Dermatomyositis is a chronic autoimmune disorder characterized by muscle and skin inflammation, a part of the idiopathic inflammatory myositis (IIM). Even though the disease is idiopathic, there are multifactorial factors related to dermatomyositis. This care report aimed to describe a DM case in a male patient to increase the knowledge and management of DM patients.

**Case description:** A 44-year-old Filipino male was referred to the emergency department (ED) of Siloam Hospital Mataram complaining of muscle pain and weakness with skin rashes 3 weeks before admission. Before the skin rash started, he had enlarged femoral lymph nodes in both thighs. The patient was afebrile with normal vital signs and was prescribed ibuprofen and amoxicillin. After that, he experienced skin rashes around his neck and the back of his ears with minimal pruritus. The symptoms worsened, making him unable to open his mouth and hard to breathe. In the ED, he also threw up dark-colored blood twice. Supporting examination showed elevated transaminase, increased LDH, and creatinine kinase. Biopsy results showed a histologic pattern of dermatomyositis. During hospitalization, he received a high-dose systemic steroid, antibiotic, and symptomatic treatment. He was discharged with a good outcome and planned to continue medical treatment in his country.

**Conclusion:** Dermatomyositis is an idiopathic autoimmune disease involving skin and internal organs. It is a multifactorial disease yet with unclear etiopathogenesis. Specific treatment guidelines for DM are not yet established, but initial systemic corticosteroid and additional steroid-sparring agents may exhibit good outcomes.

**Keywords:** autoimmune, dermatomyositis, idiopathic, myositis, skin rash.

---

**INTRODUCTION**

Dermatomyositis (DM) is a chronic autoimmune disorder characterized by muscle and skin inflammation, a part of the idiopathic inflammatory myositis (IIM). Although the disease is idiopathic, there are multifactorial factors related to dermatomyositis, such as genetic, environmental, and immunologic factors. The disease presents with proximal skeletal muscle weakness and skin manifestation, which differ from other types of myositis clinically and histopathologically. However, around 20% of cases present without muscle weakness, which is then referred to as clinically amyopathic dermatomyositis (CADM). The CADM is further classified into two subcategories: hypomyopathic and amyopathic dermatomyositis. The cutaneous findings include violaceous erythema in different sites such as elbows, knees, lateral hips, upper chest and back, multiple papules over the finger, heliotrope rash around the eyelid, telangiectasia, and proximal nail fold capillary dilatation. Moreover, the pathognomonic manifestations consist of Gottron's papule, Gottron's sign, V-neck sign, shawl sign, Holster sign, calcinosis, and mechanic's hand may occur in patients with DM. Dermatomyositis usually occurs in children or adults with a bimodal distribution and affects two to three times more women than men. The first age peak is 5-14, and the other group is 45-64. Nonetheless, the prevalence of DM is not quite representative of the true population because comprehensive data are lacking. A 32-retrospective study from Minnesota showed the incidence of DM based on age adjustment was 9.63 per 1,000,000 per decade, and the prevalence was 21.4 per 100,000 people. Meanwhile, juvenile dermatomyositis in the United States among children 2-17 years old was 2.1 – 4.5 per million. The length of inflammation in dermatomyositis could affect internal organs such as the pulmonary, cardiovascular, and gastrointestinal systems. Furthermore, 10-20% of cases are associated with underlying malignancy, which could alter the prognosis. The most common pulmonary manifestation is interstitial lung disease (ILD), the leading cause of death in patients with dermatomyositis. Establishing the diagnosis of dermatomyositis requires clinical judgment based on the patient's history and physical examination, thus making it challenging because some cases are not classical. Detection of several specific autoantibodies could help diagnose DM, such as anti-melanoma differentiation-associates genes 5 (MDA5), which is correlated with an increased risk of...
interstitial lung disease. However, there are no specific well-established diagnostic criteria for diagnosing dermatomyositis. Therefore, this case will explain a dermatomyositis case in our hospital with several internal organ involvements.

**CASE REPORT**

A 44-year-old Filipino male was referred to the emergency department of Siloam Hospital, Mataram, via the hospital ambulance call facility, complaining of muscle pain and weakness with skin rashes (Figure 1A-E). The symptoms started to appear 3 weeks before admission when the patient felt weak and muscle pain in both of his thighs and upper arms with stiffness and limited movement.

Along with his symptoms, there was a significant appearance of enlarged femoral lymph nodes in both of his inner thighs. The patient was afebrile with normal vital signs. The patient was prescribed ibuprofen and amoxicillin. The next day, he still felt pain and weakness but started to experience skin rashes around his neck and the back of his ears. The rashes were slightly painful and pruritic. Additional symptoms, such as diarrhea and stomachache, were denied. Two weeks before admission, the swelling of his femoral lymph nodes began to improve. However, the rashes spread to his upper chest, back, and face even more. He then received cetirizine taken orally once before bedtime. The muscle pain and weakness began to worsen with spreading rashes. One day before admission, he experienced facial pain and difficulty opening his mouth, thus making him unable to eat properly. Oedema occurred in both of his arms with stiffness. He also complained of breathing difficulty because he felt pain in both of his side chests. His blood pressure started to increase but without fever.

On his arrival at the ED, he felt nauseous and threw up dark-colored blood vomit twice. He had no history of cough or flu. His blood pressure was 130/90, heart rate 82 times per minute, temperature 36.7°C, and respiratory rate 16 times per minute. According to his previous medical history, he had high blood pressure and had routinely taken 10 mg of amlodipine before bedtime. He had no history of diabetes, food or medication allergy, asthma, or history of surgery. He did notice slight weight loss just because he had been eating a little. He had been working on the ship as a helmsman for 3 years and had been exposed to moderate ultraviolet exposure ever since. Despite this, he wore a full-body suit during his work. According to his familial history, both his mother and father were deceased due to leukemia and stroke, respectively. Based on the physical examination result, he had edema around the buccal area, limitation to open his mouth fully, normal cardiopulmonary examination, and generalized ill-defined bronchovascular pattern (A). Histopathology examination. Histopathological results show focal vacuolar interface dermatitis (yellow arrow) and mild hyperkeratosis (red arrow) (hematoxylin & eosin staining, 100x objective magnification) (B). Perivascular lymphocyte infiltrate (black arrow) (hematoxylin & eosin staining, 400x objective magnification) (C).

**Figure 1.** Generalized violaceous and erythematous patches, varying from ill to well-defined border, with thin scales (A) on both arms (B), on the thighs (C), and the presence of multiple erosion and thin scales on the lower back (D). Note the finger marks on the patient's stomach blanched with pressure (E).

**Figure 2.** Supporting examination results. Chest X-ray showing increased bronchovascular pattern (A). Histopathology examination. Histopathological results show focal vacuolar interface dermatitis (yellow arrow) and mild hyperkeratosis (red arrow) (hematoxylin & eosin staining, 100x objective magnification) (B). Perivascular lymphocyte infiltrate (black arrow) (hematoxylin & eosin staining, 400x objective magnification) (C).

**Figure 3.** Clinical manifestation on the 6th day of hospitalization. Macular violaceous erythema on the face with perioral bright pink rash and thin scales (A) and jaw stiffness (B). V-neck sign (C); Gottron sign or slight ill-defined erythema on the knees (D); Gottron papules (multiple light erythema, ill-defined, multiple papules over the metacarpophalangeal, proximal, and distal interphalangeal joints shown in blue head arrow) (E); and violaceous erythema on the upper arms (F), thighs (G), and lower abdomen (H) were observed.
macular rashes on his face, neck, upper arms, thighs, back, and lower abdomen with slight scaling on top of his rash and multiple erosions. The rash was blanched with pressure (Figure 1E). In the ED, the patient received fluid resuscitation with 1L of ringer lactate for 2 hours, a loading dose of 80 mg pantoprazole, and 1 gr of meropenem.

Supporting examination was carried out for the patient in the ER. His electrocardiography (ECG) result was normal. Laboratory results showed hyperhemoglobinemia, increased hematocrit, hyperleukocytosis, elevated liver enzymes, hypoalbuminemia, and electrolyte imbalance. His urinalysis showed mild ketonuria, positive urobilinogen, and occult blood in the urine. Several radiological examinations were done. Abdominal ultrasound showed non-specific hepatomegaly (suspected parenchymal liver disease), cholelithiasis 0.6 cm with sludge, suspected bowel inflammation, and non-specific lymphadenopathy of bilateral inguinal. Neck ultrasound revealed non-specific lymphadenopathy at the submental, bilateral submandibular, bilateral carotid space, and posterior bilateral cervical space regions. Edema in the sternocleidomastoid and trapezius muscles was found, and dermatomyositis was suspected.

His chest X-ray showed an increased bronchovascular pattern (Figure 2A). Next, a lumbosacral X-ray was done with paralumbar muscle spasms without any compression. Doppler ultrasound revealed valve incompetency at the median cubital vein. Furthermore, musculoskeletal ultrasound showed signs of myositis, tendinosis, and subcutaneous edema. The patient was then screened for COVID-19 infection with an antigen swab test. However, the result was positive. Polymerase chain reaction (PCR) was done twice to confirm the COVID-19 infection. Both PCR results were detected as negative. This case was handled by a multidisciplinary team, which included the internal medicine doctor, neurologist, dermatologist, and pulmonologist.

Furthermore, a blood culture was done with negative bacterial growth. Since he had leukocytosis and a familial history of leukemia, a blood smear morphology test was carried out. The result was interpreted as a sign of inflammation without any suspicion of malignancy. The level of creatinine kinase (7,519 U/L) and lactate dehydrogenase (593 U/L) was highly elevated. Moreover, the patient was screened for thyroid disease. The TSH level appeared normal. The ANA profile was conducted to rule out any underlying autoimmune disease, revealing mild positive for AMA-M2, Scl-70, and DFS70 antigens. A punch biopsy was also executed. The biopsy result showed mild hyperkeratosis in the epidermis, mild perivascular lymphocyte infiltrates, collagen thickening, and erythrocyte extravasation (Figure 2B-C). Those findings established the diagnosis of dermatomyositis with transaminitis, electrolyte imbalance, hypoalbuminemia, hematemeses due to probable erosive gastritis, controlled hypertension, and lung involvement. Electromyography (EMG) and muscle biopsy were planned to be conducted; however, the patient was not suitable as the EMG candidate, and he did not give his consent for muscle biopsy.

During hospitalization, the patient received a high dose of steroid (1 gr of methylprednisolone intravenously (IV) per day) for 3 days, albumin transfusion for correction, furosemide IV 20 mg per day, diphenhydramine IV twice per day, pantoprazole IV 40 mg twice a day, meropenem IV 1 gr thrice a day, IVFD 3% normal saline with 0.9% saline for maintenance, topical ceramide, and topical antibiotic for his skin. On the 4th day of his admission, the patient was put on a central venous catheter because his extremities had become too swollen to put on a peripheral IV line. The rashes started to be less pruritic and painful. The jaw stiffness and erythema around the face remained (Figure 3A-B). Besides, the generalized rash, V-neck sign, Gottron papules, and Gottron sign could still be observed (Figure 3C-H).

He was hospitalized for 9 days, and his condition improved. There was no additional appearance of skin rash, and the rashes started to darken compared to the initial condition, with significant muscle strength improvement. The swelling had diminished, and his electrolyte level, as well as liver enzymes, improved better. There was no adverse event from the treatment. After his condition was stabilized, he was discharged from the hospital to continue further medical examination and treatment in his country. He was prescribed an oral antibiotic, steroid, topical antibiotic, and symptomatic medication.

**DISCUSSION**

Dermatomyositis is a chronic acquired immune-mediated disease that presents muscle weakness and skin rash. In around 50-70% of cases, patients with DM have myositis-specific autoantibodies. Nonetheless, the etiopathogenesis of dermatomyositis remains questionable.

The risks of developing dermatomyositis are multifactorial and include genetics, immunology, and environmental factors.

Genetics plays a role in the etiology of dermatomyositis. Human leukocyte antigen (HLA) polymorphism increases the risk of developing the disease. The first identified allele was HLA-B8, harbored in 75% of juvenile dermatomyositis patients.

Several high-risk haplotypes of HLA also contribute to disease occurrence. These haplotypes include HLA-A*68, HLA-DRB1*0301, HLA-DQA1*0104, HLA-DRB1*07, DQA1*05, and DQB1*02, 24 Besides, the innate and adaptive immune response also partake in the pathogenesis of DM. The histological and molecular features of the disease prove this evidence.

The activity of CD4+, CD8+ T cells, B cells, dendritic cells, and macrophages causes the direct inflammatory effect. In contrast, the indirect effect involves several cytokines such as interferons (IFNs), interleukins (ILs), and tumor necrosis factors (TNF).

Higinterferon (IFN) levels can induce DM-autoantigen, such as MDA5, which then accounts for humoral response-producing autoantibodies. Those autoantibodies help identify the diagnosis of dermatomyositis and are associated with the risks of systemic disease. For example, anti-tRNA synthetase and anti-melanoma differentiation-associated gene 5 (MDA5) are associated with an increased risk of interstitial lung disease; anti–transcriptional intermediary factor (TIF1)-g and anti-nuclear matrix protein 2 (NXP2) are correlated with cancer risks. More than 80% of patients with myositis present with autoantibodies.
Furthermore, viral infection may also trigger the disease, including coxsackie B, enterovirus, and parvovirus. Several drugs, such as antineoplastic drugs, antibiotics (penicillin, sulfonamide, isoniazid), NSAIDs (diclofenac), and radiation could contribute to disease development.

Dermatomyositis is two times more prevalent in women compared to men, and it affects approximately 1-6 persons per 100,000 people. This disease is recognized as a disease with a bimodal age distribution. In adults, the mean age at diagnosis is 44 ± 18.3 years. Since the disease is one of the rare diseases, estimating the incidence and prevalence of true dermatomyositis becomes quite challenging. In addition, establishing the diagnosis of dermatomyositis requires precision and specific standardization.

Several countries, such as Japan and Taiwan, used the insurance claims database for the data analysis. According to the study, DM cases’ approximate annual incidence rate was 10-13 and 6-10 per million, respectively. Moreover, studies from the few-based and largest populations approximated similar results for the incidence and prevalence of dermatomyositis. The prevalence was estimated at 10-20 cases per 100,000 people, while the incidence was 5-10 per 1,000,000 per year. Based on age-adjusted incidence, there was 13.98 per 1 million (95% CI, 8.08-19.89) and 4.68 per 1 million (95% CI, 1.15-8.20) for women and men respectively. Since the disease is related to malignancy and internal organ involvement, which could cause morbidity and mortality. Additionally, clinically amyopathic dermatomyositis patients have the same risk of developing systemic disease as classic dermatomyositis. Dermatomyositis may involve extracutaneous pulmonary, gastrointestinal, cardiovascular, and musculoskeletal manifestations. In our case report, the patient had classic dermatomyositis with gastrointestinal, musculoskeletal, and pulmonary involvement based on clinical approach and supporting exams.

**Internal Organ Involvement**

**Gastrointestinal (GI) manifestation**

The most common GI symptom is dysphagia in juvenile dermatomyositis; meanwhile, in adults, patients complain of abdominal pain (65%), vomiting (16.6%), diarrhea (4.5%), and bleeding (12.5%). The symptoms may appear acute or subacute, ranging from 2 days to 6 months, and are mostly present in women. Even though GI tract involvement is rare in DM patients, it can cause life-threatening problems. The manifestation includes perforation and ulcer formation. Moreover, upper and lower GI tracts are commonly affected. If perforation occurs, the duodenum is the most prevalent affected site. Some studies explained the main cause of GI manifestation was vascular compromise. Histopathologically, several patients also presented with vasculitis. The vascular compromise happens due to chronic arteriopathy, caused by an activated complement and membrane attack complex that destroys the capillary system. Several studies also found that myositis-specific autoantibodies (MSA) (e.g., NXP-2) were likely positive in DM patients with GI manifestation. However, there was no clear explanation regarding the key role of MSA in DM patients with GI complications. The treatment for DM patients with GI involvement should consider more aggressive treatments because of the tendency for life-threatening events. Pulsed intravenous corticosteroids may be considered initially for patients with GI complications. Moreover, additional intravenous immunoglobulin (IVIG) or immunosuppressant medication such as methotrexate and azathioprine may be good choices for treatment.

Our patient presented with nausea and hematemesis, which is a high suspicion of GI involvement of dermatomyositis.

**Pulmonary manifestation**

In patients with DM, interstitial lung disease (ILD) is the common manifestation that causes morbidity and mortality in patients. In addition, ILD mostly occurs as an internal organ complication in DM patients and is a leading cause of hospitalization and cause of death, with a mortality rate between 7.5%-44%. Patients mostly complain of Interstitial lung disease, which may occur at any point of the disease course, with the median time ranging from 16.9 to 18 months. Several MSA also correlate with ILD occurrence. For example, anti-Mi-2, anti-TIF-1-gamma, and NXP-2 lower the risk of developing ILD, while anti-MDA-5 increases the risk. Additionally, palmar papules, punched-out ulcers, and skin necrosis are the pathognomonic manifestations of the presence of MDA-
5. In some cases, interstitial lung disease will progress and cause pulmonary hypertension with symptoms of increased fatigue, shortness of breath, dyspnea on exertion, palpitation, chest pain, edema, and lightheadedness. Along with anti-synthetase syndrome, it will decrease the chance of survival. Usually, the pulmonary screening must be conveyed for all DM patients, regardless of the symptoms. Pulmonary function test (PFT) may be useful but not an adequate screening tool for ILD detection. The common finding is a restrictive pattern with decreased vital capacity (FVC). Moreover, a high-resolution CT scan may be considered a valuable diagnostic test that may present with nonspecific interstitial pneumonia.\(^\text{2,4}\) In this case, the patient had an increased bronchovascular pattern on the chest X-ray without obvious pulmonary symptoms. During treatment, he never had breathing difficulty or cough.

**Musculoskeletal manifestation**

Typically, DM patients complain of muscle weakness in the extensor muscle forming the shoulder, pelvic girdle, and proximal limbs. Myositis in DM presents symmetrically in the proximal extremities. However, around 20% of patients are clinically amyopathic without evidence of muscle weakness. Furthermore, several patients with muscle pain may be present without muscle weakness. Respiratory muscles may also be affected, causing significant respiratory insufficiency and failure. Arthralgia also occurs in 30-40% of DM patients. It commonly affects small joints such as the wrist, MCP, IP, elbows, shoulder, and ankles. The symptoms should be distinguished from rheumatoid arthritis.\(^\text{4}\)

**Malignancy**

Various cancers, such as breast, lung, ovarian, hematologic, and nasopharyngeal cancers, are linked to dermatomyositis. Malignancy in dermatomyositis occurs in around 10-20% of DM cases and usually happens at 1-2 years of disease onset. Myositis-specific autoantibodies related to increased risk of malignancy include anti-(TIF1)-g and anti-(NXP2). Several risk factors for developing malignancy in dermatomyositis involve male gender, older age, absence of ILD, presence of specific autoantibodies, severe skin manifestation, and dysphagia. Protective factors of malignancy include ILD, Raynaud phenomenon, and arthritis.\(^\text{5}\)

**Treatment**

The first line of initial treatment for classic dermatomyositis is a systemic corticosteroid, prednisone, at a dose of more than 0.5 mg/kg/day. However, corticosteroid sparing agents may be necessary to treat myositis and extracutaneous manifestation. Besides, these agents are important to minimize the side effects of systemic corticosteroids, which may cause induced myopathy.\(^\text{4}\) Antimalarial medication (e.g., hydroxychloroquine) is considered the first-line treatment of DM. However, patients treated with antimalarial drugs are more likely to have flare-ups.\(^\text{11}\) The first-line treatment for cutaneous disease includes photoprotection and topical steroids. Topical steroids help reduce erythema, pruritus, and scales, and they are in adjunct with systemic corticosteroids.\(^\text{4}\) Common corticosteroid-sparing agents are methotrexate and azathioprine.\(^\text{6}\) Intravenous immunoglobulin is effective for severe manifestations such as dysphagia and respiratory muscle involvement.\(^\text{4}\) As observed in this patient, a high dose of corticosteroid and other treatments gives a good outcome.

**CONCLUSION**

Dermatomyositis is an idiopathic autoimmune disease that involves the skin and internal organs. Classic dermatomyositis presents with muscle weakness, and it is a multifactorial disease with unclear etiopathogenesis. Several myositis-specific autoantibodies are associated with the increased risk of internal organ manifestation and cancers. Extracutaneous manifestations of DM include pulmonary, gastrointestinal, cardiovascular, musculoskeletal, and malignancy. Interstitial lung disease is the leading cause of mortality in DM patients. Moreover, gastrointestinal involvement also increases the risk of morbidity and mortality due to GI perforation or ulcer. Specific treatment guidelines for DM are not yet established, but initial systemic corticosteroid and additional steroid-sparing agents may exhibit good outcomes.

**CONFLICT OF INTEREST**

The author reports no conflicts of interest in this work.

**ETHICS IN PUBLICATION**

The patient had agreed to and signed an informed consent form for the study's purpose and publication while maintaining the patient's confidentiality. This study was approved by the Research Ethics Committee of the Faculty of Medicine of Universitas Mataram (No: 310/UN18.F8/ETIK/2023).

**FUNDING**

This case report does not receive any funds from the third party.

**AUTHOR CONTRIBUTION**

The first and second authors contributed substantially to the case report concept and carried out the working draft, data acquisition, data analysis, manuscript preparation, and review. For final approval, the third, fourth, and fifth authors conducted data acquisition, analysis, and manuscript review. The sixth author was responsible for data analysis.

**REFERENCES**


This work is licensed under a Creative Commons Attribution