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Pulmonary Vein Stenosis as a Rare Cause for Pulmonary Hypertension, Documented by Three-Dimensional Echocardiography

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Abstract

Introduction: Pulmonary vein stenosis includes narrowing of one or more pulmonary veins that may have congenital or acquired causes. Congenital pulmonary vein stenosis is a rare condition and also a rare cause for pulmonary hypertension. Since identifying the etiology of pulmonary hypertension plays a crucial role in medical and surgical planning, we decided to introduce a rare case of pulmonary hypertension due to pulmonary vein stenosis.

Case Presentation: A 29-year-old white female patient who was suffering from exertional dyspnea was referred to our center for further evaluation. Our findings in transthoracic and transesophageal echocardiography showed pulmonary arterial hypertension with maximum gradient of 65 - 70 mmHg and moderate to large size secondom type oval shaped atrial septal defect (2 cm - 1.2 cm) with bidirectional shunt, in a predominantly left to right direction. Atrial septal defect rims were suitable for device closure. Due to out of proportional pulmonary hypertension with atrial septal defect, further evaluations were conducted and pulmonary vein stenosis was confirmed. In this case, the echocardiography findings were consistent with right upper pulmonary vein stenosis and justified severe pulmonary hypertension with the anomaly of atrial septal defect. Device closure of atrial septal defect and balloon angioplasty of pulmonary vein stenosis were performed for our patients. Three months later, in a follow up evaluation, it was revealed that the severity of pulmonary hypertension was declined from 65 - 70 mmHg to 40 mmHg.

Conclusions: Only few cases of pulmonary hypertension secondary to congenital pulmonary vein stenosis in infancy were reported until now, and it is very uncommon in adult patients without any prior history of surgery. Patients with pulmonary vein stenosis often present with shortness of breath and may mimic symptoms of Chronic Lung Disease or pneumonia which result in misdiagnosis and treatment. Therefore, in patients with underlying congenital heart disease and out of proportional pulmonary hypertension, other potential and rare causes for pulmonary hypertension such as pulmonary vein stenosis should be considered and evaluated. It is recommended that cardiologists do not directly attribute pulmonary hypertension to the underlying heart disease without sufficient assessment to rule out other possible causes for it.

Keywords: Pulmonary Vein, Atrial Septal Defect, Echocardiography, Pulmonary Hypertension

1. Introduction

Pulmonary vein stenosis includes narrowing of one or more pulmonary veins that may have congenital or acquired causes.

The most common cause of pulmonary vein stenosis in adult patients is radiofrequency ablation procedures for treatment of atrial fibrillation (1).

Congenital pulmonary vein stenosis is a rare condition and cause of pulmonary hypertension. This cardiac malformation results from abnormal developmental process of the left atrium to connect to the pulmonary veins. In 50% of cases, this malformation coexists with the associated defects such as atrial and ventricular septal defects (2, 3).

We found a rare case of pulmonary hypertension due to pulmonary vein stenosis and decided to present it as a case-report.

2. Case Presentation

A 29-year-old white female patient, who was suffering from exertional dyspnea was referred to our center for further evaluation.

Over the past year she had experienced increasing dyspnea with functional capacity class II. The patient's past medical, family and drug history was negative.

In her initial assessment: O_2 saturation was 98% at room air. II-III/VI systolic murmur was heard over pulmonic area without any thrill and splitting of second heart sound was also found.

Patient's electrocardiography revealed normal sinus rhythm and incomplete right bundle branch block.

Our findings in transthoracic echocardiography showed: Normal left ventricle size and systolic function, severe right ventricular enlargement with mild systolic

dysfunction, mild to moderate tricuspid regurgitation with peak gradient of 60 mmHg and pulmonary pressure gradient estimated 65 - 70 mmHg. Moderate to large size secondum type oval shaped atrial septal defect (2 cm - 1.2 cm) was also noted with bidirectional shunt, in a predominantly left to right direction.

Due to out of proportional pulmonary hypertension with atrial septal defect, we decided to continue our evaluations to possibly pinpoint other possible causes for the severe pulmonary hypertension.

Other possible cardiac causes for her pulmonary hypertension included: primary pulmonary hypertension, pulmonary hypertension associated with the left heart disease, chronic pulmonary emboli, TAPVC and PAPVC, PV stenosis and pulmonary hypertension associated with intracardiac shunts.

In view of the right pulmonary vein in transesophageal echocardiography, doppler study revealed flow turbulency, continuous high peak velocity pattern with peak velocity of 226 cm/sec (Figure 1) and a web causing a blockage with a central opening that had a diameter of 0.8 cm by 0.9 cm which was located at the entrance of the right upper pulmonary vein (Figures 2 and 3).

In this case, the echocardiographic findings confirmed by three- dimensional study were consistent with RUPV stenosis; these findings illustrated an association between severe pulmonary hypertension and anomaly of atrial septal defect. Atrial septal defect had suitable rims for device closure.

Device closure of atrial septal defect and balloon angioplasty of pulmonary vein stenosis were performed for our patients. Three months later, in a follow up evaluation it was revealed that severity of pulmonary hypertension gradient was declined from 65mmHg to 40 mmHg and pulsed Doppler pulmonary vein flow pattern in transesophageal echocardiography changed from continuous high peak velocity pattern to normal, well-defined systolic and early diastolic flow pattern with peaks of _60 cm/second.

Although there is little evidence on the results of transcatheter techniques for treatment of congenital pulmonary vein stenosis, balloon angioplasty of the involved vessels acquired after radiofrequency ablation usually leads to a reasonably good initial result. However, restenosis occurs in 50% of the patients within one year (2). Considering the presence of a web as the cause of pulmonary vein stenosis in our patient, we expected the possibility of restenosis be low. Our follow up evaluation in 3 months later was also normal; however, longer term follow up is needed to ensure the maintenance of our acceptable outcome.

3. Discussion

Only few cases of congenital pulmonary vein stenosis with pulmonary hypertension were reported in infancy; however, it is very uncommon in adult patients without any prior history of surgery (4, 5).

Pulmonary vein stenosis in adult patients is commonly associated with prior radiofrequency ablation procedures for atrial fibrillation (1).

Patients with pulmonary vein stenosis often present with shortness of breath and may mimic symptoms of Chronic Lung Disease or pneumonia which result in misdiagnosis and treatment (2, 6).

Therefore, in patients with underlying congenital heart disease and out of proportion pulmonary hypertension, we should consider and evaluate other potential causes of pulmonary hypertension, such as pulmonary vein stenosis; however, it is considered to be a rare reason for it. It is recommended that cardiologists do not directly attribute pulmonary hypertension to the underlying disease without sufficient assessment to rule out other possible causes.

To the best of our knowledge, this is the first case report of pulmonary vein stenosis which was diagnosed through confirmation of the three- dimensional echocardiography. On the other hand, short-term follow up is considered as a limitation in our study, as restenosis is expected to occur within months to few years after the procedure.

Footnote

Conflict of Interest: Hedieh Alimi and Afsoon Fazlinezhad report no biomedical financial interests or potential conflicts of the interest.

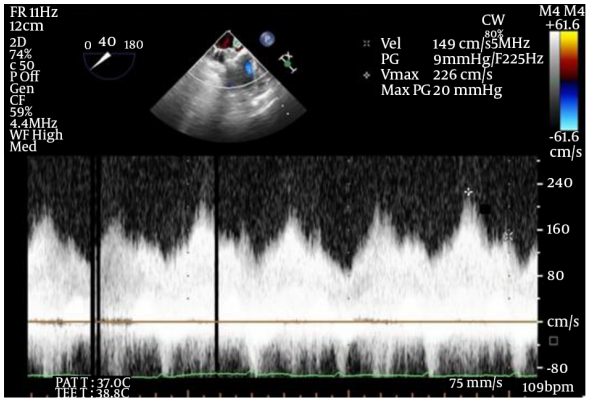
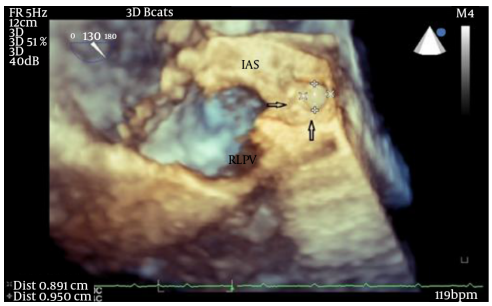


Figure 1. A, typical pulsed Doppler flow pattern of the pulmonary vein stenosis. The flow pattern is turbulent, continuous, and has an abnormally high peak velocity (maximum peak velocity = 226 cm/sec); B, normal pulsed Doppler pulmonary vein flow pattern after balloon angioplasty of the pulmonary vein. There are well-defined systolic and early diastolic peaks of _60 cm/second, and the flow reaches the baseline.

Figure 2. Three-Dimensional Transesophageal Echocardiogram



Marked area with plus sign indicates the orifice of the web in right upper pulmonary vein entrance with diameter of 0.8cm to 0.9 cm. the surrounded area, marked with arrows indicates the native orifice of the pulmonary vein entrance and margins of the web, inter atrial septum (IAS), and right lower pulmonary vein (RLPV).

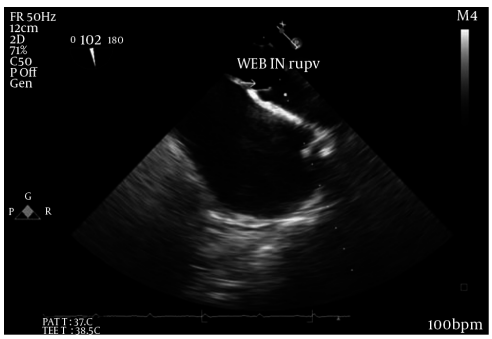


Figure 3. Web at The Entrance of Right Upper Pulmonary Vein (RUPV) in Two Dimensional Tranesophageal Echocardiography

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