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Isolated Pulmonary Hypertension with Anti-SS-A/Ro Antibodies Positivity - A Rare Case

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1. Introduction

Pulmonary Hypertension is an entity with varied types of causes. Multiple studies have also suggested an autoimmune component to the development of Pulmonary Hypertension. [1] Here is a described case of a patient with Pulmonary Hypertension and positive antinuclear antibodies (ANA) with high anti-SSA/Ro titers without associated Sjögren's Syndrome. Anti-SSA/Ro antibodies have been described in pulmonary disease in the literature, but rarely in pulmonary hypertension. This case is a rare presentation of Pulmonary Hypertension presenting with symptoms of right heart failure in conjunction with otherwise asymptomatic Sjögren's syndrome with elevated ANA and anti-SSA/Ro antibodies.

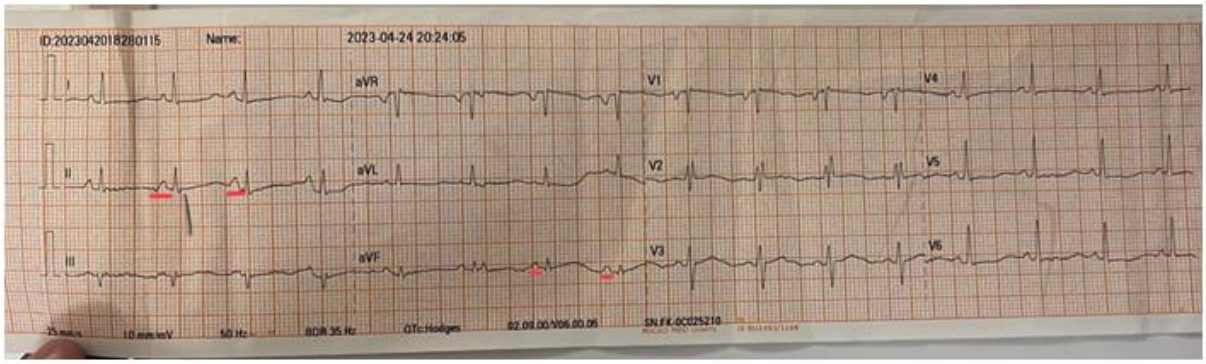
2. Case Report

A 45 years old female presented with complaints of dyspnea, abdominal pain, fatigue and bilateral lower limb swelling since 15-20 days. There was no history of cough, fever, productive sputum, reduced urine output or facial swelling. Patient had no similar history in past. There was no history of Diabetes Mellitus, Hypertension or pulmonary diseases. On examination positive findings were raised JVP measured as 12 cm of water, Bipedal pitting oedema up to mid-calf, abdominal distension with clinically detectable ascites and liver was enlarged 4 cm below right subcostal margin which was smooth tender and soft in consistency and preliminary diagnosis of Right heart failure was made. On further investigations possible causes of right heart failure like left heart failure due to valvular heart disease septal defects or isolated right heart failure due to pulmonary conditions, chronic thromboembolism were ruled out and diagnosis of Pulmonary Hypertension suspected [3]. ECG was suggestive of P pulmonale with amplitude of p wave 3mm in inferior leads [4], 2D Echo findings were suggestive of PASP (Pulmonary Arterial Systolic Pressure) 85 mm of Hg, grossly dilated Right Atrium and Right Ventricle, Normal valves except severe tricuspid regurgitation, Left Atrium and Left Ventricle were normal in dimension, No septal defect were observed and mild pericardial effusion were noted. Chest X ray and HRCT revealed no pulmonary abnormalities with cardiomegaly and pericardial effusion were only significant findings. CT-pulmonary angiography was done to rule out chronic thromboembolism. Hence diagnosis of isolated pulmonary hypertension was made. Patient had undergone HIV test which was negative. ANA profile was done to rule out autoimmune disease which was suggestive of strong anti-SS-A/Ro positivity. Patient did not have any symptoms suggestive of Sjogren syndrome like xerophthalmia, xerostomia or exocrine gland deficiency.

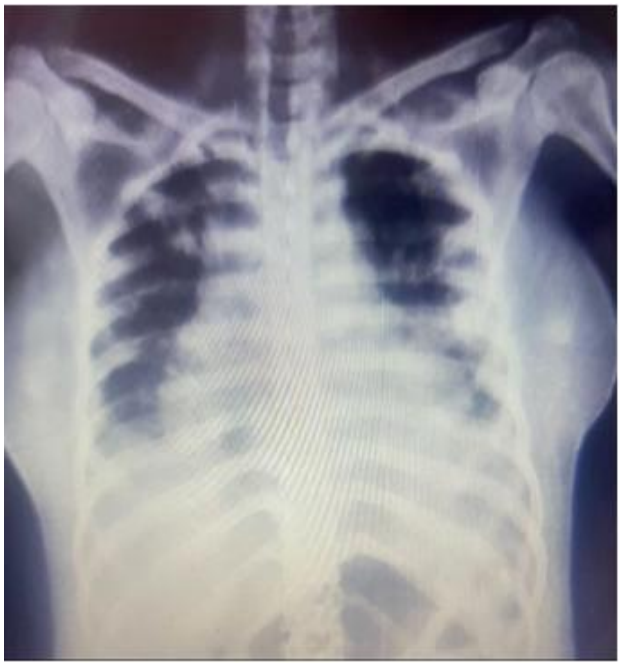
3. Discussion

Pulmonary Hypertension has been classified into 5 classes [2] recently with class 2 due to cardiac causes, class 3 due to pulmonary causes and class 4 due to pulmonary vasculature causes were easy to diagnose and treat hence were more common identified causes of pulmonary hypertension. But in recent era due to development of newer modalities it has become easier to diagnose pulmonary hypertension at earlier stages and in population without underlying diseases belonging to class 2/3/4. Isolated pulmonary hypertension belonging to class 1 seen commonly in young females with high propensity towards autoimmunity. Till now isolated Pulmonary Hypertension has been observed in patients of Scleroderma and patients infected with HIV. Further research is needed to prove firm association of Anti-SSA/Ro with isolated Pulmonary Hypertension which will help in early detection and also to halt progression and improve prognosis of the patient. If proven strong association this antibody can be used to assess chances of developing pulmonary hypertension in patients with symptoms pertaining to rheumatological diseases.

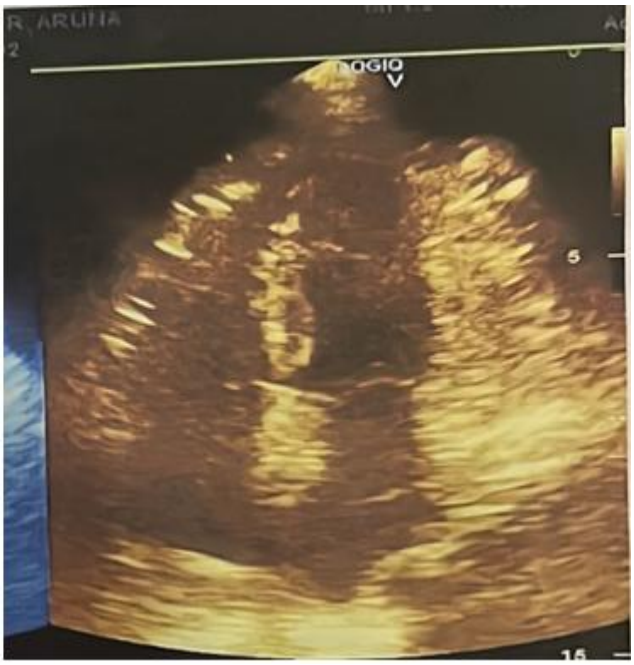
1) ECG



2) Chest X ray



3) 2D echo-



4. Conclusion

This case describes a unique association between isolated pulmonary hypertension and anti-SSA/Ro antibody, thereby illustrating the need to investigate this autoantibody and others in the pathogenesis of autoimmune pulmonary hypertension. Hence this antibody can be useful in suspecting possibility of developing pulmonary hypertension in young females presenting to OPD with symptoms pertaining to rheumatological diseases and if found with high titre of same; patient can be monitored frequently for changes of pulmonary hypertension which will be helpful for intervention and arrest of progression of disease. [5]

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