Case Report

Unusual Pulmonary Venous Return Obstruction Associated with Atrio-Ventricular Septal Defect

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Abstract

Venous pulmonary return obstruction associated with atrio-ventricular septal defect (AVSD) has been already described. The anatomic substratum of this obstruction may vary from a patient to another.

We report the case of a 20-month old infant who presented with signs of congestive right heart failure. The diagnosis of AVSD associated with unusual form of pulmonary venous return obstruction due to deviated interatrial septum was confirmed by imaging modalities.

Treatment consisted on surgical repair of the AVSD and the obstruction removal to allow the restitution of the left ventricular volume.

INTRODUCTION

Atrio-ventricular septal defect (AVSD) is a congenital heart disease generally well tolerated during the first years of life in absence of large ventricular septal defect (VSD) or important mitral valve regurgitation.

We report the case of an infant with a poorly tolerated AVSD due to pulmonary venous return obstruction caused by deviated interatrial septum.

CASE PRESENTATION

A 20-month old female infant was referred to our department for breath shortness and frequent respiratory infections.

At admission, physical examination showed a poor weight gain (height=75 cm, weight=7 kg; <3rd percentile), and polypnea (55 cpm). The heart rate was 160 bpm, while blood pressure was 125/60 mmHg. Cardiac auscultation revealed a grade 3/6 systolic murmur with S2 split at left sternal border. Furthermore, right heart failure signs were observed (jugular turgescence, abdomino jugular reflux and peripheral edema).

Electrocardiogram showed a sinus rythm, a QRS axis at −30° and rsR’ morphology in V1 lead. Chest X-ray found a cardiothoracic ratio of 0.6, with dilated right cardiac chambers and pulmonary trunk, and increased pulmonary vasculature.

Echocardiography revealed a restrictive small primum atrio-septal defect (ASD) of 4 mm of diameter, a small restrictive VSD and two separate atrio-ventricular valves (AVV) with a cleft in the anterior leaflet of the left-sided AVV (mitral valve). Importantly a left deviation of interatrial septum was observed, leading to malalignment of interatrial and interventricular septums. The left AVV was overriding the interatrial septum. As a consequence, the venous pulmonary flow had to cross the restrictive ASD to reach the level of the left atrioventricular orifice.

In addition, we found a persistent left superior vena cava draining into the coronary sinus with normal connected pulmonary veins. The right ventricle was dilated, hypertrophied and clearly dominant. The left ventricle was smaller with diastolic and telesystolic diameters of 34 mm and 15 mm, respectively.

The left and right atrioventricular annulus diameters were 15 mm and 18.5 mm respectively, with an AVV index of 0.78.

Doppler showed an accelerated flow through the ASD (maximum velocity of 2 m/s), a mild mitral regurgitation, and an important tricuspid regurgitation with a severe pulmonary arterial hypertension (75 mmHg).

Thus, we concluded to the diagnosis of AVSD associated with venous pulmonary return obstruction secondary to deviated interatrial septum with restrictive ASD, responsible for severe pulmonary hypertension.

Cite this article: Drissa M, Hakim K, Msaas H, Ouarda AF (2018) Unusual Pulmonary Venous Return Obstruction Associated with Atrio-Ventricular Septal Defect. J Cardiol Clin Res 6(3): 1138.
Patient was referred to cardiac surgery. Intraoperative findings confirmed echocardiographic data. There was a left deviation of the lower part of the interatrial septum that directed the pulmonary venous flow towards the restrictive interatrial septal defect before reaching the mitral orifice causing pulmonary vein return obstruction.

The surgical procedure consisted of the resection of the lower part (juxta valvular) of the interatrial septum. The atrial septal defect so enlarged was then closed by a patch in a manner to obtain the alignment of interatrial and the interventricular septums. Finally, the cleft of AVV was repaired by a direct suture.

Post operative echocardiogram showed trivial mitral regurgitation, free pulmonary venous return and normal pulmonary arterial pressure. Six-month follow up was uneventful.

DISCUSSION

The association of AVSD with an obstruction of pulmonary venous return has been reported through literature. In a multi-institutional study, by Kort et al. [1] showed that 13.7% of patients with pulmonary venous stenosis had AVSD; whereas, 0.9% of patients with AVSD had pulmonary venous return obstruction.

The originality of our case consisted in the anatomic substratum of the pulmonary venous obstruction that appeared to be the consequence of a malalignment of the interatrial septum associated with restrictive atrial septal defect. To the best of our knowledge, such an anatomy is reported for the first time. Indeed, in previously published cases [2-5], a supramitral membrane, a Cortri atriatum, or primitive anomalies of pulmonary veins were reported to be the common causes of pulmonary venous return obstacle.

Woong-Han [6] reported a case of malalignment of interatrial septum associated with complete ASVD. In this latter case, septal deviation did not cause any hemodynamic disturbance since the ASD was large, providing a normal flow to the mitral valve. In our case, the AVSD was poorly tolerated because of the small diameter of the ASD that caused restriction of the pulmonary venous flow with post capillary hypertension.

Regarding surgical repair, we opted for a similar strategy than that reported by van – son [7]. In our patient, a biventricular repair was indicated despite the presence of a dominant right ventricle. This was justified by a favorable AVV index [0.78] and especially the fact that the disproportion between left and right ventricles’ size might be functional and reversible after treating pulmonary venous obstruction.

To date], it remains difficult to decide of single or biventricular repair in some borderline cases of AVS with poorly balanced ventricles [8-10]. Cohen et al. [11] proposed algorithm summarizing different AVSD management strategies. The choice of two or single ventricular repair essentially depends on the AVVI, the presence or not of ductal-dependent circulation and VSD.

In our point of view, we should take into account different parameters and analyze each case separately, considering both morphological (diameters, length and ventricular volumes, measurement of atrioventricular orifices) and hemodynamic features (ventricular load conditions).

CONCLUSION

Pulmonary venous return obstruction associated with AVSD is most commonly due to supra-mitral membrane, triatrial heart, or pulmonary veins stenosis.

We reported a case illustrating an unusual anatomic presentation of venous pulmonary obstruction associated with AVSD that consisted on a malalignment of the interatrial and interventricular septum with a restrictive ASD. The surgical removal of the deviated septum allows the normalization of pulmonary hypertension and the restitution of the left ventricular volume, often underestimated preoperatively because of its reduced preload.

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