

**SINUS OF VALSALVA ANEURYSM DIAGNOSED IN YOUNG ADULT WITH  
MARFAN'S SYNDROME**Dr. Ravi Verma<sup>1</sup>, Dr. Chandan Bhatia<sup>2</sup> and Dr. Shikha Bhatia<sup>3\*</sup>Room No 102, 1<sup>st</sup> Floor Civil Hospital Palampur, Distt. Kangra, H.P.-176061.

\*Corresponding Author: Dr. Shikha Bhatia

Room No 102, 1<sup>st</sup> Floor Civil Hospital Palampur, Distt. Kangra, H.P.-176061.

Article Received on 29/10/2021

Article Revised on 19/11/2021

Article Accepted on 09/01/2022

**ABSTRACT**

Sinus of Valsalva (SOV) aneurysm is the dilatation of one or more of the aortic sinuses. These are rare and can be congenital or acquired. The cases of unruptured aneurysms may be asymptomatic and incidentally discovered, or they may be symptomatic with acute presentation due to mass effect on adjacent cardiac structures. Ruptured sinus of Valsalva aneurysms may result in an aortocardiatic shunt or manifest as progressive congestive heart failure, severe acute chest pain or cardiac arrest. Sinus of Valsalva aneurysms may have fatal complications however after treatment the prognosis is significantly improved. Thus, prompt and accurate diagnosis is important for treatment purpose. In the present case we illustrate the CT imaging features of sinus of Valsalva aneurysm in young adult with Marfan's syndrome.

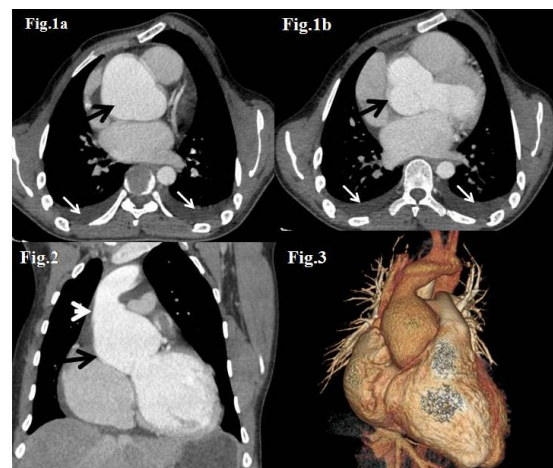
**KEYWORDS:** Sinus of Valsalva, aortic sinus, acute chest pain, aneurysm, Marfan's syndrome.**INTRODUCTION**

The sinuses of Valsalva are three small dilatations of wall of the aortic root arising between the annulus and the sinotubular ridge. Each sinus is associated with a corresponding cusp of the aortic valve namely right, left, or noncoronary cusp. The sinus of Valsalva (SOV) aneurysms are rare thoracic aortic aneurysms and can be either congenital or acquired. These were first described in 1839 by Hope et al.<sup>[1]</sup> The congenital aneurysms may result from an underlying deficiency of normal elastic tissue or focal elastic lamina weakness whereas acquired aneurysms are caused by infections, degenerative conditions i.e, atherosclerosis and cystic medial necrosis and trauma. True SOV aneurysm occurs above the aortic valve annulus and it has to be differentiated from a prolapsing aortic cusp, which occurs below the annulus.<sup>[2]</sup> In this case we illustrate the contrast enhanced CT imaging features of SOV aneurysm in correlation with echocardiography findings.

**CASE STUDY**

27years old male presented to the medicine department with the complaints of chest pain for past one month which was episodic and relieved at rest. It was associated with shortness of breath. There was no history of fever. On clinical examination the features of Marfan's syndrome were found. On echocardiography there was severe aortic regurgitation, aneurysmal dilation of aorta and moderate to severe left ventricular systolic dysfunction. For further workup aortic angiography was done with 64 slice multi-detector CT scanner which revealed aneurysmal dilatation of sinus of Valsalva

involving all the cusps measuring 65mm in diameter (black arrow in fig 1 and fig 2) with effacement of the sinotubular junction (arrowhead in fig2). No e/o any contrast leakage or thrombus was seen. Note was made of bilateral pleural effusion. Therefore, the final diagnosis was unruptured sinus of Valsalva aneurysm.



Contrast enhanced axial (fig 1a&b), oblique coronal (fig2) and volume rendered (fig 3) images of thorax showing aneurysmal dilation of sinus of Valsalva (black arrows) with effaced sinotubular junction (arrowhead). Note is made of bilateral pleural effusion (white arrow).

**DISCUSSION**

The sinus of Valsalva aneurysms have been found in 0.09% of 8138 autopsies and in 0.15%–3.5% of patients

who had undergone open heart surgery.<sup>[3-7]</sup> These aneurysms are three to four times more common in men than women.<sup>[4,6-8]</sup> The SOV aneurysms can be congenital or acquired. The congenital aneurysms occur due to localized weakness of the elastic lamina at the aortic root.<sup>[9,10]</sup> This is seen in connective tissue disorders such as Marfan's and Ehlers-Danlos syndromes as seen in our case. Acquired aneurysms are most commonly caused by infections such as bacterial endocarditis, syphilis, and tuberculosis; degenerative disorders like atherosclerosis and cystic medial necrosis; and trauma.

The clinical features of SOV aneurysms may be seen at any age.<sup>[9]</sup> It ranges from 4 to 96 years with mean age at presentation is 35.4 years. The symptoms at presentation are variable and depend on the size of the aneurysm. These include severe dyspnea, chest pain, and progressive congestive cardiac failure with signs of volume overload. However, the unruptured cases may remain asymptomatic and diagnosed incidentally for example due to abnormal cardiac contour on chest x-ray or abnormal murmurs during clinical examination.

Both ruptured and unruptured sinus of Valsalva aneurysms are associated multiple complications and can be potentially fatal. Therefore, early and accurate diagnosis may improve the prognosis. The echocardiography is the initial imaging modality used in the screening of cardiac abnormality. It is non invasive, cost effective and readily available, however for the detailed anatomy, extent of the aneurysm and its associated complications the cross sectional imaging modalities like cardiac CT or cardiac MR are done. These help in treatment planning which further improve the prognosis. Therefore, the detailed knowledge of the anatomy of the aortic root and understanding of its pathological features leads to prompt and accurate diagnosis.

## REFERENCES

1. Hope J, ed. A treatise on the diseases of the heart and great vessels. 3rd ed. Philadelphia, Pa: Lea and Blanchard, 1839; 466–471.
2. Ring WS. Congenital heart surgery nomenclature and database project: aortic aneurysm, sinus of Valsalva aneurysm, and aortic dissection. *Ann Thorac Surg*, 2000; 69: S147–S163.
3. Smith WA. Aneurysm of the sinus of Valsalva with report of two cases. *JAMA*, 1914; 62: 1878.
4. Mayer ED, Ruffmann K, Saggau W, et al. Ruptured aneurysms of the sinus of Valsalva. *Ann Thorac Surg*, 1986; 42: 81–85.
5. Dong C, Wu Q-Yu, Tang Y. Ruptured sinus of Valsalva aneurysm: a Beijing experience. *Ann Thorac Surg*, 2002; 74: 1621–1624.
6. Takach TJ, Reul GJ, Duncan JM, et al. Sinus of Valsalva aneurysm or fistula: management and outcome. *Ann Thorac Surg*, 1999; 68: 1573–1577.
7. Chu SH, Hung CR, How SS, et al. Ruptured aneurysms of the sinus of Valsalva in Oriental patients. *J Thorac Cardiovasc Surg*, 1990; 99: 288–298.
8. Goldberg N, Krasnow N. Sinus of Valsalva aneurysms. *Clin Cardiol*, 1990; 13: 831–836.
9. Ott DA. Aneurysm of the sinus of Valsalva. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu*, 2006; 165–176.
10. Edwards JE, Burchell HB. Specimen exhibiting the essential lesion in aneurysm of the aortic sinus. *Proc Staff Meet Mayo Clin*, 1956; 31: 407–412.