World Journal of Advanced Research and Reviews
eISSN: 2581-9615 CODEN (USA): WJARAI
Cross Ref DOI: 10.30574/wjarr
Journal homepage: https://wjarr.com/

(CASE REPORT)

Severe pulmonary hypertension as the initial manifestation of systemic lupus erythematosus: A case report a 36 years old man

Imane ESSAKET *, Imane KATIF, Meriam ATIDI, Mohamed EL JAMILI and Mustapha EL HATTAOUI

Department of cardiology, hospital university Mohmmed VI, Marrakech, Morocco.

World Journal of Advanced Research and Reviews, 2024, 21(03), 1447–1450

Publication history: Received on 12 November 2023; revised on 20 December 2023; accepted on 22 December 2023

Article DOI: https://doi.org/10.30574/wjarr.2024.21.3.2398

Abstract

Severe pulmonary hypertension as the first sign of systemic lupus erythematosus (SLE) is uncommon, but has a considerable influence on prognosis compared to other published cases of SLE. However, with the development of new therapeutic alternatives, patient mortality has decreased significantly in recent years. This case, involving a 36-year-old man whose initial presentation of SLE was severe PAH, illustrates the importance of early detection in these individuals to achieve better outcomes.

Keywords: Man; Pulmonary; Hypertension; Lupus Erythematosus

1. Introduction

SLE (Systemic Lupus Erythematosus) is a form of autoimmune disease that affects multiple organ systems in the body. SLE manifestations may differ from simple musculoskeletal concerns to serious involvement of important organs such as the kidneys, central neurological system, respiratory system, and hematological system. These possibly fatal repercussions may have an important effect on an individual's health and well-being.

However, because to developments in diagnostic procedures and treatment treatments for SLE, overall mortality rates have decreased significantly [1].

Pulmonary hypertension (PH) is a serious and sometimes fatal consequence that can arise in people with systemic lupus erythematosus (SLE) [2]. According to a recent literature review [3], the prevalence of PH in SLE ranges from 0.5% to 14%. Despite extensive research, the specific mechanisms underlying the development of PH in SLE patients remain unclear.

To fully understand the complex relationship between these antibodies and the pathophysiology of PH in SLE, further studies are required. As knowledge of the disease pathways progresses, we can look forward to better care and appropriate drugs to treat this potentially fatal consequence in SLE patients. [2]

2. Case report

A 36-year-old man presented with shortness of breath that progressively worsened over the course of a year. He also reported fatigue during this period, but denied any fever, chills, orthopnea, joint pain, myalgia or arthralgia. He noted occasional chest pain on exertion during a similar period. He had no previous medical history. Physical examination revealed no edema, no jugular venous distension, a regular rhythm with no murmur, and bilateral air entry into the lungs. There was no evidence of peripheral cyanosis, arthritis, rash, jaundice or skin telangiectasias. Initial workup

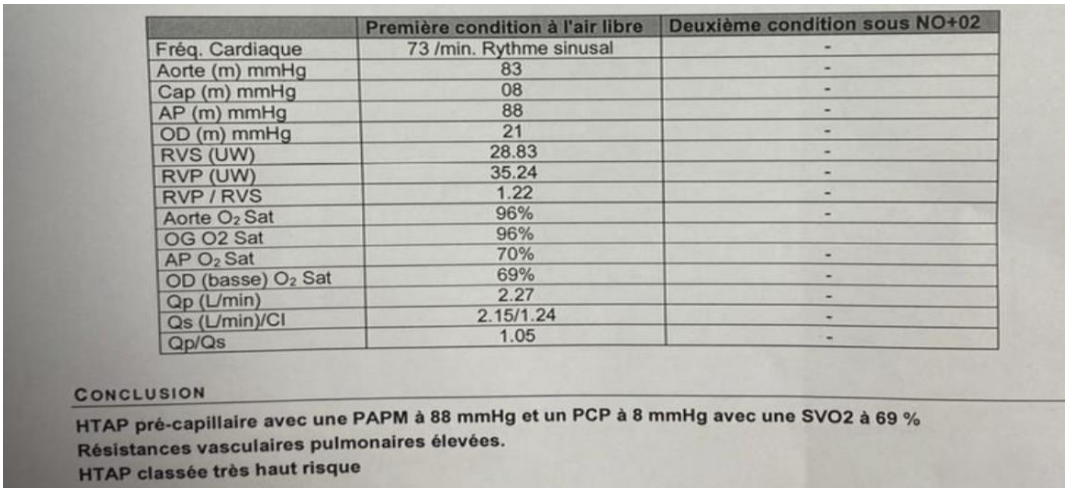
* Corresponding author: Imane ESSAKET.

revealed hemoglobin 13.8 g/dL, hematocrit 41.1%, white blood cell count 6.9 K/µL (lymphocytes 0.8 K/µL), platelet count 243 K/µL, CRP 6.7 mg/L (reference range < 5.0 mg/L), proteinuria (1+) on urinalysis. Chest X-ray revealed interstitial infiltrates suggestive of pulmonary edema and cardiomegaly. A non-contrast chest CT scan showed moderate pericardial effusion, hypertrophy of the central pulmonary arteries, with the pulmonary trunk measuring 4.1 cm, and 3 mm non-calcified pulmonary nodules in the right and left upper lobes.

On admission, an echocardiogram was performed, revealing an ejection fraction (EF) of 55-60%, significant dilatation of the right ventricle and right atrium, with a pulmonary artery systolic pressure (PASP) of 126 mmhg.

To investigate the underlying cause of his symptoms, the patient underwent emergency right heart catheterization (RHC). RCC results indicated severe pulmonary arterial hypertension (PAH) with a pulmonary artery pressure (PAP) of 97/52 mmHg, a normal pulmonary artery wedge pressure (PAWP) of 14 mmHg, and a reduced cardiac output (CO) of 2.26 L/min, confirming a primary PH (Table 1).

Table 1 Results of the right KT performed on our patient confirming pre-capillary pulmonary hypertension



	Première condition à l'air libre	Deuxième condition sous NO+02
Fréq. Cardiaque	73 /min. Rythme sinusal	-
Aorte (m) mmHg	83	-
Cap (m) mmHg	08	-
AP (m) mmHg	88	-
OD (m) mmHg	21	-
RVS (UW)	28.83	-
RVP (UW)	35.24	-
RVP / RVS	1.22	-
Aorte O_2 Sat	96%	-
OG O2 Sat	96%	
AP O_2 Sat	70%	-
OD (basse) O_2 Sat	69%	-
Qp (L/min)	2.27	-
Qs (L/min)/CI	2.15/1.24	-
Qp/Qs	1.05	-

CONCLUSION

HTAP pré-capillaire avec une PAPM à 88 mmHg et un PCP à 8 mmHg avec une SVO2 à 69 %
Résistances vasculaires pulmonaires élevées.
HTAP classée très haut risque

Considering The initial autoimmune work-up revealed a high level of antinuclear antibodies (ANA) positive for double-stranded DNA 106 (normal <16). Anti-SSA, anti-SSB and anti-Smith antibodies were negative, but the antiphospholipid antibody syndrome work-up revealed nothing significant. On the basis of the patient's clinical presentation, proteinuria, l, ANA positivity and anti-DS-DNA antibodies, a diagnosis of systemic lupus erythematosus (SLE) was suspected, in accordance with the 1997 American College of Rheumatology criteria. Unfortunately, the patient developed an arrhythmia (VT, BAV??) that did not respond to any medication and led to his death.

3. Discussion

Pulmonary arterial hypertension (PAH) represents a specific type of pulmonary hypertension (PH) distinguished by the presence of precapillary pulmonary hypertension. PAH is defined by a pulmonary artery end-expiratory wedge pressure (PAWP) of ≤15 mmHg and a pulmonary artery resistance greater than >3 Wood units, measured during right heart catheterization. To accurately diagnose PAH, it is essential to exclude other potential causes of PH, such as left heart failure, primary lung disease and venous thromboembolic disease.

The prevalence of pulmonary arterial hypertension (PAH) in patients with systemic lupus erythematosus (SLE) remains uncertain, with various studies reporting percentages ranging from 0.5% to 43% [3].However ,more recent studies have reported difference prevalence can be attributed to the diagnostic methods employed, in particular the use of echocardiography versus the gold standard of right heart catheterization (RHC).

Echocardiography is a non-invasive screening tool frequently used to identify potential cases of PAH. However, its accuracy in estimating right ventricular systolic pressure (RVSP) is around 50% [5]. While echocardiography proves to be a useful initial screening method, it is essential to recognize that its margin of error and range of sensitivities in estimating RVSP may have a significant impact on the ability to determine the true prevalence of SLE-associated PAH, particularly in the relatively small population affected by this condition.

The molecular mechanisms underlying pulmonary arterial hypertension (PAH) involve dysfunction of fibroblasts and endothelial cells, leading to impaired production of vasodilators such as nitric oxide (NO) and prostacyclin, and increased expression of vasoconstrictors, including endothelin. These molecular imbalances disrupt vascular tone and contribute to pathological changes in blood vessels, leading to pulmonary arterial vasoconstriction, in situ thrombosis (blood clot formation in blood vessels) and sometimes the development of complex plexiform lesions [6], as the disease progresses, vascular remodeling and fibrosis contribute to right ventricular (RV) dilatation and ultimately RV failure [7].

In patients with PAH associated with systemic lupus erythematosus (SLE), histological examinations have revealed the presence of macrophages, lymphocytes, antinuclear antibodies and complement in the pulmonary vasculature [8]. These immune components play a role in the inflammatory response and contribute to endothelial damage and vascular remodeling in the pulmonary arteries, adding to the complexity of PAH pathophysiology in the context of SLE. The exact interplay of these cellular and molecular mechanisms requires further investigation in order to gain a comprehensive understanding of SLE-associated PAH and develop targeted therapies to address the underlying causes of the disease [9].

The presence of pulmonary arterial hypertension (PAH) in patients with systemic lupus erythematosus (SLE) can have a significant impact on prognosis, and early recognition and treatment are crucial. In the management of SLE-associated PAH, three main molecular pathways are targeted: the nitric oxide (NO) pathway, the endothelin-1 pathway and the prostacyclin pathway:

The NO pathway plays a crucial role in vasodilation by inducing vascular smooth muscle relaxation through the production of cyclic guanosine monophosphate (cGMP). Phosphodiesterase (PDE) inhibitors are used to target this pathway. These drugs prevent the degradation of cGMP by inhibiting the PDE, thereby enhancing the vasodilatory effects of NO. In addition, there are cGMP stimulators which can also promote vasodilation.

Endothelin is a potent vasoconstrictor that is overexpressed in PAH. To counter its effects, endothelin receptor antagonists are used to block endothelin receptors, resulting in vasodilation and improved blood flow.

The prostacyclin pathway involves prostacyclin I2, which causes vasodilation of vascular smooth muscle by converting adenosine monophosphate (AMP) into cyclic AMP (cAMP). Prostanoids or prostacyclin I2 analogues can be administered to activate adenylate cyclase, mimicking the function of prostacyclin I2 and resulting in vasodilation.

It's important to note that these targeted therapies, such as PDE inhibitors, endothelin receptor antagonists and prostacyclin analogues, have been approved by the FDA specifically for the treatment of PAH and not for other World Health Organization (WHO) pulmonary hypertension classes. Prompt and appropriate use of these drugs can help manage SLE-associated PAH and improve patient outcomes. However, treatment must always be tailored to the patient's individual needs, and close monitoring is essential to assess treatment response and potential side effects.

Unfortunately, our patient presented with severe pulmonary hypertension associated with systemic lupus erythematosus (SLE), with significant functional limitations and cardiovascular involvement. Management with prostacyclin is in line with current treatment strategies for pulmonary hypertension associated with systemic lupus erythematosus. The use of prostacyclin analogues is particularly important, as they play an essential role in vasodilation and can improve symptoms and quality of life in some patients. However, despite efforts and appropriate therapies, it seems that the severity of her disease was too advanced, and the patient had to make do with prostacyclin-based treatment.

4. Conclusion

Early detection, a better understanding of the disease and the availability of more effective therapies have contributed to improved outcomes and quality of life for people living with SLE. Despite the complexity of the disease, ongoing research and medical advances offer hope for improved prognosis and management of SLE in the future.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

References

[1] A. Doria, M. Gatto, M. Zen, L. Iaccarino, et L. Punzi, « Optimizing outcome in SLE: treating-to-target and definition of treatment goals », *Autoimmun Rev*, vol. 13, nº 7, p. 770-777, juill. 2014

[2] T. Yoshio, J. Masuyama, M. Sumiya, S. Minota, et S. Kano, « Antiendothelial cell antibodies and their relation to pulmonary hypertension in systemic lupus erythematosus », *J Rheumatol*, vol. 21, nº 11, p. 2058-2063, nov. 1994.

[3] K. Tselios, D. D. Gladman, et M. B. Urowitz, « Systemic lupus erythematosus and pulmonary arterial hypertension: links, risks, and management strategies », *Open Access Rheumatol*, vol. 9, p. 1-9, 2017.

[4] L. Arnaud, C. Agard, J. Haroche, P. Cacoub, J.-C. Piette, et Z. Amoura, « Hypertension artérielle pulmonaire associée au lupus systémique », *La Revue de Médecine Interne*, vol. 32, nº 11, p. 689-697, nov. 2011.

[5] T. M. Winslow, M. A. Ossipov, G. P. Fazio, J. S. Simonson, R. F. Redberg, et N. B. Schiller, « Five-year follow-up study of the prevalence and progression of pulmonary hypertension in systemic lupus erythematosus », *American Heart Journal*, vol. 129, nº 3, p. 510-515, mars 1995.

[6] R. M. Tuder *et al.*, « Relevant Issues in the Pathology and Pathobiology of Pulmonary Hypertension », *Journal of the American College of Cardiology*, vol. 62, nº 25_Supplement, p. D4-D12, déc. 2013.

[7] M. Humbert *et al.*, « Cellular and molecular pathobiology of pulmonary arterial hypertension », *Journal of the American College of Cardiology*, vol. 43, nº 12_Supplement, p. S13-S24, juin 2004.

[8] X. Jais *et al.*, « Immunosuppressive therapy in lupus- and mixed connective tissue disease–associated pulmonary arterial hypertension: A retrospective analysis of twenty-three cases », *Arthritis & Rheumatism*, vol. 58, nº 2, p. 521-531, 2008.