

Dermatomyositis: An Acute Flare and Current Treatments

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ABSTRACT: The purpose of this case report is to assess and review the literature to determine the frequency of occurrence of dermatomyositis (DM). Dermatomyositis is a rare autoimmune condition that disproportionately affects adolescence and pediatric patients. The symptomatology experienced in this condition includes but not limited to fatigue, reduced mobility, and dysphagia. Symptoms of dysphonia and dyspnea have been reported due to weakened esophageal and respiratory muscle. Another major complication seen in DM is calcinosis. Calcinosis is a calcium deposit on soft tissue. This is mostly been attributed to late diagnosis or use of ineffective treatment regimen. Systemic corticosteroid is the first-line treatment for DM; however, other agents such as anti-malaria, IVIG, and immunosuppressive therapies have been used successfully.

KEYWORDS: dermatomyositis, autoimmune disease, rare skin condition, calcinosis, polymyositis

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Introduction

Dermatomyositis (DM) is a rare autoimmune condition that affects children and adults and is one of the many idiopathic inflammatory myopathies (IIM) with cutaneous involvement (predominately affects the skin and muscles). Organs such as the lungs, heart, and esophagus may be affected but to a lesser extent.^{1,2} The estimated incidence of DM is less than 10 cases per million population.^{1–4} Juvenile DM is a subset of DM and is usually diagnosed between the ages of 5 and 15 years, while the diagnosis in adults may occur between the ages of 40 and 60 years.³ DM is more common in women and maybe diagnosed in both children and adults.⁵ The disease presents as a purple-red skin rash, which is primarily seen on the chest, knuckles, neck, face, and back.³

Common characteristics of DM include proximal muscle weakness, muscle inflammation, and skin rash.^{1,2} One of the common complications of DM is dystrophic calcinosis, which is a very painful condition often seen in both children and adolescents but rare in adults.^{1,2,6} Calcinosis is a calcium deposit found in soft tissue in DM patients. Most cases of calcinosis develop within the first 3 years of diagnosis. The presence of calcinosis is indicative of delayed diagnosis, inadequate drug therapy or resistance to drug treatment, a longer duration of untreated disease, and chronic course or disease severity.^{1,2,5,6} The rate of calcinosis is up to 40% in children or adolescent who are diagnosed with DM.⁷ In pediatrics population, DM resembles the adult disease except for the frequency of the extra muscular activity.² The abnormality that occurs in childhood disease is called “misery.” Misery is an uncomfortable, irritable condition associated with facial flush, fatigue and inability to socialize, and some degree of muscle weakness and gait.²

There are 6 different types of DM; they are classic dermatomyositis (CDM), amyopathic dermatomyositis (ADM), hypomyopathic dermatomyositis (HDM), and clinically amyopathic

dermatomyositis (CADM), which evolves into classic DM (CADM → CDM) and juvenile dermatomyositis (JDM).⁸ CDM is defined as the hallmark cutaneous manifestation with signs of proximal muscle weakness after the onset of skin disease within the first 6 months.⁸ ADM is also associated with cutaneous involvement, it may occur within 6 months or greater of DM diagnosis, without any clinical or laboratory evidence of any skin or muscle disease.⁸ In HDM, there is no subjective muscle weakness especially after the first 6 months. However, a more subjective data may show subclinical evidence of disease process, and such evidence will include abnormal muscle enzyme and signs of myopathy on electromyography of muscle biopsy.⁸ Clinical DM is typically used to describe the subclinical evidence of ADM and HDM.⁸ Therefore, in clinical DM, there is evidence of skin and muscle disease. Clinically amyopathic DM evolving into classic DM is typical in patients with cutaneous DM with disease onset in the muscle 6 months prior to clinical presentation. Juvenile dermatomyositis is a subset of DM occurring in patient 18 years old or less.⁸

Polymyositis is a disease state that shares similar pathophysiology with DM. Polymyositis is an idiopathic inflammatory myopathy; however, it lacks skin involvement that is seen in DM.⁵ Polymyositis is defined as a subacute myopathy, which takes more than 4 months to manifest. Polymyositis is common in adult not in children; patients commonly present with weakness at the proximal muscles.^{2,5} Unlike DM that occur in both children and adults, polymyositis rarely affects children.⁵ Both polymyositis and DM involve primarily striated muscles, which underscores the fact that they are both systemic inflammatory connective tissue diseases.⁵ The diagnosis of both polymyositis and DM is mostly based on clinical manifestation and laboratory data that reflect muscle damage.⁹



Table 1. Bohan and Peter criteria.⁴

ITEMS/FEATURES	DIAGNOSIS	
Symmetrical proximal muscle weakness	Polymyositis Definite: all 1 to 4 Probable: any 3 of 1 to 4 Possible: any 2 of 1 to 4	Dermatomyositis Definite: item 5 plus any 3 of 1 to 4 Probable: item 5 plus any 2 of 1 to 4 Possible: item 5 plus and 1 of 1 to 4
Muscle biopsy evidence of myositis		
Evaluation in serum skeletal muscle enzyme		
Characteristic electromyogram pattern of myositis		
Typical rash of dermatomyositis		

Clinical manifest of heart problems are relatively uncommon in patients with polymyositis and DM. The commonly reported cardiac problem is congestive heart failure seen in 3% to 45% of myositis patients.¹⁰ Left ventricular diastolic dysfunction was observed in 12% to 42% of myositis compared with estimated 30% of the population.¹⁰

The actual cause of DM is unknown. However, factors such as genetic, immunologic, infectious, and environmental have all been considered as part of the cause of DM.² The pathophysiology of DM is as a result of humoral attack against the muscle capillaries and arterioles. The disease process is initiated when complement factor-3 (C3) is activated to form C3b and C4b fragments. This activation process leads to further formation of C3bNEO (C3bNEO is a neoantigen expressed on the surface of activated C3 component) and membrane attack complex (MAC), which is then deposited on the vascular tissues.² Other complement factor such as C5b-9 MAC is needed for antibody-mediated cell destruction. It is also important to note that other inflammatory markers such as B-cell and CD4 (helper) cells are present in the blood vessels. As the capillaries and blood vessels are destroyed, the muscles undergo perifascicular apoptosis, with the advancement of the disease process—necrotic and degenerative fibers formed throughout the muscles.²

Polymorphisms of tumor necrosis factor may be implicated in DM especially in European population. Abnormal T-cell has been implicated in both skin and muscle disease.² In addition, antinuclear antibodies (ANA) and antibodies to the cytoplasm may also be involved in the DM. Bacterial and viral infection human T-cell lymphotropic virus type 1 (HTLV-1), *Toxoplasma specie*, and *Borrelia specie* have been associated with DM. Drugs such as hydroxyurea, statins, interferon, cyclophosphamide, quinidine, and anti-tumor necrosis factor drugs (but not limited to this list) have been linked to induce DM.²

Literature Review

The estimated incidence of DM for all subtypes was 9.63 per 1 000 000 adjusting for age and sex.⁸ Based on the study by Reeder and colleagues, the prevalence of DM was estimated to be 21.42% per 100 000 of all subtypes, adjusting for age and

sex.⁸ The reported incidence for new cases ranges from 1.1 to 17 new cases per 1 000 000 inhabitants.¹¹ The incidence of malignant disease in DM varies between 6% and 60%, with men and older population reporting higher occurrence. It has also been observed that polymyositis carries slightly higher rate of malignancy compared to DM.⁷

Some of the general clinical symptomology for most patients presenting with DM involve muscle weakness of the trunk and hip girdles. An erythematous, scaly rash usually precedes muscle weakness or accompanies it.¹ The common complaints reported in this disease are fatigue and reduced endurance.⁴ In some severe cases of DM, dysphagia, dysphonia, and dyspnea have been reported due to weakened esophageal and respiratory muscle.⁴ However, these rarely lead to respiratory failure.⁴ As the disease process progresses, some patients may develop difficulties with activities of daily living.¹

Other dermatologic features associated with DM include erythema in sun-exposed skin, Gottron papules (raised erythematous papules on the surface of the skin), or a heliotrope rash (a bluish-purple eruption affecting the periorbital region). Gottron papules and a heliotrope rash are diagnostic features of DM.^{1,3,4} Another cutaneous finding seen in most patients is mechanic's hands (hyperkeratotic, fissured skin on lateral fingers and palms), which is expressly associated with antisynthetase antibodies in myositis patients.^{1,3,4} The thickened cuticles and periungual erythema are related to small vessel inflammation. The inflammatory process may be due to dilated capillary loops, which is identified on nail-fold capillaroscopy.⁵ Panniculitis, erythroderma, and diffuse or patchy alopecia with erythema are other cutaneous manifestations associated with DM.¹²

Diagnostic criteria

The Bohan and Peter¹³ criteria are the most widely used criteria for the diagnosis of DM; however, it has minor drawback of being too rigid (Table 1). Today, only a few of these criteria are used in the diagnosis of DM in addition to abnormal lab values. As a complement to the Bohan and Peter Criteria, a nail-fold capillaroscopic (NFC) test may be used to assist with the diagnosis of JDM.⁶ Scleroderma (SD) alterations consist of the

presence of capillary deletion associated with capillary ectasia and or giant capillaries; note that alterations in the SD constitute an NFC change.⁶

The gold standard of the imaging study of muscle is the use of magnetic resonance imaging (MRI). The MRI gives a detailed anatomic overview of muscle damage or disease and is very sensitive in detecting edematous muscular changes in active myositis. The use of contrast-induced ultrasound (US) may be useful tool in detecting muscle lesion and other complications such as fibrosis, cystic hematomas, or ossification.¹¹ High serum levels of muscular enzyme such as serum creatine kinase which is released during muscle damage is the most sensitive muscle enzyme during the acute phase of the DM disease process.¹¹ Nonspecific inflammatory biomarkers such as erythrocyte sedimentation rate and C-reactive protein may be elevated during the acute phase of the DM disease process.¹¹ Generally, pathological finding may consist of some inflammatory infiltrates such as B-cell, macrophages, and CD4+ cells. Autoantibodies, such as the anti-Mi-2 antibodies, have been shown to be closely associated with dermatomyositis. Anti-Mi-2 is the most common myositis-specific autoantibodies found in patients diagnosed with DM.¹¹

Goal of therapy

The goal of treatment is to reduce inflammation and vasculitis and invariably minimize symptomatology and improve quality of life of the patients. To achieve this goal, treatment regimen must be initiated early in the disease process and will require an interprofessional approach to achieve the goal of therapy.^{1,14}

Prognosis

Prior to the use of steroids, the prognosis of patients with DM is poor. The morbidity due to DM is widespread and has increased over the years. Only 20% to 40% of patients achieve remission, whereas 60% to 80% of patients with DM experience polycyclic or chronic continuous course of the disease.³ The overall mortality ratio is threefold higher in patients with DM versus the general population.³ Death may result from prolonged muscle weakness and malnutrition.³ The most common causes of death are cancer, lung, cardiac complications, and infections. Older age, lung and cardiac system involvement, cancer, and dysphagia are predictive factors for a poor prognosis.¹⁵ There is a well-established link between DM and malignancy.¹⁵ The most commonly reported malignancies associated with DM are ovarian, gastric cancer, and lymphoma.¹⁴ Other associated malignancies include lung, male genital organ, nonmelanoma skin, Kaposi sarcoma, mycosis fungoides, and melanoma.¹⁴

Pharmacotherapy

The mainstay of therapy for dermatomyositis is the administration of steroids.^{2,16-18} In addition, dose range for prednisolone is

often between 0.5 and 2 mg/kg/day for initial treatment.¹⁹ The treatment of choice is high dose of oral prednisone which must be initiated early to improve muscle weakness.^{1,15} Steroid therapy should result in symptom relief within 4 weeks of treatment after which time the steroid dosage should be slowly tapered over a 10-week period to 1 mg/kg every other day.^{1,16-19} In situations where prednisone cannot be used, second-line agents such as methotrexate and azathioprine will be appropriate. Rituximab, intravenous immunoglobulin (IVIG), and other biologics are useful in patients who developed resistance to therapy. Antipruritics, topical steroids, hydroxychloroquine, and steroids may be employed to treat superficial skin disease.¹⁴

However, for patient experiencing steroid-related toxicity, a steroid-sparing agent has been found to be beneficial in this group of patients.^{2,19-23} Some examples of the steroid-sparing agent are methotrexate, azathioprine, cyclophosphamide, cyclosporine, mycophenolate mofetil, leflunomide, chlorambucil, and tacrolimus.² Table 2 contains the list of pharmacotherapy for managing dermatomyositis.

It is important to note that the first presentation of dermatomyositis is a skin disease. It is therefore important to initiate skin protective agents, which include the use of protective clothing and the application of sun screen with at least 15 sun protection factor (SPF). This is because exposure to sunlight worsens the cutaneous aspect of the disease.² In a small study, antimalarial medicines such as hydroxychloroquine and chloroquine have been shown to be useful in the management of dermatomyositis.^{2,26,27} Unfortunately, some patients may develop drug eruption with antimalarial agents.²⁸ In the scenario of drug eruption, methotrexate has shown to be helpful in patient refractory to antimalarial agents.²

Azathioprine and IVIG are effective agents in the management of muscle involvement of dermatomyositis.² A retrospective study compared subjects treated with conventional immunosuppressive therapy plus IVIG (as an add-on therapy) on one arm, while the other arm comprised subjects treated with conventional immunosuppressive therapy alone.^{2,29} The study showed that at 6 months, both cutaneous and muscle involvement significantly improved in the IVIG add-on therapy compared with the conventional treatment alone. The study further showed that the modified Cutaneous Dermatomyositis Area and Severity Index (CDAI) also improved significantly.^{2,29}

Calcinosis complication is a very painful complication, but early diagnosis and management may help alleviate the suffering associated with it.² Calcium channel blockers especially non-dihydropyridine such as diltiazem has been beneficial in the management of calcinosis.² Tender lesions of calcinosis may be surgically removed especially if the calcinosis is localized. A retrospective study of small groups of patients found that the use of surgical and pharmacotherapy (diltiazem) was effective in reducing dystrophic calcinosis.³⁰

Table 2. Treatment modalities for dermatomyositis.^{7,14,16-24}

TREATMENT MODALITY	MECHANISM OF ACTION	DOSAGE	SIDE EFFECTS	COMMENTS
Oral prednisone	Synthetic glucocorticoid analog, used for anti-inflammatory effects and modifies immune response to diverse stimuli	0.5 to 1.5 mg/kg by mouth daily until serum creatine kinase normalizes, and then slowly taper over 12 months	Gastrointestinal symptoms, adrenal suppression, immunosuppression, avascular necrosis, and osteoporosis	Initial pharmacologic agent considers adjunctive therapy if no improvement is seen in using objective data. For example, muscle strength after 3 months of therapy
Methotrexate oral (Rheumatrex)	Antimetabolite that interferes with DNA synthesis, repair and cellular replication by inhibiting dihydrofolate reductase	7.5 to 10 mg per week, increased by 2.5 mg/week by mouth, total of 25 mg/week Intravenous: 10 mg/week, increased by 2.5 mg/week to total of 0.5 to 0.8 mg/kg Children: 1 mg/kg. As dosage increases, taper off steroid dose. Patients should be given 3 mg daily by mouth of folic acid to minimize side effects of methotrexate	Stomatitis, hepatic fibrosis, cirrhosis, nausea, abdominal pain, neutropenia, thrombocytopenia, pruritus, fever, pneumonitis, and gastrointestinal symptoms	First-line adjuvant therapy in patients unresponsive to steroids
Azathioprine (Imuran)	Suppresses cell-mediated hypersensitivities and causes alterations in antibody production. Acts as an immunosuppressive antimetabolite	2 to 3 mg/kg/day tapered to 1 mg/kg/day once steroid is tapered to 15 mg/day. Reduce dosage monthly by 25 mg intervals Maintenance dosage is 50 mg per day	Lymphoma, nausea, vomiting, hepatotoxicity, leukopenia, oral ulcers, thrombocytopenia	Screen patients for thiopurine methyltransferase deficiency before therapy (usually seen in 0.3% to 11% of White population)
Cyclophosphamide (Cytoxan)	Nitrogen mustard-type alkylating agent whose cytotoxic action is primarily due to cross-linking DNA and RNA strands and inhibits protein synthesis	Oral: administer 1 to 3 mg/kg/day Intravenous: administer 2 to 4 mg/kg/ day, in conjunction with prednisone	Increased risk for malignancy, leukopenia, thrombocytopenia, hemorrhagic cystitis, anorexia, nausea, vomiting, alopecia, sterility, congestive heart failure, and stomatitis	In refractory cases only

Table 2. (Continued)

TREATMENT MODALITY	MECHANISM OF ACTION	DOSAGE	SIDE EFFECTS	COMMENTS
Cyclosporine (Sandimmune)	Inhibition of production and release of interleukin-2, a proliferative factor necessary for the induction of cytotoxic T lymphocytes in response to alloantigenic reaction	2.5 to 10 mg by mouth per kilogram per day ^a	Impaired T-cell proliferation, nephrotoxicity, lymphoma, hypertension, hypertrichosis, gingival hyperplasia, hepatotoxicity, paresthesias, fatigue, hyperesthesia, depression, and seizures	Maintaining whole blood level of 200 to 300 ng/mL may have rapid response to therapy
Hydroxychloroquine (Plaquenil)	Interferes with antigen processing by increasing the pH where acidity is required to assemble chains of MHC class II proteins, thereby diminishing the formation of peptide-MHCs resulting in the downregulation of immune response against autoantigenic peptides	Adult: 200 mg twice daily by mouth Children: 2 to 5 mg/kg/day by mouth	Myopathy, hematologic toxicity, hepatotoxicity, antimalarial retinopathy, dizziness, ataxia, and weight loss	Adjunctive topical steroid to treat psoriasis rash. A moderate potency corticosteroid (IV) such as Desoximetasone cream. ²⁵
Intravenous immunoglobulin	Immunomodulatory effects on T cells, macrophages, and B-cell immune function and its regulatory action on membrane-damaging components of the complement system. Specific antibodies that are capable of neutralizing bacterial or even viral toxins that can have profound effects on the host's immune and inflammatory systems	2 g/kg in divided doses once per month for 3 months	Pancytopenia, death, lymphoma	Showed improvement in 70% of patients; limited by high cost
Topical steroids	Induction of phospholipase A2 inhibitory proteins which control biosynthesis of mediators of inflammation such as prostaglandins and leukotrienes by inhibiting their precursor arachidonic acid	Class I (super-high potency) or class II (high potency) topical steroid is recommended		For further control of the erythematous and pruritic skin changes

Abbreviations: MHC, major histocompatibility complex; IV, intravenous.

^aCyclosporine dosing is highly subjective; it is used only as an adjunct to oral steroid therapy in a dosage of 2.5 to 10 mg/kg/day and then tapered to the lowest effective dosage over 2 weeks (information from Kovacs and Kovacs²⁴).

A case series report by Shehata et al showed a 69% rate of inflammatory infiltrate which included intestinal vasculitis as one of the complications found in juvenile dermatomyositis. It was also found that diffuse vasculitis was common in patients who had complications of calcinosis especially in patients who had 6 months of active disease.³¹ In this case series study, there was no death as a result of intestinal vasculitis. In another case report of 48 patients with the diagnosis of DM, 6% of the study population was identified with severe gastrointestinal tract disease which carries high rate of fatal complication. However, the study did not conclude that gastrointestinal complication was the exact cause of death.³² A retrospective chart review of 676 patient record with idiopathic inflammatory was performed, 49 of those patients were diagnosed with dermatomyositis while 10 patients had the diagnosis of polymyositis.³³ An analysis of cause of death was performed using clinical and pathological symptoms. Based on the analysis, the major cause of death was associated infection (34%), tumor (4%), viral hepatitis (3%), and acute intestinal lung disease (2%). Therefore, infection is considered to be the primary cause of death in idiopathic inflammatory myopathy.³³

Patient Presentation

A 22-year-old African American female presents with juvenile dermatomyositis (diagnosed since the age of 6 years). On arrival at the emergency department (ED), her chief complaints were fatigue, inability to ambulate, and a nonhealing leg wound. Past medical histories include hypertension and a heart murmur.

Home medication regimens are hydroxychloroquine 200 mg by mouth twice daily, prednisone 50 mg by mouth daily, folate 3 mg by mouth daily, and methotrexate 7.5 mg by mouth weekly. All the home medications were continued at the hospital. In addition, the dermatologist ordered the following topical medications: hydrocortisone butyrate cream 0.1%, to apply to all affected areas of the face daily; triamcinolone acetonide lotion 0.1%, to apply to the affected skin or the trunk daily; and fluocinonide acetonide ointment 0.025%, to apply to scalp daily. Scalp oil was also applied nightly to the scalp. The patient was also encouraged to maintain moisturization with Eucerin or Aquaphor. The nutritionist added Boost and Juven supplements to her nutrition.

Her vital signs on admission were as follows: blood pressure 122/68 mmHg, pulse 77 beats per minute, temperature by oral source 97.6°F (36.4°C), respiratory rate 16 breaths per minute, height 1.575 m (5'2"), weight 45.13 kg (99 lb 8 oz), and SpO₂ 98%.

On physical examination, patient appeared sick and cachectic and stated that she had fatigue, right ear pain, and sore throat. She admitted experiencing shortness of breath (SOB), chest pain, and diarrhea. She had a splitting of the second heart sound and 2/6 systolic murmur. Other findings include rash, tingling, visual change, weakness, and headaches.

There were some facial skin rash and alopecia, which was of concern to the patient. There was a "box"-shaped rash on the

chest. The rash was red and scaly covering the central chest region from right to left clavicle extending down below her right breast dissecting her right areola across the sternum to her mid left breast up to her clavicle. Other than tenderness in the suprapubic region, her abdominal exam was unremarkable. The patient displayed atrophy and exhibited abnormal muscle tone with worsening weakness to immobility mainly in the lower extremities.

She had a 3 cm × 7 cm, nonhealing, and nonpruritic ulcer on the dorsal left thigh. In addition, the patient presented with diffused xerosis, hypopigmentation periorally along with linear streaks of hypopigmentation on her face, and some areas of hyperpigmentation on her cheeks, periorbital heliotrope sign. Her scalp displayed erythema and some focal areas of alopecia. The patient's chest demonstrated poikilodermatous changes, erythematous, and atrophic. Her hands had Gottron papules and dilated nail-fold capillary loops without clubbing.

Three years before this admission (see Figure 1), the patient had a calcium (calcinosis) deposit removed from her dorsal left thigh that produced a nonhealing wound and received wound care with multiple treatments without resolution. Three weeks before the admission, the patient coughed up a calcium deposit and was then taken to ED and later released. Intravenous antibiotics she received at that time resulted in the healing of the wound on her dorsal left thigh for the first time in 3 years, but it was inadvertently reopened due to the use of compression cuffs on her legs while in physical therapy. At that time she was issued walker for her mobility.

The patient stopped taking her medications due to pregnancy and at the recommendation of her rheumatologist. During this time, she was diagnosed with preeclampsia, underwent a Cesarean-section, and was intraoperatively transfused with platelets and blood. After she had her baby, she was then restarted on prednisone and hydroxychloroquine.

Four months before her current presentation in the ED, the patient began feeling weak and started experiencing SOB on exertion. She had also exhibited intermittent chest pain throughout the day which was not associated with exertion. She described the pain as paralyzing when lying on her back, and lasting roughly 15 minutes each time. Shortly after the chest pain subsided, she began experiencing episodes of muscle stiffness and cramping that lasted approximately 30 minutes in duration. She also began losing strength, having falls due to weakness, and SOB at rest. Eventually, the patient lost her ability to ambulate due to muscle weakness, and she began experiencing severe throbbing right ear pain. Three days before this admission, the patient was advised by her new rheumatologist to go to the ED for wound management. As she was leaving the ED, she fell and developed a large lump on her right hip.

Cancer is considered one of the common complications of dermatomyositis. During this hospital stay, the patient had a skin punch biopsy on the left axilla to rule out skin cancer which was negative. The patient experienced new onset

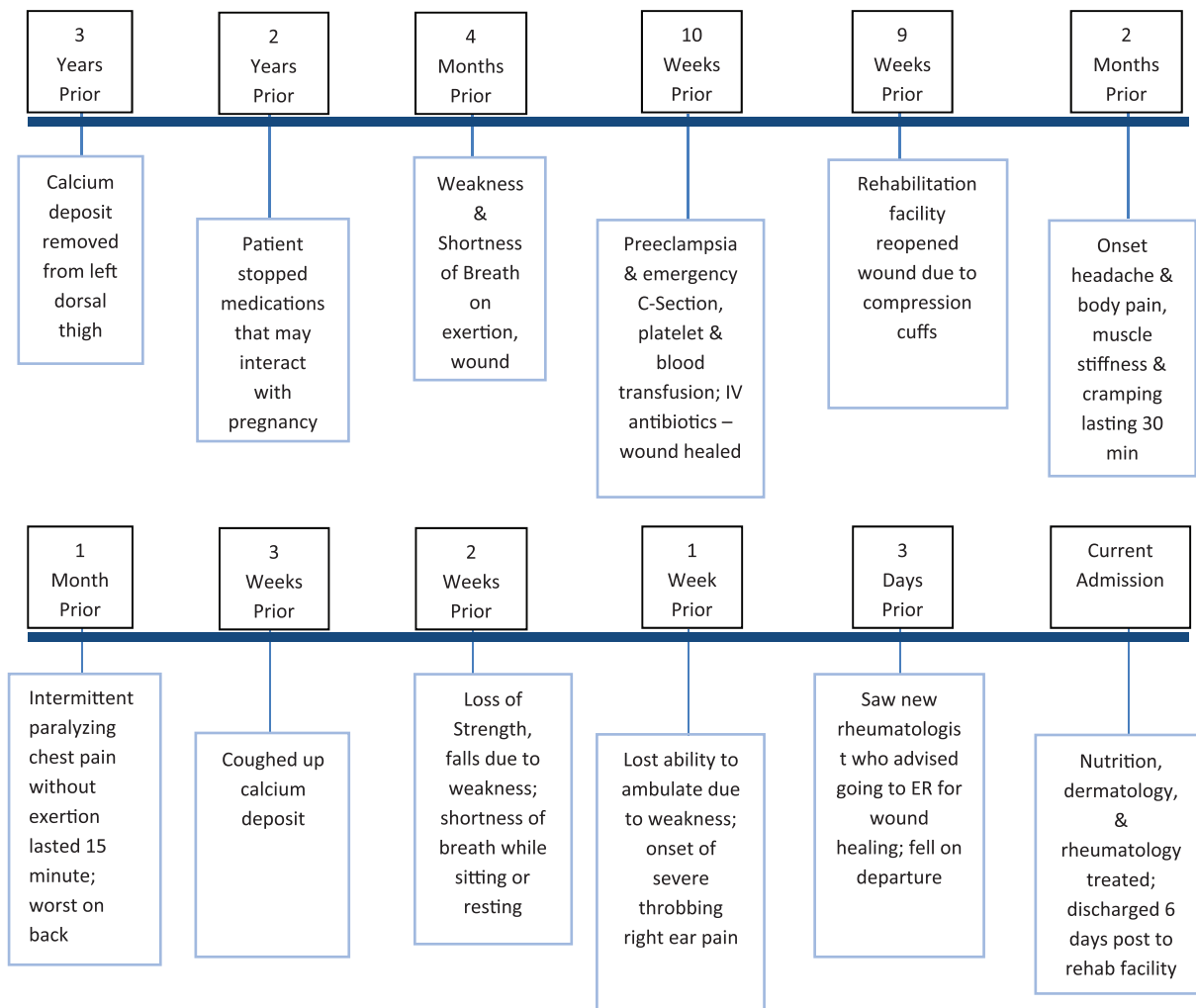


Figure 1. Events leading up to current admission. ER indicates emergency room.

pancytopenia, which the hematologist attributed to methotrexate and Plaquenil toxicities.

Summary and Conclusions

The patient was diagnosed with DM at the age of 6 years, at which time, she recalled loss of ambulation. Based on her medical history, patient has shown significant hallmark signs of dermatomyositis such as calcinosis, heliotrope rash, hypopigmentation, and hyperpigmentation. In addition, she exhibited substantial decrease in activities of daily living with DM. As seen in Figure 1, there were multiple episodes of weakness leading to this patient's hospitalization. Daily tasks that involve the use of proximal muscles such as climbing stairs, combing hair, lifting objects, and getting up from a chair all became increasingly more difficult and impossible.

The treatment of dermatomyositis involves a multidisciplinary approach, as evidenced in this case. When patients are appropriately managed using pharmacologic and nonpharmacologic therapy, the outcome and prognosis are better. Currently, there is no single pharmacologic or nonpharmacologic therapy for the management or treatment of dermatomyositis. It is

important to note that this rare condition is very debilitating to patient and care givers.

Author Contributions

JO: wrote and edited the case report and the entire body of the article, guided the original literature search. JO: provided the necessary recommendations and corrections by the reviewers. LB: did the original literature search and summary of the search.

Informed Consent

Consent was obtained from the patient and hospital for this case report.

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