. The patient was discharged in stable condition on day 21. At a 3-month follow-up, the patient remained asymptomatic and doing well with TTE findings of further reduction in the size of residual thrombosed SOV pouch with normal left ventricular function (Figs 8 to 12).

**Discussion**

Unruptured aneurysm of sinus of Valsalva is asymptomatic usually and often found incidentally. However, the condition can manifest by distortion or compression of adjacent structures as aortic insufficiency, right ventricular outflow tract obstruction, tricuspid stenosis and regurgitation, myocardial ischemia or infarction, conduction disturbances with septal penetration, mediastinal mass, infective endocarditis, thromboembolism, and rupture. In our patient, the unruptured aneurysm had extended into the interventricular septum and, presumably through compression, had...
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Fig. 7: Continuous-wave Doppler evaluation across aortic valve in deep transgastric long-axis view at sector angle of 0° showing a gradient of 23/9 mm Hg

Fig. 6: A two-dimensional deep transgastric long-axis view at sector angle of 0° showing partially thrombosed residual sinus of Valsalva pouch protruding into the left ventricle cavity with minimal turbulence across the aortic valve. LA, left atrium; LV, left ventricle; AV, aortic valve

Fig. 8: Apical four-chamber view in transthoracic echocardiography showing residual sinus of Valsalva pouch seen in basal and mid-region of the interventricular septum, protruding into the left ventricle and right atrium, on postoperative day 2. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle

Fig. 9: Apical four-chamber view in transthoracic echocardiography showing substantial reduction of thrombosed residual sinus of Valsalva pouch on postoperative day 4. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle

Fig. 10: Parasternal long-axis view in transthoracic echocardiography showing a substantial reduction in the size of residual sinus of Valsalva pouch with laminar flow across the mitral valve and mild aortic regurgitation. LA, left atrium; LV, left ventricle; MV, mitral valve; AV, aortic valve; AA, ascending aorta
compromised normal AV nodal/his bundle function and resulted in complete heart block. Case reports of the latter association are very rare. Therefore, the sinus of the Valsalva aneurysm deserves to be considered as a rare cause of complete heart block. The diagnostic modalities include TTE, TEE, multidetector cardiac computed tomography (MDCT), angiography, cardiac catheterization, and CMR imaging. Sinus of Valsalva aneurysm can be well visualized with TTE provided that acoustic windows are adequate. However, in patients with poor acoustic windows and equivocal findings on TTE, TEE is required in up to 25% of patients to further delineate the anatomy of the sinuses and the relationship of the aneurysm to surrounding structures. Multidetector cardiac computed tomography and CMR are used in conjunction with echocardiography for a comprehensive assessment of the entire aorta to confirm aortic measurements and evaluate the relationship of the aneurysm with the cardiac chambers and coronary arteries. It also enables the assessment of SVA complications (e.g., chamber compression caused by SVAs). An advantage of CMR over MDCT is that it can provide a functional assessment of cardiac function and flow, in addition to cardiac and aortic anatomy. Transesophageal echocardiography is the imaging modality of choice during definitive intervention for aneurysms.

Precardiopulmonary bypass multiplane TEE provides useful information to confirm the preoperative findings, provide conclusive information regarding aneurysm, and allow precise identification of structural anomalies and shunt locations for appropriate surgical management. The most useful TEE views are mid-esophageal aortic valve short-axis view, mid-esophageal aortic valve long-axis view, mid-esophageal five-chamber view, mid-esophageal right ventricular inflow outflow view, and deep transgastric long-axis view. Two-dimensional TEE provides us information regarding the single or multiple sinus involvement, location and size of the aneurysm, identification of cardiac chamber where aneurysm expands into, presence of wristock deformity of the aneurysm (which expands during systole), the sign of rupture (echo dropout at the tip of the wristock), presence of sinus thrombus in addition to dilatation and function of the LV and RV. Discontinuity of the wall is a sign of rupture, and color Doppler imaging helps delineate the fistulous connection. Furthermore, the color Doppler will give us valuable information regarding the flow characteristics (turbulent or laminar), shunt direction and help us in the assessment of associated valvular regurgitation and stenosis. Spectral Doppler helps us to determine peak/mean pressure gradients across intracardic fistulae, the timing, and the duration of fistula flow. In addition, the spectral Doppler gives us valuable information regarding peak/mean pressure gradients across the mitral valve, tricuspid valve, and aortic valve to assess and quantify the inflow and outflow obstruction of the LV and RV caused by the aneurysm. Doppler imaging is very useful to discriminate associated cardiac structural abnormalities such as ventricular or atrial septal defect patent foramen ovale, coarctation of the aorta, right ventricular outflow tract obstruction, Pulmonary valve stenosis and, diagnosing functional AR, compressive effects on cardiac structures, thrombus or vegetation formation or significant coexisting valvular dysfunction. The addition of 3D imaging enables better evaluation of the size and location of the aneurysm, relationship with adjacent cardiac structures, assessment of associated cardiac defects, as well as potential complications of the aneurysm that otherwise are not easily recognized by conventional 2D imaging. Real-time 3D TEE imaging has an important role in guiding surgical intervention/repair of SVAs, in terms of delineating the extent of repair required, helping ensure optimal surgical results, and avoiding complications.

Surgical repair of unruptured ASV should be advised in symptomatic cases, in cases of rupture, and in incidentally discovered cases at the time of surgery for other cardiac pathology unless specific contraindications exist. Surgical repair may involve adjuvant procedures like aortic valve-sparing procedures or aortic valve or aortic root replacement in cases of associated AR or aortic root distortion.

Post repair TEE examination helps us in the assessment of the adequacy of the surgical repair, stenosis or regurgitation of the valves, optimization of atroventricular delay using transmirtal and pulmonary venous flow with sequential ventricular pacing, evaluation of LV and RV function which guides to optimize hemodynamics by titration of inotropes and vasopressors, in addition, to assist in adequate dearing.

**Conclusion**

We emphasize the rarity of this pathology and the value of multiplane TEE providing conclusive information regarding
aneurysm, allowing precise identification of structural anomalies, shunt locations, valve abnormalities, and cardiac function for comprehensive perioperative assessment and guiding appropriate surgical management.

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