Acta Scientific Clinical Case Reports

Volume 3 Issue 7 July 2022

Case Report

A Rare Case of Clinically Amyopathic Dermatomyositis in a Filipino Female

Rene A Amadore*, Ramar John Bangayan and Therese Eileen L Natividad

Department of Internal Medicine, East Avenue Medical Center, Philippines

***Corresponding Author:** Rene A Amadore, Department of Internal Medicine, East Avenue Medical Center, Philippines.

Received: May 05, 2022

Published: June 03, 2022

Abstract

This is a case of a 59-year-old female who presented with a two-year history of heliotrope rash, Gottron's papules, shawl sign, V-neck sign, and muscle weakness. She was previously managed as a case of systemic lupus erythematosus, who initially responded to unrecalled corticosteroids. She was, however, admitted due to a one-month of progressively enlarging sacral mass which eventually turned out to be an abscess. While the abscess was being treated, her autoimmune condition was worked up and she was then managed as a case of Clinically Amyopathic Dermatomyositis (CADM) with Interstitial Lung Disease (ILD). She received corticosteroids and underwent the first cycle of cyclophosphamide infusion prior to discharge.

Keywords: Clinically Amyopathic Dermatomyositis; Interstitial Lung Disease

Introduction

Dermatomyositis (DM), along with polymyositis (PM) and inclusion body myositis (IBM), belongs to a group of heterogeneous disorders called idiopathic inflammatory myopathies (IIMs) which is characterized by muscle weakness and muscle inflammation [1]. DM most commonly occurs between ages 40 to 60 years old, with an estimated incidence of 9.63 cases per million people. Females are affected twice as often as males [2]. Characteristic manifestations of DM include Gottron's papules, heliotrope rash, nail telangiectasia, non-erosive arthritis, and symmetric proximal muscle weakness. Muscle biopsy may reveal the presence of mononuclear cell infiltrates [3]. Anti-Jo-1 antibody, an immunological marker for DM, has a high diagnostic specificity, but is only present in 30% of patients [3].

Being a systemic disease, DM can affect other organs of the body. Dermatomyositis may present with interstitial lung disease (ILD), cardiac arrhythmias, and motility disorders [3]. ILD is a common manifestation of DM, and it can even precede the onset of characteristic muscle or skin manifestations [4]. DM also increases the likelihood of developing malignancy by approximately 6-fold. DM classification can be further subdivided into classic DM or clinically amyopathic DM (CADM). Between the two, CADM has a lower proportion of patients developing malignancy at around 14% to 20%, as opposed to 20% to 25% in classic DM [20].

Case Report

This is a case of a 59-year-old Filipino female who was admitted due to a one-month history of enlarging, non-moveable, nonpruritic, and nontender sacral mass. She was initially worked up for possible malignancy, however, pelvic CT scan revealed a sacral abscess (Figure 1). Incision and drainage was performed and culture studies revealed a pansensitive growth of *Proteus mirabilis.* Patient was treated with Piperacillin-Tazobactam for 14 days with noted resolution of the abscess.

During her course of admission, the patient was noted to have cutaneous rashes and with complaints of occasional joint pains. Upon re-history, she claimed to have had pink to reddish, violaceous papules over the metacarpophalangeal and interphalangeal

Figure 1: Sacral abscess prior to incision and drainage.

joints of both hands, erythematous rashes on both eyelids, and maculopapular rashes on the upper chest which began almost two years prior to admission. No consultation or intervention was done. Nine months prior to admission, the above condition persisted, now associated with generalized, non-specific muscle weakness. It was accompanied by arthralgia, arthritis, and digital ulcerations. Patient consulted at a local clinic and was diagnosed with systemic lupus erythematosus (SLE) for which she was given unrecalled steroids for two weeks with noted improvement on joint pains. Patient was then lost to follow up.

Patient is a known hypertensive for 8 years, maintained and controlled with Losartan. She was treated for pulmonary tuberculosis in 1977 for which she received anti-koch's medications for six months. She has no diabetes mellitus, asthma, allergies, history of atopy, or known malignancies. She is a nonsmoker, nonalcoholic beverage drinker, and previously worked as a security guard.

The patient was alert, coherent, with stable vital signs, and not in respiratory distress. She has anicteric sclerae, pink palpebral conjunctivae, no palpated cervical lymphadenopathies, has clear breath sounds, no breast or axillary masses, normal cardiac rhythm with no murmurs, has flabby nontender abdomen without organomegaly, no inguinal lymph nodes, has full and equal pulses with no bipedal edema. Neurologic physical exam revealed intact higher cortical functions as well as cranial nerves, normal motor strength on all extremities, normal sensory examination, +2 reflexes on brachial, patellar, and knees, no meningeal irritation signs, and no pathologic reflexes.

Examination of the skin reveals heliotrope rash (Figure 2A), coalesced hyperpigmented maculopapular rash on the upper chest (Figure 2B), hyperpigmented linear rashes over the extensor surface of both arms (Figure 2C), Gottron's papules (Figure 2D), and with digital ulcerations (Figure 2E). The impression was dermatomyositis, and the patient received prednisone at 1 milligram per kilogram per day (mkd).

Figure 2: A. Note the subtle periorbital erythema. B. V sign: hyperpigmented (previously erythematous) irregularly shaped patches on the anterior chest. C. Hyperpigmented linear rashes over the extensor surface of both arms. D. Gottron papules: multiple linear irregularly shaped plaques on the proximal interphalangeal, metacarpophalangeal and distal interphalangeal joints. E. Minute ulcerations on the digits of both toes.

Serum tests were done revealing a positive antinuclear antibody (ANA) at 1:100 dilution, with fine speckled pattern. Her creatine phosphokinase (CPK), CPK-MB, CPK-MM were all normal with values of 31 U/L [30-135 U/L], 23 U/L [<25 U/L], and 7.77 U/L [30-110 U/L], respectively. Liver function tests are unremarkable as well: AST 56 [5-34 U/L], ALT 31 [10-55 U/L], and LDH 203 [125-220 U/L].

Neck to chest CT scan, whole abdominal to pelvic CT scan, and breast ultrasound were done and did not show any evidence of nodularities or masses that may point to a malignancy.

Esophagogastroduodenoscopy, and colonoscopy were also performed during her admission. Biopsy of a gastric polyp and duodenal tissue were done revealing chronic inflammation without any evidence of malignancy. Chest CT scan, however, revealed multiple patchy ground glass opacities with peripheral distribution in both lungs, suggestive of interstitial lung disease (Figure 3).

Figure 3: Plain Chest CT scan showing multiple patchy peripheral ground glass opacities in both lungs.

With an impression of Connective Tissue Disease-associated Interstitial Lung Disease (CTD-ILD), the patient underwent pulmonary function test which revealed a low forced vital capacity (FVC) and low FEV (forced expiratory volume). The findings were interpreted as probable moderate restrictive ventilatory defect with no significant bronchodilator response (Figure 4). Lung volume studies (LVS) and diffusing capacity of lung for carbon monoxide (DLCO) were suggested to confirm and evaluate the restrictive lung disease.

Figure 4: Pulmonary Function Test showing moderate restrictive ventilatory defect with no significant bronchodilator response.

DLCO revealed the following results: DLCO 18.61 mL/min/mmHg (14.57-26.05 mL/min/mmHg) and Total Lung Capacity (TLC) 3.31L (3.26-5.23L). Findings revealed a normal DLCO but with mild restrictive ventilatory defect with underlying obstructive

Figure 5: LVS and DLCO results showing values within normal range.

Dermatology service also saw and examined the patient. They noted multiple, welldefined, round to irregularly shaped, erythematous to hyperpigmented patches on bilateral postauricular areas, preauricular areas, bilateral upper extremities, and bilateral upper extremities. Heliotrope sign, Gottron's papules, V-neck sign, and Shawl sign were also noted. Systemic corticosteroids were continued and they started the patient on clobetasol propionate 0.05% ointment two times a day on affected erythematous areas for two weeks. Mild soap was advised for bathing.

Electromyography (EMG) and nerve conduction velocity (NCV) of both upper and lower extremities revealed abnormal results: 1) Sensory amplitudes of the median nerves are reduced with slowing of sensory conduction velocities more on the right 2) Motor amplitudes are moderate to severely reduced. Distal latencies are prolonged only on both median nerves and right peroneal nerve. 3) The right median F-waves are prolonged while the rest of the F-waves are normal. Tibial H-reflex responses show normal latencies 4) Needle EMG showed highly polyphasic motor units, some with early recruitment mostly in the proximal muscles of lower extremity more than the upper extremity. No spontaneous activity was seen. There is subtle evidence for a non-inflammatory myopathic process mostly involving the proximal lower extremity muscles. Considerations include metabolic vs steroid-induced myopathies.

Anti-Jo1 was done prior to the patient's discharge which revealed a result of 0.4 U/L [<7: negative, 7-10 equivocal, >10 positive]. With complete resolution of the sacral abscess, improvement in muscle weakness and arthritis, and infusion of first cycle of cyclophosphamide, the patient was sent home well and improved with the following take-home medications: Prednisone at 1 mkd and calcium + vitamin D tablet once a day.

Discussion and Conclusion

Patient was diagnosed with dermatomyositis using the American College of Rheumatology-European League Against Rheumatism Classification Criteria for Adult and Juvenile Idiopathic Inflammatory Myopathies (IIM). She satisfied the following criteria, with corresponding probability scores: (1) age of onset of first related symptoms at 40+ years (2.1), (2) Heliotrope rash (3.1), (3) Gottron's papules (2.1), (4) Gottron's sign (3.3). We were not able to obtain a muscle biopsy mentioned in the criteria because of the patient's financial difficulties. Anti Jo-1 turned out negative. However, despite not having a muscle biopsy and a negative autoantibody result, the patient had a total score of 10.6 which has 99% probability for IIM. Furthermore, we were able to classify the patient as having DM based on the following features: (1) age at onset of first symptoms by >18 years-old, and (2) the presence of Heliotrope rash, Gottron's papules and Gottron's sign. These criteria have a specificity of 82% and sensitivity of 87% when used without a muscle biopsy [5]. We also subclassified the condition as Clinically Amyopathic Dermatomyositis (CADM) on the basis that the cutaneous manifestations occurred for more than 6 months without clinical, laboratory, and muscle testing evidence of myopathy.

Endoscopy, colonoscopy, and multiple imaging studies we performed in order to determine possible malignancy of the patient. Patients with DM have a three to eight times increased risk for developing an associated malignancy compared with the general population, and therefore, all patients with DM should be evaluated at the time of diagnosis for the presence of an associated malignancy [6]. The most common cancer associated with DM in Southeast Asia is nasopharyngeal cancer, while breast, lung, and colon cancers are the three most common from patients in the West [7]. A local case report from Philippine General Hospital (PGH) presented a 40-year old male with typical cutaneous manifestation of DM with no muscle weakness and normal creatine phosphokinase. He was diagnosed with CADM and workup for malignancy was done including chest xray, whole abdominal ultrasound, urinalysis, fecal occult blood test, and prostate specific antigen. The patient then developed cervical lymphadenopathies and underwent biopsy revealing a metastatic undifferentiated carcinoma [20].

Neurophysiological studies have high sensitivity and specificity in diagnosing myopathies and are able to exclude other causes of weakness such as myasthenic syndromes, neuropathies with motor involvement, or motor neuron diseases [8]. These tests, according to Bohan and Peter, have a sensitivity of 89% [9]. In acute myopathies, it is recommended to conduct the study 3 weeks after symptom onset in order to ensure a higher sensitivity [8]. Another thing to consider when doing EMG is that it may cause elevation of creatine phosphokinase (CPK), which is also a blood test used in the classification criteria for DM. It is therefore recommended to do CPK determination before or three days after the EMG [10]. Findings in EMG for DM include fibrillation and positive waves, high frequency, or myotonic discharges [8]. The patient`s EMG study did not show any of these. It revealed a non-inflammatory myopathy involving proximal lower extremities muscles, with considerations of metabolic vs steroid-induced myopathies. Myopathic diseases are usually progressive, may present with atrophy in advanced diseases, and with elevated creatine kinase [15]. Patient also has been taking prednisone for three weeks when the EMG was done.

The usual dermatologic manifestations of DM are present in our patient: the Heliotrope rash, Gottron's papules, and the V-neck sign. Other characteristic rashes that were not observed include the shawl sign which refers to erythema on upper back, posterior neck, and shoulders, and the Holster sign which is a violaceous rash over the lateral hip. They can also present with scalp disease presenting with erythematous scaling plaques on the scalp. (6) On nailfold capillaroscopy, DM patients can have prominent dilated and tortuous blood vessels with accompanying avascular areas. The degree of vessel drop out as well as telangiectasias is a reflection of ongoing disease activity [11]. Exposure to sunlight worsens skin disease of DM, hence, protective clothing and application of sun block with at least 15 sun protection factor (SPF) is recommended [19].

ILD can affect 35-40% of patients with IIMs such as DM and is usually associated with the presence of antisynthetase antibody [12]. The presentation of ILD in DM patients can present with one of these three: (1) an acute, severe involvement, (2) a chronic gradually progressive signs and symptoms, or (3) asymptomatic disease in which lung disease incidentally diagnosed by imaging studies, just like how our patient presented [14]. Pulmonary function test (PFT) in ILD reveals a restrictive pattern described as low forced vital capacity (FVC) or total lung capacity (TLC) <80% predicted for age [6]. Characteristic features on high resolution chest CT scan include ground glass opacities (GGO), linear opacities, fibrosis with or without honeycombing, and bronchiectasis [6]. In a study published in European Respiratory Journal, they noted that the most frequent CT findings are reticular and ground glass opacities [16]. The patient's chest CT scan scan revealed GGOs with peripheral distribution and PFT showed a low FVC suggestive of ILD. In the same study, they found out that respiratory symptoms occurred 100 days after muscular symptoms and 340 days after the skin symptoms [16]. Early diagnosis and intervention are necessary to improve the prognosis. No standard treatment has been established yet, although combination therapy with high dose glucocorticoid (1mg/kg), a calcineurin inhibitor (tacrolimus or cyclosporine A), and cyclophosphamide should be considered as first-line therapy [17].

Progressive ILD is more frequently observed in patients with CADM [13]. CADM comprises 10% to 20% of all DM patients with increased risk for rapid progressive ILD (RP-ILD) resulting in higher rate of mortality [21]. CADM patients who have anti-melanoma differentiation-associated gene 5 (MDA-5) antibodies are more likely to develop RP-ILD [22]. In a study done by Pedram., *et al*. (2011), it was found that CADM patients can eventually develop muscle weakness from 15 months to 6 years after the onset of their skin disease [23].

The prognosis of DM patients is determined by ILD, hence it is important to control ILD to avoid poor outcomes and help maintain patient's quality of life [18]. Since our patient has CADM with CTD-ILD, tight monitoring of clinical conditions is required. Monitoring of ILD is three-tiered: (1) disease activity monitoring which include determination of anti melanoma differentiation associated gene 5 (anti-MDA5) antibody titer, serum ferritin, and clinical signs such as skin rashes, (2) respiratory condition monitoring which includes observation for development of symptoms such as dyspnea, cough, as well as progression of GGOs on HRCT, and (3) monitoring for adverse events including opportunistic infections since the patient is on immunosuppressive therapy. In Japan, routine monitoring for cytomegalovirus, pneumocystis pneumonia, and other fungal infections is done [18].

The patient was discharged well and improved after infusion of cyclophosphamide with the following take home medications: prednisone 50 mg/day (1 mg/kg/day) and calcium + vitamin D tablet once a day. She already followed up at the Rheumatology outpatient clinic for her due cyclophosphamide infusion to complete six cycles.

Recommendations

We recommend to determine other autoantibodies such as anti-MDA-5 which is usually positive in DM patients with rapidly progressive ILD. Anti-Mi-2 is not available in the Philippines but its positivity may point to classic DM, good response to treatment, and lower incidence of malignancy. One study found out that serum ferritin is a useful marker and its elevation indicates a poor prognostic factor. Lastly, two-dimensional echocardiography must be facilitated to rule out cardiac involvement.

Bibliography

1. Plotz PH., *et al*. "Myositis: immunologic contributions to understanding cause, pathogenesis, and therapy". *Annals of Internal Medicine* 122 (1995): 715-724.

2. Linos E., *et al*. "Atherosclerotic cardiovascular disease and dermatomyositis: an analysis of the Nationwide Inpatient Sample survey". *Arthritis Research and Therapy* 15.1 (2013): R7.

3. Jakubaszek M., *et al*. "Polymyositis and dermatomyositis as a risk of developing cancer". *Reumatologia* 53 (2015): 101-105.

4. Abhinav K Misra., *et al*. "Interstitial lung disease is a dominant feature in patients with circulating myositis-specific antibodies". *BMC Pulmonary medicine* 21 (2021): 370.

5. Leclair V and Lundberg IE. "New myositis classification criteria what we have learned since Bohan and Peter". *Current Rheumatology Report* 20 (2018): 18.

6. Umaima Marvi., *et al*. "Clinical Presentation and Evaluation of Dermatomyositis". *Indian Journal of Dermatology* 57.5 (2012): 375-381.

7. Wilsher ML., *et al*. "Hypercalcaemia during resolution of calcinosis in juvenile dermatomyositis". *British Medical Journal (Clin Res Ed)* 288 (1984): 1345.

8. Gerardo Gutiérrez Gutiérrez., *et al*. "Use of Electromyography in the Diagnosis of Inflammatory Myopathies". *Reumatologia Clinica* 8.4 (2011): 195-200.

9. A Bohan., *et al*. "Computer-assisted analysis of 153 patients with polymyositis and dermatomyositis". *Medicine (Baltimore)* 56 (1977): 255.

10. R Levin., *et al*. "Phillips 2nd. The time course of creatine kinase elevation following concentric needle EMG". *Muscle Nerve* 10 (1987): 242-245.

11. Smith RL., *et al*. "Skin involvement in juvenile dermatomyositis is associated with loss of end row nailfold capillary loops". *Journal of Rheumatology* 31 (2004): 1644-1649.

12. Fathi M., *et al*. "Pulmonary complications of polymyositis and dermatomyositis". *Seminars in Respiratory and Critical Care Medicine* 28 (2007): 451-458.

13. Sato S and Kuwana M. "Clinically amyopathic dermatomyositis". *Current Opinion on Rheumatology* 22 (2010): 639-643.

14. Fathi M and Lundberg IE. "Interstitial lung disease in polymyositis and dermatomyositis". *Current Opinion on Rheumatology* 17 (2005): 701-706.

15. Brent W Smith., *et al*. "Suspect myopathy? Take this approach to the work-up". *The Journal of Family Practice* 63.11 (2014): 631-637.

16. V Cottin., *et al*. "Interstitial lung disease in amyopathic dermatomyositis, dermatomyositis and polymyositis". *European Respiratory Journal* 22 (2003): 245-250.

17. Tsuji H., *et al*. "Multicenter prospective study of the efficacy and safety of combined immunosuppressive therapy with high-dose glucocorticoid, tacrolimus, and cyclophosphamide in interstitial lung diseases accompanied by antimelanoma differentiation-associated gene 5-positive dermatomyositis". *Arthritis Rheumatology* 72.3 (2020): 488-498.

18. Kazuhiro Kurasawa Satoko Arai. "Optimal management of interstitial lung disease associated with dermatomyositis/polymyositis: lessons from the Japanese experience". *Dove Press* 2014.4 (2014): 93-107.

19. John Okogbaa and Lakeasha Batiste. "Dermatomyositis: An Acute Flare and Current Treatments". *National Library of Medicine* 12 (2019): 1179547619855370

20. Felix Paolo J Lizaronda and Eileen Liesl A Cubillan. "Clinically Amyopathic Dermatomyositis Associated with a Metastatic Undifferentiated Carcinoma in an Adult Filipino Male: A Case Report". *Acta Medica Philippina* 53.2 (2019): 198-201.

21. Gan Yu-Zhou., *et al*. "Risk factors of interstitial lung diseases in clinically amyopathic dermatomyositis" 1.6 (2020): 644-649.

22. Satoshi Ikeda., *et al*. "Interstitial lung disease in clinically amyopathic dermatomyositis with and without anti-MDA-5 antibody: to lump or split?" *BMC Pulmonary Medicine* 15 (2015): 159.

23. Pedram Gerami., *et al*. "A systematic review of adult-onset clinically amyopathic dermatomyositis (dermatomyositis siné myositis): A missing link within the spectrum of the idiopathic inflammatory myopathies". *Journal of the American Academy of Dermatology* 54.4 (2006): 597-613.