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Case Report

Rupture Sinus of Valsalva into Right Ventricle with Classical Windsock Deformity on Transthoracic Echocardiography: A Case Report

Kumari Priti[1]* and Tapan K Matia[2]

[1]*MD, DM Cardiology, Consultant Interventional Cardiologist, Narayana Hrudyalaya, Durgapur, India*

[2]*MD, DNB Cardiology, Senior Consultant Interventional Cardiologist, Narayana Hrudyalaya, Durgapur,India*

***Corresponding Author**: Kumari Priti, MD, DM Cardiology, Consultant Interventional Cardiologist, Narayana Hrudyalaya, Durgapur, India.

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Abstract

We describe a case of ruptured aneurysm of sinus of Valsalva (RSOV) with classical windsock deformity diagnosed by transthoracic two-dimensional echocardiography which allowed prompt diagnosis in a patient with a recent onset of continuous murmur and acute right congestive heart failure.

Keywords: *Ruptured Sinus of Valsalva; Windsock Deformity; Transthoracic Echocardiography*

Introduction

Aneurysm of sinus of Valsalva is a rare anomaly accounting for less than 1% of congenital cardiac anomalies [1]. RSOV arises from a congenital defect of the aortic media or due to damage caused by bacterial endocarditis

Case Report

A 52-year-old man presented with a history of sudden onset dyspnoea NYHA III. Examination revealed tachycardia, a wide pulse pressure and a continuous murmur across the precordium. 12 lead ECG demonstrated LVH with non-specific ST-T changes. Chest X ray was normal. Transthoracic two-dimensional echocardiography (TTE) demonstrated ruptured sinus of valsalva (RSOV) arising from the right coronary sinus draining into the right ventricle with classical windsock deformity (Figure 1). Colour Doppler revealed continuous flow from aorta to right ventricle in systole and diastole (Figure 2) with moderate AR. Trans esophageal echocardiography (TEE) findings were consistent with TTE findings (Figure 3). Aortography showed a non-dilated aortic root and the direction of the shunt from the right coronary sinus towards the right ventricle outflow tract with 2+ aortic regurgitation. The configuration of the aneurysm was consistent with the contrast medium-filled sinus demonstrated by aortography, with a normal coronary sinus and a mobile "wind sock" aneurysm protruding into the right ventricle (Figure 4).

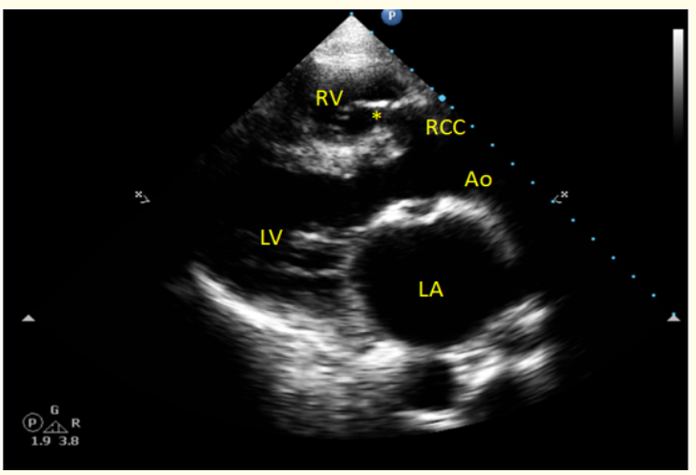

Figure 1: *2D transthoracic echocardiogram parasternal long axis view showing aneurysm of sinus of valsalva arising from right coronary sinus with classical windsock deformity. LA: Left Atrium; LV: Left Ventricle; Ao: Aorta; RCC: Right Coronary Sinus; RV: Right Ventricle; *: Classical Windsock Deformity of RSOV.*

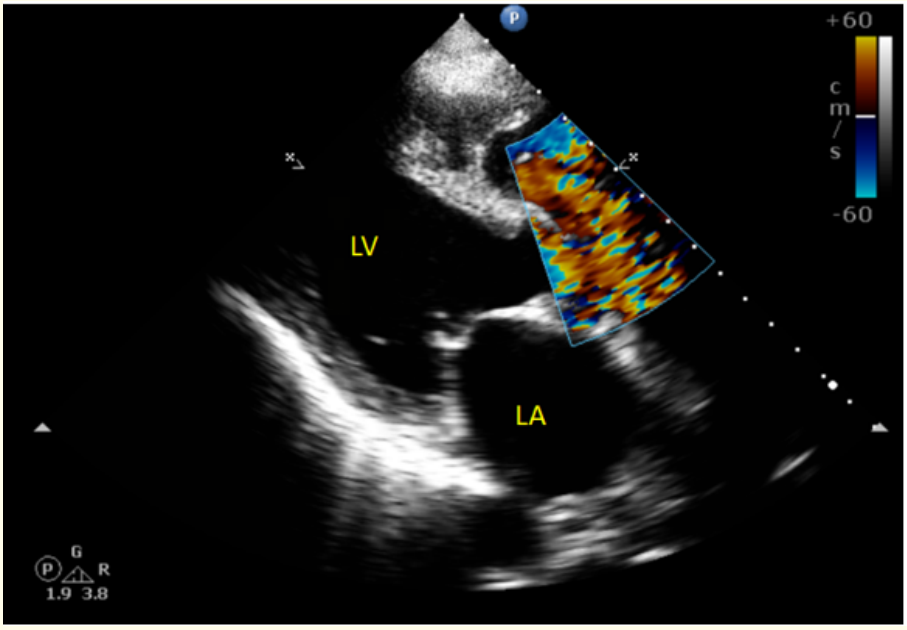

Figure 2: *2D transthoracic echocardiogram modified parasternal long axis view, colour doppler showing ruptured sinus of valsalva aneurysm arising from right coronary sinus draining into right ventricle with continuous flow in systole and diastole. LA: Left Atrium; LV: Left Ventricle.*

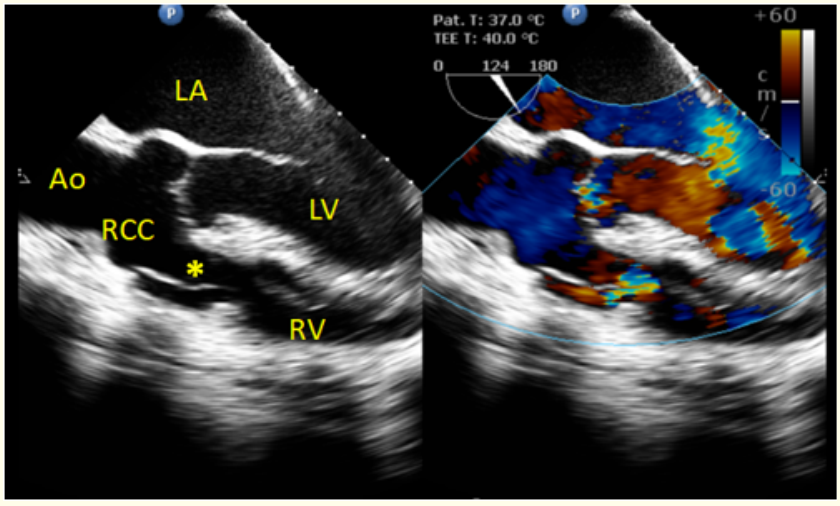

Figure 3: *Trans esophageal echocardiogram parasternal long axis view. Left panel showing 2D echo image of rupture sinus of valsalva aneurysm arising from right coronary sinus with classical wind sock deformity. Right panel- color Doppler showing shunting of blood from right coronary sinus to right ventricle. LA: Left Atrium; LV: Left Ventricle; Ao: Aorta; RCC: Right Coronary Sinus; RV: Right Ventricle; *: Classical Windsock Deformity of RSOV.*

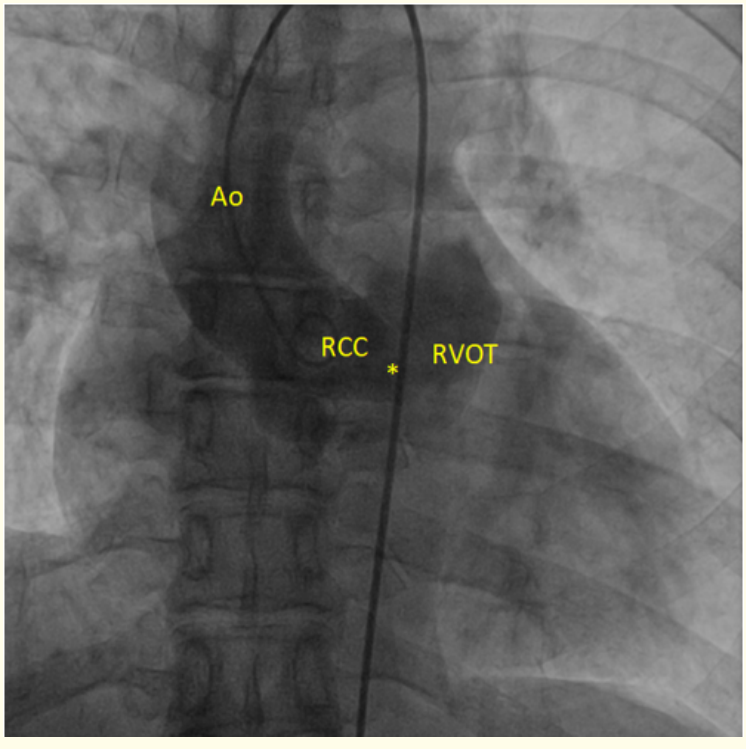

Figure 4: *Aortogram with RAO (right anterior oblique) 40° and caudal 20° view showing sinus of Valsalva aneurysm projecting into the right ventricle outflow tract with shunting of contrast from right coronary sinus into right ventricle outflow tract. Ao: Aorta; RCC: Right Coronary Sinus; RVOT: Right Ventricle Outflow Tract; *: Ruptured Sinus of Valsalva Aneurysm.*

Coronary arteries were normal. Mean pulmonary artery pressure (PAP) and Qp/Qs measured by cardiac catheterization were 34 mmHg and 1.8 respectively.

The suspicion of infective endocarditis was excluded on the basis of patient's history and laboratory analysis.

Discussion

Before the advent of echocardiography, diagnosis of RSOV in a living patient was rare, with most of the reports coming from autopsy or surgery [1]. Now a days, diagnosis is possible with both TTE and TEE. In the patient presented, TTE provided very detailed information of ruptured aneurysm of sinus of valsalva with classical windsock deformity consistent with TEE findings. Classical windsock deformity is usually very uncommon to be seen on transthoracic echocardiogram. Angiography was not done for diagnosis but to exclude coronary stenosis.

A sinus of Valsalva aneurysm (SOVA) is an enlargement of the aortic root area between the aortic valve annulus and the sinotubular ridge [2]. SOVAs can be either acquired damage or congenital defect in aortic media. In congenital SOVA, the pathology is frequently associated with Marfan's syndrome, Ehlers-Danlos syndrome, or other connective tissue disorders [3,4]. Acquired SOVA can be due to infections (infective endocarditis), chest trauma, vasculitis diseases, and iatrogenic injury during aortic valve replacement [1,5-7].

Estimated prevalence of SOVA is approximately 0.09% of the general population and comprise 0.1% to 3.5% of all congenital heart defects [5,8]. The anomaly usually occurs in isolation but may coexist with ventricular septal defect or aortic valve regurgitation in about 30 - 40% of patients [9,10]. It is more prevalent in men and people of Asian descent [9]. SOVAs usually affect right coronary sinus, noncoronary sinus, and left coronary sinus in descending order. 90 - 95% of these congenital aneurysms originates in the right or non-coronary sinus. Complications of SOVA include aortic regurgitation, coronary artery flow compromise, arrhythmias, and rupture [11].

Most non-ruptured SOVAs are asymptomatic. Ruptures typically occur between 20 and 40 years of age [12]. Ruptured aneurysm of sinus of valsalva invariably causes deterioration in heart function and has a rapid downhill course if uncorrected. They may present with substernal chest pain, abdominal pain, mild to severe dyspnea or symptoms of acute heart failure, cardiac tamponade, hemodynamic compromise, and even sudden cardiac death. Aneurysm arising in the non-coronary sinus almost always rupture into the right atrium, while those arising in the right coronary sinus generally communicate with the right ventricle and occasionally with the right atrium. Left SOVA rupture into left atrium or left ventricular outflow tract [13]. However, rupture may also occur into the pericardium, the pleural space, or the left heart chambers [14]. Sakakibara and Konno classification defines types I-IV according to the position of the aneurysm sac relative to the right or non-coronary aortic valve cusps [15].

Definitive diagnosis of RSOV is usually performed with sufficient accuracy using a TTE but sometimes requires a transoesophageal echocardiogram (TEE) or cardiac catheterization if the echo images are suboptimal or additional lesions need to be defined especially in cases caused by endocarditis. Details of involvement of other coronary sinuses and the extent of involvement and damage to the surrounding structures especially the aortic valve cusps are best obtained by a TEE. In this era when device closure of RSOV is gaining popularity, detailed TEE evaluation for assessment of device closure include assessment of the maximum diameter of the aortic end of the RSOV, the minimum diameter and the length of the windsock, distance of the aortic end of the RSOV from the coronary ostium, presence of AR or VSD. Intraprocedural TEE can also facilitate the performance of percutaneous or surgical closure of defect by assessment of any residual defects after the correction.

Early surgical intervention is the treatment of choice for ruptured aneurysms. Transcatheter device closure is emerging as a surgical alternative. The first transcatheter closure of a SOVA was reported in 1994 by Cullen., *et al.* using a Rashkind umbrella [16].

Conclusion

RSOV requires early diagnosis and treatment. TTE plays an important role in the diagnosis of RSOV.

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