narra j

Case Report

Cardiomyopathy as the forgotten symptom of systemic lupus erythematosus in children: A case report

Eric Yudhianto[1], Ririe F. Malisie[1,2,3*], Hafaz Z. Abdillah[2,3] and Rita E. Rusli[2,3]

[1]Department of Emergency Medicine, Murni Teguh Memorial Hospital, Medan, Indonesia; [2]Department of Pediatrics, Murni Teguh Memorial Hospital, Medan, Indonesia; [3]Department of Pediatrics, Faculty of Medicine, Universitas Sumatera Utara, Medan, Indonesia

*Corresponding author: ririe.fachrina.malisie@usu.ac.id

Abstract

Cardiomyopathy is a rare clinical manifestation in pediatric systemic lupus erythematosus (SLE), with only a single case reported in the literature. Its identification in pediatric SLE is challenging due to its typically subclinical presentation and low incidence, which frequently result in delayed diagnosis and management. The aim of this study was to present a unique case of dilated cardiomyopathy, a rare cardiac complication of SLE, which can be life-threatening if not promptly recognized and treated. An 11-year-old boy was admitted to the emergency department of Murni Teguh Memorial Hospital, Medan, Indonesia, diagnosed with SLE based on the 2019 European Alliance of Associations for Rheumatology (EULAR) criteria, with a total score of 30 and a Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) score of 16, indicating high disease activity. Clinical findings included oral ulcers, a non-pruritic hyperpigmented discoid macule, anemia, lymphopenia, positive both the direct and indirect Coombs tests, elevated D-dimer level, and pulmonary congestion. Initial treatment stabilized the patient condition, allowing transfer to the general ward by day five. Five days after admission, the patient developed palpitations and tachycardia, with a heart rate of 140 beats per minute. Electrocardiography showed sinus tachycardia, while echocardiography revealed all cardiac chambers dilation, left ventricular ejection fraction (LVEF) of 43%, moderate mitral regurgitation, and mild pulmonary regurgitation, subsequently diagnosed as dilated cardiomyopathy. Heart failure therapy was initiated with intravenous furosemide, oral ramipril, and digoxin. Palpitations and tachycardia resolved within two days. Following two weeks of treatment, the patient was discharged with stable vital signs. A one-month follow-up thoracic echocardiography demonstrated improved cardiomyopathy, with an LVEF of 53%. Cardiomyopathy in pediatric SLE is rare but can cause significant morbidity and mortality if undiagnosed. Its nonspecific presentation and immune-mediated pathogenesis make early detection challenging. Due to its rarity, it may be overlooked, highlighting the importance of comprehensive cardiac evaluation, including echocardiography, in children with suspected cardiac involvement.

Keywords: Pediatric, autoimmune, systemic lupus erythematosus, cardiac involvement, cardiomyopathy

Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune condition characterized by extensive inflammation due to the presence of antibodies targeting self-antigens in the

https://narraj.org/

Received: October 12, 2024 - Accepted: February 5, 2025 - Published online: February 17, 2025

bloodstream [1]. It affects individuals of all ages and genders, with a higher prevalence observed in women of childbearing age [1]. Pediatric SLE is a rare autoimmune disorder, with an incidence reported to range from 0.28 to 0.9 per 100,000 children annually and a prevalence of 3.3 to 8.8 per 100,000 in the pediatric population [2]. The condition typically manifests during the peripubertal stage, most commonly between the ages of 11 and 12, and predominantly affects females, with a female-to-male ratio of 4:1 [3].

Despite its low prevalence, pediatric SLE is clinically significant due to its potential for severe multi-organ involvement and a more aggressive disease course compared to adult-onset SLE, necessitating early recognition and prompt management to mitigate complications and improve long-term outcomes [4]. Studies reported that children demonstrated higher disease activity at the time of diagnosis than adults, which correlated with more severe clinical presentations [4,5]. Additionally, the diagnosis of pediatric SLE was frequently delayed due to the variability in symptoms and overlap with other pediatric conditions; such delays often resulted in a more advanced disease stage at the time of diagnosis, further worsening symptom severity, consequently contributing to higher morbidity and mortality rates in pediatric SLE [6]. The five-year survival rate for childhood-onset SLE was reported to be approximately 65%, reflecting the significant risks associated with this condition [5]. Approximately 27% of pediatric SLE patients died within one year of diagnosis, with infections and renal failure identified as the primary causes of death [7].

SLE is associated with significant complications involving the integumentary system, joints, kidneys, blood cells, cardiovascular system, and nervous system [1]. Among these, renal involvement is the most prevalent, occurring in 50% to 75% of cases and often manifesting as lupus nephritis [8]. Cardiovascular complications have been reported in approximately 50% of patients with SLE, with pericarditis being the most common type [9]. Other cardiovascular manifestations include coronary artery disease, myocarditis, thromboembolic events, and conduction abnormalities [10].

Coronary artery disease in SLE is primarily driven by widespread inflammatory cytokines, particularly interferon-alpha, which contribute to endothelial dysfunction and accelerate atherosclerosis [11]. This process impairs vascular repair mechanisms, leading to plaque formation and progressive narrowing of coronary arteries [11]. Immune-mediated damage to the myocardium has been associated with myocarditis, as well as valvular and conduction abnormalities [12]. Furthermore, an increased risk of thromboembolic events is attributed to the presence of antiphospholipid antibodies, which induce a hypercoagulable state [13].

The annual incidence of childhood cardiomyopathy is approximately 1.13 per 100,000, with dilated cardiomyopathy being the most common subtype in children, reported at an incidence of 0.57 cases per 100,000 per year [14]. However, the exact prevalence of cardiomyopathy directly associated with SLE remains uncertain, with only one documented pediatric case report available in the literature [15]. Cardiomyopathy with clinical manifestations in SLE is rare [15]. In children, dilated cardiomyopathy is often undiagnosed until symptoms emerge or signs of heart failure become apparent [16]. Approximately 8% to 15% of pediatric dilated cardiomyopathy patients present with cardiac arrest, sudden death, or near-arrest events requiring emergency resuscitation [17,18]. Among patients seeking hospital care for the first time, shortness of breath is the most commonly reported symptom, occurring in approximately 50% of cases [17]. Gastrointestinal complaints and fatigue are reported by one-third of patients, while cough and symptoms mimicking an upper respiratory tract infection are observed in up to 20% of cases [16].

Diagnosing SLE in pediatrics poses significant challenges due to its wide range of symptoms, which are often non-specific and may overlap with other conditions. Furthermore, the identification of cardiomyopathy in pediatric SLE is particularly complex, as it is characterized by subclinical involvement and a low incidence, frequently resulting in delayed diagnosis and management. The aim of this study was to present a case of an 11-year-old boy diagnosed with dilated cardiomyopathy, a rare cardiac complication of SLE. This report aims to highlight awareness of this rare but severe complication, which can be life-threatening if not promptly recognized and treated.

Case

An 11-year-old boy was admitted to the emergency department of Murni Teguh Memorial Hospital, Medan, Indonesia, with a primary complaint of high fever persisting for two days and seizures occurring at home. The patient experienced a 5-minute tonic-clonic seizure with eyes wide open. Following the seizure, the patient regained consciousness but reported a headache. According to the patient's parents, intermittent fever had been present for one month. The patient also complained of nausea and vomiting occurring twice a day for the past day, along with a productive cough and joint and muscle pain lasting for more than two weeks. Additionally, the patient experienced unexplained fatigue and hair loss for the past two months. There was no family history of autoimmune or heart disease.

Upon admission, a physical examination revealed that the patient was alert, with a blood pressure of 90/50 mmHg, a heart rate of 150 beats per minute (bpm) in a regular rhythm, and a respiratory rate of 24 breaths per minute, which was also regular. The patient's oxygen saturation (SaO_2) was 93% on room air; however, with the administration of 3 liters per minute via a nasal cannula, the SaO_2 stabilized at 99%. The patient's temperature was recorded at 40°C. No abnormalities were noted during the examination of the head, eyes, ears, nose, and neck. Oral ulcers and erythema were observed on the hard palate. No murmurs, rhonchi, or wheezing were detected. A non-pruritic, hyperpigmented discoid macule was noted on the trunk, thighs, palms, hands, and feet. According to the patient's parents, the rash had been red in color one month prior (**Figure 1**).

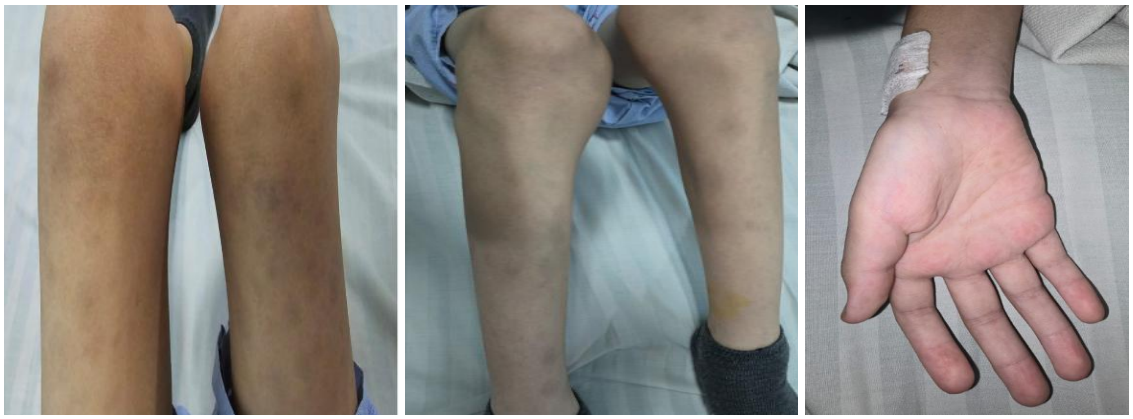

Figure 1. Non-pruritic hyperpigmented macules predominantly located on the thigh and lower leg, exhibiting irregular borders and vary in size, with no associated itching or other significant dermatological symptoms.

Laboratory examination revealed hypochromic microcytic anemia, with hemoglobin levels at 6.4 g/dL, mean corpuscular volume (MCV) at 74.1 fL and mean corpuscular hemoglobin (MCH) at 24.3 pg. Neutrophilia was observed at 79.9%, while lymphopenia was noted at 10.6%. A high monocyte counts of 9.3% were also presented. Platelet and white blood cell counts were within normal limits. The peripheral blood smear test suggested suspected autoimmune hemolytic anemia with reactive neutrophilia. Renal function (urea and creatinine) was within normal limits. The patient also showed elevated liver enzymes, with alanine aminotransferase (ALT) and aspartate aminotransferase (AST) levels at 109 U/L and 621 U/L, respectively. C-reactive protein (CRP) was measured at 10 mg/dL. Dengue NS1 and anti-Salmonella IgM tests were negative. No abnormalities were detected in the electrolyte levels, including potassium, sodium, and chloride levels. Both the direct and indirect Coombs tests were positive (+3). D-dimer levels were elevated at 6.18. Prothrombin time (PT) and activated partial thromboplastin time (APTT) tests were normal. Albumin levels were 2.7 g/dL. Serology polymerase chain reaction (PCR) for enterovirus was negative, and urinalysis exhibited protein at 2+. Blood cultures showed no microorganism growth.

Anti-nuclear antibody immunofluorescence (ANA IF) testing revealed a titer of 1:320 with a homogeneous pattern, while the qualitative rheumatoid factor (RF) test was negative. A chest X-ray demonstrated pulmonary congestion (**Figure 2**). Computed tomography (CT) imaging of the

upper and lower abdomen identified grade 1 left hydronephrosis and minimal bilateral pleural effusion (**Figure 2**).

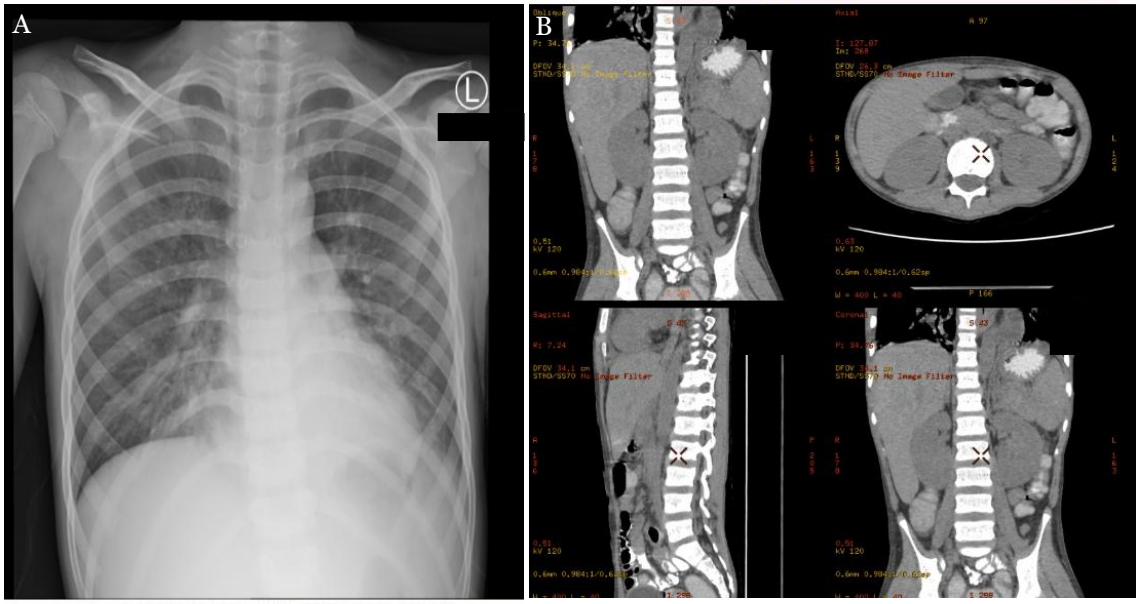

Figure 2. Imaging examination revealed cardiopulmonary and abdominal abnormalities. (A) Chest X-ray demonstrates cardiomegaly with pulmonary congestion, raising suspicion of pneumonia. (B) Non-contrast upper and lower abdominal CT scan shows grade 1 hydronephrosis with minimal pleural effusion.

Following a multidisciplinary approach involving a pediatric intensivist, cardiologist, and allergist-immunologist, SLE was diagnosed based on the 2019 European Alliance of Associations for Rheumatology (EULAR) criteria [19], yielding a total score of 30 (**Table 1**). The SLEDAI score was assessed at 16 points, with the following components: seizure (8 points), arthritis (2 points), alopecia (2 points), oral ulcers (2 points), pleural effusion (2 points), and fever (1 point), indicating high disease activity. However, immunological criterion data required for the 2019 EULAR criteria, including SLE-specific antibodies, complement levels, and antiphospholipid antibodies, were not assessed due to the unavailability of these tests at the hospital.

Table 1. Patient's scoring according to the 2019 European Alliance of Associations for Rheumatology (EULAR) criteria [19]

Entry criterion		Score	Clinical manifestation in the present patient	Score
Anti-nuclear antibody (ANA) titer ≥1:180		Mandatory	1:320	Positive
Clinical criteria				
Constitutional	Fever	2	Yes (fever, 1 month intermittently)	2
Hematologic	Leukopenia	3	No (8.57×10^3/CU mm)	0
	Thrombocytopenia	4	No (250×10^3/CU mm)	0
	Autoimmune hemolysis	4	Yes, direct comb and indirect comb test positive	4
Neuropsychiatric	Delirium	2	No	0
	Psychosis	3	No	0
	Seizures	5	Yes	5
Mucocutaneous	Non-scarring alopecia	2	Yes	2
	Oral ulcers	2	Yes	2
	Subacute cutaneous or discoid lupus	4	Yes, (rashes on hand and leg)	4
	Acute cutaneous lupus	6	No	0
Serosal	Pleural or pericardial effusion	5	Yes, bilateral pleural effusion	5
	Acute pericarditis	6	No	0
Musculoskeletal	Joint involvement	6	Yes, arthritis and arthralgia	6
Renal	Proteinuria >0.5 g/24 hour	4	Data are not available	0

Entry criterion		Score	Clinical manifestation in the present patient	Score
Anti-nuclear antibody (ANA) titer ≥1:180		Mandatory	1:320	Positive
Clinical criteria				
	Renal biopsy class II or V lupus nephritis	8	Data are not available	0
	Renal biopsy class III or IV lupus nephritis	10	Data are not available	0
Immunological criteria		Score	Patient result	Point
Antiphospholipid antibodies	Anti-cardiolipin antibodies or anti-beta2 glycoprotein antibodies or lupus anticoagulant	2	Data are not available	0
Complement	Low C3 or low C4	3	Data are not available	0
	Low C3 and low C4	4	Data are not available	0
SLE-specific antibodies	Anti-dsDNA antibody or anti-Smith antibody	6	Data are not available	0
Total score				30

SLE: systemic lupus erythematosus

While in the Pediatric Intensive Care Unit (PICU), the patient received intravenous (IV) ampicillin-sulbactam 500 mg every 8 hours for 5 days, IV paracetamol 350 mg every six hours for three days, a total of 325 cc of packed red cell (PRC) transfusion, ambroxol syrup 5 cc three times daily for five days, and IV furosemide 20 mg for one day. Post-transfusion hemoglobin levels were measured at 10.5 g/dL. Treatment for SLE was initiated once the patient's condition stabilized, and the patient was transferred to the general ward on the fifth day. The treatment regimen included intravenous methylprednisolone 750 mg in 100 mL of 0.9% NaCl for three days, followed by intravenous cyclophosphamide 750 mg in 250 mL of 0.9% NaCl. Oral methylprednisolone 28 mg twice daily was started on the ninth day. On the tenth day, oral methylprednisolone 28 mg twice daily was continued, along with oral hydroxychloroquine 100 mg daily, until discharge.

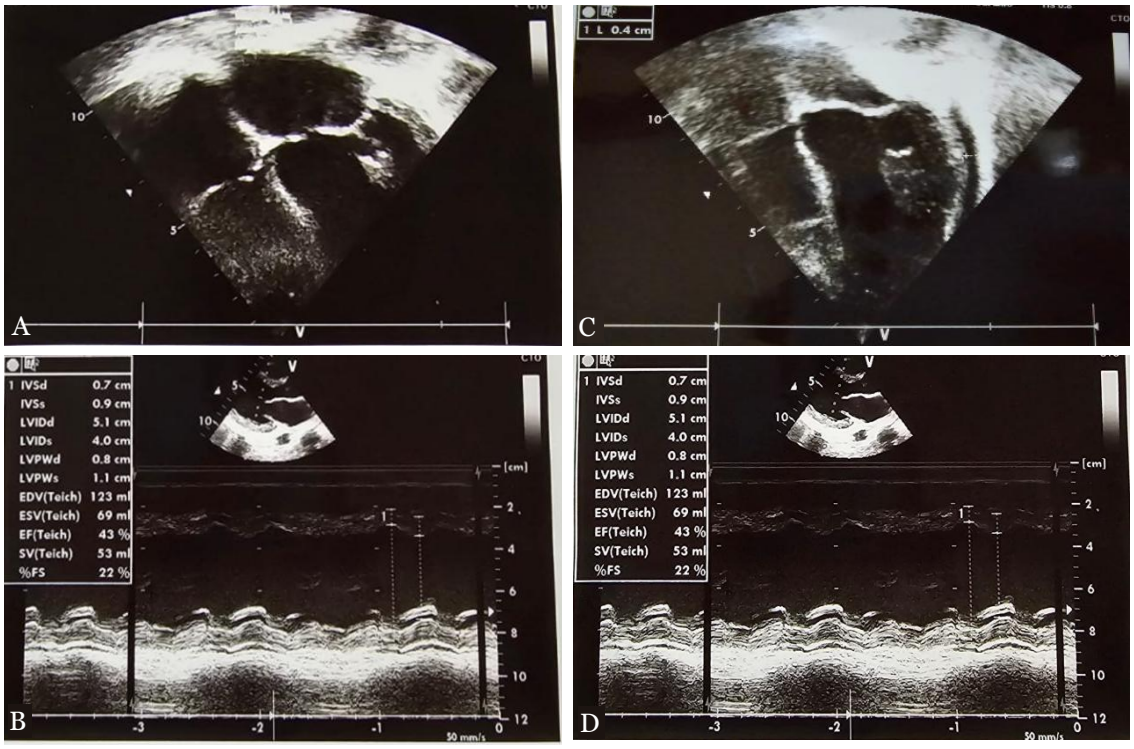

Figure 2. Echocardiography performed one month prior to treatment (A and B) demonstrated dilation of all cardiac chambers with a left ventricular ejection fraction (LVEF) of 43%. Echocardiography conducted one month after treatment (C and D) revealed an increase in LVEF to 53%, indicating improved cardiac function.

Five days after admission, the patient developed heart palpitations and tachycardia, with a heart rate reaching 140 bpm. Electrocardiography (ECG) and echocardiography were subsequently performed. The ECG demonstrated sinus tachycardia, while echocardiography revealed dilation of all cardiac chambers and a left ventricular ejection fraction (LVEF) of 43%, subsequently diagnosed as dilated cardiomyopathy. Moderate mitral regurgitation and mild pulmonary regurgitation were also noted, with no evidence of patent ductus arteriosus. Heart failure therapy was initiated, including intravenous furosemide 10 mg twice daily, oral ramipril 2.5 mg once daily, and oral digoxin 0.15 mg twice daily. Two days after drug administration, heart palpitations subsided, and tachycardia was no longer observed. After two weeks of treatment, the patient was discharged with stable vital signs and prescribed oral furosemide 10 mg twice daily, oral ramipril 1 mg once daily, oral digoxin 0.15 mg twice daily, oral methylprednisolone 28 mg twice daily, and oral hydroxychloroquine 100 mg once daily. A follow-up echocardiography one month later showed improvement in cardiomyopathy, with an LVEF of 53% (**Figure 3**).

Discussion

Diagnosing SLE in pediatric patients poses significant challenges due to its highly variable clinical manifestations and its ability to mimic other conditions, earning it the nickname "the great mimicker." In the present case, the patient initially presented with a high fever persisting for two days and a tonic-clonic seizure lasting five minutes, followed by a headache. The patient also experienced intermittent fever for one month, nausea, vomiting, productive cough, and joint and muscle pain for over two weeks. Additionally, unexplained fatigue and hair loss had been reported for two months. Notably, there was no family history of autoimmune or cardiac disorders, further complicating the diagnosis of the patient. These nonspecific and overlapping symptoms delayed the diagnostic process, highlighting the complexity of identifying SLE in pediatric populations [8].

To date, no well-validated diagnostic test or universally accepted gold standard exists for SLE diagnosis. Current clinical criteria are primarily utilized to classify cases [20]. The most recent classification system, the 2019 EULAR/ACR criteria, requires a positive ANA test as an entry criterion; once this prerequisite is met, additional weighted criteria are applied [19]. These criteria are categorized into seven clinical domains—constitutional, hematologic, neuropsychiatric, mucocutaneous, serosal, musculoskeletal, and renal—and three immunological domains, which include antiphospholipid antibodies, complement proteins, and SLE-specific antibodies [19]. Each criterion is assigned a weight ranging from 2 to 10 points, and a cumulative score of 10 or more points is required to classify a patient as having SLE, with sensitivity and specificity of these criteria reported as 96.1% and 93.4%, respectively [19]. In the present case, the patient met the mandatory ANA requirement and fulfilled the clinical domain criteria with a total score of 30 points. However, none of the immunological domain criteria were met due to the unavailability of facilities and resources for comprehensive immunological testing at the hospital.

Cardiac involvement in SLE is common and multifaceted, affecting the pericardium, myocardium, conduction system, and valves, potentially resulting in fatal outcomes [21]. Among these, dilated cardiomyopathy is rare but represents a severe manifestation of SLE. In the present case, echocardiography revealed dilation of all cardiac chambers, a reduced LVEF of 43%, moderate mitral regurgitation, and mild pulmonary regurgitation without evidence of patent ductus arteriosus. This presentation is consistent with SLE-induced dilated cardiomyopathy, emphasizing the importance of recognizing and addressing cardiac complications in SLE patients.

Myocardial disease in SLE has three primary etiologies: (1) myocarditis, the first and most common etiology which leads to cardiomyopathy specific to SLE; (2) myocardial ischemia, caused by coronary arteritis, atherosclerosis, thrombosis, or coronary embolism; and (3) myocardial dysfunction due to mitral or aortic regurgitation [22]. The precise mechanisms underlying lupus cardiomyopathy remain incompletely understood. Several hypotheses have been proposed, including immune-mediated myocardial injury [23,24]. In this mechanism, autoantibodies and immune complexes deposit in the myocardium, triggering an inflammatory response [23,24]. This inflammation can result in myocardial damage, necrosis, fibrosis, and ultimately impairing

cardiac function [23,24]. Early recognition and targeted treatment of these underlying processes are essential to prevent progression to irreversible cardiac dysfunction.

Dilated cardiomyopathy is the most common type of childhood cardiomyopathy, accounting for approximately half of all cases, characterized by left ventricular dilation and systolic dysfunction [25]. Key etiological factors in childhood include infections, toxic exposures such as chemotherapy, genetic mutations, and other conditions such as inborn metabolic errors and neuromuscular disorders [25]. In many cases, the underlying cause remains unidentified; this form, termed idiopathic dilated cardiomyopathy, has been associated with autoimmune processes that may contribute to its pathophysiology [26]. Among the identified causes of dilated cardiomyopathy, myocarditis accounts for 46%, while neuromuscular diseases, such as Duchenne muscular dystrophy, constitute approximately 25%, followed by familial cardiomyopathy [27]. In the present case, no evidence of these conditions was identified, supporting the conclusion that the dilated cardiomyopathy was attributable to SLE. This underscores the need for clinicians to consider autoimmune etiologies when other common causes are excluded, particularly in patients with systemic symptoms suggestive of SLE. Lupus myocarditis has been clinically observed in 3–9% of patients with SLE. However, post-mortem analyses reveal an incidence as high as 57%, indicating a significant prevalence of subclinical disease [28].

SLE predominantly affects females of childbearing age, with male pediatric cases being exceptionally rare. Hormonal differences between pediatric males and females may influence the severity and presentation of cardiomyopathy in SLE [29,30]. Males are hypothesized to exhibit a more robust inflammatory response due to lower levels of estrogen, a hormone with cardioprotective effects [29,30]. This disparity may result in a higher incidence of severe cardiac manifestations in male patients [29,30]. However, in the present case, the role of hormonal influence could not be determined as estrogen and testosterone levels were not evaluated. Further research is needed to elucidate the role of hormonal factors in male pediatric SLE patients.

Thorough history-taking and identification of clinical manifestations, combined with ECG, echocardiography, and supplementary tests such as angiography and biochemical assessments, are essential for the diagnosis of lupus cardiomyopathy [31]. Early symptoms of all types of cardiomyopathies often include exercise intolerance, which manifests as shortness of breath and lethargy, typically due to insufficient cardiac reserve during physical activity [32]. These symptoms are frequently overlooked or misattributed to other conditions, often of a pulmonary origin. As fluid retention progresses and resting filling pressures increase, breathlessness may occur even during routine daily activities [32]. This may present as dyspnea or cough, particularly when lying down [32].

Although peripheral edema is often considered a key sign of congestion, it may not be evident even in cases of significant fluid retention, especially in younger patients [32]. This subtle presentation sometimes makes the diagnosis of cardiomyopathy challenging. Echocardiography is the primary method for diagnosing and monitoring various heart diseases, particularly cardiomyopathy, serving as the first imaging investigation due to its favorable cost-efficiency, low risk, and ease of follow-up [31,33]. However, the gold standard for confirming the diagnosis is endomyocardial biopsy, although its invasive nature limits its use [31,33]. Delayed or missed diagnosis of SLE-induced cardiomyopathy may result in rapid progression to structural and functional cardiac abnormalities, potentially leading to heart failure and arrhythmias. These complications often developed insidiously, without prior symptoms, underscoring the importance of timely and accurate diagnosis [34].

Coronary angiography is a valuable diagnostic tool; however, it was not performed on this patient. Cardiac catheterization can aid in ruling out anomalous coronary arteries, assessing etiology and prognosis through endomyocardial biopsy, and evaluating candidacy for cardiac transplantation, including pulmonary vascular resistance measurement. However, other imaging modalities are generally preferred for diagnosing dilated cardiomyopathy. While cardiac catheterization retains a role in excluding coronary lesions, its potential benefits must be carefully weighed against procedural risks, particularly in critically ill patients with poor ventricular function, such as in the present case.

Lupus cardiomyopathy initially presenting as decompensated dilated cardiomyopathy is relatively uncommon [31]. In the present patient, the most likely cause of dilated cardiomyopathy

is autoimmune factors, supported by immunological screening results consistent with a diagnosis of SLE. A previous case reported a 7-year-old girl diagnosed with SLE who exhibited nonspecific symptoms, including prolonged fever, seizures, weight loss, hair loss, and a skin rash upon admission, similar to this patient [35]. However, unlike the present case report, the previous one included a more extensive immunological evaluation, such as complement C3 levels, anti-double-stranded DNA antibodies, antiphospholipid antibodies, and anti-SSA/Ro antibodies, which significantly facilitated the diagnostic process. Furthermore, in the previous case report, echocardiography revealed dilated cardiomyopathy with an initial ejection fraction of 35%, which improved to 71% following four months of treatment [35]. Similarly, in the present case, echocardiographic findings demonstrated improvement in LVEF from 43% to 53% after one month of treatment. These observations underscore the importance of timely intervention and comprehensive care in achieving favorable outcomes for SLE-related complications.

The present patient received furosemide 10 mg twice daily, ramipril 2.5 mg once daily, and digoxin 0.15 mg twice daily. Digoxin is generally not indicated for heart failure management in patients without atrial fibrillation; however, its use in the present case was based on specific clinical considerations. Furthermore, the decision to omit beta-blockers and mineralocorticoid receptor antagonists (MRAs) reflects an individualized treatment strategy. When no identifiable or reversible cause of dilated cardiomyopathy is detected, management remains supportive, emphasizing an anticongestive regimen, arrhythmia control, and prevention of thromboembolic events. An integral component of the anticongestive regimen for dilated cardiomyopathy includes renin–angiotensin–aldosterone system (RAAS) inhibition, diuretics, and digoxin, along with bed rest or activity restriction.

A limitation of the present case report is the unavailability of hospital resources to conduct more comprehensive immunological criterion data required for the 2019 EULAR criteria, including SLE-specific antibodies, complement levels, and antiphospholipid antibodies. Given the rarity of lupus cardiomyopathy in pediatric patients, future research should focus on the creation of multi-center, longitudinal case series that document the clinical course, laboratory findings, and therapeutic outcomes of affected individuals. This would help to identify common patterns, risk factors, and prognostic indicators. Furthermore, research exploring genetic predispositions, including polymorphisms in immune response genes, and environmental triggers (e.g., infections, medications, and stressors) that may contribute to lupus cardiomyopathy is critical.

Conclusion

Cardiomyopathy in pediatric SLE is rare but can lead to significant morbidity and mortality if not promptly diagnosed. Its presentation is often nonspecific, making early detection challenging. The pathogenesis involves immune-mediated myocardial damage and fibrosis. Due to the rarity of cases and limited pediatric studies, cardiomyopathy in SLE may be overlooked. Therefore, comprehensive cardiac evaluation, including echocardiography, is essential in children presenting with symptoms suggestive of cardiac involvement to ensure timely diagnosis and treatment.

Ethics approval

Written informed consent was obtained from the patient's parent for the publication of this case report, ensuring the confidentiality and anonymity of the patient's identity.

Acknowledgments

The authors would like to thank the staff at Murni Teguh Hospital, Medan, Indonesia, who were involved in this case report.

Competing interests

All the authors declare that there are no conflicts of interest.

Funding

This study received no external funding

Underlying data

Derived data supporting the findings of this study are available as part of the article.

Declaration of artificial intelligence use

This study used artificial intelligence (AI) tools and methodologies in the following capacities of which AI-based language models ChatGPT was employed in the language refinement (improving grammar, sentence structure, and readability of the manuscript). We confirm that all AI-assisted processes were critically reviewed by the authors to ensure the integrity and reliability of the results. The final decisions and interpretations presented in this article were solely made by the authors.

How to cite

Yudhianto E, Malisie RF, Abdillah HZ, Rusli RE. Cardiomyopathy as the forgotten symptom of systemic lupus erythematosus in children: A case report. Narra J 2025; 5 (1): e1621 - http://doi.org/10.52225/narra.v5i1.1621.

References

1. Kliegman RM, Stanton B, Behrman RE, *et al.* Nelson textbook of pediatrics. 19th edition. Philadelphia: Saunders; 2011.
2. Valenzuela-Almada MO, Hocaoglu M, Dabit JY, *et al.* Epidemiology of childhood-onset systemic lupus erythematosus: A population-based study. Arthritis Care Res 2022:74(5);728-732.
3. Nicodemus. Lupus eritematosus sistemik pada anak. J Kdokt Meditek 2024:30(1);35-44.
4. Mina R, Brunner HI. Update on differences between childhood-onset and adult-onset systemic lupus erythematosus. Arthritis Res Ther 2013;15(4):218.
5. Stokes ME, Phillips-Beyer A, Li Q. Disease features at diagnosis and changes in disease course severity among commercially insured patients with childhood-onset compared with adult-onset systemic lupus erythematosus. Lupus 2025;32(2):167-177.
6. Bundhun PK, Kumari A, Huang F. Differences in clinical features observed between childhood-onset versus adult-onset systemic lupus erythematosus: A systematic review and meta-analysis. Medicine 207;96(37):e8086.
7. Listiyono F, Murni I, Sumadiono S, *et al.* Predictors of mortality in children with systemic lupus erythematosus. Available from: https://paediatricaindonesiana.org/index.php/paediatrica-indonesiana/article/view/1967. Accessed on: 12 January 2025.
8. Levy DM, Kamphuis S. Systemic lupus erythematosus in children and adolescents. Pediatr Clin North Am 2012;59(2):345-364.
9. Doria A, Laccarino L, Sarzi-Puttini P, *et al.* Cardiac involvement in systemic lupus erythematosus. Lupus 2005;14(9):683-686.
10. Zhang H, Zhang L, Guo N. Pediatric-onset systemic lupus erythematosus with coronary artery dilation: A case report. Medicine 2020;99(5):e18946.
11. Guzmán-Martínez G, Marañón C. Immune mechanisms associated with cardiovascular disease in systemic lupus erythematosus: A path to potential biomarkers. Front Immunol 2022;13:974826.
12. Moder KG, Miller TD, Tazelaar HD. Cardiac involvement in systemic lupus erythematosus. Mayo Clin Proc 1999;74(3):275-284.
13. Yafasova A, Fosbøl EL, Schou M, *et al.* Long-term cardiovascular outcomes in systemic lupus erythematosus. J Am Coll Cardiol 2021;77(14):1717-1727.
14. Towbin JA, Lowe AM, Colan SD, *et al.* Incidence, causes, and outcomes of dilated cardiomyopathy in children. JAMA. 2006;296(15):1867-1876.
15. Moder KG, Miller TD, Tazelaar HD. Cardiac involvement in systemic lupus erythematosus. Mayo Clin Proc 1999;74(3):275-284.
16. Koenig PR, Abdulla Ri. Pediatric cardiology: A journey to the past. In: Anderson RH, Backer CL, Berger S, *et al.* Editors. Pediatric cardiology. Cham: Springer; 2024.

17. Hollander SA, Bernstein D, Yeh J, *et al*. Outcomes of children following a first hospitalization for dilated cardiomyopathy. Circ Heart Fail 2012;5:437-443.
18. Jammal AMB, Young D, McCarrison S, *et al*. Dilated cardiomyopathy in a national paediatric population. Eur J Pediatr 2019;178:1229-1235.
19. Aringer M, Costenbader K, Daikh D, *et al*. 2019 European league against rheumatism/American College of Rheumatology classification criteria for systemic lupus erythematosus. Arthritis Rheumatol 2019;71(9):1400-1412.
20. Groot N, Graeff N, Avcin T, *et al*. European evidence-based recommendations for diagnosis and treatment of childhood-onset systemic lupus erythematosus: the SHARE initiative. Ann Rheum Dis 2017;76:1788-1796.
21. Raval JJ, Ruiz CR, Heywood J, *et al*. SLE strikes the heart! A rare presentation of SLE myocarditis presenting as cardiogenic shock. BMC Cardiovasc Disord 2021;21:294.
22. Roldan CA, Crawford MH. Connective tissue diseases and the heart. In: Crawford MH, editor. Current diagnosis & treatment in cardiology. Rio de Janeiro: Prentice Hall do Brasil; 1995.
23. Zinoune L, Maazouzi M, Benbouchta K, *et al*. Dilated cardiomyopathy complicated by an intracavitary thrombus and acute heart failure: A rare presentation of systemic lupus erythematosus. Ann Med Surg 2022;82:104739.
24. Caforio ALP, Pankuweit S, Arbustini E, *et al*. 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. Eur Heart J 2013;34(33):2636-2648.
25. Rath A, Weintraub R. Overview of cardiomyopathies in childhood. Front Pediatr 2021;9:708732.
26. Lappé JM, Pelfrey CM, Tang WH. Recent insights into the role of autoimmunity in idiopathic dilated cardiomyopathy. J Card Fail 2008;14(6):521-530.
27. Park MK, Salamat M. Park's pediatric cardiology for practitioners. 7th edition. Philadelphia: Elsevier; 2020.
28. Mohanty B, Sunder A. Lupus myocarditis-A rare case. J Family Med Prim Care 2020;9(8):4441-4443.
29. Moder KG, Miller TD, Tazelaar HD. Cardiac involvement in systemic lupus erythematosus. Mayo Clin Proc 1999;74(3):275-284.
30. Corker A, Learmonth M, Patrick DM, *et al*. Cardiac and vascular complications in lupus: Is there a role for sex? Front Immunol 2023;14:1098383.
31. Loscalzo J, Hutter AM. Harrison's cardiovascular medicine. 2nd ed. New York: McGraw-Hill Education; 2013.
32. Trasca L, Popescu MR, Popescu AC, *et al*. Echocardiography in the diagnosis of cardiomyopathies: Current status and future directions. Rev Cardiovasc Med 2022;23(8):280.
33. Zinoune L, Maazouzi M, Benbouchta K, *et al*. Dilated cardiomyopathy complicated by an intracavitary thrombus and acute heart failure: A rare presentation of systemic lupus erythematosus. Ann Med Surg 2022;82:104739.
34. Okorie IJ, Appiah-Kubi E, Owusu-Antwi P, *et al*. Lupus-induced accelerated heart failure in a young African American female: Cardiovascular and systemic complications of noncompliance to maintenance therapy and the social determinants of cardiovascular disease. Cureus 2024;16(1):e51819.
35. Tiaaza F, Amine MB, Asmaa S, *et al*. Dilated cardiomyopathy in systemic lupus erythematosus: A pediatric case report. Available from: https://www.researchsquare.com/article/rs-339594/v1. Accessed: 10 January 2025.