

# Amyopathic Dermatomyositis with Interstitial Lung Disease: A Rare Type of Dermatomyositis with Accelerated Mortality Rate

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## Abstract

**Background:** It was observed that there have been cases with pathognomonic skin lesions of Dermatomyositis, but without muscle weakness and abnormal skeletal muscle enzymes, hence they did not satisfy the Peter and Bohan criteria, and management became undirected. Until in 1993, Euwer and Sontheimer published the diagnostic criteria for Amyopathic Dermatomyositis, a type of Dermatomyositis, without proximal muscle weakness and elevated skeletal muscle enzymes.

**Setting:** St. Luke's Medical Center, Quezon City, Philippines.

**Case:** We report here a case of a 59-year old female who was admitted for shortness of breath presenting with hyperpigmented papules over the metacarpophalangeal joints. One week before admission, fever, productive cough, and progressive shortness of breath developed prompting hospital admission. She arrived at the ER tachypneic with oxygen saturation of 55% at room air. She had intercostal retractions and crackles on both lung fields. There were violaceous macular rashes on the metacarpophalangeal, and proximal interphalangeal joints of both hands. Neurological examination was normal.

**Diagnostics:** Chest CT scan revealed diffuse ground glass opacities with interstitial thickening consistent with Interstitial Lung Disease (ILD). The serum Creatine Kinase level and electromyography were normal. Skin biopsy showed perivascular lymphohistiocytic infiltrates, suggestive of dermatomyositis.

**Conclusion:** Dermatomyositis is rare and the Amyopathic type is more uncommon (incidence of 2.08/1,000,000 population). Recognizing it can be a challenge. Amyopathic Dermatomyositis (AMD) patients have 80% survival in less than a year and drops to 60% if ILD develops. Hence, early diagnosis and aggressive management are imperative.

**Keywords:** Amyopathic dermatomyositis; interstitial lung disease

## Introduction

The Peter and Bohan criteria [1] (Table 1) have traditionally been used to diagnose patients with Dermatomyositis (DM). Presentation with the characteristic rash plus one other criterion, gives a diagnosis of possible DM. If with a rash plus two other criteria were met, diagnosis of dermatomyositis is probable. On the other hand, a rash plus three other criteria, is given a diagnosis of definite DM. However, clinicians have seen patients with pathognomonic skin lesions of Gottron's papules but do not fit the criteria. It was noted in the 1980s that, unless the criteria were met, a patient could not be diagnosed as such, management was delayed, and patients deteriorated. The Peter and Bohan criteria were found to have a low sensitivity in diagnosing Dermatomyositis mainly because the disease entity itself is a spectrum presenting with no muscle weakness and normal skeletal muscle enzymes to those with proximal muscle weakness and elevated skeletal muscle enzymes.

Later in 1993, several types of Dermatomyositis were recognized and characterized. This led to the emergence of what is called now the Amyopathic Dermatomyositis (ADM) [2], which at present is diagnosed using the Euwer and Sontheimer Diagnostic Criteria for ADM. Hence, diagnosis can still be made and appropriate management can be started, especially that Dermatomyositis may have a low survival rate in just one year.

## Case

This is a case of a 59-year old female presenting with rashes and shortness of breath. She was apparently well until 13 weeks before admission, when she noted appearance of multiple erythematous, macular, non-pruritic rashes over the malar and cheek areas of the face bilaterally, not precipitated nor aggravated by exposure to sunlight. These gradually increased in number and progressively involved the

neck, upper back, and upper abdomen. There was no fever, cough, colds, anorexia, headache, body malaise, dyspnea, abdominal pain, nausea, vomiting, or change in bowel habits. There was no intake of allergenic food and drugs, insect bites, exposure to allergens, nor recent travel. There was no consult done or medications taken. Above symptoms persisted until 11 weeks before admission, when erythematous, macular, mildly pruritic rashes appeared on the forearms, arms, upper chest, and lower abdomen which eventually progressed appearing as hyperpigmented patches with excoriations within one week period. This was associated with pain with a severity scale of 3 out of 10 over the lower back and proximal anterior and posterior thighs, aggravated upon standing up. There was no fever, abdominal pain, weakness, or numbness. She consulted a dermatologist who diagnosed her with allergic dermatitis. She was prescribed colloidal, oatmeal soap and hypoallergenic soap; however, there was no improvement of symptoms.

Above symptoms persisted until 9 weeks before admission, when pruritic, hyperpigmented papules with hyperkeratosis developed over the metacarpophalangeal joints of both hands. Weight loss of approximately 4 to 5 kilograms within 4 weeks was also observed. Previously noted symptoms persisted, until 7 weeks before admission

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when she noted bilateral periorbital swelling. There was no eye itchiness, redness, discharge, pain, blurring of vision, facial swelling, or fever. She consulted another dermatologist who prescribed her with Clobetasol cream and Betamethasone+Loratadine 250 mcg/5 mg ointment. Symptoms however persisted, prompting consult with a third physician who started her with Methylprednisolone 24 mg after breakfast and 16 mg after dinner and Methotrexate 7.5 mg once a day every Sunday. She was relieved of her myalgia after a week. However rashes and periorbital swelling persisted. Until 1 week before admission, she developed generalized body weakness, anorexia, intermittent fever with a maximum temperature of 38.5°C occurring anytime of the day and productive cough with whitish sputum. She self-medicated with Paracetamol. Previously noted symptoms persisted until three days before admission when she started to experience shortness of breath without associated chest pain, palpitation, orthopnea, or bipedal edema. Shortness of breath progressed occurring even at rest, prompting her to consult, which subsequently led to admission.

Above symptoms persisted until 9 weeks before admission, when pruritic, hyperpigmented papules with hyperkeratosis developed over the metacarpophalangeal joints of both hands. Weight loss of approximately 4 to 5 kilograms within 4 weeks was also observed. Previously noted symptoms persisted, until 7 weeks before admission when she noted bilateral periorbital swelling. There was no eye itchiness, redness, discharge, pain, blurring of vision, facial swelling, or fever. She consulted another dermatologist who prescribed her with Clobetasol cream and Betamethasone+Loratadine 250 mcg/5 mg. Symptoms however persisted, prompting consult with a third physician who started her with Methylprednisolone 24 mg after breakfast and 16 mg after dinner and Methotrexate 7.5 mg once a day every Sunday. She was relieved of her myalgia after a week. However rashes and periorbital swelling persisted. Until 1 week before admission when she developed generalized body weakness, anorexia, intermittent fever with a maximum temperature of 38.5°C occurring anytime of the day and productive cough with whitish sputum. She self-medicated with Paracetamol. Previously noted symptoms persisted until three days before admission when she started to experience shortness of breath without associated chest pain, palpitation, orthopnea, or bipedal edema. Shortness of breath progressed occurring even at rest, prompting her to consult thus was admitted.

Review of systems was unremarkable. She has no hypertension, diabetes mellitus, bronchial asthma, or known allergy. She had two caesarean sections in the 1970s. She was menopausal at 52 years old. She was neither a smoker nor an alcoholic beverage drinker, and denied history of illicit drug use. There was a family history of diabetes and hypertension but no family history of cancer, bronchial asthma, thyroid disorder, or autoimmune disease.



Figure 1: Skin lesions on the chest.

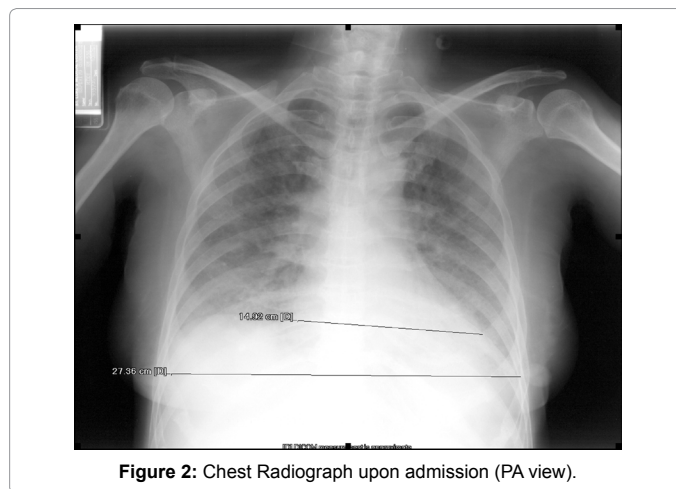


Figure 2: Chest Radiograph upon admission (PA view).

On admission, she was conscious, coherent, tachypneic, with the following vital signs: BP=120/80 mmHg, heart rate=90 beats per minute, respiratory rate of 24 cycles per minute, temperature of 37.3°C (99.14°F), and oxygen saturation of 55% at room air. Her Body Mass Index was 21.8 kg/m<sup>2</sup>. There were multiple ill-defined dusky red dry patches over the chest (Figure 1) and back, and erythematous plaques of varied sizes with lichenification over the abdomen.

There were violaceous macular rashes on the skin overlying the metacarpophalangeal and proximal interphalangeal joints of both hands. There was no joint swelling, tenderness, or limitation of movement. She had violaceous discoloration over the left upper eyelid, pink palpebral conjunctivae and anicteric sclerae. There was a circular whitish papule on the right ear. There was no tragal tenderness, aural and nasal discharge. Tonsils were not enlarged and the posterior pharyngeal walls were non-hyperemic. She had supple neck, no cervical lymphadenopathy and no palpable mass. She had equal chest expansion, with intercostal retractions, and crackles were noted on the right middle to basal lung fields, and left base. No wheezes appreciated. Cardiac and abdominal examinations were normal. Pulses were full and equal on all extremities. No bipedal edema. There were no cranial nerve deficits, muscle weakness, or sensory deficit.

## Diagnostics

Upon admission, arterial blood gas (pH 7.47, pCO<sub>2</sub>=27.2, pO<sub>2</sub>=56.8, HCO<sub>3</sub>=19.7) showed hypoxemia of 56.8 with PaO<sub>2</sub>/FiO<sub>2</sub> ratio of 95. Chest radiograph (Figure 2) showed bilateral lung haziness with slightly prominent pulmonary vascular markings, which may relate to beginning pulmonary congestion. Concomitant pneumonia cannot be ruled out. Persistent hypoxemia and bilateral lung haziness on chest radiograph prompted further evaluation with a high resolution CT scan of the chest.

Representative section of the HRCT of the chest in axial view (Figure 3) showed prominent “crazy paving pattern” in both lung fields due to diffuse ground glass opacities with interstitial thickening. Subpleural and parenchymal patchy areas of consolidation were also seen. Reticulonodular opacities were appreciated in the right upper lobe and minimally in the right lower lobe. Moreover, there were patchy areas of consolidation in the left lower lobe. Results of the HRCT of the chest were officially signed out as - *to consider interstitial lung disease may be secondary to collagen vascular disease, infectious, or inflammatory process.*

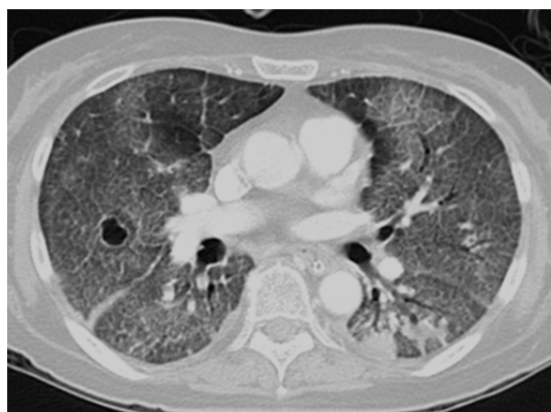


Figure 3: HRCT of the chest (axial view) showing "crazy paving pattern".

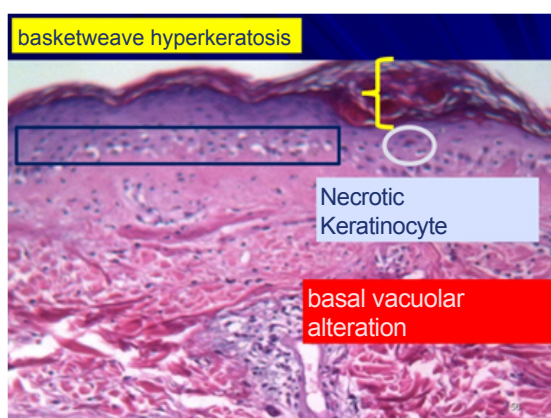


Figure 4a: Microsection of the skin biopsy specimen from the 3<sup>rd</sup> right knuckle.

Other laboratory examinations done include serum creatinine and blood urea nitrogen with normal results. Complete blood count showed anemia of 10.8 g/dL and leukocytosis with neutrophilic predominance (WBC 18,300/ Neutrophils 91%). Both Anti-dsDNA and Anti-Sm of our patient were negative. Antinuclear antibody was requested and showed insignificant titer. ECG and 2d echocardiography showed normal results.

Skin biopsy of the cutaneous lesions was done. A total of three skin punch biopsy specimens were obtained on the right elbow, dorsum of the third digit of the right hand, and abdomen. The three specimens revealed similar results. Upon closer examination of the skin punch biopsy from the third right knuckle, microsections revealed basketweave hyperkeratosis. *Basketweave hyperkeratosis* refers to thickening of the stratum corneum arranged in crisscross fashion that resembles the pattern in a woven basket. There was also basal vacuolar alteration with rare necrotic keratinocytes. In a normal skin, the basement membrane is intact, without spaces in between keratinocytes along the dermo-epidermal junction. However in this picture (Figure 4a), the white spaces or vacuoles were interspersed between the keratinocytes as seen within the blue rectangle. This can be attributed to damage on the basement membrane due to inflammation. The confluence of vacuoles in interface dermatitis may result in the formation of cleft at the dermo-epidermal junction. Necrotic keratinocytes refer to premature cornification or slow death of keratinocytes secondary to

inflammation, which may appear as dyskeratotic cells with pyknotic nuclei and eosinophilic cytoplasm.

There was markedly increased dermal mucin especially around blood vessels appearing as blue stringy materials in this section as pointed by the blue arrows. Mucin is found in between collagen bundles. The blood vessels in this section are highlighted in red (Figure 4b). There was mild superficial and mid perivascular lymphohistiocytic infiltrates with occasional extravasated erythrocytes seen. In a normal skin, infiltrates are not present or may be very minimal. Their appearance signifies changes on the papillary and reticular dermis in cases inflammation. The histopathologic diagnosis for this particular case was connective tissue disease including dermatomyositis. The three biopsies at different sites showed similar findings although varying in degrees of basal vacuolar alteration and one section showed more dyskeratotic keratinocytes. These changes however, still represented the same clinical entity.

The presence of superficial and perivascular lymphohistiocytic infiltrate, and the absence granulomatous inflammation, the skin biopsy findings were favoring with the pathognomonic skin lesions in dermatomyositis.

## Discussion

Dermatomyositis (DM) is a rare disease with overall age and sex adjusted incidence of only 9.63 per 1,000,000 populations and clinically amyopathic dermatomyositis has an overall age and sex adjusted incidence of 2.08 per 1,000,000 populations [3].

Our patient presented with rashes similar with that of DM. Muscle

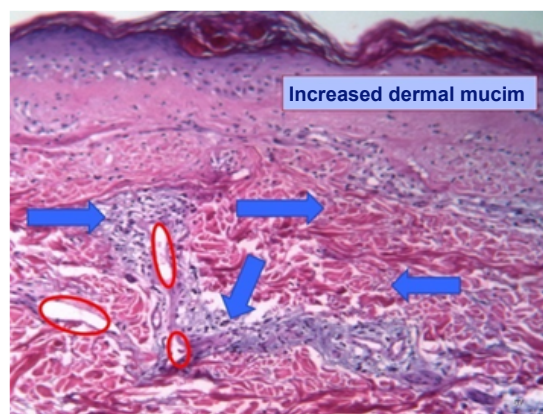


Figure 4b: Microsection of the skin biopsy specimen from the 3<sup>rd</sup> right knuckle.

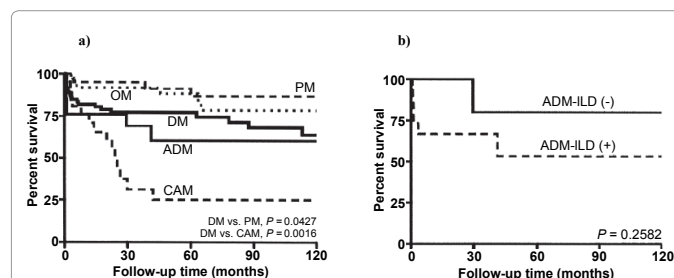


Figure 5: Survival Curves of patient with (a) Amyopathic Dermatomyositis alone and (b) Amyopathic Dermatomyositis with Interstitial Lung Disease (ILD) (Figures courtesy of Dr. Hidehiro Yamada; with permission).

biopsy, however, was not done. Creatine kinase level, the most sensitive indicator of muscle involvement in DM, was normal. Electromyography (EMG) did not show presence of myopathy. She did not present with symmetric muscle weakness. Only the pathognomonic rash was met and based on the *Peter and Bohan criteria* (Table 1), our patient cannot be diagnosed as Dermatomyositis.

In 1993, Sontheimer divided Dermatomyositis into four groups: Classic, Hypomyopathic, Amyopathic, and Juvenile [2]. Table 2 shows the symptoms, signs, and laboratory results of our patient compared with that of the different types of dermatomyositis.

According to the population-based retrospective study by Reeder et al. [4] of DM patients from 1976-2007, the following are the mean age in years at the time of diagnosis of DM. Classic, Hypomyopathic, and Amyopathic may all occur in adults, while Juvenile Dermatomyositis, could be any of the three types occurring in patients less than 18 years old. All types of DM have the hallmark skin lesions of erythematous macular rashes often with scaling over the metacarpophalangeal and proximal phalangeal joints. Proximal muscle weakness occurs in Classic Dermatomyositis, and not with Hypomyopathic and Amyopathic. Skeletal muscle enzymes particularly Total CK, is elevated with Classic and Hypomyopathic Dermatomyositis. Amyopathic patients have normal CK level. Myopathy on EMG is seen in Classic and Hypomyopathic Dermatomyositis only. Lung pathology including interstitial lung disease, respiratory muscle insufficiency, or aspiration pneumonia may occur in all types of DM.

Comparing Classic, Hypomyopathic and Amyopathic Dermatomyositis, the presentation of our patient and the laboratory

**Table 1:** Bohan and Peter Diagnostic Criteria (1975) for Dermatomyositis [1].

1	Macules and papules with erythema and violaceous discoloration on the eyelids, proximal interphalangeal joints and metacarpophalangeal joints.
2	Positive muscle biopsy
3	Elevation of muscle enzymes
4	EMG evidence with a triad of: a) short small polyphasic motor units b) fibrillation, positive sharp waves, insertional irritability
5	Proximal muscle weakness
	<i>Possible DM=rash+1 other</i> <i>Probable DM=rash+2 other</i> <i>Definite DM=rash+3 other</i>

**Table 2:** Types of Dermatomyositis and its features in comparison to that of the patient.

	Classic	Hypomyopathic	Amyopathic	Juvenile	Patient
<b>Mean age at diagnosis (years)</b>	51.9	65.9	48.5	6.8	59
<b>Hallmark skin lesions</b>	erythema of the knuckles with a raised violaceous scaly eruption, violaceous hue on the upper eyelids, a flat red rash on the face and upper trunk				+
<b>Proximal muscle weakness</b>	+	-	-	±	-
<b>Total CK level</b>	Elevated	Elevated	Normal	Normal or elevated	Normal
<b>Myopathy</b>					
	+	±	-	±	-
<b>Lung pathology</b>	Interstitial lung disease, respiratory muscle insufficiency, or aspiration pneumonia				Interstitial lung disease

Adapted from Sontheimer RD and Costner MI. Dermatomyositis in Fitzpatrick's Dermatology in General Medicine. 6th Ed. Vol 2. p1536-1553

**Table 3:** Euwer and Sontheimer Diagnostic Criteria (1993) for Amyopathic Dermatomyositis.

1.	Macules and papules with erythema and violaceous discoloration on the eyelids, proximal interphalangeal joints, and /or metacarpophalangeal joints
2.	Absence of clinical evidence of proximal motor weakness within 2 years of skin disease
3.	Normal skeletal muscle enzyme levels for 2 yrs following appearance of skin lesions
4.	Skin biopsy specimen findings compatible with dermatomyositis

results are more leaning towards the diagnosis of amyopathic dermatomyositis.

To further strengthen our diagnosis, the following are the diagnostic criteria for Amyopathic Dermatomyositis (ADM) published by Euwer and Sontheimer [2] (Table 3) in 1993. All four criteria should be fulfilled to diagnose amyopathic dermatomyositis. There are cutaneous changes pathognomonic of dermatomyositis. These are papules with a violaceous hue overlying the dorsal- lateral aspect of interphalangeal and/or metacarpophalangeal joints termed as the *Gottron's papules*, while symmetric confluent macular violaceous erythema overlying the same site of *Gottron's papules* are termed *Gottron's sign*. These skin changes were both present in our patient. She had no proximal motor weakness. Total CK level was normal. Skin biopsy was consistent with dermatomyositis, hence fulfilling the criteria for ADM.

According to Strauss et al. [5], 5-10% of patients with Dermatomyositis have pulmonary disease in the form of interstitial pulmonary fibrosis, respiratory muscle insufficiency, or aspiration pneumonia. They develop non-productive cough, shortness of breath, and dyspnea. Appearance of signs and symptoms of interstitial pneumonitis in a patient newly diagnosed with dermatomyositis should be considered a medical emergency. According to Takada et al. [6], patients with ADM may rapidly develop interstitial lung disease (ILD). Progression of ILD results to restrictive lung defect, and impaired gas exchange leading to persistent hypoxemia requiring assisted ventilation. ILD in these patients is often unresponsive to high dose corticosteroids and immunosuppressants [6]. Upon hospital admission, our patient was in respiratory distress with oxygen saturation of only 55% at room air. She was hooked to BIPAP (Bilevel positive airway pressure ventilator) and eventually intubated.

In a 30-year retrospective study by Hill et al. [3] on 618 dermatomyositis patients, patients with dermatomyositis have an increase in risk of malignant disease in the period around diagnosis and this increased risk is persistent through all years of follow-up. These findings suggest that heightened surveillance for cancer is imperative among patients newly diagnosed with dermatomyositis. According to the same study, the following are the cancer types that predominate in patients with dermatomyositis. The highest risks of developing malignancy are of the 1) ovary - during the 5-year period after the diagnosis of dermatomyositis, the risk of a female patient developing ovarian cancer was 16 times higher than the risk for women without dermatomyositis; 2) Lung, trachea, and bronchus; 3) Hodgkin's lymphoma; 4) colorectal cancer; and 5) breast.

In this present case, the patient had no palpable breast masses or skin and nipple changes. Breast mammography did not show signs of malignancy. Cancer antigen 125 (CA-125) was normal. HRCT with contrast of the chest did not show signs suspicious for respiratory tract malignancy. Our patient did not present with lymphadenopathy and CT scan of the chest and whole abdomen did not show enlarged lymph nodes. Our patient did not have changes in bowel habits [9]. Serum

Carcino-Embryonic Antigen (CEA) however was elevated. Whole abdomen CT scan only showed fatty infiltration of the liver. She was advised to undergo colonoscopy for further evaluation. However, she refused.

Our final diagnosis then is Amyopathic Dermatomyositis, Acute Respiratory Failure Type 1 secondary to Interstitial Lung Disease and Community acquired pneumonia – High Risk, to consider malignancy of the colon.

### How do we manage our patient?

Systemic glucocorticoids remain the traditional first-line therapy for dermatomyositis and early intervention with these is associated with a better overall prognosis. In adults, prednisone is given orally in divided doses of 1 to 1.5 mg/kg/day. Our patient was initially started on Methylprednisolone equivalent to Prednisone at 1 mg/kg/day [2]. In addition, the acutely ill patients, can be given intravenous pulse therapy with recommended dose of 1 gram per for 3-5 days. Our patient was given IV Methylprednisolone 1 g/day infusion for 3 days, however without significant improvement.

About 25% of patients do not respond adequately to systemic glucocorticoids and some patients cannot tolerate the side effects of prolonged high-dose glucocorticoid therapy. Therefore these patients will need additional or step-up medications. *Methotrexate* inhibits dihydrofolic acid reductase, interferes with DNA synthesis, repair and cellular replication. It is given orally starting at 7.5 mg weekly for the first 3 weeks (2.5 mg every 12 h for 3 doses), with gradual dose escalation by 2.5 mg per week to a total of 25 mg weekly. It has been successful in the treatment of patients with classic dermatomyositis and some patients affected primarily by the cutaneous manifestations of dermatomyositis. Our patient received Methotrexate 7.5mg OD every Sunday in addition to the oral Methylprednisolone, 7 weeks prior to admission.

Another immunosuppressant, Cyclophosphamide has been reported to be of benefit in treating severe complications of Interstitial Lung Disease (ILD) in patients with amyopathic dermatomyositis. It is however, given at varying doses, based on literature. Sometimes it is given together with daily prednisone, while others give it as an intermittent intravenous infusion every 3-4 weeks depending on the severity of ILD. In a study done by Yamasaki et al. [7] among polymyositis/dermatomyositis patients with progressive interstitial pneumonia, Cyclophosphamide was given 300-800mg/ m<sup>2</sup> body surface area for at least 6 times every 4 weeks, however, with inconclusive results in improving patient's symptoms such as dyspnea. According to Stone [11], among those patients with normal renal function, the recommended initial dose of intermittent Cyclophosphamide is 750 mg/m<sup>2</sup> of body surface area. The dose can even be increase up to 1 gram/m<sup>2</sup> BSA in some patients with rheumatologic disease. Our patient was given Cyclophosphamide 1000mg in 250mL 0.45% NaCl over 2-hour infusion (equivalent to about 750 mg per m<sup>2</sup> of body surface area), when there was no noted significant improvement after the IV pulse corticosteroid therapy. Cyclophosphamide was given for one dose only.

For patients with progressive ILD in spite of the combination of glucocorticoids and a second agent, a third immunosuppressive agent may be added. Intravenous immunoglobulin (IV Ig) improves not only strength and rash but also the underlying immunopathology. It has shown a significant benefit in refractory myopathy associated with dermatomyositis that is resistant to conventional therapy. However, regarding ILD, its efficiency remains unclear and only limited

information is available [9]. A dose of 2 g/kg divided over 2–5 days per course is recommended. Benefit is often short-lived (8 weeks). Repeated infusions every 6–8 weeks are generally required to maintain improvement. Our patient received intravenous immunoglobulin 2 g/kg IV infusion divided over 3 days starting on the 12<sup>th</sup> hospital day after the course of IV pulse steroid therapy and Cyclophosphamide infusion were given.

Dermatomyositis patients frequently experience the consequences of long-term use of immunosuppressive agents, including opportunistic systemic infections and even opportunistic infection-induced lymphoma (EBV). Therefore, administration of broad-spectrum antibiotics is imperative, especially in the acutely ill patients [2]. Upon admission, our patient was started on Piperacillin-Tazobactam and Levofloxacin antibiotics as treatment for pneumonia. However, she remained febrile with persistent desaturation episodes as low as 80% despite maximum ventilatory support. Antibiotics were eventually shifted to Imipenem–Cilastatin. Caspofungin was started based on blood culture growth of *Rhodotorula glutinis*.

### What is our patient's prognosis?

Both patients with clinically amyopathic dermatomyositis (ADM) and patients with primary dermatomyositis had accelerated mortality during the first year after initial presentation. ADM in particular has a drop in percent survival to 80% in less than half a year (Figure 5a) [10]. Furthermore, ADM patients with ILD deteriorate early in about 6 months with survival rate at 60% (Figure 5b). This figure shows that patients with dermatomyositis have accelerated deterioration and mortality when they develop interstitial lung disease.

In a 23-year retrospective study by Santo et al. [11] of 318 dermatomyositis related deaths in Sao Paulo, Brazil in 1985-2007, the principal associated causes of death when dermatomyositis is listed as the underlying cause of death were as follows: pneumonia (43.8%); interstitial pulmonary diseases and other pulmonary conditions (in 34.1); and septicemia (in 22.8%). Our patient expired on the 22<sup>nd</sup> hospital day due to progressive interstitial lung disease, pneumonia, and septicemia.

### Conclusion

Dermatomyositis is rare and the AMD type is even more uncommon (incidence of 2.08/1,000,000 population). Therefore, recognizing it can be a challenge. AMD patients have 80% survival in less than a year and drops to only 60% if ILD develops. And because of the long-term immunosuppression given for DM, patients often acquire infections that may further complicate management. Studies have shown that pneumonia is the leading underlying cause of death in these patients. Hence, early diagnosis and aggressive management are imperative.

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