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A RESEARCH STUDY ON INTENSE PULMONARY HYPERTENSION AS THE ADVANCED DEMONSTRATION OF SLE

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ABSTRACT

The autoimmune disease systemic lupus erythematosus (SLE) affects various organ systems. Clinical symptoms might range from moderate musculoskeletal disease to life-threatening involvement of the kidneys, central neurological system, respiratory system, and haematological system.. SLE associated with pulmonary artery hypertension, on the other hand, continues to have a greater rate (PAH).Pulmonary hypertension (PH) is a collection of illnesses with a poor prognosis, which can lead to right heart dilation and failure. It's defined as a mean pulmonary artery pressure of more than 25 mmHg at rest, as determined during a right cardiac catheterization. Based on etiologies and pathophysiology, the World Health Organization (WHO) has divided PH into five distinct categories. A 32-year-old female patient came to her for the first time. A primary care physician who has been seeing patients with progressively worsening symptoms. For a period of two months, experienced worsening shortness of breath (SOB) with exertion and bilateral lower extremities edoema. No fevers, chills, orthopnea, joint aches, myalgias, or other symptoms were mentioned. Arthralgias are pains in the joints. For a comparable duration, she experienced occasional chest pain with exertion. Her medical history included the following: Losartan was just prescribed for her hypertension. Severe PAH is a rare early symptom of SLE. The prognosis for these patients is bleak. Improvements in the availability of treatment treatments has enhanced survival; yet. The death rate is still very high. The importance of prompt recognition cannot be overstated and paramount importance, as early treatment can save lives to enhance the prognosis of these individuals.

Keywords: Severe Pulmonary Hypertension, Case Report and Literature Review, Arthralgias.

INTRODUCTION

The autoimmune disease systemic lupus erythematosus (SLE) affects various organ systems. Clinical symptoms might range from moderate musculoskeletal disease to life-threatening involvement of the kidneys, central neurological system, respiratory system, and haematological system [1].

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SLE associated with pulmonary artery hypertension, on the other hand, continues to have a greater rate (PAH).Pulmonary hypertension (PH) is a collection of illnesses with a poor prognosis, which can lead to right heart dilation and failure [2]. It's defined as a mean pulmonary artery pressure of more than 25 mmHg at rest, as determined during a right cardiac catheterization. Based on etiologies and pathophysiology, the World Health Organization (WHO) has divided PH into five distinct categories [3]. Pseudoarterial hypertension (PAH) is a set of conditions classified as idiopathic and familial, as well as disorders associated with other ailments (such as connective tissue diseases (CTD)) [4, 5]. The most prevalent cause of PAH is systemic sclerosis; however, SLE is becoming more often recognized as a cause of CTD [6, 7]. In SLE, PAH prevalence varies between 0.5 and 43 percent. SLE is rarely diagnosed with severe PAH as the initial symptom [8, 9]. As

the lone initial presentation of SLE, we offer a case of a young healthy woman who presented to the hospital with severe PAH, right heart failure, and cardiogenic shock [10, 11, 12].

Case presentation:

A 32-year-old female patient came to her for the first time. A primary care physician who has been seeing patients with progressively worsening symptoms. For a period of two months, experienced worsening shortness of breath (SOB) with exertion and bilateral lower extremities edoema. No fevers, chills, orthopnea, joint aches, myalgias, or other symptoms were mentioned. Arthralgias are pains in the joints. For a comparable duration, she experienced occasional chest pain with exertion. Her medical history included the following: Losartan was just prescribed for her hypertension. She also mentioned a two-month-old sinus illness. Antibiotics were used to treat the infection on the back. Physical examination revealed modest bilateral pitting edoema in the lower 60 limbs, no jugular venous distension, and a steady heartbeat with no irregularities. There were no murmurs and bilateral air entry in the lungs. There was no indication of peripheral cyanosis, arthritis, dermatitis, or other symptoms. Skin telengectasias, or jaundice. The preliminary investigation revealed haemoglobin 13.0 g/dL, hematocrit 43.1 percent , white blood cell count 3.0 kilobytes per litre (lymphocyte count 0.4 kilobytes per litre, reference range) BNP 492, CRP, platelet count 140 K/L, 0.7–4.2 K/L, ESR 55 mm/HR and 9.8 mg/L (reference range 5.0 mg/L). (A reference range of 0–20 mm/HR is used). There was proof of Her urinalysis revealed proteinuria (3+). LDH, sGOT, and sGPT . Creatinine levels were within normal limits. The chest x-ray was notable for Pneumoedema and interstitial infiltrates are both caused by interstitial infiltrates.

A CT scan in the chest revealed mild enlargement of the central arteries due to a massive pericardial effusion.The pulmonary trunk is measured by the pulmonary arteries. 3.4 cm of bilateral axillary lymphadenopathy and 3 mm of axillary lymphadenopathy noncalcified pulmonary nodules in the right upper lobe and noncalcified pulmonary nodules in the left upper lobe upper left lobe. Low oxygen levels are becoming more of a problem when shortness of breath worsens. On a CT scan, the patient had high blood pressure and a pericardial effusion. The patient was admitted to the hospital and an echocardiography was performed showed a significant right ventricular ejection fraction (EF) of 55–60% pericardial effusion, ventricular and right atrial dilatation, pulmonary artery systolic pressure (PASP) of 89 mmHg without the physiology of tamponade 'The patient had an urgent right heart catheterization (RHC), which revealed significant pulmonary artery hypertension (PAH) with pulmonary artery stenosis. Normal pulmonary artery pressure (PAP) of 100/60mmHg 15 mmHg wedge pressure (PAWP) and decreased cardiac output. The primary PH was confirmed by the output (CO) of 3.51 L/min. C3 and C4 complements have reduced. Her PH was categorized as group 1 PH as a result, and she was started on prostacyclin. Remodulin 19 mcg/kg/min infusion for functional class IX. For severe PAH, nitric oxide (NO) was later introduced. The concentration of NO was measured in parts per million, ranging from 20 to 60. Furthermore, inotropic support and continuous vasopressor therapy. She was also put on renal replacement therapy (CRRT).500 mg solumedrol IV BID and 200 mg hydroxychloroquine for a diagnosis of SLE-CTD, once day by rheumatologist. PAH is the result of this process.

Discussion:

Pulmonary artery hypertension (PAH) is a type of pulmonary hypertension (PH) in which precapillary pulmonary hypertension is present. On right heart catheterization, it is defined by end-expiratory pulmonary artery wedge pressure (PAWP) 15 mmHg and pulmonary artery resistance >3 Wood units. Other causes of PH must also be ruled out, such as left heart failure, primary lung disease, and venous thromboembolic disease. Several types of thromboembolic illness must be ruled out. Variability in the prevalence of PAH related with SLE has been found in a number of retrospective studies (SLE-aPAH).Various studies have shown that an initial insult to the endothelium in the form of infections, hypoxia, wall stress, or unknown stimuli causes an imbalance in the synthesis of vasoconstrictors and vasodilators, resulting in elevated levels of endothilin-1 and thromboxane A2, the primary vasoconstrictors, which are found in PAH. Therapeutic recommendations for SLE-aPAH are based on a variety of criteria, including echocardiography, WHO functional class (FC), exercise capability, and hemodynamic and laboratory data. Depending on the severity of the disease, a combination of immunosuppressants and pulmonary vasodilators is usually used. For the treatment of WHO FC II with symptoms, a number of oral treatments have been licenced. ' Tese consists of three orally active endothelin receptor antagonists (ETRA) (bosentan, ambrisentan, and macitentan), two orally active PDE5 inhibitors (sildenafil and tadalafil), and one orally active guanylate cyclase stimulator (bosentan, ambrisentan, and macitentan), and one orally active guanylate cyclase stimulator ((riociguat). If single medications do not provide enough relief, increasing dosage and combination therapy are indicated. When the diagnosis of SLE was established, she was started on high-dose steroids, Plaquenil, and prostacyclin.

Table 1: Hemodynamic parametres from right heart catherization		
Hemodynamic parametres (units)	**Reference range**	**Current PAH exaberation**
RAP (mmHg)	2 -6	RAP 18/16
PAWP (mmHg)	4 - 12	15
PAP (mmHg)	20 – 30 systolic	100/60 mmHg
Mean PAP (mmHg)	8/12 diastolic	
CO (L/min)	25	76
CL (L/min)	4 – 8	3.58

Conclusion:

Severe PAH is a rare early symptom of SLE. The prognosis for these patients is bleak. Improvements in the availability of treatment treatments has enhanced survival; yet. The death rate is still very high. The importance of prompt recognition cannot be overstated and paramount importance, as early treatment can save lives to enhance the prognosis of these individuals

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