

## Systemic Lupus Erythematosus Complicated by Dilated Cardiomyopathy and Severe Heart Failure: A Case Report

Driss Britel<sup>2</sup>, Mehdi Moujahid<sup>1\*</sup>, Laktib Nabil<sup>2</sup>, Youssef Fihri<sup>2</sup>, Soumia Faïd<sup>2</sup>, Hicham Faliouni<sup>2</sup>, Najat Mouine<sup>1</sup>, Ilyasse Asfalou<sup>3</sup>, Zouhair Lakhal<sup>2</sup>, Aatif Benyass<sup>4</sup>

<sup>1</sup>Clinical Cardiology Department, Military Hospital Mohamed V, Rabat, Morocco

<sup>2</sup>Cardiac Catheterization Unit, Military Hospital Mohamed V, Rabat, Morocco

<sup>3</sup>Non-invasive cardiology department, Military Hospital Mohamed V, Rabat, Morocco

<sup>4</sup>Cardiology Center, Military Hospital Mohamed V, Rabat, Morocco

DOI: <https://doi.org/10.36347/sasjm.2024.v10i11.001>

| Received: 24.09.2024 | Accepted: 30.10.2024 | Published: 01.11.2024

\*Corresponding author: Mehdi Moujahid

Clinical Cardiology Department, Military Hospital Mohamed V, Rabat, Morocco

### Abstract

### Case Report

Systemic lupus erythematosus (SLE), a connective tissue disease characterized by the production of autoantibodies, can affect all organ systems. Cardiac involvement in patients with systemic lupus erythematosus (SLE) has been documented since the early 20th century. The manifestations may be diverse and may affect any portion of the heart, including the pericardium, the conduction system, the myocardium, the valves, and the coronary arteries. The incidence of cardiomyopathy associated with systemic lupus erythematosus (SLE) that is clinically manifested is rare; the majority of studies have indicated a prevalence of approximately 10%. Echocardiography has offered insights into cardiac anatomy and function in patients with systemic lupus erythematosus (SLE), with and without cardiac involvement. Endomyocardial biopsy is considered as the gold standard for diagnosis; however, its low sensitivity and risk of complications have limited its application. Cardiac magnetic resonance (CMR) imaging offers a promising alternative to myocardial biopsy. We present the case of a 57-year-old female with a history of lupus since 2010, who presented with an acute inaugural decompensation of dilated cardiomyopathy, with a left ventricular ejection fraction (LVEF) of 28%.

**Keywords:** Dilated cardiomyopathy, autoimmune disorder, heart failure, lupus cardiomyopathy, systemic lupus erythematosus.

Copyright © 2024 The Author(s): This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CC BY-NC 4.0) which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use provided the original author and source are credited.

## INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disorder that is characterized by inflammation affecting multiple systems and the presence of autoantibodies that attack self-antigens [1]. The clinical presentation of the disease often follows a relapsing and remitting pattern, resulting in multisystem inflammation and a wide range of symptoms [2]. Approximately 90% of SLE cases manifest in females, frequently emerging during their reproductive years [3]. Cardiac involvement has been documented in more than 50% of patients [4, 5]. Furthermore, the disease can affect any of the three layers of cardiac tissue: the endocardium, the myocardium, or the epicardium, as well as the pericardial serosa [6]. However, while the incidence of cardiac involvement is elevated, the incidence of dilated cardiomyopathy in SLE remains unknown [5, 6]. The management of SLE

cardiomyopathy is challenging due to its rarity. Therefore, a multidisciplinary approach is essential for optimal outcomes [7]. We report the case of a 57-year-old woman treated for lupus since 2010 who presented with acute onset of decompensated dilated cardiomyopathy. The diagnosis was made on the basis of comprehensive evaluation.

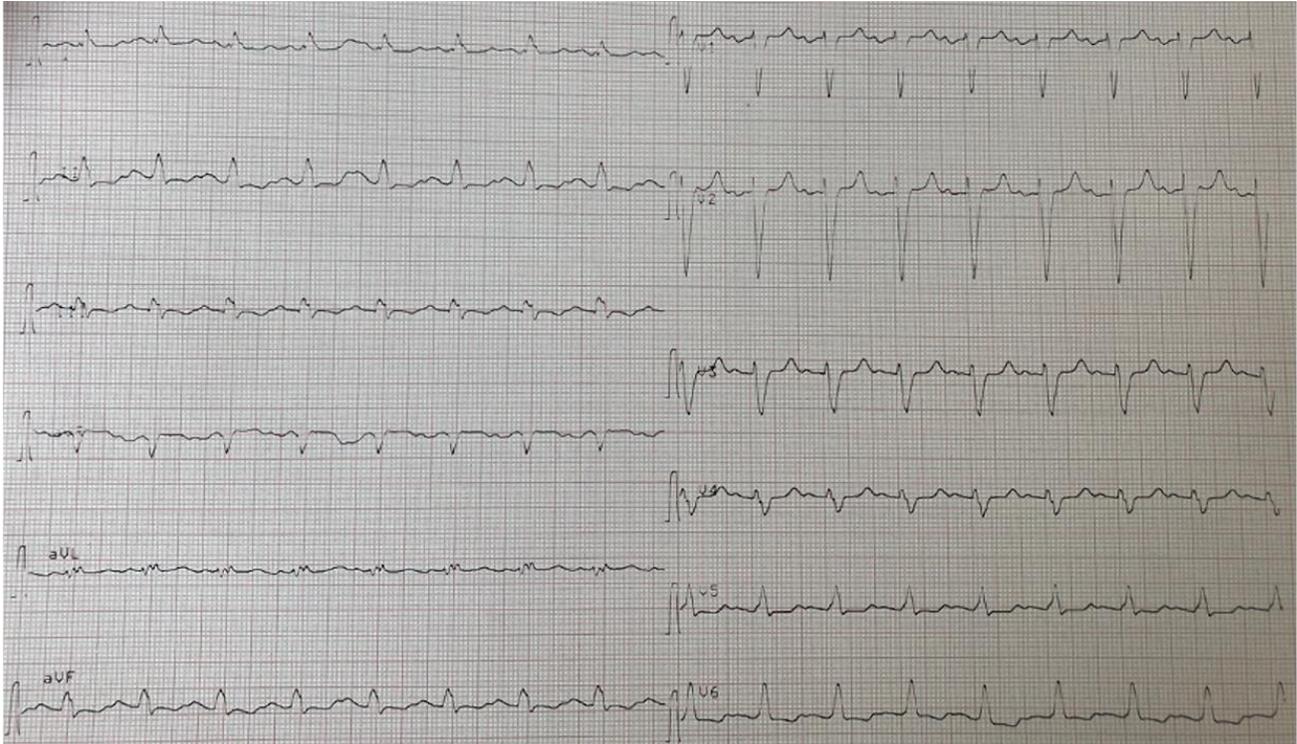
## CASE PRESENTATION

A 57-year-old female with a history of systemic lupus erythematosus since 2010, who had no cardiovascular risk factors, presented to our department with progressive dyspnea on exertion (New York Heart Association class III) and orthopnea. Her symptoms developed one month prior to presentation but worsened one week before admission. On initial evaluation, the patient was dyspneic and tachycardic at rest. Her blood pressure was 110/80 mmHg, pulse rate was 124 beats per

**Citation:** Driss Britel, Mehdi Moujahid, Laktib Nabil, Youssef Fihri, Soumia Faïd, Hicham Faliouni, Najat Mouine, Ilyasse Asfalou, Zouhair Lakhal, Aatif Benyass. Systemic Lupus Erythematosus Complicated by Dilated Cardiomyopathy and Severe Heart Failure: A Case Report. SAS J Med, 2024 Nov 10(11): N/A.

minute, respiratory rate was 32 breaths per minute, and body temperature was 36.7°C. Examination revealed signs of heart failure, including marked jugular venous distension, bilateral basal rales, and significant bilateral

leg edema. The electrocardiogram (EKG) (Figure 1) showed a regular sinus rhythm with a mean heart rate of 100 beats per minute and a complete left bundle branch block.



**Figure 1: The electrocardiogram shows a complete left bundle branch block**

The chest X-ray (Figure 2) showed symmetrical globular enlargement of the heart and pulmonary congestion with a perihilar distribution.



**Figure 2: The chest x-ray reveals enlargement of the heart and pulmonary congestion**

Her laboratory tests revealed (Table 1) elevated BNP and d-dimers level, lymphopenia and normal Troponin level. Routine blood investigations including

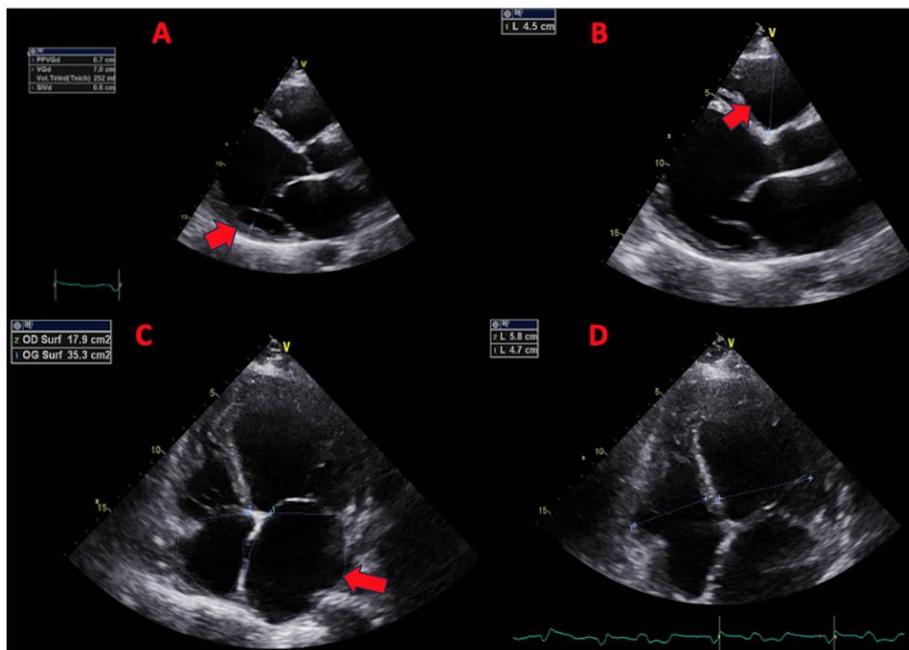
coagulation parameters, liver and renal function tests were normal.

**Table 1: Results of our patient's biological tests**

Parameters	Observed Values	Normal range
Haemoglobin	13,8 g/dl	12-16 g/dl
White blood cell	6800/mm <sup>3</sup>	4000-10 000/mm <sup>3</sup>
Lymphocytes	700/mm <sup>3</sup>	1500-4000/mm <sup>3</sup>
Platelets	215 000/mm <sup>3</sup>	150 000-400 000/mm <sup>3</sup>
Troponine	14 ng/l	2-34 ng/l
C-reactive protein	4,1 mg/l	<5mg/l
Natraemia	134 mmol/l	135-145 mmol/l
Kalemia	4,02 mmol/l	3,5-5,5 mmol/l
Magnesiumemia	16,9 mg/l	18-26 mg/l
Chloremia	106 mmol/l	100-110 mmol/l
Phosphoremia	2,9 mg/dl	2,7-4,5 mg/dl
Calcemia	89 mg/l	88-104 mg/l
NT-Pro-BNP	10 864 ng/l	< 125 ng/l
HBA1C	5,3%	<6,5%
Total cholesterol	1,36 g/l	<2g/l
LDL cholesterol	0,77 g/l	1-1,60 g/l
HDL cholesterol	0,38 g/l	0,4-0,6 g/l
Triglyceride	1,06 g/l	<1,5g/l
Uraemia	0,33 g/l	0,15-0,5 g/l
Creatinemia	8 mg/l	6-13 mg/l
Estimated glomerular filtration rate (eGFR).	78,68 ml/min/1.73m <sup>2</sup>	>60 ml/min/1,73m <sup>2</sup>
Alanine transaminase (ALT)	17 UI/l	<40 UI/l
Aspartate transaminase (AST).	13 UI/l	<35 UI/l
Total Bilirubin	8 mg/l	3-10 mg/l
Gamma-glutamyltransferase (GGT)	20 UI/l	< 32 UI/l
Prothrombin time (PT).	100 %	>70%
Alkaline phosphatase (ALP)	45 UI/l	40-150 UI/l
Albumin	38 mg/l.	>30 mg/l
Thyroid stimulating hormone (TSH)	1,23 mUI/l	0.4-4 mUI/l
Thyroxine (T4)	13 pmol/l	9-19 pmol/l

The echocardiography revealed a significant biventricular enlargement (Figure 3) with diffuse hypokinesia and a markedly depressed left ventricular

ejection fraction (LVEF = 28 %) (Figure 4). No pericardial effusion.



**Figure 3: The echocardiography demonstrates biventricular enlargement**

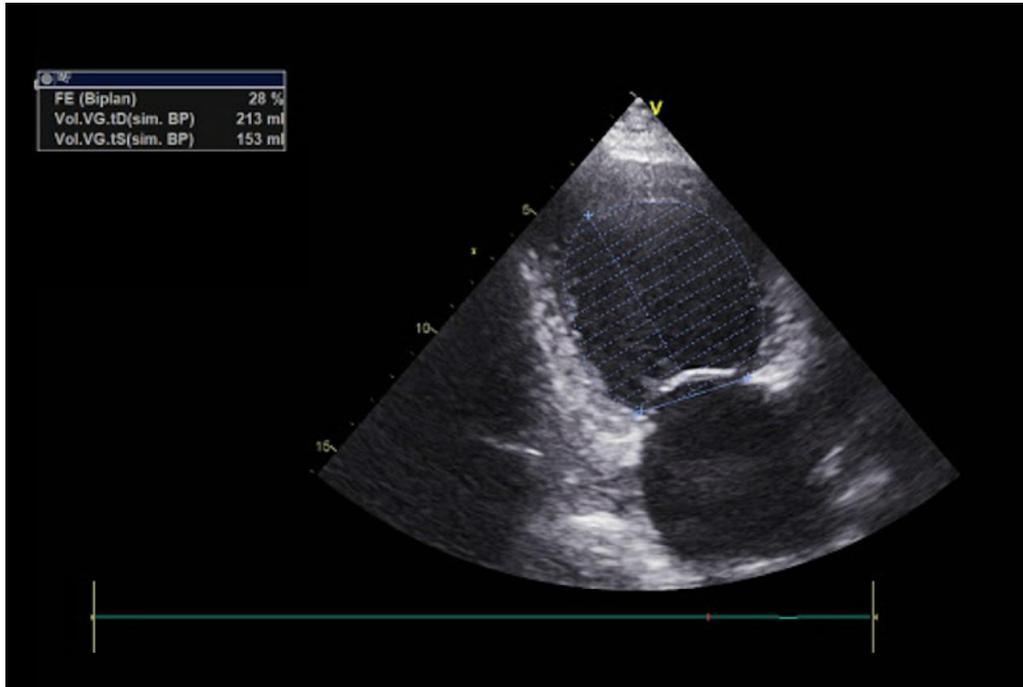
The echocardiogram reveals a dilated left ventricle (subimage A) with a telediastolic diameter of 70 mm, a dilated right ventricle (subimage B) with an antero-posterior diameter of 45 mm, and resulting in a right ventricle-to-left ventricle ratio of less than 1 (subimage D). Additionally, the left atria were observed to be dilated (subimage C), with a surface area of 35 cm<sup>2</sup>

L = Length

OG surf = Surface of the left atrium.

OD surf = Surface of the right atrium.

VGd = Telediastolic diameter of the left ventricle.



**Figure 4: The echocardiography shows a depressed left ventricular ejection fraction (LVEF=28%)**

FE = Ejection Fraction

Vol VG tD = Tele diastolic left ventricular volume.

Vol VG tS = Tele systolic left ventricular volume.

Reversible causes of cardiomyopathy were considered. Chest computed tomography angiography was not supportive of pulmonary embolism as the underlying etiology of right sided heart failure. Laboratory tests revealed normal iron and vitamins levels and normal thyroid function. Serological examination for viral infections, including SARS COV2, herpes simplex, cytomegalovirus, Epstein Barr, rubeola, hepatitis B and C, and HIV were all normal. A search for underlying malignancy was also negative. Coronary angiography revealed normal coronary arteries with no evidence of stenosis. Cardiac MRI revealed primary dilated hypokinetic cardiomyopathy, with a left ventricular ejection fraction (LVEF= 26%). There were no sequelae of infarction or myocarditis, but there was dilatation of the left atrium and mitral regurgitation, along with minimal pericardial detachment.

## DISCUSSION

Dilated cardiomyopathy (DCM) is a disease of the heart muscle defined by systolic dilation and dysfunction of one or both ventricles, frequently

presenting with symptoms of congestive heart failure [8]. Several factors can contribute to the development of dilated cardiomyopathy (DCM), such as viral infections, alcohol and drug use, exposure to toxins, pregnancy and the postpartum period, thyrotoxicosis, autoimmune collagen vascular diseases, and genetic susceptibility [8]. In our patient, most of these possibilities were ruled out. The causes were reduced to two: autoimmune and idiopathic cardiomyopathy.

Systemic lupus erythematosus (SLE) is an autoimmune inflammatory disease of unknown etiology [6], which primarily affects women of reproductive age [6]. Can damage multiple organs, including the heart [9]. The American College of Rheumatology has developed 11 diagnostic criteria for SLE [7]. A definitive diagnosis requires the presence of 4 of these criteria [10].

Cardiac manifestations in SLE are varied [7]. Pericarditis is the most common form of cardiac involvement [11]. Myocardial involvement is less common in SLE, occurring in up to 9% of patients [12], and often associated with coronary atherosclerosis [13].

Dilated cardiomyopathy is a rare and usually late clinical manifestation of SLE, rarely reported in the literature [14].

The exact pathogenesis of myocardial involvement is not fully understood [15]. Case reports of SLE patients with cardiac manifestations who underwent myocardial biopsy do not provide evidence of myocarditis [16, 17]. This suggests that myocardial inflammation may not be the pathophysiologic basis [7], and that other factors such as thrombotic or inflammatory microvascular coronary disease may be involved [7, 18].

Lupus cardiomyopathy is diagnosed based on clinical, electrocardiographic, echocardiographic, angiographic, and biochemical features [7]. Coronary angiography is essential for differentiating lupus cardiomyopathy from coronary artery atherosclerosis [7]. Endomyocardial biopsy is the gold standard for confirming the diagnosis [19]. Due to low sensitivity and potential complications, this procedure is not used [20]. Cardiac magnetic resonance (CMR) is an emerging noninvasive imaging method [7], that replace myocardial biopsy [21].

The management of cardiomyopathy associated with lupus is unclear due to its rarity. Initial and early treatment may include steroids, heart failure drugs, anticoagulants, antiarrhythmics, and cardiovascular risk factor management [12]. Immunotherapy may be effective in active inflammation [22]. In severe and advanced cases, heart transplantation is an option for SLE patients with heart failure [23].

## CONCLUSIONS

Cardiac Systemic lupus erythematosus is a complex disease with multiple possible pathological processes that requires prompt investigation and treatment. Delays in diagnosis and treatment can result in irreversible damage, as evidenced by our patient's case. The early detection of preclinical dilated cardiomyopathy has the potential to significantly reduce morbidity and mortality by initiating cardioprotective therapy at an early stage.

## REFERENCES

1. Rebecca, E. S., & Stacy, P. A. Systemic Lupus Erythematosus. Nelson Textbook of Pediatrics, chapter 199. Elsevier (ed): 2025. 2, 1490-1499.
2. Systemic Lupus Erythematosus (SLE). (2024). <https://emedicine.medscape.com/article/332244-overview>.
3. Siegel, C. H., & Sammaritano, L. R. (2024). Systemic Lupus Erythematosus: A Review. *JAMA*. Doi: 10.1001/jama.2024.2315
4. Tincani, A., Rebaioli, C. B., Taglietti, M., & Shoenfeld, Y. (2006). Heart involvement in systemic lupus erythematosus, anti-phospholipid syndrome and neonatal lupus. *Rheumatology*, 45(suppl\_4), iv8-iv13. Doi: 10.1093/rheumatology/ke1308
5. Wijetunga, M., & Rockson, S. (2002). Myocarditis in systemic lupus erythematosus. *The American journal of medicine*, 113(5), 419-423. Doi: 10.1016/s0002-9343(02)01223-8
6. Durrance, R. J., Movahedian, M., Haile, W., Teller, K., & Pinsker, R. (2019). Systemic lupus erythematosus presenting as myopericarditis with acute heart failure: A case report and literature review. *Case reports in rheumatology*, 2019(1), 6173276. Doi: 10.1155/2019/6173276
7. Zinoune, L., Maazouzi, M., Benbouchta, K., Amaqdouf, S., & Ismaili, N. (2022). Dilated cardiomyopathy complicated by an intracavitary thrombus and acute heart failure: A rare presentation of systemic lupus erythematosus. *Annals of Medicine and Surgery*, 82, 104739. Doi: 10.1016/j.amsu.2022.104739
8. Mahmaljy, H., Yelamanchili, V. S., & Singhal, M. (2023). Dilated cardiomyopathy. In *StatPearls [Internet]*. StatPearls Publishing.
9. Jain, D., & Halushka, M. K. (2009). Cardiac pathology of systemic lupus erythematosus. *Journal of clinical pathology*, 62(7), 584-592. Doi: 10.1136/jcp.2009.064311
10. Em, T. (1982). The 1982 revised criteria for the classification of systemic lupus erythematosus. *Arthritis Rheum.*, 25, 1271-1277. Doi: 10.1002/art.1780251101
11. Mohamed, A. A., Hammam, N., El Zohri, M. H., & Gheita, T. A. (2019). Cardiac manifestations in systemic lupus erythematosus: clinical correlates of subclinical echocardiographic features. *BioMed Research International*, 2019(1), 2437105. Doi: 10.1155/2019/2437105
12. Ishimori, M. L., Agarwal, M., Beigel, R., Ng, R. K., Firooz, N., Weisman, M. H., & Siegel, R. J. (2014). Systemic lupus erythematosus cardiomyopathy—a case series demonstrating a reversible form of left ventricular dysfunction. *Echocardiography*, 31(5), 563-568. Doi: 10.1111/echo.12425
13. Wilhelm, A. J., & Major, A. S. (2012). Accelerated atherosclerosis in SLE: mechanisms and prevention approaches. *International journal of clinical rheumatology*, 7(5), 527-539. Doi: 10.2217/ijr.12.46
14. Caforio, A. L., Adler, Y., Agostini, C., Allanore, Y., Anastasakis, A., Arad, M., ... & Linhart, A. (2017). Diagnosis and management of myocardial involvement in systemic immune-mediated diseases: a position statement of the European Society of Cardiology Working Group on Myocardial and Pericardial Disease. *European heart journal*, 38(35), 2649-2662. Doi: 10.1093/eurheartj/ehx321
15. Kim, C. H., Al-Kindi, S. G., Jandali, B., Askari, A. D., Zacharias, M., & Oliveira, G. H. (2017). Incidence and risk of heart failure in systemic lupus

- erythematosus. *Heart*, 103(3), 227-233. 10.1136/heartjnl-2016-309561
16. Gottenberg, J. E., Roux, S., Assayag, P., Clerc, D., & Mariette, X. (2004). Specific cardiomyopathy in lupus patients: report of three cases. *Joint Bone Spine*, 71(1), 66-69. [https://doi.org/10.1016/S1297-319X\(03\)00101-5](https://doi.org/10.1016/S1297-319X(03)00101-5).
  17. Cervera, R., Font, J., Pare, C., Azqueta, M., Perez-Villa, F., Lopez-Soto, A., & Ingelmo, M. (1992). Cardiac disease in systemic lupus erythematosus: prospective study of 70 patients. *Annals of the Rheumatic Diseases*, 51(2), 156-159. 10.1136/ard.51.2.156
  18. Murai, K., Oku, H., Takeuchi, K., Kanayama, Y., Inoue, T., & Takeda, T. (1987). Alterations in myocardial systolic and diastolic function in patients with active systemic lupus erythematosus. *American heart journal*, 113(4), 966-971. Doi: 10.1016/0002-8703(87)90058-5
  19. Caforio, A. L., Pankuweit, S., Arbustini, E., Basso, C., Gimeno-Blanes, J., Felix, S. B., ... & Elliott, P. M. (2013). Current state of knowledge on aetiology, diagnosis, management, and therapy of myocarditis: a position statement of the European Society of Cardiology Working Group on Myocardial and Pericardial Diseases. *European heart journal*, 34(33), 2636-2648. Doi: 10.1093/eurheartj/ehz210
  20. Gartshteyn, Y., Tamargo, M., Fleischer, S., Kapoor, T., Li, J., Askanase, A., ... & Geraldino-Pardilla, L. (2020). Endomyocardial biopsies in the diagnosis of myocardial involvement in systemic lupus erythematosus. *Lupus*, 29(2), 199-204. Doi: 10.1177/0961203319897116
  21. de Roos, A. (2019). Diagnosis of myocarditis at cardiac MRI: the continuing quest for improved tissue characterization. *Radiology*, 292(3), 618-619. Doi: 10.1148/radiol.2019191476
  22. Meridor, K., Shoenfeld, Y., Tayer-Shifman, O., & Levy, Y. (2021). Lupus acute cardiomyopathy is highly responsive to intravenous immunoglobulin treatment: Case series and literature review. *Medicine*, 100(18), e25591. Doi: 10.1097/MD.00000000000025591
  23. Ashrafi, R., Garg, P., McKay, E., Gosney, J., Chuah, S., & Davis, G. (2011). Aggressive cardiac involvement in systemic lupus erythematosus: a case report and a comprehensive literature review. *Cardiology Research and Practice*, 2011(1), 578390. Doi: 10.4061/2011/578390