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Case Report

Double chambered right ventricle with ruptured sinus of Valsalva: rare entity

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ABSTRACT

Valsalva sinus aneurysm (SVAs) and double-chambered right ventricle (DCRV) is an uncommon congenital heart defect and are usually associated with ventricular septal defect (VSD) in 50% to 60% of patient's population. The current case report had documented a 59 years old hypertensive asymptomatic male with a known case of SVAs presented to our hospital following device closure for 6 months. On examination, the patient was diagnosed to have DCRV induced by SVAs prior to the closure along with other factors. However, to the best of our knowledge, this is the first case report on DCRV induced by SVAs in synergy with other associated multifactors and without VSD among the Indian population.

Keywords: Cardiac MRI, DCRV, Incidence, RV, SVAs, VSD

INTRODUCTION

Valsalva sinus aneurysm (SVAs) is an uncommon congenital or acquired (endocarditis, syphilis, infective endocarditis, Bechet disease etc) cardiac abnormality and linked to a VSD in 50% to 60% of patient's population. Majority of the aneurysm instigated either involving right coronary or non-coronary sinus estimating about 97%. Amongst SVAs patients, the incidence of rupture is reported to be 34% to 47%, involving right ventricle or right atrium, and often results in hemodynamic instability due to heart failure (HF).[1] Early surgical intervention is the treatment of choice.

DCRV is a rare congenital heart disease characterized by anomalous muscle bundle that divide the right ventricle into a proximal high-pressure chamber and a distal low-pressure chamber.[2] The incidence of VSD associated with DCRV was determined to be 70% to 80% and other cardiac anomalies comprises of valvar pulmonary stenosis, atrial septal defect, tricuspid valve regurgitation, aortic valve regurgitation, persistent left superior vena cava, tetralogy of fallot, ruptured sinus of valsalva aneurysm, complete or corrected transposition of the great arteries, and Ebstein anomaly.[2] Usually, DCRV is detected at childhood or adulthood but more often misdiagnosed because right ventricular tract is not routine portion during adult echocardiographic investigation and in addition, the patients remain asymptomatic till adolescence. Recently Nakashima et al, Suzuki et al and Hu et al had reported a case report on the unusual presentation of DCRV induced by ruptured SVAs along with VSD in a 54-year-old man, 51-year-old woman and 41-years old woman respectively. Earlier report had demonstrated the occurrence of above stated three diseases in same patient.[3-4]

However, to the best of our knowledge, this is the first case report on DCRV induced by SVAs in synergy with other associated multifactor and without VSD among the Indian population. The current report describes a case of the above stated disease occurring at age of 59 years old gentleman and stresses the importance of high level of suspicion for DCRV is often required with such kind of cardiac anomalies. Moreover, the report emphasizes clinical presentation, beneficial diagnostic and treatment modalities, and the need for inter-professional team dynamics to achieve optimal outcome in such patients.

CASE REPORT

A 59 years old hypertensive male with a known case of SVAs presented to our hospital with a history of polyarthralgia for 10 years with an increase in severity for last 6 months. He was treated with non-steroidal anti-inflammatory drugs (NSAIDS) for long term or chronically. Patient had undergone device closure for ruptured SVAs before 5 months. Patient had no chest pain, dyspnea or any of the cardiac symptoms. Pansystolic murmur was heard on the left parasternal area during auscultation. His physical examination revealed sinus rhythm with normal pulse rate of 94 beats per minute, elevated blood pressure with a systolic blood pressure of 150 and diastolic pressure of 90 mmHg. He was on antihypertensive drug (Tablet amlodipine 5 mg once daily). Right knee was found to be warm and tender. Chest radiography was suggestive of mild cardiomegaly whereas increased amplitude of the R wave on V1 and inverted T waves on V1-V3 was observed which indicated the presence of right ventricular overload. Routine blood investigations showed increased erythrocyte sedimentation rate (ESR) (120 mm/hr), C-reactive protein (CRP of 223 mg/dL) and serum creatinine (1.7 mg/dL). Elevated total count (TC) and differential count (DC) was noted with synovial fluid analysis. Investigation of ESR, CRP, TC and DC were indicative of inflammatory arthritis. The result of blood culture and anti-cyclic citrullinated peptide (anti-CCP) negative. Rheumatologist, nephrologist and cardiologist were consulted.

Two-dimensional (2-D) transthoracic echocardiography (TTE) exhibited the findings of severe right ventricle (RV) sub infundibular obstruction, concentric left ventricle hypertrophy (LVH), and ruptured SVAs device closure with no residual leak, and a peak pressure gradient of 70 mmHg between inflow and outflow tract of RV indicating the feature of DCRV as demonstrated in Figure 1 A and B.

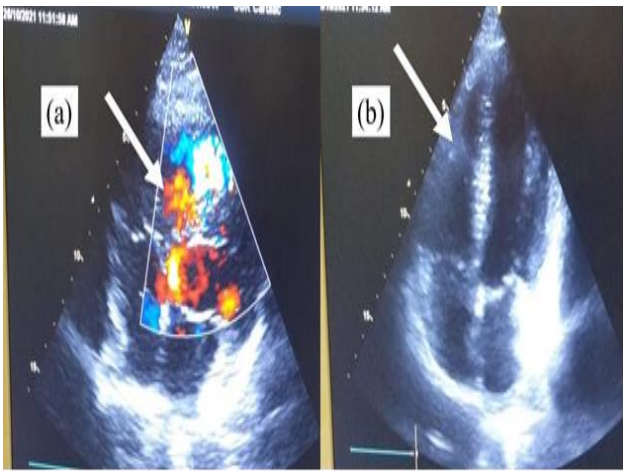

Figure 1 (A and B): TTE. No residual leak seen in ruptured SVAs device closure and severe RV sub infundibular obstruction. A marked muscle band protruding from right ventricular free wall to interventricular septum suggesting features of DCRV.

Cardiac magnetic resonance imaging (MRI) was used to rule out any other associated cardiac anomalies as well as quantify and evaluate RV pressure gradient and RV wall thickness. Cardiac MRI confirmed the findings of 2-D TTE which once again showed significant right ventricular cavity gradient (Figure 2) and the patient required surgical resection for the same.

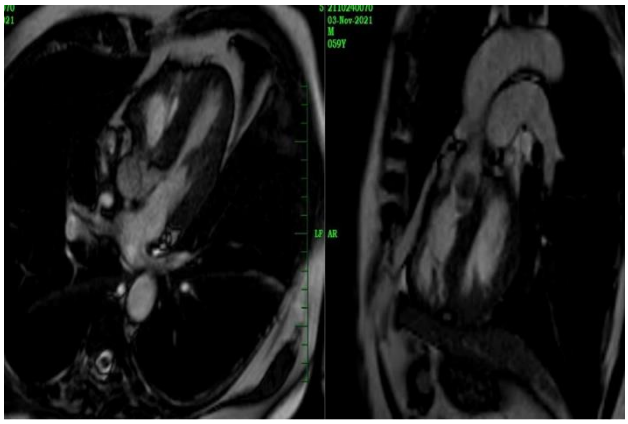

Figure 2: Cardiac MRI confirmed the features DCRV and ruled the absence of other associated cardiac anomalies. Demonstrated protruded right SVA's toward the right ventricle.

Patient advised for the surgical resection of muscular band and discharged with beta-blockers (Tablet metoprolol 50 mg), calcium channel blockers (Tablet cilnidipine 10 mg) along with disease-modifying antirheumatic drug (DMARD) such tab leflunomide 10 mg etc.

DISCUSSION

SVA is an unusual cardiovascular condition that usually results from a congenital aortic media defect or bacterial endocarditis. It accounts for 0.09 to 0.15 percent of all congenital cardiac anomalies and up to 3.5 percent of all congenital heart defects.[6] In nearly 30 to 40% of patients, concomitant heart abnormalities, such as a VSD or aortic valve regurgitation (AR), are present.[1,7] Aneurysms in the right coronary sinus account for 70% of all aneurysms, and they are more common in men and people of Asian descent.[1] Aneurysms can cause compression of an adjacent chamber, a coronary artery, or the conduction system, which can result either alone in myocardial ischemia or conduction abnormalities combined.

Clinical signs manifestations occur in 80% of patients, with the higher percentage of them being between the ages of 30 and 45.[1] In agreement to our report, the age of the patient was more than 45 years. Aneurysms of the sinus of Valsalva rupture in 35% of patients, causing acute symptoms in a quarter of them.[8] Shortness of breath, chest pain, and weariness are all symptoms of rupture.[9,10] The degree of symptoms is determined by the severity of the shunt, the presence of concomitant lesions, and the age at presentation.[1] With divergence, our patient had no symptoms in spite of having severe right RV sub

infundibular obstruction with a peak gradient of 70 mmHg following device closure.

The majority of aneurysms primarily explode into the right-sided heart chambers than other reported parts of heart as coinciding with our report.[8,10] In our case, diagnosis of ruptured SVAs was made with TTE and cardiac MRI was employed to rule out other cardiac anomalies. In contrast, studies have used Transoesophageal Echocardiography (TEE) for diagnosis of ruptured and non-ruptured aneurysms.

To accord with Nakashima et al study, our patients also had similar DCRV hemodynamics but in disagreement to their findings, our case had demonstrated the presence of abnormal muscular band or moderator band identified by TTE and cardiac MRI modalities.[3] Moreover, our study had elucidated the diagnosis of SVAs and DCRV morphology with non-invasive methods as in identical to Hu et al.[5] The publication of case report literatures concerned with DCRV in SVAs association either in the presence or absence of VSD are summarized in the Table 1.

Table 1: List of publication available in DCRV and SVAs.

Author (year)	Age and sex/place of study (Years)	Diagnosis	Imaging modalities used	Findings (Pressure gradient between RV inflow and outflow tract)
Ogata et al[11] (1999)	45, F	DCVR, SVAs, VSD	NA	NA
Hu et al[5] (2015)	41, F China	DCVR, SVAs, VSD	TTE	90 mmHg
Suzuki et al[4] (2019)	51, F, Japan	DCVR, SVAs, VSD	TTE, TEE, computed tomography, cardiac catheterization	49 mm Hg
Nakashima et al[3] (2020)	54, M, Japan	DCVR, SVAs, VSD	TTE, cardiac catheterization,	36 mmHg
Our case report	59, M, India	DCVR, SVAs	TTE, cardiac MRI	70 mmHg

Note: F: Female; M: Male; NA: Not Available.

DCRV has been almost associated with other congenital anomalies.[2] But this is the first paper that has documented the presence of DCRV due to ruptured SVAs pathology and combined factors in the absence of VSD in the Indian national. Previously, Nakashima et al study had suggested the three possible mechanism for the development DCRV because of SVAs pathology as follows: protruding of right SVA into RV; presence of jet lesion and thickened RV in identical to our opinion in this case.[3] We believe that our patients should have developed DCRV morphology even before the device closure itself because elevated RV stroke volume owing to left to right shunt might have resulted in stenosis and increased pressure gradient across RV mid portion. Arai et al had established the benefits of using beta-blockers in DCRV as supported in our case study.[12]

CONCLUSION

Thus, the present case report concludes that DCRV is rare entity, and it is mandatory to have high level of suspicion for DCRV when often such kind of cardiac anomalies is encountered during cardiac diagnosis since the obstruction progress is very slow. Simultaneously, the study had demonstrated the use of beta-blockers in DCRV patients. In addition, the study confirmed the proposed mechanism of protruding SVA into RA, presence of jet lesion, thickened RV, and increased RV stroke volume apart from other factor might be a responsible factor for development of DCRV in SVAs patients. Moreover, our study had reported development of DCRV morphology in SVAs among Indian national for the first time.

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Ethical approval: Not required

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