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A Rare Case of Pulmonary Arterial Hypertension as the Initial Presentation of Systemic Lupus Erythematosus in a Pediatric Patient

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Pulmonary hypertension (PH) diagnosis can be challenging, particularly in young patients with systemic lupus erythematosus (SLE). We report a 10-year-old female presenting with suspected sinus venosus atrial septal defect (ASD) with Eisenmengerisation, later diagnosed with primary pulmonary arterial hypertension (PAH) secondary to SLE. She received vasodilators and immunosuppressants, showing partial treatment response.

Keywords: Sinus venosus; pulmonary hypertension; antibodies; atrial septal defect; SLE.

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1. INTRODUCTION

Pulmonary hypertension (PH) poses diagnostic challenges in pediatric patients, often linked to structural cardiac abnormalities leading to Eisenmenger syndrome. SLE is a chronic autoimmune disease with diverse pulmonary manifestations, including PH, a rare and life-threatening complication. In Patients of SLE occurrence of pulmonary complications doesn't correspond to the duration of the disease [1]. The usual prevalence of PAH in patient of SLE is 2.8 to 23.3 % [2]. Pulmonary arterial hypertension (PAH) is a rare and a life-threatening complication and its diagnosis is usually delayed to due rarity and more than 40 percent patients with early PAH remains asymptomatic [3-7].

Here we describe a rare case of a young female who presented as a case of suspected sinus venosus atrial septal defect (ASD) with Eisenmengerisation and on thorough investigations was found to be suffering from PH secondary to SLE.

2. CASE PRESENTATION

A 10-year-old female presented with progressive exertional dyspnea (NYHA II-III) over three years. No signs or symptoms suggestive of SLE.

Initial evaluation revealed:

- **Pulse:** 96/min; BP: 90/60 mmHg; Respiratory rate: 24/min

- No pallor, icterus, clubbing, lymphadenopathy, or edema

- **Cardiovascular examination:** Loud P2, early diastolic murmur of grade III in pulmonary area and grade III parasternal heave.

- **Other systemic examination:** Normal.

Diagnostic Workup:

- **Routine blood and urine tests:** Normal.

- Thyroid function, renal function and hepatic function tests were within normal limits.

- **Chest radiograph:** Cardiomegaly and bilateral hilar prominence.

- Antinuclear antibody profile was strongly positive for PCNA antibody (1:320 titre).

- **Electrocardiogram (ECG):** Sinus tachycardia, P pulmonale, right ventricular hypertrophy with strain and right axis deviation.

- **Echocardiography:** Dilated right atrium and ventricle with severe pulmonary hypertension. Interatrial septum drop out of 2.5 mm was noted. Bubble contrast was suggestive of right to left shunt through PFO.

- **Transesophageal echocardiography:** Could not be done as the patient was uncooperative.

- **Cardiac catheterization:** MVO2 52.4%, Qp/Qs 0.6, PVR 34.26 WU.

- Cardiac CT done to rule out sinus venosus ASD was suggestive of an oblique slit like defect between the septum primum and septum secundum of 2 mm in mid portion of interatrial septum without any obvious jet suggestive of patent foramen ovale. Also, cardiomegaly with dilated right atrium (6.0 X 5.2 cm) and right ventricle (4.9 cm in mid ventricle level in diastole) with reflux of contrast in hepatic veins.

- CT pulmonary angiography was done to rule out any pulmonary AV malformations and AV fistulas and any extracardiac shunts and it was suggestive of dilated main pulmonary artery, left and right branch of pulmonary artery with P/A ratio of > 1 suggestive of PAH.

- Ultrasound of the abdomen and hepatoportal doppler done to look for intrahepatic shunts was within normal limits.

- CECT abdomen and pelvis done to look for any portosystemic shunt revealed normal study.

3. DISCUSSION

SLE is an autoimmune disease characterized by spectrum of abnormalities [1]. Occurrence of PAH in SLE is not uncommon but the occurrence of PAH in patients of SLE in younger age group is a rare entity [1,8,2]. According to French registry mean age of PAH is 52 ± 15 years, with 25% of patients with > 60 years [3,2]. In majority of children PAH is associated with structural cardiac abnormality leading to Eisenmenger syndrome [8]. Clinically differentiating patients with PPH with Eisenmenger syndrome is difficult [8]. There is no significant association between the duration of SLE and the occurrence of pulmonary complications [1]. Proliferating cell nuclear antigen (PCNA) is an intranuclear protein and has a role in DNA repair and replication.

Anti-PCNA antibodies are considered a rare but highly specific marker for SLE [9, 10, 2].

Our patient, a young female presented with increasing intensity of dyspnea on exertion which initially appeared to be of cardiac origin (Pretricuspid shunt with Eisenmengerisation) from the history, physical examination and echocardiography. But further evaluation revealed primary PH due to SLE.

4. CONCLUSION

Differentiating PH from primary PAH in young patients can be challenging, leading to delayed diagnosis and treatment. A high index of suspicion is required as PAH can manifest without typical SLE symptoms. A Multidisciplinary approach with collaboration between rheumatologists, cardiologists, and pulmonologists ensures comprehensive care. Despite the rarity of PAH in young SLE patients, early diagnosis and treatment significantly improve outcomes. Comprehensive workup is essential to facilitate timely intervention and prevent disease progression.

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc) and text-to-image generators have been used during writing or editing of manuscripts.

CONSENT

As per international standards, parental written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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