VENTRICULAR SEPTAL DEFECT WITH SEVERE AORTIC REGURGITATION, SUB AORTIC STENOSIS, AORTIC AND PULMONARY ROOTS DILATION IN AN ADULT: A CASE REPORT

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ABSTRACT

A case of 30 years’ male, a farmer by occupation who was a diagnosed case of a ventricular septal defect (VSD) since the age of seven, but refused for surgical closure back then, came in the emergency room (ER) with decompensated congestive cardiac failure and peripheral edema. After initial stabilization, he underwent transthoracic echocardiography (TTE) that showed small perimembranous VSD with the left to right shunt, a subaortic membrane with severe left ventricular outflow tract (LVOT) obstruction and moderate aortic and mitral regurgitation. TTE was followed by Computed Tomography (CT) thoracic aortogram that revealed small VSD with significant aortic regurgitation (AR). Subaortic membrane, aneurysmal dilated aortic root and ascending aorta and aneurysmal dilated pulmonary arteries. Cardiac surgeons were taken onboard and advised for VSD closure with aortic valve replacement, subaortic membrane resection and aortic root replacement. Patient and family were counseled regarding the procedure but refused again for surgical closure.

Key Words: Ventricular Septal Defect, Aortic Regurgitation, Subaortic Membrane, Pulmonary Roots Dilation, Adult

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Contribution
DN conceived the idea of the case report. Data collection and manuscript writing was done by AM and PA. All the authors contributed equally to the submitted manuscript.

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INTRODUCTION

Peri-membranous ventricular septal defect (VSD), synonymous with type-2 VSD, is the most common type of VSD (that is 80% of the cases). 1 This defect is in the membranous septum and is adjacent to the septal leaflet of the tricuspid valve. 1 Aortic regurgitation is usually associated with supra-cristal VSD and is rare with type 2 VSD. 2 VSD may develop into sub-aortic stenosis in 20% of cases and in these; it is frequently membranous and fixed. 3 There are many studies about the ascending aorta dilation with tetralogy of fallot (TOF), transposition of the great arteries (TGA) and truncus arteriosus but few reported it with VSD.

However, scarce data are available in literature with respect to clinical features and surgical management of ascending aorta dilation with congenital VSD. 4 VSD’s due to the left to right shunting yields increased pulmonary artery blood flow and shear stress leading to pulmonary artery dilation (mostly with patent ductus arteriosus (PDA) and atrial septal defect (ASD)). 5

CASE REPORT

A 30 years old male, presented in the emergency room (ER) with symptoms of dyspnea (New York Heart Association (NYHA) Functional Classification III) for last four months. He was previously treated for bronchial asthma with cough and bipedal edema since 15 days that was up to mid-shin. He has been having the above symptoms since the age of seven and was diagnosed to have a VSD. He was advised surgical closure back then, but the family was reluctant for it despite having symptoms.

On arrival in ER, he was in decompensated cardiac failure, BP= 100/60 PULSE =100/min and spO2 89%, with bipedal edema up to mid-shins and bilateral pulmonary crepitations up to mid-chest. A pansystolic murmur of four/six intensity heard all over precordium more on left lower sternal edge not radiating to axilla with loud palpable P2. After written informed consent, chest radiograph revealed signs of pulmonary edema and electrocardiograph showed normal sinus rhythm with left ventricular hypertrophy (LVH). After initial treatment with diuretics, he was clinically stabilized and moved to the floor.

Later he underwent trans-thoracic echo that showed EF 36%, perimembranous VSD (11 mm size) with the left to right shunt, subaortic membrane (17 mm) with severe left ventricular outflow tract (LVOT) obstruction, severely dilated left atrium (LA) with severe left ventricular Hypertrophy (LVH). LVOT gradient= PPG 70mmhg and MPG=43mmhg, pressure gradients across VSD= 70 mmhg. Mild to moderate mitral regurgitation (MR), moderate aortic regurgitation (AR) and mild tricuspid regurgitation (TR) (Figure 1).

Considering the above findings CT- angiography was performed that showed small perimembranous VSD (size 4 mm), non-coapting tricuspid and aortic valve. Concentric subaortic membrane narrowest part 11 mm. Aneurysmal...
dilated aortic root and ascending aorta (37.2 mm). Aneurysmally dilated pulmonary arteries, main pulmonary artery (MPA) = 54 mm, right pulmonary artery (RPA) = 36.2 mm, and left pulmonary artery (LPA) = 27 mm. Evidence of significant AR (Figure 2).

Figure 2: A) A cross-sectional view showing pulmonary trunk dilation. B) Cross-sectional view with the finding of VSD. C) Cross-sectional view showing sub-aortic membrane

Cardiac surgeons were taken on board considering the above findings and advised for VSD closure with aortic valve replacement (AVR) and sub-aortic membrane resection with aortic root replacement. Patient and family were counseled regarding the surgery, but despite explaining later consequences of not undergoing correction they again refused for surgery. Currently, he is at home, is clinically stable and is doing well on medications but the family has been counseled regarding the high-risk status of the patient.

**DISCUSSION**

Subaortic membrane, pulmonary artery dilatation, aortic root dilatation, and AR all have been reported as complications of perimembranous VSDs. But the last two ones are rarely seen. The association of VSD and AR was first described by Laubry and Pezzi in 1921. AR results due
to prolapse of the aortic leaflets and is most commonly reported with supra cristal VSD and is an associated lesion in approximately three to eight percent of the patients with overall VSD and with perimembranous type, it is rarely seen.\(^2\)

Aortic dilation is most commonly reported with tetralogy of fallot (TOF), transposition of the great arteries (TGA) and truncus arteriosus and few reports are related to VSD.\(^4\) According to one study, there were 0.89% of adult patients with VSD among all those suffering from ascending aorta dilation (AAD).\(^5\) The outcomes of AAD may be a rupture, aortic dissection, and death.\(^5,6\) The annual probability of the above complications is 14.1% once the aneurysm reaches a maximum diameter of 6 cm.\(^5\) Hence it is vital to address it in congenital heart defect (CHD) patients. To avoid the complications it should be treated by the replacement of ascending aorta or remodeling/replacement of aortic root and the results are good enough. Many AAD patients are asymptomatic and are incidentally found on chest X-ray or TTE.\(^6\) Echocardiography also has a diagnostic value for AAD and VSD, but it is far less reliable for AAD and is the most widely used imaging modality for VSD.\(^6\) Computed tomography angiography (CTA) is another important tool which provides a rapid and precise evaluation of ascending aorta but less valuable for VSD, CTA is more accurate though.\(^6\)

The most common cause of pulmonary artery dilation is pulmonary hypertension but it is also related to PDA, ASD, and VSD.\(^3\) Normal pulmonary artery diameter is less than 29 mm for males and less than 27 mm for females.\(^9\) The pulmonary artery size may be more accurately measured by CT and magnetic resonance imaging (MRI) scans. It can be assessed by plain radiograph but the above-mentioned technologies are more accurate.\(^9\) The cause of pulmonary artery (PA) dilation with VSD is due to increased or turbulent blood flow due to the left to right shunting.\(^9\) Dilated pulmonary artery could be life-threatening but patients with increased pulmonary artery pressures are at higher risk. The outcome may be left main coronary compression, PA dissection or rupture with cardiac tamponade.\(^9\) Compression of the left main coronary artery (LMCA) may result in chest pain, left ventricular (LV) dysfunction, arrhythmias and less commonly sudden cardiac death.\(^9\)

Subaortic stenosis (membrane) occurs in the natural history of VSD usually after the first year of life and it is progressive and requires surgery in most cases.\(^3\) The advent of echocardiography enabled us to understand that subaortic stenosis is not present in the first year of life but arises generally when the VSD shows signs of decreased size and spontaneous closure.\(^3\) The association of subaortic stenosis with perimembranous VSD is due to the chance for spontaneous closure because most of them were small and had formed of tricuspid tissue in their border.\(^3\) It doesn't cause symptoms often but some authors report symptoms in adulthood and depend on severity of obstruction and ventricular hypertrophy and it is usually an incidental finding in echocardiography.\(^3\) Surgery is indicated when gradients are > 40 mmHg.\(^3\) Some indicate surgery immediately after diagnosis regardless of gradients, others with gradients either between 20-80 mmHg.\(^3\)

The association of peri-membranous VSD with AR and aortic dilation is rarely reported, but many cases have been seen with sub-aortic stenosis and pulmonary artery dilation. Usually, one or two of the above-mentioned complications are seen but our case outstands in a way that all the mentioned rare complications progressively developed as he grew older. Surgical correction is mandatory in order to prevent further complications regarding which patient was counseled thoroughly.

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