Gac Méd Caracas 2023;131(Supl 2):S212-S219
DOI: 10.47307/GMC.2023.131.s2.18

A Classic Dermatomyositis: A Case Report of Rare Idiopathic Inflammatory Myopathy

Una dermatomiositis clásica: reporte de un caso de miopatía inflamatoria idiopática rara

Satyawardhana[1], Awalia[2*]

SUMMARY

Background: *Dermatomyositis (DM) is a rare autoimmune disease and is one of the idiopathic inflammatory myopathies which predominately affects the skin and muscles. Its estimated incidence is less than 10 cases per million population with an overall female/male ratio is approximately 2:1. This case report presents a classic case of dermatomyositis in a male in Indonesia.*
Case Presentation: *A 38-year-old male was admitted to the Dr. Soetomo Hospital for weakness in all four extremities and dysphagia. Initial symptoms included a red-purplish rash around the eyes, further tests showed an elevation of a serum muscle enzyme and abnormal electromyography. The symptoms did not improve with a systemic corticosteroid but improved with an immunosuppressive agent. This patient had a typical clinical manifestation of classic DM.*
Conclusion: *Immunosuppressive agents including cyclophosphamide should be considered in refractory cases with corticosteroids.*

Keywords: *Myositis, dermatomyositis, cyclophosphamide.*

RESUMEN

Antecedentes*: La dermatomiositis (DM) es una enfermedad autoinmune rara y es una de las miopatías inflamatorias idiopáticas que afecta predominantemente la piel y los músculos. Su incidencia estimada es inferior a 10 casos por millón de habitantes con una relación global mujer/hombre de aproximadamente 2:1. Este reporte de caso presenta un caso clásico de dermatomiositis en un hombre en Indonesia.*
Presentación del caso*: Un hombre de 38 años ingresó en el Hospital Dr. Soetomo por debilidad en las cuatro extremidades y disfagia. Los síntomas iniciales incluyeron una erupción de color rojo púrpura alrededor de los ojos, las pruebas posteriores mostraron una elevación de una enzima muscular sérica y una electromiografía anormal. Los síntomas no mejoraron con un corticosteroide sistémico, pero*

DOI: https://doi.org/10.47307/GMC.2023.131.s2.18

ORCID: 0000-0002-6057-4714[1]
ORCID: 0000-0002-4232-384X[2]

[1] Department of Internal Medicine, Faculty of Medicine Universitas Airlangga - Dr. Soetomo Hospital, Surabaya, Indonesia.
[2] Division of Rheumatology, Department of Internal Medicine, Faculty of Medicine, Universitas Airlangga - Dr. Soetomo Hospital, Surabaya, Indonesia.

Recibido: 23 de febrero 2023
Aceptado: 6 de marzo 2023

*Corresponding author: Department of Internal Medicine, Faculty of Medicine, Universitas Airlangga – Dr. Soetomo Hospital, Jl. Mayjend Prof. Dr. Moestopo No. 6-8, Airlangga, Gubeng, Surabaya, East Java 60286, Indonesia
E-mail: awalia@fk.unair.ac.id (Awalia)

mejoraron con un agente inmunosupresor. Este paciente tenía una manifestación clínica típica de la DM clásica.

Conclusión*: Los agentes inmunosupresores, incluida la ciclofosfamida, deben considerarse en casos refractarios a los corticosteroides.*

Palabras clave: *Miositis, dermatomiositis, ciclofosfamida*

.

INTRODUCTION

Dermatomyositis (DM), an idiopathic inflammatory myopathy, manifests as inflammation of the skin, muscles, and joints, and sometimes involves other organs such as the lungs, heart, joints, and gastrointestinal tract. Skin manifestation of DM occurs in patients with classic myositis or clinically amyopathic dermatomyositis (1). DM affects both children and adults with approximately a 2:1 ratio of female to male. In the last two decades, the number of DM has been reported more frequently compared to previously due to more precise diagnostic criteria and better health access and services. Although, the incidence of SM varies depending on study methodologies, female gender, and urban life are consistent risk factors (2). DM is a rare disease with an incidence of approximately 10 cases per million adults (3).

The pathophysiology of DM involves idiopathic inflammation mediated by muscle and/or connective tissue damage, along with the involvement of other organs. The causative factor is the idiopathic activation of the immune system, which causes immunological attacks on muscle fibers and endomysial capillaries (2). In this present case report, we report a male DM patient from Indonesia with classic presentations.

CASE PRESENTATION

A man, 38 years old, married, was referred to the Emergency Department of Dr. Soetomo, Surabaya, Indonesia with a chief complaint of weakness in all four extremities accompanied by swallowing difficulty. The patient's complaint began with weakness in both legs that started in the patient's thigh in the last two months and was getting weaker until he was unable to walk. Weakness in both arms was felt not long after and since the last month, the patient complained of difficulty swallowing and slurred speech in the past two weeks before hospital admission. At hospital admission, the patient was only able to drink fluid.

Five months before the muscle weakness, there were red-purple rashes around the eyes that were getting darker over time. The rashes were also found on the patient's chest and their appearance was not related to sun exposure. No complaint of numbness in the patient skin, hair loss, or pain in the joints previously. The patient admitted that within six months, he had intermittent fever. There were no complaints related to defecation or urination.

There was no history of hypertension or diabetes. The patient never had a history of stroke, history of trauma, or previous history of autoimmune disease. The patient worked as a driver for 18 years and is a heavy smoker (12 cigarettes per day) and currently lives at home with the patient's nephew. There was no history of similar illness or history of autoimmune disease in the patient's family.

On physical examination, the patient was weak, well aware of GCS E4V5M6, his body weight was 55 kg, height 165 cm with blood pressure 110/62 mmHg, pulse 82x/min, respiratory rate 22x/min, temperature 36.8 ºC, and oxygen saturation was 99 % with free air. Purplish red lesions were found around both eyes, pink papules over the metacarpal and interphalangeal joints, and erythematous patches were also seen in the upper chest and upper back (Figure 1). Examination of the thorax was found to be normal both in the lungs and heart. The abdomen was flat with normal bowel sounds, without tenderness. The extremities were warm, dry, and red with weakness in all four extremities with muscle strength in the upper extremities being 3/3 and the lower extremities being 2/2 without any sensory abnormalities.

Initial laboratory examinations revealed a level of hemoglobin 11.3 g/dL, leukocytes 11 450/mm^3 with 79 % neutrophils and 8 % lymphocytes, platelets 174 000/mm^3, sodium 13^9 mEq/L, potassium 4 mEq/L, chloride 10^3 mEq/L, blood glucose 10^9 mg/dL, creatinine 0.69 mg/dL, blood urea nitrogen (BUN) 25 mg/dL,

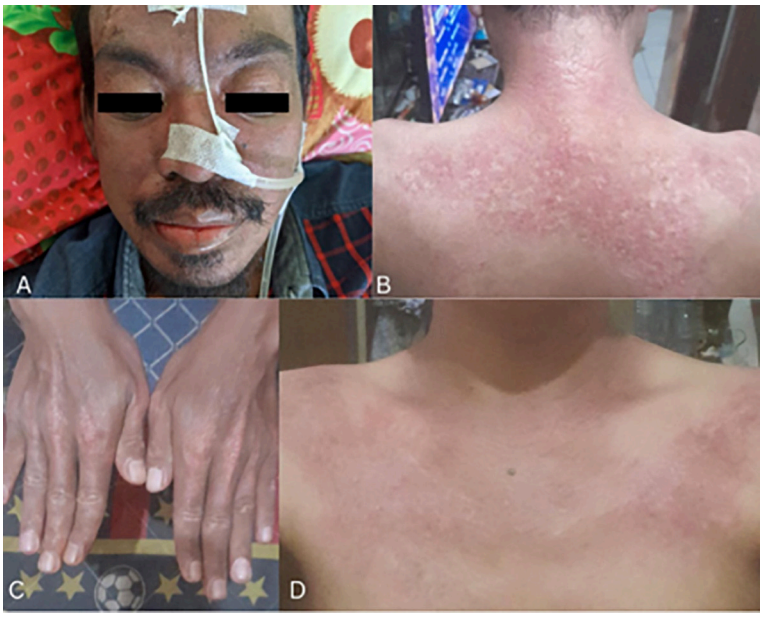

Figure 1. Pathognomonic skin manifestation of dermatomyositis. (A) Heliotrope rash, red-purplish erythema around both eyes involving upper eyelids, in the patient. (B) Shawl sign, erythematous macules, and patches over posterior shoulders, neck, and upper back. (C) Gottron's papules are pink papules over the dorsal side of the metacarpal or interphalangeal joints. (D) V-like sign, erythematous macules, and patches over the lower anterior neck and upper chest.

SGOT 121 U/L, SGPT 64 U/L, albumin 3.06 mg/dL, C-reactive protein (CRP) 0.2 mg/L, partial thromboplastin time (PPT) 27 sec, and activated partial thromboplastin time (aPTT) 14 sec. The chest X-ray showed no abnormalities in the lungs and heart.

Based on the above findings, the patient was diagnosed with susp dermatomyositis dd chronic inflammatory demyelinating polyradiculoneuropathy (CIPD) with elevated transaminase enzymes and hypoalbumin. The patient was planned to undergo further examinations including of anti-nuclear antibody (ANA) test, C3, C4, and creatine kinase. The patient was also planned to undergo skin biopsy, electromyography (EMG), and fiberoptic endoscopic evaluation of swallowing (FEES) examination.

The patient was given an infusion of NaCl 0.9 % 1 500 mL for 24 h, oral vipalbumin 500 mg every 8 h, and planned to receive an IV pulse dose of methylprednisolone 1 g every 24 h for 3 days. The high protein diet was given via nasogastric tube (NGT), Entrasol 100 mL each 2 h and increased gradually. In addition, the neurologist provided the IV mecobalamin 500 μg every 12 h and IV fursulthiamine 2.5 mg every 12 h.

The Antinuclear antibodies (ANA) test yielded negative results with an increased erythrocyte sedimentation rate (ESR) of 41 mm/h procalcitonin 0.32 ng/mL, slightly low C3 level (80 mg/dL), normal C4 (23 mg/dL), elevated creatinine kinase level (2,251 U/L) and negative for both HBsAg and anti-HIV screening test. The Electromyography (EMG) examination suggested demyelinating motor polyneuropathy with suspicion of muscle disease (Figure 2).

After receiving an IV pulse dose of 1 g methylprednisolone every 24 h for 3 days, no improvement was observed. The therapy continued with IV methylprednisolone 62.5 mg every 24 h, infusion of Asering: D5 %: Kalbamin 1: 1: 1 every 24 h, and B complex 1 tablet every 24 h.

Since no clinical improvement in the patient on the 6th day of admission (i.e., IV three days

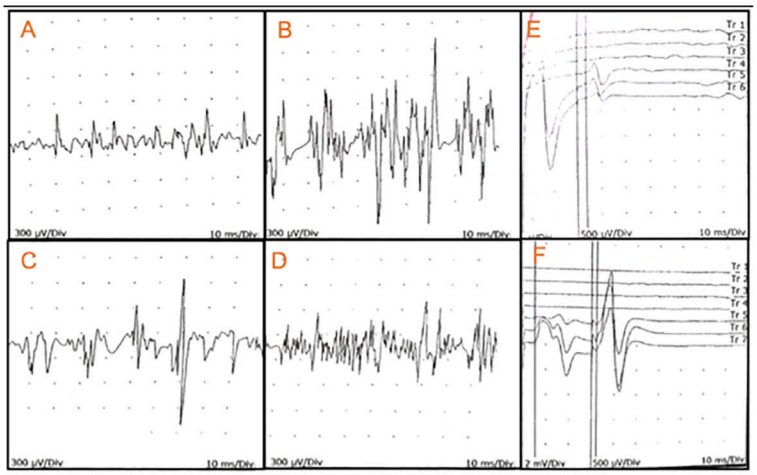

Figure 2. EMG showing polyphasic motor unit potential (A-D) and early recruitment (E, F). The EMG's impression was suspecting a muscle disease.

with pulse dose of 1 g methylprednisolone and three days of IV methylprednisolone 62.5 mg), the patient was treated with 1 g cyclophosphamide immunosuppressant. However, the patient refused to be treated and asked to be discharged. The patient was discharged with oral methylprednisolone 16-16-0 mg.

One day after the patient was discharged from the hospital, the patient came back to the Emergency Department of Dr. Soetomo Hospital with a chief complaint the body felt sore and could not eat because the a nasogastric tube (NGT) was detached. The patient was re-hospitalized with the plan of administering 1 g cyclophosphamide. The patient has been treated with the NGT diet with Entrasol 6x200 mL, infusion of Asering: D5 %: Kalbamin 1: 1: 1 every 24 h, IV cyclophosphamide 1 g within 4 h for 1 day, IV ceftriaxone 1 g every 12 h and vitamin B complex 1 tablet every 24 h.

At hospital admission, a skin biopsy was conducted on the skin lesion on the chest. The result showed a skin tissue lined with the atrophic epidermis, with flattened ridges, showing vacuolar degeneration in some of the basal cells. The dermis layer showed a mild infiltration of lymphocyte cells at the dermo-epidermal junction and was accompanied by the distribution of melanophages with a biopsy conclusion consistent with systemic lupus erythematosus. Initially, a muscle biopsy was planned, but the patient refused due to fear.

After being admitted for six days, the weakness was still present, the patient also complained that sometimes the body felt sore but improved. The patient was discharged with oral methylprednisolone 16-16-0 mg, oral hydroxychloroquine 200 mg every 8 h, and folic acid 1 mg every 24 h. The patient still had to use the NGT and was planned to be treated for six cycles of cyclophosphamide for six months and was scheduled for a cervical magnetic resonance imaging (MRI) examination and a fiberoptic endoscopic evaluation of swallowing (FEES).

The results of the cervical MRI of the patient revealed no lesion or mass in the nasopharynx, oropharynx, or hypopharynx. There was also no visualization of lesions or changes in the intensity of the neck muscles. The FEES examination concluded oropharyngeal dysphagia with severe penetration and aspiration. One month after the first cycle of cyclophosphamide, the weakness and swallowing difficulty remained unchanged. The patient returned to the hospital for the second cycle of cyclophosphamide, and the dose of methylprednisolone was tapered down to 8 mg every 24 h.

After the second cycle of cyclophosphamide, the patient was able to eat fine porridge, but the complaint of limb weakness still presented but improved slightly. The oral drugs included methylprednisolone 8 mg every 24 h, hydroxychloroquine was reduced to 200 mg every 24 h, folic acid 1 mg every 24 h, and $CaCO_3$ 500 mg every 24 h.

The patient was able to walk slowly with help after the administration of the third cycle of cyclophosphamide. The patient's condition continued to improve and after the fifth cycle of cyclophosphamide, the patient was able to walk without help and was able to eat soft rice. The muscle strength of the upper extremities was 4/4 and the lower extremities were 4/4. After the sixth cycle of cyclophosphamide, the patient was provided with oral azathioprine 50 mg every 12 h, methylprednisolone 8 mg every 24 h, folic acid 1 mg every 24 h, and $CaCO_3$ 500 mg every 24 h. During follow-up in the outpatient clinic, the patient said that the condition was not recovered as before, but gradually improved and was being still routinely monitored by the rheumatologist.

DISCUSSION

DM is one of the subgroups of idiopathic inflammatory myopathy (IIM) (5). Patients with DM are subclassified according to the disease subtype into classic dermatomyositis (CDM), amyopathic dermatomyositis (ADM), clinically amyopathic dermatomyositis (CADM), CADM that progressed to CDM, dermatomyositis hypomyositis (HDM), and juvenile dermatomyositis (JDM) (6,7). Our patient can be classified as a CDM of which five months before the muscle weakness onset, the patient had a red-purplish rash around the eyes and on the patient's chest (Figure 1). A CDM is characterized by skin manifestation with evidence of proximal muscle weakness occurring within six months of the onset of the skin (5). According to the patient history and clinical data, our patient can be classified into definite DM based on Bohan and Peter Diagnostic Criteria (6): 1) typical rash of DM; 2) symmetrical weakness of limb-girdle; 3) elevated CK enzyme; and 4) EMG pattern suspecting a muscle disease. The EMG of the patient had early recruitment and a polyphasic motor unit potential supporting the confirmation of the diagnosis. The patient also can be classified as a definite case of idiopathic inflammatory myositis (IIM) according to EULAR/ACR classification criteria for adult and juvenile IIM with a score of 8.1 without a muscle biopsy. Its classification can be further sub-grouped into DM based on EULAR/ACR classification. The patient presented pathognomonic DM clinical and laboratory symptoms, including symmetric proximal muscular weakness with pathognomonic cutaneous signs and a significant increase of muscle enzymes, which satisfied the needed criteria for DM diagnosis (8).

The exact pathomechanism of DM is remained unknown and traditionally has been viewed as a humoral-mediated vasculopathy disease. Similar to most other autoimmune disease, there is a combination of certain genetic predisposition and an exogenous factor (including infection) that trigger the disease. The strongest genetic risk for DM susceptibility is presumed to be the human leukocyte antigen (HLA), specifically HLA-D related (DR) antigen. Exogenous factors for DM include ultraviolet (UV) radiation, medication (NSAID), smoking, and viral infections (9).

The immune activation in DM is not completely understood, but it is likely to be the result of inappropriate complement activation resulting in capillary destruction in the endothelial cells near the endomysium, leading to muscle ischemia and damage (9). The initiating event is the activation of the completer-3 factor (C3), which forms C3b and C4b. This is followed by the formation of the C3bNEO neoantigen and the C5b-C9 membrane attack complex (MAC). This membrane attacks the complex deposits on the walls of blood vessels and causes inflammation. Hypoxic injury to muscle fibers leads to atrophy of muscle fibers, especially muscle fibers in the periphery that are distant from the vascular supply. Over time, the density or density of the capillaries decreases, and muscle fibers begin to undergo necrosis and degeneration (10).

The skin manifestations of DM include the pathognomonic finding of Gottron's papules and heliotropic rashes. Other skin manifestations are the "shawl sign" of the upper back and "V sign" of the anterior neck and upper chest and the "holster sign" of lateral thighs and psoriasiform erythema and scaling of the scalp (11). The

muscle weakness gradually worsens over weeks to months but is usually symmetrical and proximal, with distal muscle weakness occurring late in the course of the disease. A head drop occurred if the neck extensor muscles are affected and in more severe diseases, patients may experience dysphagia, dysphonia, and weakness of the respiratory muscles (12). Our patient had a presentation of heliotropic rash, which is a pathognomonic finding of DM. The muscle weakness started from both thighs, gradually worsening to both legs, which is consistent to DM characteristics which are symmetrical and proximal. Two weeks before admission, there was a complaint of dysphagia which indicated a progressing muscle weakness.

One of the most frequently performed laboratory tests in patients with suspected autoimmune disease is serum complement level. Serum complement exists in the circulation in an inactive state and can be activated when there is a formation of auto-antibodies and immune complexes, resulting in consumption and decreased levels of the complement system in the serum (13,14). ANA status is also tested in patients with suspected autoimmune diseases including DM, although the clinical significance of this status in DM is still unknown. More than 50 % of confirmed cases may have a positive ANA test. However, the serum of patients with an autoimmune disease may produce a negative result while having clear signs and symptoms of the disease. This can be due to current immunosuppressive treatment, the influence of antigenic deficiency in a testing substrate, or loss of IgG through the kidney (15,16). The more specific autoantibody for DM, called myositis-specific autoantibodies (MSA), is currently identifiable in 80 % of adult cases. The frequency of the MSA varies depending on the population studied (3). In our patient, the result of the ANA test was negative with a decreased serum complement level. The patient has not been tested for MSA due to limited funds. The symptoms of dysphagia and heliotrope rash can be observed clearly in this patient and are consistent with a previous cohort study which stated that its frequency is higher in ANA-negative patients (15,16).

Skin biopsy findings in dermatomyositis are similar to those found in systemic lupus erythematosus (10). The most consistent histologic findings of dermatomyositis include an increase in dermal mucin, vacuolar changes of the basal cell layer, and a mild to moderate inflammatory mononuclear cell infiltrate (17).

The goals of DM management are focused on treating muscle weakness, skin disease, and treating other underlying complications (10). There is still no treatment recommendation for DM based on randomized controlled trials (RCTs). The treatment of DM is mainly based on expert opinion or consensus. The current choice of first-line therapy is glucocorticoids (14,18). A high dose of prednisone (1-1.5 mg/kg/day), dexamethasone, or methylprednisolone can be used for autoimmune disease including all IIM subtypes. An intravenous pulse dose of methylprednisolone (500 – 1 000 mg IV daily for 2-4 consecutive days) may be necessary if the disease activity is severe, in the case of IIM is when muscle involvement is severe. A combination with steroid-sparing agents like methotrexate and azathioprine is often used for additional immunosuppressive action and to facilitate steroid tapering (11,19).

A similar case of classical DM in Nepal showed a good clinical response to steroids, the patient was given 50 mg of prednisolone and has shown muscular improvement and a decrease in muscle enzyme (20). Our patient failed to show adequate response to the steroid. Patients who do not respond satisfactorily to steroid therapy and azathioprine or methotrexate are considered refractory. Treatment options for resistant cases include rituximab, mycophenolate mofetil, calcineurin inhibitors, intravenous immunoglobulin, and cyclophosphamide (10). Cyclophosphamide has strong cytotoxic and immunosuppressive effects and has been reported to be useful in severe DM. A cohort study of 123 DM patients has shown a significant improvement in disease activity through the administration of cyclophosphamide (14).

Most patients with DM will require lifelong treatment. DM has a 63 % of 5-year survival rate and a 53 % of 10-year survival rate. Adult-onset DM is said to be associated with underlying malignancy (around 25 % of cases). With an increased risk for lung, breast, ovarian, nasopharyngeal, cervical, colorectal,

or esophageal malignancies reported among DM patients. The mortality of this disease will increase when there is a known malignancy. Our patient didn't have any signs or symptoms of an underlying malignancy. The chest x-ray showed no suspicious lesion and the FEES examination revealed no evidence supporting nasopharyngeal malignancy, which may indicate a better prognosis. However, a thorough scan for malignancy such as a PET scan should be performed to predict the prognosis better, but the test was not available in our hospital. In patients receiving immunosuppressant therapy, the risk of infection is increased. In a previous study, respiratory distress due to pneumonia infection contribute as the third most common cause of death in DM patients (21). This consequence is linked to an increased risk of infection or aspiration as a result of dysphagia, which can lead to pneumonia.

Unfortunately, we didn't evaluate the enzyme creatine kinase (CK) after the course of cyclophosphamide. However, our approach to this case has shown that in the case of DM with steroid resistance, a stronger immunosuppressant drug should be considered to achieve a better clinical response.

CONCLUSIONS

A 38 years-old male with a chief complaint of weakness on all four extremities and difficulties in swallowing was reported. Five months before those complaints, skin rashes appeared on my face and chest. The patient had a typical clinical manifestation of classic DM and was later supported by the laboratories, EMG, and skin biopsy findings. An immunosuppressive agent such as cyclophosphamide therapy could improve the clinical manifestations in the patient when there is steroid resistance. This case highlights the complexity of the diagnosis and management of the DM and early diagnosis of the cases could prevent the progression of the disease.

Acknowledgments

We would like to thank the patient and all the hospital staff involved in the patient's care.

Conflict of interest

The authors have no conflict of interest.

Sponsor

This case report received no external funding.

REFERENCES

1. Wolstencroft PW, Chung L, Li S, Casciola-Rosen L, Fiorentino DF. Factors associated with clinical remission of skin disease in dermatomyositis. JAMA Dermatology. 2018;154(1):44-51.
2. Yang SH, Chang C, Lian ZX. Polymyositis and dermatomyositis – challenges in diagnosis and management. J Transl Autoimmun. 2019;2(October):100018.
3. Tansley SL, McHugh NJ, Wedderburn LR. Adult and juvenile dermatomyositis: Are the distinct clinical features explained by our current understanding of serological subgroups and pathogenic mechanisms? Arthritis Res Ther. 2013;15(2).
4. DeWane ME, Waldman R, Lu J. Dermatomyositis: Clinical features and pathogenesis. J American Academy of Dermatology. American Academy of Dermatology, Inc. 2020;82:267-281.
5. Lundberg IE, Miller FW, Tjärnlund A, Bottai M. Diagnosis and classification of idiopathic inflammatory myopathies. J Intern Med. 2016;280(1):39-51.
6. Okogbaa J, Batiste L. Dermatomyositis: An Acute Flare and Current Treatments. Clin Med Insights Case Reports. 2019;12.
7. Bendewald MJ, Wetter DA, Li X, Davis MDP. Incidence of Dermatomyositis and Clinically Amyopathic Dermatomyositis. Arch Dermatol. 2010;146(1):26.
8. Bottai M, Tjärnlund A, Santoni G, Werth VP, Pilkington C, De Visser M, et al. EULAR/ACR classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups: A methodology report. RMD Open. 2017;3(2).
9. Waldman R, DeWane ME, Lu J. Dermatomyositis: Diagnosis and treatment. J American Academy of Dermatology. Am Acad Dermatol, Inc. 2020;82:283-296.
10. Qudsiya Z, Waseem M, Cornell W, New M. Dermatomyositis. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan. 2022.
11. Cobos GA, Femia A, Vleugels RA. Dermatomyositis: An Update on Diagnosis and Treatment. Am J Clin Dermatol. 2020;21(3):339-353.

12. Marvi U, Chung L, Fiorentino DF. Clinical presentation and evaluation of dermatomyositis. Indian J Dermatol. 2012;57(5):375-381.
13. Abas IH, Tambunan BA, Awalia A. The Correlation between Serum C3 and C4 Complement Levels with Disease Activity Systemic Lupus Erythematosus Patients In Dr. Soetomo Hospital, Surabaya. Curr Intern Med Res Pract Surabaya J. 2021;2(1):1.
14. Kamperman RG, van der Kooi AJ, de Visser M, Aronica E, Raaphorst J. Pathophysiological Mechanisms and Treatment of Dermatomyositis and Immune Mediated Necrotizing Myopathies: A Focused Review. Int J Mol Sci. 2022;23(8).
15. Marfu'ah NO. Diagnostic Problems of a Male Patient With Mixed Connective Tissue Disease. Curr Intern Med Res Pract Surabaya J. 2021;2(1):11.
16. Hoesly PM, Sluzevich JC, Jambusaria-Pahlajani A, Lesser ER, Heckman MG, Abril A. Association of antinuclear antibody status with clinical features and malignancy risk in adult-onset dermatomyositis. J Am Acad Dermatol. 2019;80(5):1364-1370.
17. Smith ES, Hallman JR, Deluca AM, Goldenberg G, Jorizzo JL, Sangueza OP. Dermatomyositis: A clinicopathological study of 40 patients. Am J Dermatopathol. 2009;31(1):61-67.
18. Gunawan PI, Saharso D. A case of juvenile dermatomyositis responding to methotrexate and steroid. Bangladesh J Med Sci. 2018;17(04):675-677.
19. Felani R, Awalia. An Indonesian pregnant woman with systemic lupus erythematosus and cardiac tamponade: A case report. Int J Surg Case Rep. 2022;94(6):107159.
20. Pokhrel S, Pardhe BD, Giri N, Pokhrel R, Paudel D. Classical dermatomyositis: A case report. Clin Cosmet Investig Dermatol. 2020;13:123-126.
21. Airio A, Kautiainen H, Hakala M. Prognosis and mortality of polymyositis and dermatomyositis patients. Clin Rheumatol. 2006;25(2):234-239.