

## CASE REPORTS

# Ruptured Sinus of Valsalva Aneurysm with Subaortic Membrane causing Severe Left Ventricular Outflow Tract Obstruction

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## Abstract

Rupture sinus of Valsalva aneurysm (SVA) is an uncommonly encountered condition. It can present with wide range of manifestations from an asymptomatic murmur to cardiogenic shock. The case discussed in this report had a rare combination of ruptured SVA with subaortic membrane. Corrective cardiac surgery was advised, but due to financial constraints, the patient was not willing for surgery.

## Introduction

Sinus of Valsalva aneurysm is a rare cardiac anomaly with a prevalence of 1.09% of all congenital heart diseases in the Oriental population and 0.2% in the Western population.<sup>1</sup> Subaortic membrane is a fibrous membrane below the aortic valve with prevalence of 6.5% among acyanotic congenital heart disease patients.<sup>2</sup> Ruptured SVA and subaortic membrane individually being very rare, their combined presence is extremely rare. We present a rare case of a 27-year-old female with ruptured right SVA into right ventricular outflow tract (RVOT) with left to right shunt

and subaortic membrane causing severe left ventricular outflow tract obstruction.

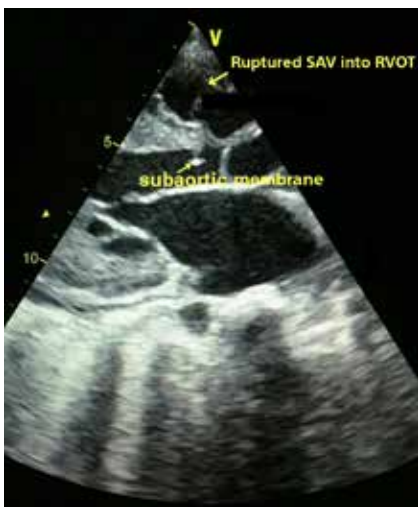
## Case Report

A 27-year-old female patient was admitted with complaints of shortness of breath, palpitations and retrosternal non radiating chest pain for seven days. She also had swelling both feet and paroxysmal nocturnal dyspnea. She did not have any significant past medical history. On examination she had respiratory rate of 22/min, regular bounding pulse of rate 112/min, blood

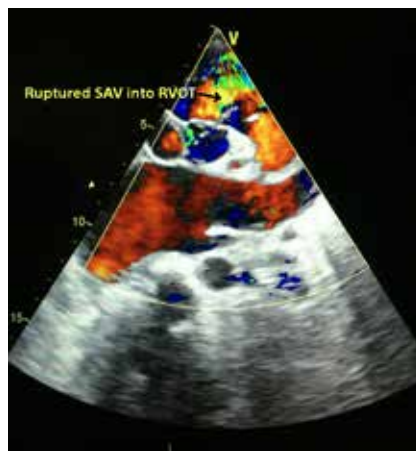
pressure 140/50 mm of Hg, prominent carotid pulsation, elevated jugular venous pressure, apical impulse in left 6<sup>th</sup> intercostal space (ICS), lateral to mid clavicular line and continuous thrill over left 3<sup>rd</sup> and 4<sup>th</sup> ICS. On auscultation there was a normal S1, normally split S2 with loud P2, and loud continuous murmur heard along upper left sternal border. Laboratory findings revealed WBC count 12500/ $\mu$ L with 70% neutrophils and 28% lymphocytes, hemoglobin 9.5 gm%, total bilirubin of 1.96 mg/dl, direct bilirubin of 0.64 mg/dl, AST 78 U/L, ALT 69 U/L, normal renal function and electrolytes. Electrocardiogram revealed left ventricular hypertrophy and X-ray chest showed cardiomegaly with left ventricular configuration. Transthoracic echocardiogram revealed 14 mm ruptured right SVA into RVOT causing left to right shunt. There was presence of subaortic membrane causing severe left ventricular outflow tract obstruction with peak pressure gradient of 117 mm of Hg, concentric left ventricular hypertrophy, minimal pericardial effusion, normal left ventricular systolic function, normal right atrial and right ventricular size (Figures 1 and 2). Patient was managed conservatively with diuretics, ACE inhibitors and digoxin as she refused for surgery due to financial constraints and was discharged in stable condition on medications.

## Discussion

Sinus of Valsalva aneurysms are rare but well described clinical entity. It is more common in Asians in whom it typically presents in adolescence and young adulthood. It is usually congenital in origin, most often caused by weakness at the junction



**Fig. 1: Two-dimensional transthoracic echocardiographic parasternal long axis view showing ruptured SVA into RVOT with subaortic membrane**



**Fig. 2: Two-dimensional transthoracic echocardiographic colour flow Doppler showing ruptured SVA into RVOT**

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of the aortic media and the annulus fibrosus. Unruptured SVA usually remain asymptomatic and are found incidentally during diagnostic studies. As the aneurysms are frequently clinically silent, their exact prevalence is unknown; however an autopsy study of 8,138 individuals suggests a prevalence of 0.09% in the general population.<sup>3</sup> Approximately 65-85% of SVA originate from the right sinus of Valsalva, while SVA originating from non-coronary (10-30%) and left sinuses (<5%) are less common.<sup>4</sup> SVA are usually diagnosed in the setting of clinical sequelae of a rupture. Majority of SVA arising from right coronary sinus rupture into right ventricle (RV) and less commonly to right atrium (RA), left ventricle (LV) or left atrium (LA). Those arising from non-coronary sinus rupture mostly into RA and less commonly in RV. Left aortic sinus aneurysms commonly rupture into LV or LA. Most ruptures develop well after puberty, between 20 and 40 years of age. The consequences of rupture depend on the size, the rapidity of the process and the chamber into which rupture occurs.

Sinus of Valsalva aneurysms may be associated with other cardiac anomalies

which include ventricular septal defect (30-60%), aortic insufficiency (25-45%), bicuspid aortic valve (10%), pulmonary stenosis, coarctation of the aorta, atrial septal defect, and subvalvular aneurysms.<sup>5,6</sup> Discrete subaortic membrane is a rare cardiac anomaly and its combined presence with SVA is extremely rare. There is no data on prevalence of subaortic membrane in SVA patients. Jain et al.<sup>7</sup> described a case of subaortic membrane with rupture of SVA presenting with infective endocarditis and its thromboembolic complications. Among 234 SVA patients who underwent surgical repair between January 1999 and December 2009, Guo et al.<sup>8</sup> from China described discrete subaortic membrane in seven (2.99%) patients. The resultant effects of subaortic membrane are essentially the same as valvular aortic stenosis: left ventricular hypertrophy from the pressure overload, myocardial ischemia, heart failure, and sudden death. In addition, a subaortic membrane may cause aortic insufficiency and permanent structural damage to the aortic valve due to alteration of left ventricular outflow dynamics. Hence surgical management with resection of the

subaortic membrane and repair of SVA is recommended. The rare combination of rupture of right SVA and subaortic membrane was found in our case. Our patient refused for surgery and was discharged on conservative management.

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