RESEARCH PAPER

Classic Dermatomyositis with Rapidly Progressive Muscle Weakness and Extensive Cutaneous Involvement: A Case Report

Running title: *Rapidly Progressive Dermatomyositis: A Case Report*

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ABSTRACT

Dermatomyositis is a systemic autoimmune disorder characterized by distinctive cutaneous manifestations and variable muscle involvement. We report the case of 50-years old female who developed progressive reddish-violaceous patches over the neck, chest, back, and arms, followed by involvement of the eyelids, elbows, and dorsal fingers, as well as spontaneous ulcerations on the right elbow and upper arm. Over the subsequent months, she experienced progressive muscle weakness on both extremities that caused her unable to lift her limbs or ambulate independently. Physical examination revealed heliotrope rash, shawl and V-neck signs, and Gottron's papules. Imaging studies suggested suspected interstitial lung disease, while abdominal ultrasonography identified hepatomegaly and multiple uterine fibroids. Histopathological evaluation showed an atrophic epidermis with vacuolar interface changes and perivascular lymphocytic infiltrates, features compatible with dermatomyositis or acute cutaneous lupus erythematosus. The patient received systemic and topical corticosteroids, topical antibiotics, supportive medical therapy, and strict photoprotection. This case highlights the complex presentation of dermatomyositis with rapidly progressive muscle weakness and multisystem involvement, emphasizing the importance of early recognition, thorough evaluation, and multidisciplinary management to prevent long-term morbidity.

Keywords: Dermatomyositis; Heliotrope rash; Gottron's papules; Muscle weakness; Cutaneous ulceration; Interstitial lung disease; Histopathology.

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Introduction

Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by a combination of pathognomonic cutaneous manifestations and varying degrees of muscle weakness.[1] Classic dermatomyositis includes three major criteria such as heliotrope sign, Gottron papules and Gottron sign and 14 additional minor criteria that facilitate clinical recognition, although some patients may initially present with subtle or atypical skin lesions. Muscle involvement may also vary widely, and up to 20% of patients exhibit clinically amyopathic disease, contributing to diagnostic challenges in early stages.[2,3]

Worldwide, dermatomyositis is considered a rare disease, with an estimated annual incidence ranging from 1–10 cases per million population and a prevalence of 5–22 cases per 100,000 persons. It typically affecting adults aged 40–60 years and children aged 5–15 years. Women are disproportionately affected, with a female-to-male ratio of approximately 2:1.[4,5] Specific data on the prevalence and incidence in Indonesia is not detailed, but a study from RSUPN dr. Cipto Mangunkusumo in Jakarta reported six cases of dermatomyositis over a five-year period from 2017 to 2022.[6] Case reports from dr. Soetomo General Hospital in Surabaya had also reporting a case of classic dermatomyositis in adult and juvenile dermatomyositis.[7,8]

Dermatomyositis is frequently associated with systemic involvement, particularly muscle and lung.[9] Other systemic manifestations may include

gastrointestinal, hepatic, and cardiac abnormalities, further broadening the disease spectrum.[10] Histopathological examination plays a supportive role in diagnosis, typically demonstrating interface dermatitis, vacuolar alteration of basal keratinocytes, and perivascular lymphocytic infiltrates, although overlap with acute cutaneous lupus erythematosus may reduce diagnostic specificity.[6,7] Given this variability, integrating clinical features, laboratory findings, imaging results, and histopathology is essential for accurate diagnosis.

Early recognition and prompt treatment of dermatomyositis are critical to preventing irreversible muscle damage, functional decline, and systemic complications. This case highlights the importance of a comprehensive approach in evaluating patients with progressive muscle weakness and characteristic skin lesions, particularly when multisystem involvement raises concern for more severe disease

Case Report

A fifty-year-old female presented with a four-month history of progressively worsening reddish-violaceous patches on the neck, chest, upper back, and arms. Approximately one month after onset, the lesions expanded to the eyelids, elbows, and dorsal aspects of the fingers. The patient also developed spontaneous ulcerations on the right elbow and upper arm without any preceding trauma. She reported increased warmth and burning sensations on the face and neck following sun exposure. Concurrently, she experienced pain and progressive weakness affecting both upper and lower extremities over a three-month period, eventually becoming unable to lift her arms or ambulate independently.

The patient had been diagnosed with spondyloarthritis three months earlier and was under routine follow-up at the rheumatology clinic. She had no history of diabetes mellitus, hypertension, allergies, or similar complaints in family members.

On examination, vital signs were stable. Dermatologic evaluation revealed violaceous erythematous patches on the neck, chest, and back; heliotrope rash involving the upper eyelids; characteristic Gottron's papules and plaques over the dorsal finger joints and elbows; and ulcerations with crusting on the right elbow and upper arm. A shawl sign and V-neck sign were clearly observed (**Figure 1**). Musculoskeletal assessment demonstrated marked reduction in proximal muscle strength in both upper and lower extremities, with the patient unable to lift the limbs against gravity.

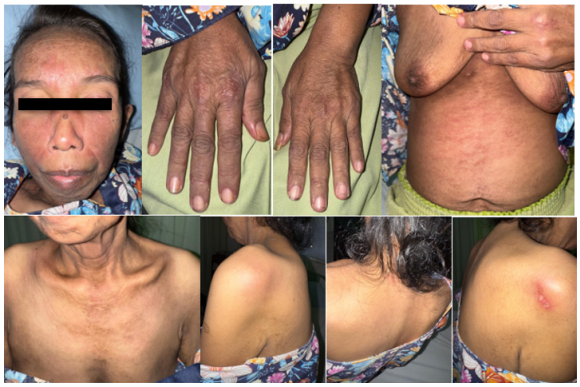

Figure 1. Dermatologic status of patient on day 1

Laboratory evaluation demonstrated normocytic anemia (hemoglobin level 9.9 g/dL) and markedly elevated liver enzymes (AST/ALT), which supported the presence of systemic inflammation and possible hepatic involvement often seen in dermatomyositis. The patient also showed a critically elevated LDH (577 U/L) and markedly increased ferritin (1519 ng/mL), both reflecting significant tissue injury and active inflammatory myopathy. These findings aligned with her rapidly progressive muscle weakness and suggested a high inflammatory burden.

The ANA test was negative, which does not exclude dermatomyositis, as up to half of patients may remain ANA-negative. Viral hepatitis and HIV serologies were non-reactive, ruling out infectious etiologies of elevated transaminases and muscle injury, and supporting autoimmune inflammatory myopathy as the primary process. Mild hyperbilirubinemia also reflected hepatic involvement likely secondary to systemic inflammation rather than viral disease.

Chest radiography showed bilateral parahilar infiltrates, an air-containing thin-walled lesion consistent with bullae in the right suprahilar region, and evidence of left pleural effusion. High-resolution computed tomography performed earlier confirmed the presence of pneumonia with suspected interstitial lung disease, which commonly found in dermatomyositis. Abdominal ultrasonography revealed hepatomegaly with increased parenchymal echogenicity, mild splenomegaly, parenchymal liver disease, and multiple uterine fibroids.

Electromyography (EMG) performed on July 30th, 2025 demonstrated right C8 nerve root irritation, while needle EMG findings were consistent with a myopathic process, a pattern commonly seen in inflammatory muscle disease. These results further supported the presence of an underlying myositis.

A skin biopsy was obtained from an erythematous lesion. Histopathology demonstrated

epidermal atrophy, vacuolar degeneration of basal keratinocytes, follicular plugging, and perivascular and periappendageal lymphocytic infiltrates (**Figure 2**). These microscopic findings were consistent with dermatomyositis but showed overlap with acute cutaneous lupus erythematosus, making definitive distinction challenging.

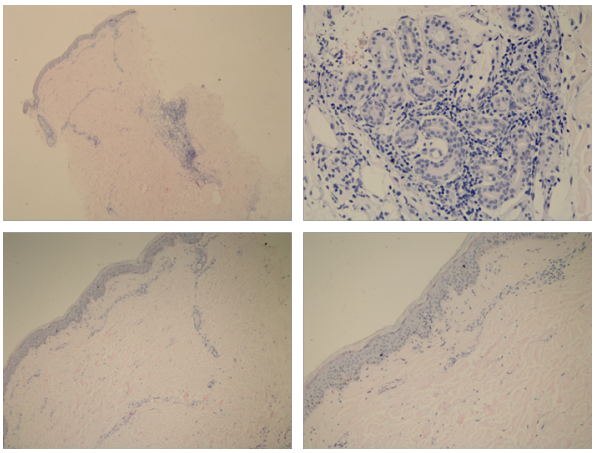

Figure 2 Histopathological appearance from the skin biopsy

Based on the findings of classic cutaneous signs, rapidly progressive proximal muscle weakness, elevated muscle and inflammatory markers, and supportive histopathology, we assess this patient with dermatomyositis, with differential considerations including mixed connective tissue disease (MCTD), accompanied by suspected interstitial lung disease (ILD) and hepatomegaly. The initial dermatologic therapy included topical mometasone cream for facial lesions, desoxymethasone for body lesions, and natrium fusidate for areas of erosion. Strict photoprotection was advised. The internal medicine team initiated systemic corticosteroid therapy (oral methylprednisolone) as first-line management for inflammatory myopathy. N-acetylcysteine (NAC) was administered as an antioxidant adjunct to reduce oxidative stress associated with muscle inflammation. Gastroprotective therapy (omeprazole) and calcium supplementation were provided to mitigate corticosteroid-related adverse effects. Supportive measures included nutritional optimization to address catabolic burden and supplemental oxygen for suspected interstitial lung involvement.

Discussion

Dermatomyositis is an idiopathic inflammatory myopathy (IIM) characterized by distinctive cutaneous manifestations, variable muscle involvement, and damaged internal organs. The present case demonstrated a classic yet unusually aggressive form of dermatomyositis, with extensive cutaneous involvement including heliotrope rash, shawl and V-neck signs, and Gottron's papules combined with rapidly progressive proximal muscle weakness and early multisystem abnormalities, which strongly supported the diagnosis of classic dermatomyositis. These hallmark cutaneous lesions result from immune-mediated microangiopathy affecting skin and muscle, which is considered a central mechanism in dermatomyositis pathogenesis.[1,2] Combination of the findings put the the patient at higher risk for severe disease progression, systemic complications, and poorer outcomes. Understanding the interplay between the clinical presentation, underlying pathomechanisms, laboratory abnormalities, and organ involvement is essential for appropriate diagnostic evaluation and timely therapeutic intervention.

The exact pathophysiological mechanisms causing DM remain unknown. In general, similar to most other auto-immune disorders, an environmental trigger is presumed to be required in combination with a genetic predisposition. DM is multifactorial, complex, and it involve dysregulation of both innate and adaptive immunity activation.[1,11]

The strongest genetic risk for disease susceptibility is presumed to localize to specific leucocyte antigen (HLA) alleles. High-risk haplotypes include HLA-A*68 in North American Whites, HLA-DRB1*0301 in African Americans, HLA-DQA1*0104 and HLA-DRB1*07 in Han Chinese, DQA1*05 and DQB1*02 in people from the UK. Also, DRB1*03-DQA1*05-DQB1*02 haplotype is strongly associated with the development of interstitial lung disease in dermatomyositis.[10,11]

From the immune-mediated mechanism, it complies both innate and adaptive immune activation. Muscle and skin biopsies show infiltration of CD3+ T cells, plasmacytoid dendritic cells, and macrophages as well as B cells (especially in muscle).[1] The muscle microenvironment overexpress innate immune receptors, including Toll-like receptors, which may lead to nuclear factor kappa B (NF-B) and pro-inflammatory cytokine and chemokine signaling. This leads to interferon (IFN)-1 overexpression in dermatomyositis.[1,11]

High levels of IFN-induced genes and proteins found in blood, muscle, and skin and have been shown to correlate with disease activity. In both skin and muscle, the cells expressing these IFN-induced gene products appear most concentrated in the

areas of tissue damage and may play a role in recruiting and activating cytotoxic effector cells, which may cause a pro-inflammatory state, MHC-class I overexpression, and non-immune mediated toxicity via mitochondrial damage and endoplasmic reticulum stress. [1,11]

Activation of the type I interferon (IFN-1) pathway is now recognized as a hallmark of dermatomyositis. IFN-α and IFN-β stimulate the JAK–STAT pathway, leading to overexpression of interferon-stimulated genes (ISGs), which is strongly expressed in perifascicular regions and contributes to microangiopathy. Microangiopathy in DM is also one of the hallmark features, which characterized by endothelial damage, capillary dropout, and perifascicular ischemia. Deposition of the complement membrane attack complex (C5b–9) within small vessels is considered one of the earliest events in dermatomyositis and correlates with muscle fiber necrosis. This microvascular injury contributes to perifascicular atrophy, a classic histologic feature of DM.[1,11]

Plasmacytoid dendritic cells, potent producers of IFN-1, are consistently found in DM muscle biopsies, supporting local IFN-driven inflammation. Another key IFN-inducible protein, ISG15, is markedly increased and may amplify tissue injury through conjugation with muscle proteins. Transcriptomic studies confirm that DM shows the strongest IFN-1 gene signature among inflammatory myopathies, correlating with disease activity and cutaneous severity. These findings reinforce that IFN-1 overexpression plays a central role in the immune dysregulation and tissue damage seen in DM.[11]

Multiple environmental factors may trigger chronic immune activation in genetically susceptible individuals. Proposed triggers for DM include ultraviolet (UV) radiation, viral infections, medications, and smoking. UV exposure has been linked with DM and anti-Mi2 antibodies in adult women and with juvenile dermatomyositis (JDM) and anti-TIF1 antibodies in children. Viral infections may play a role in triggering immune activation or disrupting immune tolerance, but attempts to isolate viruses from DM muscle samples have been unsuccessful. Earlier study found that DM/JDM flares were associated with UV exposure, infections, and some medications, although only sun exposure (OR 2.2) and recent NSAID use (OR 1.9) remained significant predictors in multivariable analysis. Smoking has been associated with DM and the development of ILD, dysphagia, malignancy, and cardiac involvement. Other potential environmental triggers are less well established.[12,13] The strong photosensitive distribution of cutaneous lesions in this patient may be explained by ultraviolet (UV)–induced enhancement of IFN pathways, which amplifies keratinocyte injury and triggers immune activation.

Cutaneous findings are central to the diagnosis of dermatomyositis, and Sontheimer's criteria emphasize the importance of pathognomonic and characteristic skin lesions in establishing classic dermatomyositis (CDM). According to these criteria, diagnosis can be established by the presence of two major criteria or one major and two minor criteria followed by consistent skin biopsy result.[1,14] In this patient, multiple Sontheimer-defined pathognomonic features were present, including a prominent heliotrope rash, well-formed Gottron papules on the dorsal fingers, and violaceous erythema on the elbows and extensor surfaces. Additional characteristic lesions such as malar eminence on the eyelids, V-neck sign, shawl sign, and violaceous patches on the elbow and upper arm further supported the diagnosis of classic dermatomyositis. The presence of spontaneous ulcerations on the elbows and upper arm, although not part of Sontheimer's original criteria, is recognized in more severe or vasculopathic variants of dermatomyositis and may indicate a higher-risk disease phenotype commonly associated with systemic involvement such as interstitial lung disease.[15]

Proximal muscle weakness is a defining feature of DM, affecting activities such as lifting the arms, climbing stairs, or rising from a seated position. This patient developed progressive symmetrical muscle weakness over a three-month period and ultimately became unable to lift her limbs, indicating severe and rapidly evolving myopathy. Laboratory results showed markedly elevated LDH, AST and ALT levels, which reflect significant muscle fiber injury and systemic inflammation. These markers are often highly sensitive for myositis and may remain elevated even when creatine kinase (CK) is normal or unavailable, making them valuable supportive marker.[1,16]

The patient's EMG findings showing both C8 root irritation and needle EMG features suggestive of myopathy, which offer additional support for inflammatory muscle involvement. Although radiculopathy can coexist, the presence of myopathic motor unit potentials aligns with the clinical presentation of rapidly progressive proximal muscle weakness. EMG abnormalities in dermatomyositis frequently correlate with active muscle inflammation, and in settings where CK levels are unavailable, EMG

can serve as a useful adjunct in confirming muscle disease and guiding treatment decisions.[10]

Skin biopsy remains a valuable diagnostic tool in dermatomyositis. The patient's biopsy revealed epidermal atrophy, vacuolar degeneration of basal keratinocytes, follicular plugging, and perivascular lymphocytic infiltration. These findings are compatible with DM but are not fully specific, as interface dermatitis with similar features can be seen in acute cutaneous lupus erythematosus.[17] The diagnostic overlap highlights the importance of correlating histopathology with clinical and laboratory data.

The absence of ANA positivity does not exclude DM; 40–50% of patients may have negative ANA, particularly those with MSA-driven phenotypes.[17] Due to the unavailability of MSA testing, diagnosis in this case relied on the presence of cardinal cutaneous findings, muscle weakness, laboratory abnormalities, and supportive histopathology.

Management of dermatomyositis generally follows a stepwise treatment ladder that addresses both cutaneous and muscular involvement. First-line therapy for skin disease includes strict photoprotection and topical corticosteroids, consistent with the initial approach in this patient who presented with extensive photo-distributed lesions and ulcerations. For muscle involvement, systemic corticosteroids remain the standard first-line treatment, and were appropriately initiated due to her rapidly progressive proximal muscle weakness. In patients with severe or multisystem disease—such as suspected interstitial lung disease—early consideration of second-line immunosuppressive agents (e.g., methotrexate, mycophenolate mofetil, azathioprine, or IVIG) is warranted to achieve better disease control and reduce steroid exposure. Third-line therapies such as rituximab, cyclophosphamide, or JAK inhibitors may be reserved for refractory or rapidly progressive cases. Tailoring therapy based on disease severity and organ involvement, supported by multidisciplinary management, is essential for optimizing outcomes.[1]

Taken together, the clinical, laboratory, and histopathological elements discussed above illustrate the multifactorial nature of dermatomyositis and the need to interpret findings within a unified immunopathogenic framework.

Conclusion

This case underscores the clinical importance of recognizing classic dermatomyositis even when it presents with an unusually rapid and aggressive course. Unlike many reported cases that highlight either predominant cutaneous or muscular involvement, this patient presented with a combination of extensive photo-distributed lesions, spontaneous ulcerations, and swiftly progressive proximal muscle weakness, accompanied by multisystem abnormalities, including suspected interstitial lung disease, hepatic dysfunction, and markedly elevated inflammatory biomarkers. The coexistence of severe cutaneous features with early systemic deterioration is relatively uncommon and raises concern for a potentially high-risk phenotype. The diagnostic overlap on histopathology with acute cutaneous lupus erythematosus further highlights the challenges clinicians may encounter, reinforcing the need for integrated interpretation of clinical, laboratory, imaging, and biopsy findings.

This report emphasizes the variability and potential severity of dermatomyositis. It also highlights the importance of timely recognition and multidisciplinary management to prevent irreversible morbidity. This case contributes meaningful insight into the spectrum of dermatomyositis presentations, particularly those that deviate from the more indolent patterns commonly described in the literature.

Declaration of patient consent Author certify that they had obtained all appropriate patient consent.

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Author's contribution

ADK: Identification and management of the case, literature review, manuscript writing.

IC: Substantial contribution to management of the case, critical review of the manuscript writing.

MAU: Substantial contribution to management of the case, critical review of the manuscript writing.

All authors have given final approval of the version to be published and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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