CASE REPORT

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An Interesting Association with Pulmonary Hypertension: Swyer-James-Macleod Syndrome

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Abstract

Pulmonary hypertension (PH) is a chronic disease that has increased awareness in recent years and has high morbidity and mortality. There are many unexplained points outside the current PH classification. Congenital heart diseases and lung pathologies are important causes of PH. Swyer-James-Macleod syndrome is a syndrome characterized by unilateral pulmonary arterial flow reduction due to involvement mostly in childhood and contralateral lung hyperlucency in imaging methods. It is rare for this syndrome to be observed together with congenital heart diseases and to be defined in a patient with PH.

Keywords: Atrial septal defect, congenital heart disease, pulmonary hypertension, Swyer-James-Macleod syndrome

INTRODUCTION

Pulmonary hypertension (PH) is a disease that can include more than one clinical condition and is closely related to cardiovascular and respiratory system diseases. The definition of PH is made by right heart catheterization. In right heart catheterization measurements, the disease is diagnosed when the mean pulmonary artery pressure is >20 mmHg. Pulmonary vascular resistance (PVR) and pulmonary artery wedge pressure measurements are also essential for disease classification.[1]

In the 2022 European Society of Cardiology (ESC) PH guidelines, the disease was evaluated in 5 groups. Cardiovascular diseases are the most common in the etiology of the disease. In addition, lung pulmonary obstructive, and systemic diseases are among the causes of PH. However, isolated pulmonary arterial hypertension is the focus of PH treatment. Congenital heart diseases are also included in the group 1 PH classification according to the ESC 2022 PH guidelines. Swyer-James-Macleod syndrome (SJMS), which we have diagnosed, is a disease that is generally thought to be related to childhood infections and is evaluated within the classification of developmental lung diseases and group 3 PH related to it.[2] The coexistence of SJMS and congenital heart diseases is rare.

In our study, we present a case of a patient with SJMS who was diagnosed with PH and had a history of atrial septal defect (ASD) closure.

CASE REPORT

A 22-year-old female patient presented to us with increasing difficulty in breathing for the last two years. In addition to breathing difficulties, the patient had long-standing stabbing chest pains. There was no sputum or cough complaint. In the patient's history, it was learned that she had undergone ASD closure surgery 11 years ago. The patient had no history

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of recurrent lung infections. Physical examination of the patient revealed that blood pressure was 110/60 mmHg, heart rate was rhythmic 65 beats/min, oxygen saturation was 96%, and cardiac auscultation had a 2/6 systolic murmur. On the chest X-ray of the patient, increased vascularity in the right lung, heart, and mediastinal structures were directed to the left (Figure 1). Sinus rhythm and incomplete right bundle branch block morphology were observed on electrocardiography.

The ejection fraction was evaluated as 60% in the patient's echocardiography. No serious pathology was observed in the left-sided valves. The RV basal diameter was measured as 4.2 cm, RV FAC: 50%, TAPSE: 19 mm, and RV SM: 10.9 cm/s. The patient's systolic pulmonary artery pressure was 55 mmHg, TR velocity was 3.8 m/s, and the TAPSE/sPAP ratio was 0.34 mm/mmHg. It was also noted that the patient did not have a serious lung infection during childhood. The 6-min walking test was found to be 460 m. On the patient's complaints, he was hospitalized for right heart catheterization and medical treatment. The patient's laboratory parameters were found to be normal. The N-terminal pro-brain natriuretic peptide (NT-proBNP) level was measured as 174 ng/L. In the right heart, the mean pulmonary artery pressure was measured as 32 mmHg. The vasoreactivity test was negative. Cardiac output: 4 lt/min, PVR was calculated as 5.4 wood units, Qp/Qs: 1.1. The pulmonary capillary wedge pressure was measured as 8 mmHg. Then, ventilation-perfusion scintigraphy was performed with Tc99m. Because of scintigraphy, a hypoplasic left lung with subsegmental hypoperfusion was observed, and a compensatory hypertrophic right lung with normal perfusion was observed (Figure 2). Pulmonary computed tomography angiography of the patient revealed a hypoplastic left main pulmonary artery (Figure 3, 4) Our patient was diagnosed with SJMS. Our patient, who also had a history of ASD closure, was evaluated as group 1 PH in the low-risk group, and sildenafil 20 mg 3*1 treatment was started. During the follow-up of the patient, systolic pulmonary artery pressure was measured as 35 mmHg on transthoracic echocardiography, and functional capacity was evaluated according to NYHA 1.

Informed consent was obtained from the patient.

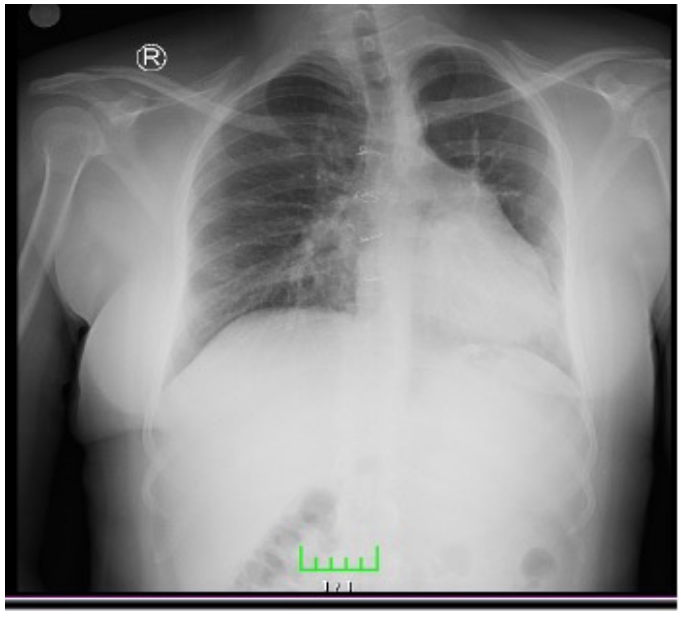

Figure 1: On chest X-ray, the vascularity of the right lung was increased, and the heart and mediastinal structures were observed to be deviated to the left

Discussion

PH is a chronic disease that can affect all age groups, and its prevalence is estimated to be 1% in the global population. Cardiovascular diseases are the most common causes of PH. The second most common is chronic obstructive pulmonary disease.[3] Although the prevalence of PH increases with age, it is seen especially in the younger patient group due to congenital heart diseases. It would also be useful to investigate systemic diseases in young patients with PH, human immunodeficiency virus, and infective diseases such as schistosomiasis in endemic areas.[4]

The diagnosis of PH is made by right heart catheterization. After the diagnosis is made, the etiology is clarified using catheter measurements and different imaging methods. Clarification of

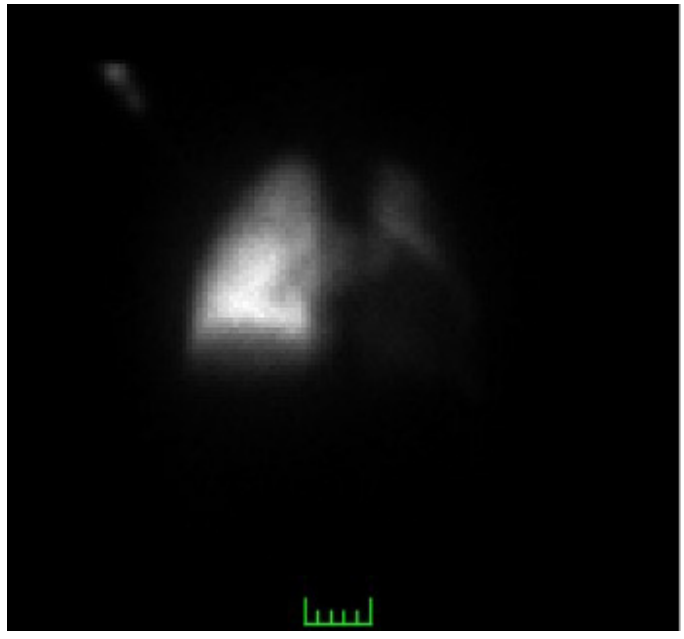

Figure 2: In the scintigraphy images, hypoplastic left lung with subsegmental hypoperfusion and compensatory hypertrophic right lung with normal perfusion were observed

the etiology and evaluation of hemodynamic measurements play a role in arranging the treatment plan, as classification and risk assessment play an important role for treating PH. In the follow-up of the patients, treatment arrangements are made with a four-stage evaluation system, including World Health Organization functional classification, 6-min walk test, and BNP or NT-proBNP levels.

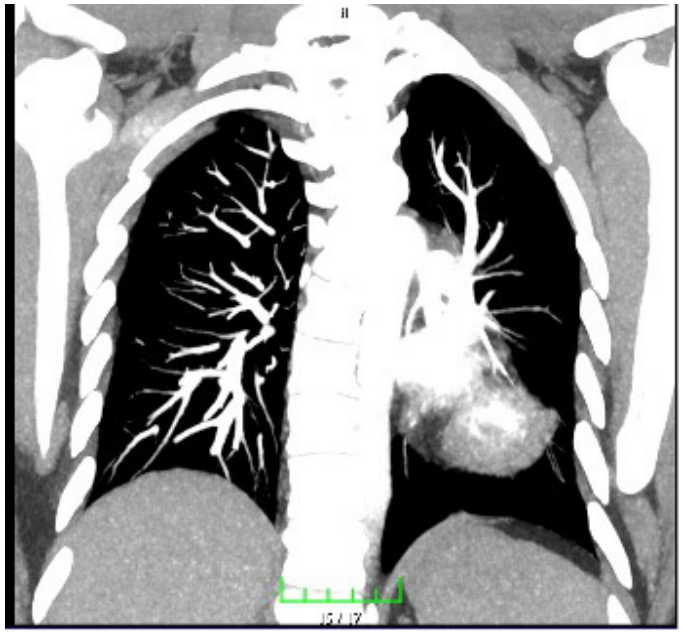

Figure 3: Compensatory increase in the right air spaces was observed in the patient's pulmonary computed tomography angiography

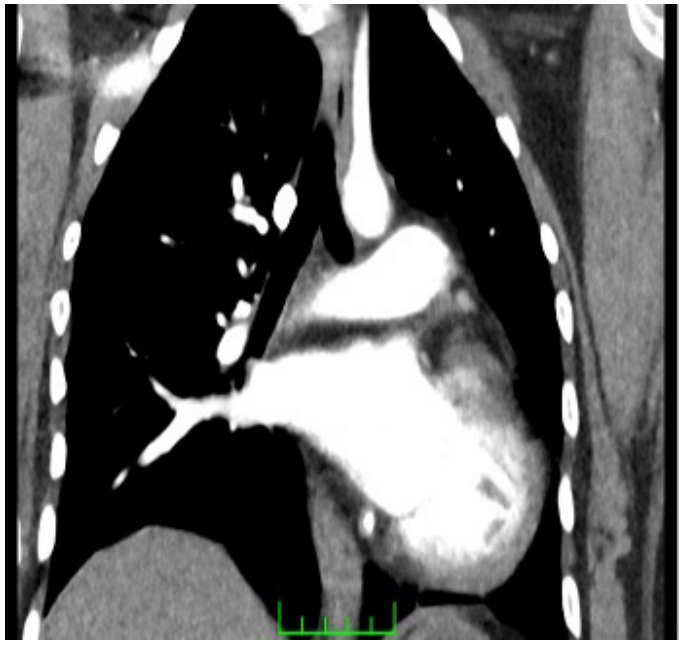

Figure 4: Pulmonary computed tomography angiography of the patient showed hypoplasia of the left main pulmonary artery

SJMS is a rare clinical condition. Although adult patients are generally asymptomatic, some patients may develop exertional dyspnea, cough, sputum production, and hemoptysis.[5] It is thought that the most likely mechanism causing the disease is the underdevelopment of bronchioles and terminal branches secondary to bronchiolitis obliterans, which occurs in early life.[6] There are rare reports of its association with SJMS and congenital heart diseases. The association between coronary outflow anomaly and ventricular septal defect has been shown previously.[7] It remains a mystery whether it causes PH together with these. It is important to prevent and treat recurrent infections during the follow-up of the disease. However, there are no definitive data on the clinical follow-up of patients with PH and their prognosis.

SJMS can be evaluated within the scope of developmental lung diseases and can be evaluated in group-3 PH class.[8] The presence of ASD as a congenital heart disease in our patient and its correction, the absence of echocardiographic measurements because the patient had no complaints for 10 years after the repair and did not come to follow-ups, weakens our ability to reveal the etiology. In our patient, who also had a history of ASD closure, it can be thought that the mechanism of PH development combined group 1 and group 3, depending on the underlying lung disease. Measurements of lung functional capacity in our patient could guide us in the differentiation. Our patient had no history of recurrent lung infections. There was no evidence of bronchiectasis in the lung evaluation. Diffusing capacity of the lung for carbon monoxide measurements are a useful test in this sense, and it is a shortcoming that it could not be performed in our patient.

In case our patient had a combination of group 1 and group 3 PH, it was decided to start monotherapy first because he had dyspnea complaints, had a mean pulmonary artery pressure of 32 mmHg, and was in the low-risk group. However, because the patient was known to have persistent PH after possible congenital heart disease and the prognosis in this case was worse, combination therapy was considered in the follow-up. Endothelin receptor antagonists were considered to be added to the treatment if the patient's complaints continued during the follow-up. In this patient, considering the pathogenesis of group 4 PH and flow-limiting obstructive pulmonary disease, adding riociguat to the treatment may also be beneficial.

Conclusion

In this case report, a patient with SJMS and ASD coexistence, which is rare in the etiology of PH, and our treatment management are presented.

Ethics

Informed Consent: Informed consent was obtained from the patient.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: A.T.Ş., Concept: Y.A., Design: A.T.Ş., A.A., Data Collection or Processing: Y.A., M.A.D., Analysis or Interpretation: M.A.D., Literature Search: A.T.Ş., C.K., Writing: A.T.Ş.

Conflict of Interest: No conflict of interest was declared by the authors.

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