

What a coincidence to be pregnant: partial pulmonary venous return abnormality, iatrogenic atrial septal defect, rheumatic mitral stenosis

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Abstract

Herein we present a case of concomitant congenital anomalies with an iatrogenic defect. The female patient underwent a percutaneous mitral balloon valvuloplasty due to rheumatic mitral stenosis. Unfortunately, an iatrogenic atrial septal defect (ASD) occurred and also, partial anomalous pulmonary venous return was observed at post-procedure evaluation. The patient had severe symptoms and the right heart chambers were dilated on imaging. But perhaps, the most crucial point was that the patient was planning a pregnancy. After a difficult and patient-involved decision process, the patient underwent to successful robotic surgery for iatrogenic ASD and partial anomalous pulmonary venous return. After operation, the patient was asymptomatic and right heart chambers normalized.

Introduction

With the development of diagnostic methods, the incidence of some congenital anomalies is increasing. One of them is the anomalous pulmonary venous return (APVR). This congenital problem, first described by Winslow in 1739, is characterized by the drainage of all or part of the pulmonary veins into the systemic venous circulation or the right atrium (1). This anomaly, which creates a volume load in the right chambers due to the left-right shunt, is frequently detected together with other congenital heart anomalies, especially atrial septal defect (2). However, in this case, we will try to present the coexistence of iatrogenic, not congenital, ASD and PAPVR in an adult female patient with rheumatic mitral stenosis.

Case Report

A 26-year-old female patient was referred to our tertiary center because her dyspnea did not decrease 2 months after percutaneous balloon mitral valvuloplasty (PBMV) for rheumatic mitral stenosis. She had no family history of CHD. At the admission, her functional capacity was NYHA-III. Arterial blood pressure was 125/80 mmHg, peripheral oxygen saturation was 97%, and heart rate was 75/minute. There was a 2/6-degree systolic murmur on auscultation besides a wide split-second heart sound (S2). ECG showed sinus rhythm and did not show any specific changes. Two months ago, just after the PBMV procedure, mitral valve area (MVA) was reported as 1.8 cm². The patient was admitted to the emergency department several times with palpitation attacks after discharge and had some transthoracic echocardiography (TTE) reports performed by different cardiologists during this period. Her MVA measurements ranged between 1.2 cm² and 1.9 cm² on TTE.

On our TTE examination, ejection fraction (EF) was normal, mitral valve area (MVA) was 1.72 cm^2 , and we observed a secundum type atrial septal defect (ASD) accompanied by dilatation in the right heart chambers (Figure-1) with moderate tricuspid regurgitation (TR). Pulmonary arterial systolic pressure (PASP) was also measured as 48 mmHg. Transesophageal echocardiography (TOE) showed that MVA was 1.65 cm^2 by pressure half time on 2-D evaluation and a diastolic D-sign on the interventricular septum (IVS) (Figure-2). With the 3-dimensional Multiplanar reconstruction (MPR) measurement of MVA was 1.63 cm^2 (Figure-3). The mitral valve gradient was 9/5 mmHg, and as such, it was evaluated as mild mitral stenosis. Moderate TR was observed, PASP was 50 mmHg, and tricuspid annular diameter was 37.5 mm. The interatrial septum was aneurysmatic and a $1.22 \times 0.67 \text{ cm}$ of atrial septal defect was observed with the 3-D examination (Figure-4), and left to right shunt was seen through this defect. Also, the patent foramen ovale (PFO) tunnel was observed. Although the drainage of the left upper, left lower, and right lower pulmonary veins into the left atrium were observed, a cardiac MRI evaluation was planned because the right upper pulmonary vein drainage could not be adequately evaluated. Interestingly, the patient applied PBMV two months ago, and she was assessed with TOE before; no ASD finding was found in that period. In this case, we concluded that the current ASD is an iatrogenic defect secondary to the PBMV.

On cardiac MRI, dilatation in the right chambers, diastolic D-sign on IVS, secundum type ASD (Qp/Qs ratio: 2.82) were also observed, consistent with TOE. We found that the right upper and middle lobe pulmonary veins drained into the superior vena cava on cardiac computed tomography (CT), and it was interpreted as a partial pulmonary venous return anomaly (Figure-5). Then, we performed a cardiac catheterization, and our main findings are followings; 1) Qp/Qs: 6.28, 2) pulmonary vascular resistance was 1.04 wood unit, 3) systemic vascular resistance was 10.42 wood unit, 4) pulmonary capillary wedge pressure was 7 mm Hg, 5) mean pulmonary artery pressure was 15 mm Hg. Hence, the results of catheterization consisted of left-to-right shunt. At this stage, the evaluation made with the patient gained importance, and the heart team decision was determined because the patient was young and considering pregnancy.

Surgical treatment for pulmonary venous return anomaly and ASD was decided in the heart team council for the patient. A follow-up with medical treatment was planned for the mitral valve depends on low pulmonary capillary wedge pressure. The PAPVR was repaired by robotic intracardiac routing using the da-Vinci system, and the ASD was closed successfully.

In the TTE evaluation performed 2 months after the surgery, the right heart chambers were in normal size (Figure-6), MVA was 1.63 cm^2 , mitral valve gradient was 9/6 mmHg, mild tricuspid regurgitation, and PASP was 24 mmHg. The patient was symptom-free, and her functional capacity was NYHA-I.

Discussion

To the best of our knowledge, this is the first case in which partial anomalous pulmonary venous return (PAPVR) anomaly, iatrogenic ASD and rheumatic mitral stenosis were found together. Anomalous pulmonary venous return (APVR) is a rare congenital anomaly seen between 0.4% and 0.7% in the autopsy series (3). The main pathophysiologic problem in APVR is the direct or indirect opening of the pulmonary veins into the right atrium (1,4). While one or more pulmonary veins are defective in the partial type, all of the pulmonary veins are unrelated to the left atrium in total anomaly (5). PAPVR is most commonly encountered in the supracardiac form in which the right upper pulmonary vein drains into the right atrium or superior vena cava (6), as in our case. The abnormal pulmonary venous return may be supracardiac, cardiac, infracardiac, or mixed type. Although an anomaly in the drainage of a single vein usually does not cause hemodynamic problems, an anomaly in more than one vein or other congenital or acquired diseases may cause the symptoms to occur (4,7). PAPVR is often accompanied by atrial septal defect, which has been reported to be 82% in some publications (8). There are also some publications on its association with rheumatic mitral stenosis (9,10).

PAPVR usually has a silent clinical course and varies depending on the degree of left-to-right shunt and other associated cardiac anomalies (11,12). The clinical problem in our patient was accompanying mitral stenosis. Diagnosis of PAPVR was missed during the evaluation period for mitral stenosis, and therefore

the treatment was determined as percutaneous mitral balloon valvuloplasty. However, with the effect of iatrogenic ASD due to PBMV, unfortunately, the volume load in the right chambers increased. Thus, the patient's symptoms continued as before PBMV, although mitral stenosis was treated.

The incidence of ASD following PBMV ranges from 4% to 53% (13). This high variability is related mainly to the diagnostic method used. In a study by Arora et al., in which TOE was used in the assessment, the rate of ASD was observed as 92% immediately after PBMV, 80% after 72 hours, and 10% in the evaluation 3 months later. So, most of the iatrogenic ASD close spontaneously (14).

In our case, the iatrogenic atrial septal defect was too large to be associated with only transseptal puncture. Most likely, the inappropriate withdrawn of the percutaneous mitral balloon delivery system damaged aneurysmatic septum and caused it to tear. Otherwise, such a large atrial septal defect was unlikely to be overlooked in TOE assessment before PBMV. The abnormally connected pulmonary vein was drained to the superior region of SVC, so it cannot be clearly detected with TOE. Also, the presence of diastolic D-sign has forced us to find the cause of a volume load other than iatrogenic ASD. As a matter of fact, while the diastolic d-sign indicates a significant volume load, it was not possible to explain this with only 2-monthly iatrogenic ASD. however, we would expect to see systolic D-sign secondary to pressure increase in patients with severe mitral stenosis diagnosed late.

The crucial question to be answered in our case was which strategy should be chosen in a patient with diastolic-D sign, dilatation in the right heart cavities, mild mitral stenosis, and especially planning pregnancy. Cardiac MRI and CT demonstrated a partial pulmonary venous return anomaly associated with an isolated single vein located superiorly. In cardiac catheterization, significant left-right shunt was observed, and pulmonary capillary wedge pressure was within normal limits. These findings proved that mitral stenosis was not serious and the main problem was left-to-right shunt. It was decided by the heart team to perform PAPVR and ASD repair with robotic method. Close follow-up was planned for mitral valve stenosis. Thus, the patient would have the chance to be treat with PMBV or mitral valve replacement after pregnancy. In addition, a treatment without sternotomy would have reduced the risk of cardiac surgery that could be performed in the follow-up. After all, the patient was successfully treated with the robotic surgery (da-Vinci system), and normalization was observed in the right heart chambers and pulmonary artery pressure in the second month control TTE.

The take-home messages that we want to give in our patient, who was a very important example of the individualized treatment option:

- 1- It should never be forgotten in clinical practice that an acquired disease such as rheumatic mitral stenosis may be accompanied by a congenital disease.
- 2- Even if the partial pulmonary venous return anomaly is isolated, it should always be kept in mind in the causes of volume overload and pulmonary hypertension in adults.
- 3- Iatrogenic atrial septal defect sizes can sometimes be much higher than expected. The operation team should definitely check the IAS just after the procedure.
- 4- Some individual indications such as pregnancy request may force the physician to completely review patient management, surgical indications and methods.
- 5- The optimum treatment option for each patient should be evaluated in their own circumstances.

Conclusion

While evaluating structural diseases, whether congenital or acquired, other accompanying anomalies should definitely be kept in mind. Structural interventional procedures can cause some complications and it is important to always control this. At the same time, the clinical and social process of each patient is different, and the patient should be included in the treatment planning and the best decision should be made as a team.

Data Availability Statement: The data that demonstrate the findings of this case are available from the corresponding author upon reasonable request.

Ethical approval statement and informed consent are not applicable for my case report.

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Figure Legends

1. Dilatation of right heart chambers is shown on Transthoracic Echocardiography Apical 4-Chamber window. (RA: Right atrium, LA: Left atrium, RV: Right ventricle, LV: Left ventricle)
2. Diastolic D-sign on the Transthoracic Echocardiography PSAX window, red arrow shows flattening of the interventricular septum in diastole. (PSAX: Parasternal Short Axis, RV: Right ventricle, LV: Left ventricle, MV: Mitral Valve)
3. Measurement of mitral valve area with 3D multiplanar reconstruction method on Transesophageal Echocardiography. As seen in the figure, the mitral valve area was measured as 1.63 cm^2 .
4. Iatrogenic Atrial septal defect image on Transesophageal Echocardiography aortic short axis window, red arrow shows the defect on the interatrial septum. (A: Right atrium, LA: Left atrium)
5. Evaluation of pulmonary veins with Cardiac Computed Tomography is shown. The red dashed lines represent the area that we can see on transesophageal echocardiography, the blue arrow represents the drainage of the right middle lobe pulmonary vein, and the green arrow the right upper pulmonary vein to the superior vena cava. (The red star indicates the superior vena cava.)
6. Normalization in the right heart chambers is observed on Transthoracic Echocardiography Apical 4-Chamber window, 1 month after the surgery. (RA: Right atrium, LA: Left atrium, RV: Right ventricle, LV: Left ventricle)







