BJMHR
British Journal of Medical and Health Research
Journal home page: www.bjmhr.com

Pulmonary artery hypertension as Initial presentation of SLE: A Case Report

Sakshi Jain*, Charles Abraham E, Dharmendra Vatsaraj, Palak Doshi
Department of medicine, Smt. B. K Shah Medical Institute and Research Centre, Sumandeep Vidyapeeth, Vadodara- 391760

ABSTRACT

Pulmonary artery hypertension (PAH) is a rare initial presentation of connective tissue disorder (CTD) like systemic lupus erythematosus (SLE). SLE associated with PAH has worse prognosis than isolated SLE. However, there has been improvement in mortality of the patients in the recent years due to newer treatment modalities. Still early recognition is the key step. Here we present a case of young female with pulmonary hypertension associated with right heart failure and on further investigation was found to have CTD associated PAH. This case represents the importance of early recognition and treatment of SLE-associated PAH which will improve the survival rate in the patients.

Keywords: Pulmonary artery hypertension, systemic lupus erythematosus, connective tissue disorder

*Corresponding Author Email: krpatil22786@gmail.com
Received 04 August 2022, Accepted 30 August 2022

Please cite this article as: Jain S *et al*., Pulmonary artery hypertension as Initial presentation of SLE: A Case Report. British Journal of Medical and Health Research 2022.

INTRODUCTION

Systemic lupus erythematosus (SLE) is a disease of reproductive age group women. It is an autoimmune disease due to dysregulated activation of innate immune system. The clinical manifestations vary from musculoskeletal disease to renal, central nervous system, respiratory, cardiovascular system involvement. Genetic and environmental factors trigger the activation of innate immune system leading to defective clearance of apoptotic debris. With the advancement in treatment strategies for SLE, mortality has reduced[1]. PAH is defined as mean pulmonary artery pressure >20 mmHg measured with right heart catheterization[2]. PH is classified into 5 different categories. Category 1 includes pulmonary arterial hypertension (PAH), which has different groups as idiopathic, familial, and associated with connective tissue diseases (CTD)[3]. SLE is recognized as an emerging cause of PAH. We hereby discuss a case of young woman who presented with PAH leading to right heart failure associated with SLE.

Case Presentation

A 40 -year-old female patient who initially presented with complaints of progressively worsening shortness of breath (SOB) on exertion and mild bilateral lower extremity oedema for a duration of 2 months. She also endorsed fatigue during that time associated with myalgia and arthralgia; however, she denied any history of fevers, chills, orthopnoea, or PND. She did notice occasional chest pain with exertion for a similar period. She complains of alopecia, recurrent oral ulcers, decreased mouth opening, darkening, and stretching of skin. Her past medical history suggested hypothyroidism for 5 years, for which she was on regular medication. She also had a history of 2 abortions in the past.

Physical examination showed mild bilateral pitting oedema in the lower extremities, no jugular venous distension, regular rhythm with no murmurs appreciated, and bilateral air entry in the lungs. Multiple hyper pigmented macules and patches on the forehead, nose and cheeks. Beaked nose and purse string mouth were seen. Diffuse hyperpigmentation seen over bilateral arms, forearms, thighs and back associated with thickening of skin over hands.

Table 1: Biochemical Markers

CBC	**8.8/4000/1.59**
ESR	12
CRP	4.94
URINE Routine Micro	+1 Albuminuria
Electrolyte	136/4.2/103
Sr. Creatinine	1.0
Total bilrubin	2.0 (1.1/0.9)
SGOT	164
SGPT	67
Uric acid	4.4
RA factor	<10

DCT	Positive
CK total	91
TSH	11
Anti- TPO ab	26.1
Stool for Ova or cyst	No evidence of schistosomiasis
BNP	482

Table 2: Autoimmune workup

ANA TITER- IFA	1:100 (normal <1:40)
Pattern	Speckled with fine cytoplasmic positivity seen
SS-A	Positive
Sm (U1-nRNP)	Positive
ds DNA	Positive
Rib P- Protein	Positive
Complement C3 levels	64 mg/dl (79-152)
Complement C4 levels	20 mg/dl (16-38)

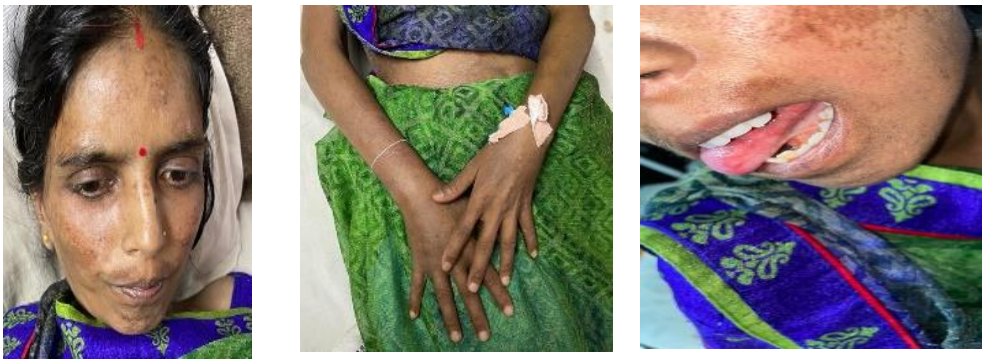

Figure 1: Cutaneous Manifestation Of SLE

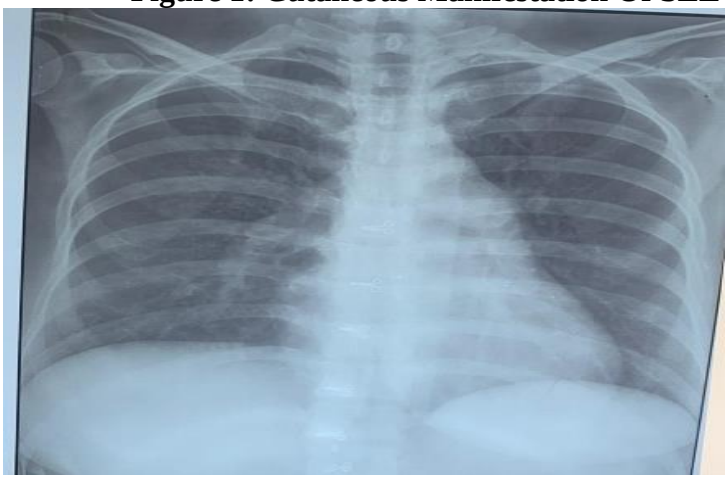

Figure 2: Chest X-Ray PA View

There is evidence of prominence of main pulmonary trunk with straightening of left heart border. Dilatation of descending pulmonary artery noted in right side. Chest x-ray was remarkable for interstitial infiltrates concerning for pulmonary edema and cardiomegaly. Findings suggest possibility of pulmonary artery hypertension

Renal Biopsy Suggestive of-

The salient features include mesangial hypercellularity along with mesangial deposits of all immunoglobulins and complements. There is no obvious evidence of thrombotic

microangiopathy. The above features are likely to be due to the autoimmune condition (equivalent to Class II Lupus nephritis).

The Patient was newly diagnosed as SLE-related connective tissue disease (SLE-CTD). Given this, her PH could be classified as group 1 PH, and she was started on tab bosentan, sildenafil with diuretic support, methotrexate and steroids were given for the management of SLE- CTD leading to PAH. On follow up the patient's PASP reduced from 84 mmHg to 40 mmHg.

Table 3: Radiological Investigations

CT Pulmonary Angiography	**Trans Esophageal Echo**
IMPRESSION 1. Enlargement of right atrium and Right ventricle with relatively small left atrium and left ventricle. 2. Dilatation of pulmonary trunk and right and left main pulmonary arteries with pulmonary trunk measuring 35.4 mm in diameter - compatible with pulmonary arterial hypertension. 3. No obvious pulmonary thromboembolism or anomalous pulmonary arteriovenous communication. 4. Mild pericardial effusion.	IMPRESSION Dilated RA/RV. Moderate TR, Severe PAH (PASP~84mmHg). RV dysfunction present. TAPSE~12mm. Mild pericardial effusion present. Intact IAS. No evidence of LA, LAA or LV clot.

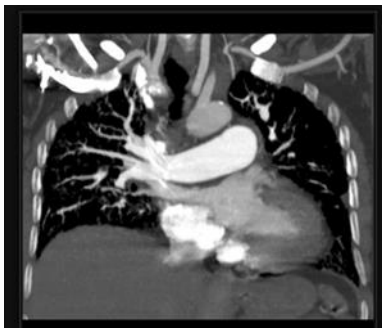

Figure 3A Dilatation of Pulmonary Trunk

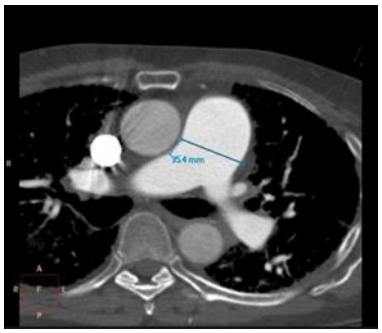

Figure 3B Pulmonary Trunk Measures 35.4 mm

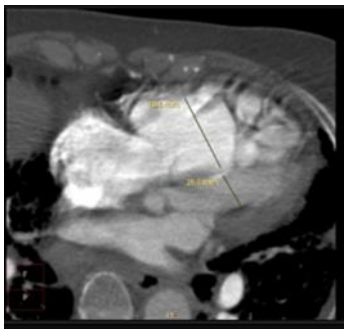

Figure 3C Enlargement of Right Atrium and Right Ventricle

Figure 3: CT Pulmonary Angiography

DISCUSSION

Pulmonary artery hypertension is a small vessel vasculopathy of the pulmonary arteries. It is a subclass of pulmonary hypertension (PH). It is defined by end-expiratory pulmonary artery wedge pressure (PAWP) as ≤15 mmHg and pulmonary artery resistance [3] Wood units on right heart catheterization and mPAP < 20 mmHg. Other causes of PH like left heart failure, primary lung disease, and venous thromboembolic disease need to be excluded. Other causes of type 1 PH are idiopathic, BMPR 2 (bone morphogenic protein receptor 2) mutation, drugs (Dasitinib, aminorex), HIV, and schistosomiasis. Some studies have noted the prevalence between 0.5% and 43% [4]. The pathophysiological mechanisms linking PAH to SLE are complex and still under investigation. Studies have proposed that an initial insult in the form

of infections, hypoxia, wall stress, or unknown stimuli to endothelium leads to an imbalance between production of vasoconstrictors (elevated levels of endothilin-1 and thromboxane A2) and vasodilators (decreased levels of prostacyclin), seen in PAH. Production of hypoxia inducible factor and erythropoietin also leads to proliferation of smooth muscles in pulmonary vessels and remodelling of vasculature [6]. Plexiform lesions: destruction of endothelial cell integrity and infiltration of inflammatory cells lead to obstruction of the lumen [7]. Another process is thromboembolic phenomenon seen in patients with positive anti-phospholipid antibodies leading to hypercoagulable state. Three main molecular pathways are targeted in the treatment of SLE-aPAH: the nitric oxide (NO) pathway, the endothilin-1 pathway, and the prostacyclin pathway. Drugs targeting these pathways are only FDA approved for PAH [5]. In treatment of SLE-aPAH, therapeutic decisions are based on variable factors including echocardiography, WHO classification functional class (FC), 6 minute walk test, and hemodynamic and laboratory parameters. Treatment modalities for PAH are: Multiple oral therapies include orally active endothelin receptor antagonists (ETRA), orally active PDE5 inhibitors, orally active guanylate cyclase stimulator (riociguat), and parental prostonoid therapy. Combination therapies are recommended. Our patient was started on low-dose steroids along with bosentan, sildenafil and methotrexate. SLE associated with PAH has worse prognosis than isolated SLE. There is a paucity of literature regarding the survival of patients with SLE who present initially with PAH. The 5-year survival in these patients is 60.2%, compared to 97.8% in isolated SLE [8]. Although SLE-aPAH carries poor prognosis, with the development of newer therapeutic options, the median survival of patients has improved from less than 3 years to a 2- year survival of more than 90% [9].

CONCLUSION

Severe PAH is a rare initial presentation of SLE. The prognosis in these patients is very grim. Advancement in therapeutic options has improved survival; however, the mortality is still very high. There is further need to research on the drugs which has mortality benefit and prompt recognition remains of utmost importance as early institution of treatment might improve the survival in these patients.

REFERENCES

1. A. Doria, M. Gatto, M. Zen, L. Iaccarino, and L. Punzi, “Optimizing outcome in SLE: treating-to-target and definition of treatment goals,” Autoimmunity Reviews, vol. 13, no. 7, pp. 770–777, 2014.
2. M. M. Hoeper, H. J. Bogaard, R. Condliffe et al., “Definitions and diagnosis of pulmonary hypertension,” Journal of the American College of Cardiology, vol. 62, no. 25, pp. D42–D50, 2013.

3. G. Simonneau, M. A. Gatzoulis, I. Adatia et al., “Updated clinical classification of pulmonary hypertension,” Journal of the American College of Cardiology, vol. 62, no. 25, pp. D34–D41, 2013.
4. J. Wang, J. Qian, Y. Wang et al., “Serological biomarkers as risk factors of SLE-associated pulmonary arterial hypertension: a systematic review and meta-analysis,” Lupus, vol. 26, no. 13, pp. 1390–1400, 2017.
5. K. Tselios, D. Gladman, and M. Urowitz, “Systemic lupus erythematosus and pulmonary arterial hypertension: links, risks, and management strategies,” Open Access Rheumatology: Research and Reviews, vol. 9, pp. 1–9, 2016.
6. L. C. Huber, H. Bye, and M. Brock, “'e pathogenesis of pulmonary hypertension—an update,” Swiss Medical Weekly, vol. 145, p. w14202, 2015.
7. S. S. Pullamsetti, R. Savai, W. Janssen et al., “Inflammation, immunological reaction and role of infection in pulmonary hypertension,” Clinical Microbiology and Infection, vol. 17, no. 1, pp. 7–14, 2011.
8. K. Kang, S.-K. Kwok, J. Ju et al., “'e causes of death in Korean patients with systemic lupus erythematosus over 11 years,” Lupus, vol. 20, no. 9, pp. 989–997, 2011.
9. W. H. Fares and T. K. Trow, “Targeted approaches to the treatment of pulmonary hypertension,” . Therapeutic Advances in Respiratory Disease, vol. 6, no. 3, pp. 147–159, 2012.