AN ASSESSMENT OF A 72 YEARS MAN WITH DERMATOMYOSITIS

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ABSTRACT
Dermatomyositis is an uncommon inflammatory disease marked by muscle weakness and a distinctive skin change. There is still few cases of this disease reported in Indonesia. It is not only because of its low prevalence rate, similarity manifestation with other connective tissue disease, but also its diagnosis work ups such as Myositis-Sppecifc Antibodies (MSAs) is not widely available and quite expensive in Indonesia. We report a case about a man with pathognomonic skin manifestations and symmetrical proximal muscle weakness, along with laboratory work up, pet scan, muscle skin biopsy, and positif anti-Mi2 supporting diagnosis of dermatomyositis.

Keywords: Dermatomyositis, Myositis-Specific Antibodies (MSAs), anti-Mi2

I. INTRODUCTION
Dermatomyositis is an idiopathic inflammatory myopathy (IIM) that involves various organs and is characterized by changes in the skin. Based on epidemiological data, the incidence of IIM cases is 2.47-7.80 per 100000 individuals yearly with a prevalence rate of 9.54-32.74 per 100,000. Particularly, in the United States, the prevalence rate is 1-6 per 100,000 adults. Dermatomyositis affects all races, however, it is more prevalent in African Americans, and usually appears at the age of 40-60 years, and it is known to affect twice as many women as men.[1–3]

The cases of this disease are still not widely reported in Indonesia due to minimal data records and uneven distribution of supporting examinations for the diagnosis with an expensive cost. Therefore, further discussion is required for insight into dermatomyositis, its diagnosis, and management.

II. CASE REPORT
This study examined a 72-years-old Chinese married man that lives in Surabaya named Mr. J, with 3 children, and unemployed. The patient came to the Emergency Unit with the main complaint of feeling weak, which was deteriorating 3 days before hospitalization. For the previous 2 weeks, the patient complained of a little difficulty swallowing food or drink, but was still able to eat porridge and drink slowly. Consequently, the patient went to the internal medicine polyclinic, was given oral medication, and was advised to come to the ER for inpatient care on the condition that the complaints were getting worse. However, the patient denied complains of nausea and vomiting, pain when swallowing, fever, cough, shortness of breath, urination, and defecation.

About 6 months earlier, there was complaint of redness on the face, chest, elbow, and knee area without itching or pain, which improved after the administration of ointment. Furthermore, complaints of reddish patches reappeared on the face, about 2 months earlier, especially around the eyes, which also looked puffy. The redness was accompanied by small bumps behind the right and left hands, the arm area around the elbow, thigh, and knee. The skin feels thickened and stiff, but not itchy or painful. The patient also complains that the upper arm strength feels somewhat weak, hence, was unable to lift heavy objects above the head.

About 1 month earlier, the patient was treated as an outpatient at a Singapore hospital, where blood tests, pet scans, skin, and muscle biopsies were performed on the right upper arm. However, the patient was unable to perform check-ups again due to the pandemic.

Mr. J had a 3-year history of high blood pressure and diabetes and was placed under medication, which includes the intake of caduet 10/40 1x1 tablet, eclid 100mg 1x1 tablet, and Trajenta duo 2.5/500 1x1 tablet. Furthermore, the patient denied a history of heart disease, stroke, asthma, drug, and food allergies, as well as no family history of similar diseases.

The results of the physical examination include moderate general condition, had consciousness of compost mentis, height 170cm, weight 68kg, blood pressure 120/70mmHg, pulse 85times/minute, respiration 20 times/minute, temperature 36.6°C (axillary). Further examination showed no anemic conjunctiva in the head, no enlarged lymph nodes in the neck, and no abnormalities on the chest. The condition of the abdomen was within normal limits and the
examination of the extremities found warm, dry, and red acral. Hence, the strength of muscle tone in the upper and lower extremities ensures the active movement of the joint against resistance (scale 4).

Red patches were found in the periorbita, forehead, and cheek area, accompanied by minimal periorbital edema (heliotrope rash). Gottron's sign was found on the extensor surfaces of both the right and left elbow joints, hand, and knee extensors, with minimal scaling. Furthermore, Gottron's papules were found on the dorsal surfaces of the metacarpophalangeal and interphalangeal joints of both hands (Figs 1 and 2).

An examination in Singapore regarding a similar case of dermatomyositis found Na 141mmol/L, K 3.4mmol/L, Cl 103mmol/L, urea 9.52mg/dl, albumin 3.6g/dl, SGOT 41 U/L, SGPT 33U/L, GDP 74mg/dl, uric acid 6.41mg/dl, cholesterol 138mg/dl, triglycerides 158mg/dl, HDL 46mg/dl, LDL 61mg/dl. Non-reactive HBsAg, Rheumatoid factor <3.5IU/mL, CRP <5.0mg/L, ANA IF <80, Anti ds DNA <25IU/mL, positive anti-Mi-2 alpha and beta, but anti-MDA5, anti-TIF1 gamma, anti-NXP2, anti-SAE1, anti-Ku, anti-Scl-100, anti-Scl-75, anti-Jo-1, anti-SRP, anti-PL-7, anti-PL-12, anti-EJ, anti-OJ, and anti-Ro-52 were negative.

**Pet scan:** the result showed a slight increase in FDG (F-18 Fluorodeoxyglucose) activity in the circumference of bilateral shoulder muscle. This is due to muscle tension, myositis, or an inflammatory process such as polymyalgia rheumatica, therefore, further examination was recommended.

**Skin Biopsy:** hyperkeratosis occurred on the stratum corneum, inflammation on the perivascular area with lymphocytic infiltration.

**Muscle Biopsy:** lymphocytic infiltration was found in the perimysal area, the muscle was degenerating, and mild necrosis occurred in the perifascicular area.

**Result:** dermatomyositis.

Examinations on the first day of treatment showed Hb 12.0g/dl, leukocytes 5960/uL, platelets 176000/uL, neutrophils 72.6%, lymphocytes 6.5%, ESR 20mm/hour, GDS 105mg/dl, SGOT 136U/L, SGPT 39 U/L, BUN 15mg/dl, creatinine 0.58mg/dl, Na132 mmol/L, K 3.05mmol/L, Cl 97mmol/L, CRP 11.6mg/L, CK 1791.9U/L, albumin 2.7g/dl. Therefore, the x-ray of the thorax showed an atherosclerotic aorta, and pulmo showed no abnormalities.

The patient was diagnosed with Dermatomyositis, Hypoalbuminemia 2.7, Hypokalemia 3.05, controlled hypertension, controlled type 2 DM, planned albumin and potassium evaluation, GDP, GD2JPP, HbA1C. Then, the patient was treated with a refined porridge diet of 1700kcal/day, Asering Infusion 1000cc/24hours; Drip Albumin 25% 100cc in 4 hours; Injection of Methyl Prednisolone 500mg iv every 24 hours for 3 days; Lansoprazole injection 30mg iv every 24 hours. Also, Mycophenolate sodium 2x360mg, KSR 3x600mg, Amlodipine 1x10mg, Atorvastatin 1x20mg, Linagliptin 1x2.5mg, and Metformin 2x500mg were administered orally.
DISEASE PROGRESS

On the 3rd day of treatment, there was an improvement in the symptoms of weakness, redness on the face, elbows, backs of hands, and swallowing difficulty. On physical examination, the general condition was adequate with blood pressure 120/70mmHg, pulse 80 beats/minute, respiration 20 breaths/minute, temperature 36.6ºC (axillary), and improved skin lesions. The result of the laboratory examination showed Na 139mmol/L, K 3.9mmol/L, Cl 97mmol/L, albumin 3.1g/dL, SGOT 115U/L, SGPT 41U/L, GDP 98mg/dL. Furthermore, the patient was diagnosed with dermatomyositis, improved hypoalbuminemia, resolved hypokalemia, control hypertension, and Type 2 diabetes melitus. Afterward, the patient was given therapy including soft diet 2100kcal/day, aminofluid infusion 500cc/24hours, 500mg methyl prednisolone injection every 24 hours for 3 days, and 30mg lansoprazole injection every 24 hours. However, the therapy administered orally include mycophenolate sodium 2x360mg, amloidipine 1x10mg, atorvastatin 1x20mg, linagliptin 1x2.5mg, and metformin 2x500 mg.

On the 4th day of treatment, the patient received 500mg of cyclophosphamide in 30 minutes and methylprednisolone injection was reduced to 62.5mg intravenously every 24 hours. However, the patient and his family decided to return home due to improved health condition and was then scheduled for examination at the outpatient polyclinic of the hospital in Singapore three days later.

III. DISCUSSION

Dermatomyositis is an idiopathic inflammatory myopathy (IIM) characterized by a skin lesion and a diverse collection of systemic clinical manifestations. The pathogenesis of dermatomyositis involves many factors and is not fully understood to date. Some of the factors that are believed to play an essential role in the development of this disease include genetic, environmental, and immune system mechanisms.[1,4,5] Myositis-Specific Antibodies (MSAs) such as anti-Mi2, anti-MDA5, anti-NXP2, anti-TIF1, and anti-SAE are specifically associated with the diagnosis of IIM. Besides anti-Jo1, other MSAs were not included in the IIM diagnostic criteria.[6,7]

Mi-2 antibodies directly inhibit the action of nuclear DNA helicase, which is involved in the transcription process. The patient in this study has “classic dermatomyositis” characterized by pathognomonic skin manifestations, and symmetrical proximal muscle weakness. However, clinically the myopathy is mild, the increase in creatine kinase is usually disproportionate to the severity of the myopathy. Furthermore, dermatomyositis with anti-Mi-2 usually responds well to therapy, has a good prognosis, and is not associated with malignancy or ILD.[1,3]

In this case, the result of the laboratory examination showed a positive Anti-Mi-2 autoantibody. This corresponded to the appearance of pathognomonic clinical manifestations of skin lesions, mild myositis with elevated creatine kinase, and a good response to therapy.

Skin lesions play an important role in the diagnosis of dermatomyositis; however, they are sometimes difficult to distinguish from other connective tissue diseases, particularly lupus erythematosus. To facilitate the diagnosis, skin manifestations in dermatomyositis are divided into seven groups, namely pathognomonic, characteristic, compatible, less common, rare, recent-described, and non-specific. Pathognomonic skin disorders in dermatomyositis include Gottron's papules and signs. Gottron's papules are skin lesions in the form of purplish, flat, sometimes scaly erythematous papules on the dorsal surface of the metacarpophalangeal and interphalangeal joints. On the other hand, Gottron's sign is an erythematous macule, which sometimes accompanied by desquamation, and usually appearing along the extensor surfaces of the joints of the extremities, especially on the dorsal and lateral parts of the hand and fingers.[5,3]

Characteristic skin manifestations include heliotrope rash, which is a purplish or erythematous patch on the eyelids or periorbital area and is often accompanied by periorbital edema. It also includes the presence of changes in the nail fold in the form of telangiectasia and periangual erythema, dystrophic cuticles, and hemorrhagic infarcts. There are purple erythematous macules behind the hand, the extensor parts of the upper and forearm, and the nape of the neck (Shawl sign), V area of the upper chest (V sign), and forehead. Compatible skin manifestations include poikiloderma, which may be hypo or hyperpigmented. Telangiectasia and atrophy are usually found on the upper chest or lateral forearm. Also, the holster sign is a symmetrical poikiloderma found on the hip and lateral thigh below the greater trochanter. Furthermore, periorbital edema and swelling of the face occur with or without erythema.[2,3]

In this case, the patient complained of skin symptoms for the previous 6 months. Based on physical examination, red spots were found in the periorbital area (heliotrope rash), forehead and cheek areas. Gottron’s sign was found on the extensor surfaces of the right and left elbow joints, left and right hand and knee extensors, with minimal scaling. While gottron's papules were found on the dorsal surfaces of the metacarpophalangeal and interphalangeal joints of both hands.

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Muscle involvement in dermatomyositis occurs before, concomitantly, or weeks to months after the skin manifestations appear. Muscle weakness is usually symmetrical having a mild to severe effect on the shoulders and is detected by laboratory or radiological examination. One of its major symptoms is the difficulty in lifting the arm, which results in daily movement limitations. Muscle weakness involves the neck, pharynx, or esophagus causing dysphonia or dysphagia, as well as the diaphragm causing dyspnea.[6,9]

In this case, complaints of mild dysphagia and muscle weakness in both shoulders were felt about 4 months after the appearance of skin lesions. It was then accompanied by increased CK and SGOT, which were determined on laboratory examination and mild bilateral shoulder muscle myositis on a pet scan.

Systemic manifestations of dermatomyositis involve abnormalities in the lungs, heart, joints, and digestive tract. Furthermore, pulmonary abnormalities occur in 10-45% of patients, with the most common manifestation of interstitial lung disease (ILD). The clinical symptoms include severe shortness of breath, leading to fatal pulmonary insufficiency. Consequently, pulmonary function tests and high-resolution CT thorax need to be performed in all patients with suspected pulmonary involvement.[5,6]

The initial evaluation of a patient with suspected dermatomyositis should include a full-body skin examination, such as muscle strength and laboratory tests. In difficult cases, a skin and muscle biopsy, or muscle imaging examinations are needed to confirm the diagnosis. Initially, the diagnosis of dermatomyositis was made based on the criteria established by Bohan and Peter in 1975. Subsequently, many new classification systems were proposed, including EULAR/ACR (2017).[10]

In this case, based on the 2017 EULAR/ACR criteria, a total score of 16 was obtained, therefore, it was included in the definite IIM category. Then in the criteria tree, the final diagnosis of dermatomyositis was obtained.

The management of dermatomyositis aims to reduce inflammation and vasculitis as well as minimize symptoms and improve the patient's quality of life. Therefore, to achieve this goal, therapeutic regimens need to be initiated as early as possible and an interprofessional approach is required. Furthermore, systemic corticosteroids are the mainstay of initial therapy in dermatomyositis with myopathy. However, corticosteroids should not be given to CADM and as monotherapy due to their ineffectiveness and the associated long-term adverse effects. In early-onset patients with myopathy or other systemic symptoms, a combination of systemic corticosteroids with immunosuppressants or oral biologic therapy is recommended. Some examples of steroid-sparing agents are methotrexate, azathioprine, cyclophosphamide, cyclosporine, mycophenolate mofetil, leflunomide, chlorambucil, and tacrolimus.[4,10]

In contraindicated corticosteroids, the second-line agents such as methotrexate and azathioprine are appropriate. However, when resistance to therapy is established, then rituximab, intravenous immunoglobulin (IVIG), and other biologic agents may be administered. Other therapies used for the treatment of superficial skin lesions include antipruritics, topical steroids, hydroxychloroquine, and steroids.[4,10]

In this case, the patient was given systemic corticosteroid therapy and oral mycophenolic acid. The patient showed a good response to therapy and returned home on the third day.

IV. CONCLUSION

This study examines a 72-years-old man with complaints of weakness, skin lesions on the eyelids, face, elbows, knees and hands, accompanied by mild dysphagia and muscle weakness in both shoulders. The patient was diagnosed with dermatomyositis based on different examinations including anamnese, physical, CK, SGOT, anti-Mi-2 enzymes, and pet scan, as well as skin and muscle biopsy. The patient was given systemic corticosteroid therapy and oral mycophenolic acid. Until this report was made, there was a good response to therapy.

REFERENCES
