Classic dermatomyositis in a 36-year-old Filipino female: A case report with emphasis on the early recognition of cutaneous findings of dermatomyositis*

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ABSTRACT

Introduction: Dermatomyositis is a rare idiopathic inflammatory myopathy with characteristic skin manifestations and proximal muscular weakness. In 30 percent of classic dermatomyositis, skin findings precede muscle weakness. Since the initial skin lesions are not always highly characteristic of dermatomyositis, there may be a delay in diagnosis and treatment.

Case report: This is a case of a 36-year-old Filipino female who initially presented with erythematous patches and plaques on the face, arms, and thighs. One week later, she developed multiple, well-defined, erythematous to violaceous, edematous, tender patches and plaques on the face, V of the neck, upper back, proximal extremities and buttocks. Seven weeks later, she developed proximal muscle weakness described as difficulty in raising her arms and difficulty in standing up from a sitting position. Histopathology was consistent with dermatomyositis. SGPT, C3, ANA, and anti-ds-DNA were normal. SGOT and creatine kinase were 5 and 15 times the normal value, respectively. She was treated with prednisone from the first week of illness and hydroxychloroquine from the fifth week of illness. Her condition greatly improved with no progression of the disease for the succeeding 3 years.

Conclusion: Even in the absence of muscle weakness, there should be a high index of suspicion for dermatomyositis in patients with confluent, erythematous patches and plaques on the face, trunk and proximal extremities. Adequate work-up and clinical monitoring will pave the way for early diagnosis and consequently early treatment and a better patient outcome.

***Keywords**: dermatomyositis, inflammatory myopathy, corticosteroids*

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INTRODUCTION

Dermatomyositis is a rare autoimmune disorder that targets the skin, skeletal musculature, and other organs. Little is known about the basic mechanisms underlying the pathogenesis. Skin findings are characteristic and play a significant role in its diagnosis. These include violaceous patches and edematous plaques in the periorbital area (heliotrope sign), V of the neck (V-sign), shoulders (shawl sign), proximal extremities and buttocks (holster sign). In classic dermatomyositis, proximal muscle weakness also develops. In majority of the cases, cutaneous manifestations and proximal muscle weakness present simultaneously, making it easy to diagnose dermatomyositis. However, in 30% of the cases, patients initially present with cutaneous manifestations only. Proximal muscle weakness presents several weeks to months after the onset of the condition.[1] To the untrained eye, these skin lesions can be mistaken for skin conditions such as eczema, contact dermatitis and psoriasis. Hence, early and proper recognition of the skin findings of dermatomyositis is crucial.

Advances in the management of dermatomyositis have improved the prognosis of the patients. Early recognition and aggressive treatment are important in decreasing the mortality and morbidity of this disease. This case illustrates an example of dermatomyositis initially presenting with skin manifestations only, followed by muscle weakness after 7 weeks. Early recognition and treatment resulted in a good patient outcome.

CASE REPORT

A 36-year-old Filipino female presented with a one -week history of erythematous patches and plaques on the glabella, periorbital and nasolabial area, arms, and thighs. One week later, the lesions became edematous and painful bruise-like lesions. The lesions were also seen on the V of the neck, upper back, proximal extremities and buttocks. There was no associated fever or weakness; no medication taken nor consultation sought. The review of systems and past medical history and family history were unremarkable.

Physical examination revealed multiple well-defined erythematous to violaceous, tender patches and edematous plaques on the eyelids, glabella, periorbital area, nasolabial area, upper chest, V of the neck, upper extremities (Figure 1), lower extremities (Figure 2) and buttocks. Working diagnosis was dermatomyositis. Differential diagnoses considered were lupus erythematosus, contact dermatitis and photocontact dermatitis. She was given prednisone at a dose of 0.5mg/kg/day (30mg/day), loratadine 10mg/day, betamethasone dipropionate 2% lotion twice a day and sunscreen SPF 30.

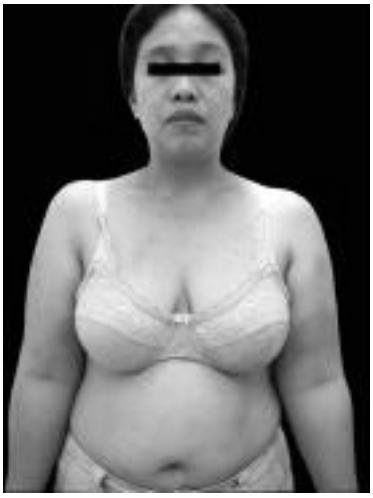

Figure 1. Multiple, erythematous to violaceous patches and plaques on the eyelids, glabella, periorbital area, nasolabial area, upper chest, V of the neck and upper extremities on the first week of illness

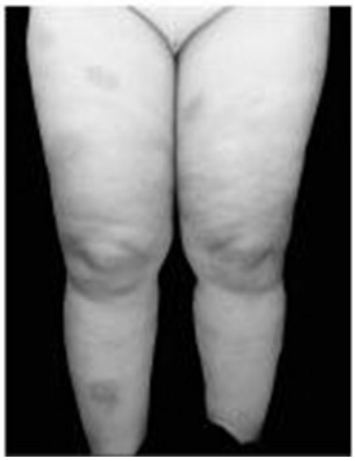

Figure 2. Multiple, violaceous patches and plaques on the lower extremities during the 1st week of illness

Skin punch biopsy was done on the first day of consult and the result came out after two weeks. The biopsy finding was consistent with the diagnosis of dermatomyositis; compact hyperkeratosis, flattened epidermis, and vacuolar interface change with lymphocytes appearing within the dermis. There was superficial and deep perivascular and peri-adnexal infiltrate of lymphocytes, histiocytes, neutrophils, and melanophages with leukocytoclastic vasculitis. (Fig 3).

On the third week of illness, the lesions worsened despite the consistent use of advised medications. She had periorbital edema of both eyelids, edema and blisters on the extremities, fever, generalized weakness, easy fatigability, and muscle pains. Complete blood count, BUN, creatinine, and lipid profile were normal, while urine WBC and LDH were slightly increased. She was maintained on prednisone, loratadine, betamethasone dipropionate lotion and sunscreen.

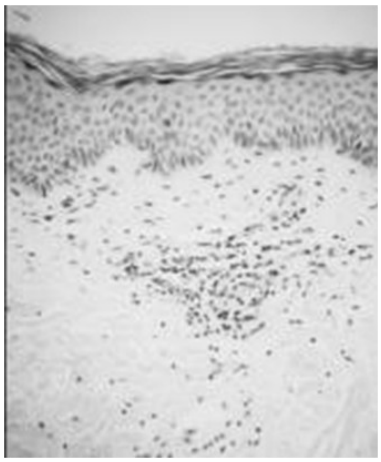

Figure 3. Photomicrograph of the skin punch biopsy specimen showed interface vacuolar dermatitis and leukocytoclastic vasculitis, consistent with dermatomyositis (hematoxylin-eosin stain, 10x)

On the fifth week of illness, 12-lead echocardiogram, kidney, urethra and bladder ultrasound, urine culture and sensitivity, SGPT, C3, ANA and anti-ds-DNA showed normal results. However, SGOT was elevated five times the the normal value. She was started on hydroxychloroquine 200mg twice a day and prednisone was increased to 50mg/day.

On the seventh week of illness, there were no new lesion, however she complained of difficulty in raising her arms and standing up from a sitting position. Her proximal muscles had fair contraction with the ability to overcome gravity, while her distal muscles had good contraction with ability to offer some resistance, and muscle strength tests were 3/5 and 4/5, respectively (Figure 4). During this time, she had multiple well -defined erythematous to violaceous, edematous plaques on the of the forehead, eyelids, malar area (Figure 5), V of the neck (Figure 6), trunk (Figure 7), buttocks, and upper and lower extremities (Figure 8), consistent with the characteristic distribution of dermatomyositis. Creatine kinase was elevated fifteen times the normal value, further supporting the diagnosis of dermatomyositis. Over a span of 4 weeks, her skin symptoms and muscle weakness resolved leaving poikilodermatous skin changes with areas of atrophy. Hydroxychloroquine 200mg twice a day was continued while prednisone was tapered off at 10mg/week on the 11th week of illness. She was referred to other specialties and there were no signs of cardiac, respiratory, gastrointestinal, gynecological and ophthalmologic involvement.

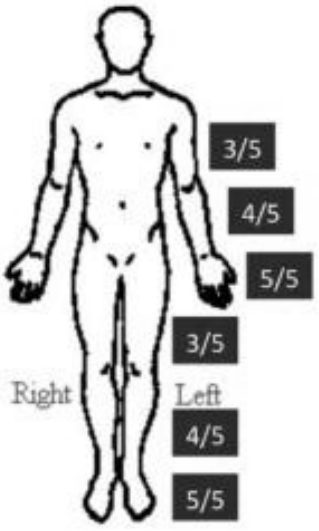

Figure 4. Muscle strength test showed proximal muscle weakness as compared to the distal muscle

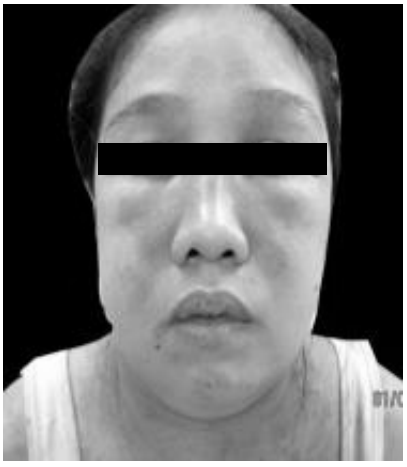

Figure 5. Heliotrope rash manifests as erythematous to violaceous edematous plaques on the eyelids. She also had multiple erythematous to violaceous plaques on the forehead, malar area and during the 7th week of illness

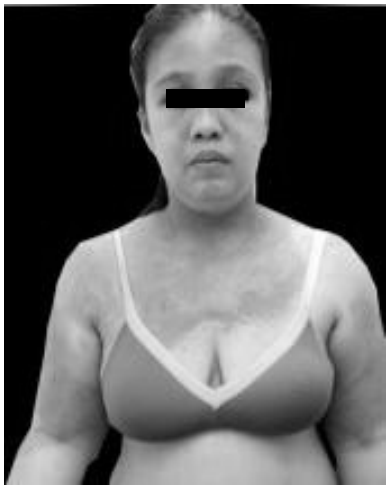

Figure 6. V-sign is seen as multiple, violaceous patches and plaques on the V of the neck and upper chest

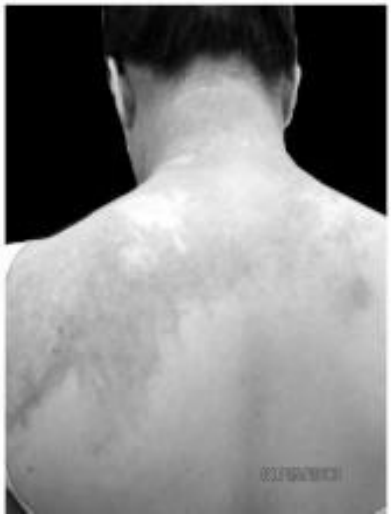

Figure 7. Multiple, violaceous patches and plaques the upper back and shoulders define shawl sign, a characteristic finding in dermatomyositis.

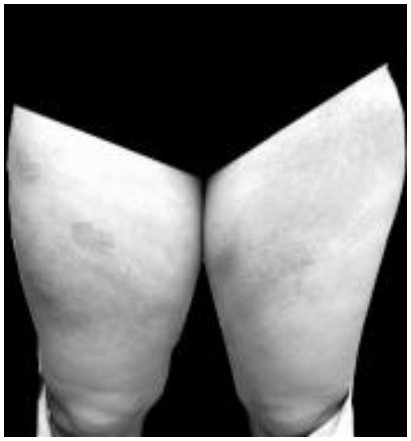

Figure 8. Holster sign manifests as violaceous patches and plaques on the lateral aspect of the hips and thighs.

Dermatomyositis is an idiopathic inflammatory myopathy that targets the skeletal musculature and the skin.[2-4] The etiology remains uncertain. There is a biphasic age of distribution with an initial peak at age 5-14 years and second peak at 45 to 54 years old, and is twice more common in females.[5] The prevalence rate ranges from 9.54 to 32.74 cases per 100,000 individuals.[6] Based on the Philippine Dermatological Society Health Information System database, there were 90 new cases of DM among 398,455 new patients seen from 2011 to February 2017.[7]

The pathogenesis of dermatomyositis begins with exposure of genetically predisposed patients to ultraviolet light or a virus. This leads to the loss of tolerance to self-antigens that cause the specific clinical manifestations.[1] Myositis-specific autoantibodies are found in 50-70 percent of patients with dermatomyositis which likely cause the tissue destruction.[3]

Skin findings are the distinguishing clinical feature of dermatomyositis. The classic skin lesions are symmetric, confluent, macular, violaceous to erythematous patches and plaques. These are seen on the central aspect of the face, periorbital areas, forehead and scalp, V of the neck and upper chest, posterior shoulder, extensors of the fingers, hands, forearms, arms, deltoid areas, and lateral hips and thighs. Gottron's sign, a pathognomonic feature of dermatomyositis, presents as violaceous papules on the interphalangeal or metacarpophalangeal joints.[1] Since the skin lesions of dermatomyositis gradually appear and may not be highly suggestive of the disease from the beginning, a high index of suspicion is important in diagnosing and treating dermatomyositis. Our patient developed these skin lesions over 7 weeks before muscle weakness became evident.

Other common skin lesions include: excoriation with secondary infection and ulceration; subepidermal vesicles and bullae in areas of intense inflammation; post inflammatory hyperpigmentation; deep, irregular ulcers; and non-scarring alopecia.[1,5]

Muscle weakness affecting the shoulders and hip girdle muscle present as difficulty raising the arms and standing up from a sitting position, respectively. In 60 percent of patients, cutaneous lesions and muscle weakness are present concomitantly, while in 10 percent, muscle weakness precedes the cutaneous findings.[1] In these groups of presentation, dermatomyositis can easily be diagnosed clinically. However, in 30 percent, cutaneous findings present without

muscle weakness initially. Muscle weakness only develops after weeks to months. In these cases, clinical diagnosis alone is more difficult and proper treatment may be delayed. Our patient initially had skin lesions alone.

In case of doubt, a skin biopsy is advisable. Characteristic histopathology of dermatomyositis shows hyperkeratosis, basement membrane degeneration, vacuolar alteration of basal keratinocytes, melanin incontinence dermal edema, and interstitial mucin deposition with sparse perivascular lymphocytic infiltrate, acanthosis and epidermal atrophy.[5]

Table 1. Classification Criteria for Polymyositis and Dermatomyositis[8]

Patients presenting with at least one finding from item 1 and four findings from items 2 through 9 are said to have dermatomyositis (sensitivity, 94.1% and specificity, 90.3%)

1. Skin lesions: heliotrope sign, Gottron's sign, or erythema on the extensor surface of extremity joints, slight raised red-purple erythema over elbows or knees
2. Proximal muscle weakness (upper or lower extremity and trunk)
3. Elevated serum creatine kinase or aldolase level
4. Muscle pain on grasping or spontaneous pain
5. Myogenic changes on electromyography
6. Positive anti-Jo-1 antibody test
7. Nondestructive arthritis or arthralgias
8. Systemic inflammatory signs (temperature > 37° C, elevated serum C-reactive protein level or accelerated erythrocyte sedimentation rate >20 mm/hr)
9. Pathologic findings compatible with inflammatory myositis

Our patient fulfilled the classification criteria for dermatomyositis by Tanimoto et al.[8] She had 1 of the 3 skin lesions: heliotrope sign. She also had 4 findings from items 2 to 9: proximal muscle weakness from the 7th week of illness, elevated serum creatine kinase which was 15 times the normal value, spontaneous muscle pain and pathologic findings compatible with inflammatory myositis.

Organ systems can also be involved and present as weakness of the upper third of the esophagus, diffuse interstitial fibrosis (15-30 percent), non-erosive arthritis of the knees, wrists, elbows, and fingers (20-65 percent); and arrhythmia or conduction defect.[1]

Non-invasive procedures such as magnetic resonance imaging and ultrasound are currently being utilized to localize affected muscles. More invasive procedures, such as muscle biopsy and electromyography, are performed in cases that are difficult to diagnose. However, practitioners defer invasive procedures in patients with no muscle symptoms due to sampling errors and low specificity in such patients.[9]

Since dermatomyositis involves internal organs, imaging studies including chest x-ray, CT scan, esophageal motility study, and electrocardiogram are also utilized. During treatment, it is important to monitor muscle enzymes, such as creatine kinase, aldolase, lactate dehydrogenase, SGOT and SGPT to monitor response to treatment.[1]

Adult-onset dermatomyositis has been associated with increased risk of malignancy specially those who are older than 40 years old.[10] In a study by Mebazaa et al., among 130 dermatomyositis cases, 20 were associated with cancer (15.38%); of which, breast cancer (35%) and nasopharyngeal cancer (25%) predominated.[11] In another study by Airio et al., they concluded that the patients with dermatomyositis had a 6.5-fold risk of malignancy like ovarian, stomach, non-melanoma skin, lung and male genital organ cancers.[12] Diagnosis can be made by physical examination, routine laboratory and radiologic examination while imaging techniques like CT scan and magnetic resonance imaging are not routinely done.[1] Since approximately 15% of adult dermatomyositis patient have either a pre-existing malignancy or will develop a malignancy in the future, they should be screened for malignancy and regularly monitored.[10]

In treating the patients, rapid aggressive treatment is indicated in early, active dermatomyositis. Delayed initiation of treatment will result in worsening of the patient's condition and a worse outcome. The mainstay for therapy of dermatomyositis is immunosuppression. Prednisone, the most widely used treatment, ranges from a dose of 0.5 to 1.5 mg/kg/day and is gradually tapered.[5] Non-steroid agents such as hydroxychloroquine 400 mg/day, azathioprine 50mg/day, methotrexate 15mg/week, cyclosporine 3-4 mg/kg/day, cyclophosphamide 300-800 mg/m^2 IV every four weeks and human immunoglobulin 1 g/kg/day over two days can be used as an adjunct to oral steroids.[13]

In a study by Dalakas et al., 15 patients on prednisone 25 mg/day were randomly assigned to receive one infusion of immune globulin (IV Ig) 2mg/kg/day or placebo for three months. Eight patients on IV Ig had significant improvement in scores of muscle strength and neuromuscular symptoms, while seven patients on placebo did not improve. This suggests that high-dose IV Ig is a safe and effective adjuvant treatment for refractory dermatomyositis.[14]

Dinh *et al.,* report three cases of dermatomyositis refractory to prednisone. The patients were given rituximab at 375 mg/m^2 weekly for four consecutive weeks. Lesions significantly improved eight weeks after the last dose of treatment.[15]

Long term effects of dermatomyositis vary from mild rash to life threatening muscle weakness or lung involvement.[16] Mobidity and mortality of dermatomyositis are worsened by poor prognostic factors include the following: progressive disease, presence of malignancy, older age, delayed initiation of therapy, esophageal, cardiac and pulmonary involvement, and extensive cutaneous lesions on the trunk. The one-year survival rate ranges from 83-95 percent, while the 5-year survival ranges from 63-95 percent. Before the advent of corticosteroid treatment, survival rate was at 50%.[17] Initiation of treatment is the only factor that the physicians can control, hence early recognition and treatment of dermatomyositis is imperative. Our patient was treated with prednisone starting on the first week of illness and hydroxychloroquine starting on the fifth week of illness. She has been regularly monitored for the past 3 years and she has not developed any signs of progression of the disease and systemic manifestations. The plan for the patient is regular biannual checkup for 2 more years then yearly to monitor for any progression of the disease and systemic involvement, internal medicine and obstetrics and gynecology checkups for any malignancy and ophthalmologic checkup for retinal toxicity secondary to hydroxychloroquine.

CONCLUSION

This case illustrates that even in the absence of muscle weakness, there should be a high index of suspicion for dermatomyositis specially in cases where confluent, erythematous patches and plaques present on typical areas of involvement such as the face, chest and the proximal extremities. A high index of suspicion with adequate work-up will pave the way for early diagnosis and, consequently, early treatment and a better patient prognosis.

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