Comprehensive Case Analysis: Diagnosing and Managing Myositis in Newly Diagnosed Systemic Lupus Erythematosus Patients in Indonesia

*Nabila Abiyasa Putri[1,2], Awalia[3]**

[1]Internal Medicine Specialist Study Program, Department of Internal Medicine, Faculty of Medicine, Universitas Airlangga, Surabaya, Indonesia.
[2]Internal Medicine Specialist Study Program, Department of Internal Medicine, Dr. Soetomo General Academic Hospital, Surabaya, Indonesia.
[3]Division of Rheumatology, Department of Internal Medicine, Dr. Soetomo General Academic Hospital, Surabaya, Indonesia.

Corresponding Author:
Awalia, MD. Rheumatology Division, Department of Internal Medicine, Faculty of Medicine of Universitas Airlangga - Dr. Soetomo General Hospital. Jl. Mayjend. Prof. Dr. Moestopo No. 6-8, Gubeng, Surabaya 60286, Indonesia. Email: awalia_nov74@yahoo.com

ABSTRACT

An 18-year-old female with systemic lupus erythematosus (SLE) presented with bilateral thigh pain, fever, and diarrhea three days before admission. Diagnosed with SLE one month earlier, she reported prior symptoms, including joint pain, malar rash, hair loss, and hyperpigmented lesions. Initial investigations revealed elevated transaminase levels (AST 355, ALT 132), positive ANA, decreased complement levels (C3 68, C4 16.8), and raised creatine kinase (619). A muscle biopsy confirmed myositis. The patient was treated with immunosuppressant (a combination of steroids and hydroxychloroquine) and supportive therapy. By the eighth day of hospitalization, her symptoms, especially thigh pain, resolved. Electromyocardiography was done, and the results were normal, indicating therapeutic success. This case highlights the importance of prompt diagnosis and management of myositis as a rare SLE manifestation to achieve favorable outcomes.

***Keywords**: Systemic Lupus Erythematosus (SLE), Myositis, Muscle Biopsy, Electromyocardiography.*

INTRODUCTION

SLE is a chronic systemic autoimmune disease that affects various organ including the skin, lungs, kidneys, joints, blood, and the peripheral and central nervous systems.[1] It has an estimated national incidence of 5.1 per 100,000 person-years, with significantly higher rates among women (8.7 compared to 1.2 in men) and the highest incidence observed in Black women (15.9), followed by Asian/Pacific Islander (7.6), Hispanic (6.8), and White (5.7) women.[2]

Skeletal muscle involvement in SLE, typically manifesting as myositis, is reported in 4%–16% of patients. Myositis prevalence studies across Asia, Europe, and North America estimate an incidence of 11 to 660 new cases per 1,000,000 person-years, with 2.9 to 34 cases per 100,000 population. While regional and ethnic differences in incidence remain unclear, a north-to-south gradient in the prevalence of myositis subtypes, such as dermatomyositis, has been suggested in Europe and North America.[3] SLE myositis is strictly defined as elevated creatine kinase together with evidence of muscle edema on MRI, myopathic electromyography, and/or myopathic muscle biopsy. Muscle pathology findings include perifascicular atrophy, venulitis, perimyseal inflammation, neurogenic muscle injury, and muscle fiber necrosis.[4] The overlap

of symptoms with other conditions, variability in clinical presentations, and limited research on muscle biopsy findings make the diagnosis of inflammatory myositis in SLE particularly challenging.

CASE ILLUSTRATION

Ms. D, an 18-year-old female, presented to the ER with a chief complaint of bilateral thigh pain three days before admission, accompanied by diarrhea and fever. Pain was felt throughout the day, most notably in the morning. She had been diagnosed with systemic lupus erythematosus (SLE) one month prior and was undergoing therapy. Her initial symptoms included joint pain, malar rash, hair loss, and hyperpigmented lesions.

On physical examination, notable findings

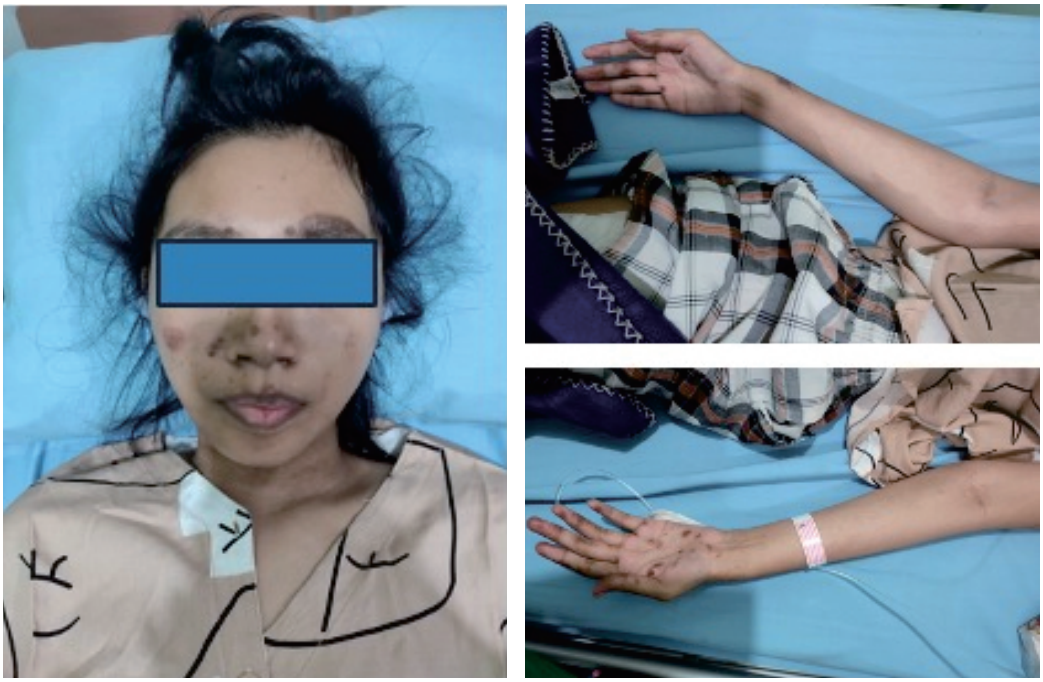

Figure 1. The patient was hospitalized and underwent further diagnostic evaluations, including creatine kinase measurement and a muscle biopsy (**Figure 2**). The creatine kinase level was elevated at 619 U/L (normal <167 U/L). Muscle biopsy findings revealed striated muscle tissue with round-to-spindle nuclei, fine chromatin, and extensive cytoplasm. Additional areas showed mature adipose tissue, fibrous connective tissue, and inflammatory lymphocytes and histiocytes, with no evidence of malignancy.

Figure 2. The patient received comprehensive treatment during her hospitalization, including: 1500 ml of intravenous fluids every 24 hours; Metoclopramide 10 mg intravenously every 8 hours; Omeprazole 40 mg intravenously every 12 hours; Cefoperazone-sulbactam 1 g intravenously every 12 hours; Hydrocortisone 100 mg intravenously every 8 hours; Paracetamol 500 mg orally every 8 hours; Hydroxychloroquine 200 mg orally once daily; N-acetylcysteine 200 mg orally every 8 hours and Attapulgite (2 tablets everytime diarrhea occured, up to 10 tablets daily)

included tachycardia, discoid lupus lesions on the face, hands, and feet, as well as tenderness in both thighs, predominantly on the right (**Figure 1**). Initial laboratory investigations revealed leukocyte levels within normal limits (7380) but with neutrophil predominance (81.5%), elevated transaminases (SGOT 355, SGPT 132), increased procalcitonin (31.43), a positive ANA test, and decreased complement levels (C3: 68, C4: 16.8). Based on these findings, the patient was provisionally diagnosed with suspiscion of lupus myositis, sepsis and acute gastroenteritis with moderate dehydration.

After eight days of treatment, the patient showed significant improvement, particularly in thigh pain. An electromyography conducted on the final day of hospitalization yielded normal results, indicating favorable therapeutic outcomes. The patient was discharged in stable condition with ongoing follow-up planned to monitor her progress.

DISCUSSION

The diagnosis of SLE has evolved significantly over the past decades, transitioning from the 1982 American College of Rheumatology (ACR) criteria, which were revised in 1997, to more comprehensive diagnostic frameworks. In 2012, the Systemic Lupus International Collaborating Clinics (SLICC) introduced updated criteria that incorporated lupus nephritis and required a total of four or more positive criteria, with at least one clinical and one immunological criterion. More recently, the European League Against Rheumatism (EULAR) developed the 2018 criteria, often used in research, which include a prerequisite antinuclear antibody (ANA) titer of 1:80 or higher and emphasize severe lupus manifestations, requiring a total score of at least 10.[5] In this case, the patient presented with characteristic clinical and laboratory findings aligning with the SLICC criteria for SLE. Clinically, she reported joint pain, malar rash, hair loss, and hyperpigmented lesions, while laboratory findings included a positive ANA test and reduced complement levels. These findings fulfill both the clinical and immunological requirements of the SLICC criteria, confirming the diagnosis of SLE.

Myopathy, derived from the Greek words "myo" (muscle) and "pati" (disease), refers to a spectrum of muscle diseases typically characterized by symptoms such as weakness, stiffness, cramps, and spasms. Idiopathic inflammatory myopathies (IIM) represent a subgroup of myopathies defined by inflammatory infiltration of skeletal muscle, and occasionally cardiac muscle, resulting in muscle weakness, pain, and associated extramuscular manifestations.[6,7] IIM can be classified into several subtypes, including dermatomyositis (DM, with juvenile and adult-onset forms), polymyositis (PM), inclusion body myositis (IBM), and amyopathic or clinically amyopathic dermatomyositis (ADM or CADM). While all subtypes share common symptoms of muscle weakness, they vary clinically based on the specific muscle groups involved and their histopathological characteristics.[6,7] In this case, the patient presented with bilateral thigh pain persisting for three days, with the pain being most intense in the morning and persisting throughout the day. This presentation aligns with typical symptoms of inflammatory myopathies. The patient's history of joint pain further supports the likelihood of an underlying systemic inflammatory condition, consistent with her prior diagnosis of SLE.

Elevated serum creatine kinase (CK) levels are a sign of significant muscle involvement, as CK is a highly sensitive marker of muscle damage, particularly in the acute phase of disease. Muscle injury leads to CK release into the bloodstream, making it a key indicator in diagnosing myositis. Other enzymes, such as aldolase, myoglobin, lactate dehydrogenase (LDH), and transaminases (AST and ALT), may also be elevated in cases of muscle damage. Additionally, inflammatory biomarkers, including erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), tend to rise during the active phases of systemic inflammatory diseases.[8] In this patient, initial laboratory results demonstrated significantly elevated transaminase levels (AST 355 and ALT 132), raising suspicion of muscle damage. Further testing confirmed raised CK levels (619 U/L), consistent with muscle injury and supporting the diagnosis of lupus myositis.

Muscle biopsy is a crucial diagnostic tool for confirming myositis, providing valuable histopathological insights into muscle involvement. Typical findings in myositis cases include myofiber necrosis, perifascicular atrophy, perivascular inflammation, lymphocytic invasion of non-necrotic muscle fibers (indicative of primary inflammation), and endomysial or perimysial inflammation. To enhance the clarity of these findings, various staining techniques can be employed, including H&E staining, modified Gomori trichrome, myosin adenosine triphosphatase (pH 4.3, 4.6, and 9.4), NADH-tetrazolium reductase, acid phosphatase, succinate dehydrogenase (SDH), cytochrome oxidase, alkaline phosphatase, PAS, PAS-diastase control, and Congo red staining.[1]

In this case, the patient underwent a muscle biopsy to confirm the diagnosis of myositis. Histopathological examination revealed striated muscle tissue characterized by round-to-spindle nuclei, fine chromatin, and extensive cytoplasm. In addition, mature adipose tissue and fibrous connective tissue were observed, with significant infiltration of inflammatory cells, including lymphocytes and histiocytes. Importantly, no evidence of malignancy was found, supporting the diagnosis of lupus myositis. These findings align with previously described muscle pathology in myositis, further substantiating the clinical suspicion.

The primary goal of therapy in systemic lupus erythematosus (SLE) is to manage disease flares, alleviate symptoms, and prevent recurrence.[9] According to the 2019 EULAR Guidelines, all lupus patients should receive hydroxychloroquine at a dose not exceeding 5 mg/kg of actual body weight. Chronic glucocorticoid use should be minimized to less than 7.5 mg/day (prednisone equivalent), with discontinuation as the ultimate goal when feasible. Steroids remain the first-line therapy for SLE-associated myositis.[10]

In this case, the patient initially received oral methylprednisolone (8 mg daily), which was transitioned to intravenous hydrocortisone (100 mg every 8 hours) during hospitalization. Additionally, hydroxychloroquine (200 mg daily) was administered throughout the patient's treatment. This approach yielded satisfactory results, particularly in resolving the bilateral thigh pain.

Comparable findings have been reported in the literature. Gading et al. (2019) observed the successful use of prednisone (2 mg/kg/day for two weeks) in SLE patients with myositis, combined with ibuprofen (10 mg/kg per dose) for pain relief and azathioprine for long-term management.[11] Similarly, Paramasivam et al. (2022) reported the use of hydrocortisone, just like our case, although the therapeutic response was poor, necessitating a switch to high-dose methylprednisolone.[12] In contrast, the patient in this case achieved significant improvement with hydrocortisone therapy, demonstrating variability in individual responses to treatment and underscoring the importance of tailored therapeutic strategies in managing SLE-associated myositis.

The needle electromyography (EMG) examination is a crucial component of electrodiagnostic (EDX) studies for assessing myopathic disorders. It provides valuable information by confirming active myopathy, refining differential diagnoses, and identifying suitable sites for muscle biopsy. The selection of muscles for examination depends on the patient's pattern of weakness, with analysis focusing on motor unit action potential (MUAP) morphology and recruitment patterns to aid in diagnosing myopathic processes. These disorders typically involve a decrease or dysfunction in individual muscle fibers.[13]

In this case, the patient's condition improved significantly after eight days of treatment, particularly in alleviating thigh pain. The needle EMG performed on the final day of hospitalization revealed normal results, suggesting effective therapeutic outcomes. The patient was discharged in stable condition, with follow-up care planned to ensure continued recovery and monitor for any recurrence.

CONCLUSION

The patient was treated with immunosuppressant (a combination of steroids and hydroxychloroquine) and supportive therapy. By the eighth day of hospitalization,

her symptoms, especially thigh pain, resolved. Electromyocardiography was done, and the results were normal, indicating therapeutic success.

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CONFLICT OF INTERESTS

All of the authors declare that there is no conflict of interest.

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