



The systemic management of cutaneous dermatomyositis: Results of a stepwise strategy☆☆☆



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ABSTRACT

Treatment of dermatomyositis (DM) is often achieved with a stepwise algorithm. However, the literature lacks quality evidence to support the use of this therapeutic strategy. The result of a stepwise therapeutic strategy in the management of skin-only DM is presented to better understand the clinical outcomes and allow for future studies. A cohort of 102 patients with DM, 41 of whom had skin-only disease, were seen between July 2009 and April 2013 at a referral-based connective tissue disease clinic. The Cutaneous Dermatomyositis Disease Area and Severity Index was used to prospectively assess disease severity and the outcomes in 41 adult patients with skin-only DM were analyzed. Of the 41 patients with skin-only DM, 23 patients (56.1%) received antimalarial medications alone and 18 patients (43.9%) received second- or third-line agents. Ten patients (24.4%) remained at the first level of the treatment algorithm and received only hydroxychloroquine. Prednisone was included in the treatment regimen for 11 patients with skin-only disease (26.8%). The results show that management of cutaneous DM often requires second-line agents because antimalarial medications alone are insufficient to treat most patients with skin-only disease.

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Introduction

Dermatomyositis (DM) is an idiopathic inflammatory myopathy that is characterized by varying degrees of skin disease with or without muscle involvement (Sontheimer 1999). Classic DM presents with typical skin findings, proximal muscle weakness, and evidence of myositis. Clinically amyopathic DM refers to patients with the characteristic skin findings but the absence of muscular weakness and evidence of myositis. The term “cutaneous DM” refers to the skin findings in patients with either classic or clinically amyopathic DM and “skin-only DM” refers to the subset of patients with clinically amyopathic DM who have no lung involvement. Patients with skin-only DM made up the final cohort in our study with 41 participants.

DM requires a multifaceted approach to treatment that considers the involved organs, potential adverse effects of medications, patient preference, and comorbidities. Treatment of the myositis component is accomplished with systemic corticosteroid medications, typically combined with a cytotoxic drug. The response of muscle and skin disease to systemic therapy is often discordant. Cutaneous manifestations of DM appear more refractory to treatment (Sontheimer 2004). Although the disability that is associated with myositis causes great morbidity, the effects of pruritus, visible skin lesions, and photosensitivity on quality of life (QoL) correlates with the severity of disease and is greater than in other chronic dermatologic and non-dermatologic conditions (Goreshi et al. 2011; Hundley et al. 2006).

The literature lacks strong evidence for the use of most agents in the treatment of patients with cutaneous DM. A therapeutic ladder on the basis of retrospective studies, case reports, and expert opinions proposes aggressive sun protection, topical approaches, and antipruritic and antimalarial medications as a first-line therapy for patients with cutaneous DM (Dawkins et al. 1998; Iorizzo and Jorizzo 2008; Lam and Vleugels 2012; Sontheimer 2004). If adequate control is not achieved with this first step, second line agents such as methotrexate (MTX), mycophenolate mofetil (MMF), or azathioprine (AZA) are added. Patients who are refractory to treatment with these agents

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may receive intravenous immunoglobulin (IVIg) treatment. Dapsone, thalidomide, rituximab, and calcineurin inhibitors may also be used. There are a few reports on the use of nondrug therapies such as stem cell transplantation, plasmapheresis, and total body irradiation for patients with refractory cutaneous DM (Lam and Vleugels 2012). This study employs a prospective database and the Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI) to report on the quantitative results with this therapeutic strategy.

Methods

Patient population

Patients at a referral-based dermatology clinic were screened and data from eligible patients were added to a DM database. Patients over age 18 years with clinical and histological evidence of adult-onset DM and who provided written consent were included regardless of their treatment status. At each study visit, patient clinical information was collected and a skin assessment was completed with the CDASI. Prospectively gathered CDASI scores and historically gathered treatment data were stored in a

Research Electronic Data Capture online database. This study was approved by the University of Pennsylvania Institutional Review Board.

Treatment algorithm

Patients with cutaneous DM were evaluated by the senior author (VPW) and the data managed with a stepwise algorithm (Fig. 1). Patients with predominantly skin disease initially received antimalarial medications. Most patients initiated hydroxychloroquine (HCQ) at a dose of ≤ 6.5 mg/kg/day for 8 weeks. Quinacrine 100 mg/day was added if an adequate trial of HCQ was ineffective to control disease activity. Chloroquine ≤ 3.5 mg/kg/day was used in lieu of HCQ for patients who had a previous reaction to HCQ or for those patients in whom HCQ was ineffective. If symptoms progressed despite use of antimalarial medications for 8 weeks, treatment was escalated to include a cytotoxic agent such as MTX, MMF, or AZA. If adequate control was not achieved with one cytotoxic agent at a maximum dose for 8 weeks, another was substituted. The use of IVIg, dosed at 2 g/kg administered over two to five days each month, was considered if antimalarial medications and cytotoxic drugs were ineffective. For refractory cases, oral calcineurin

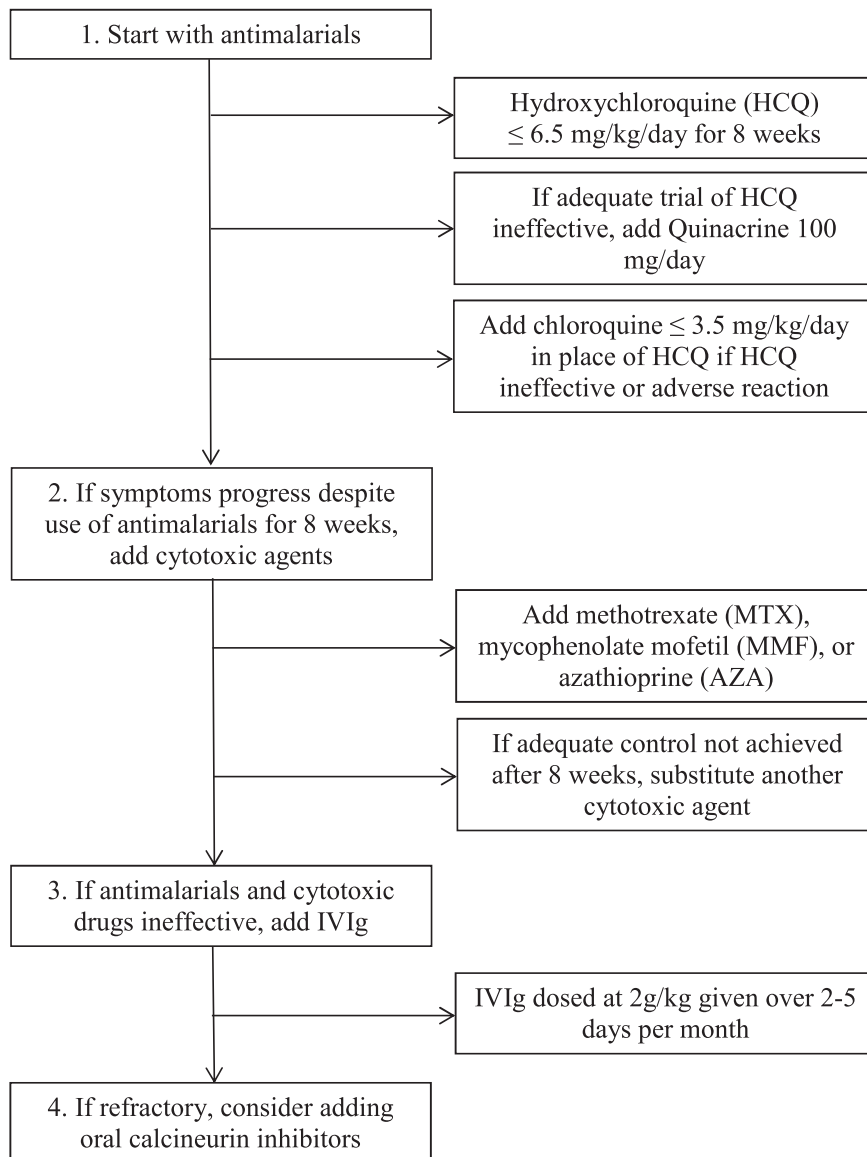


Fig. 1. Treatment algorithm.

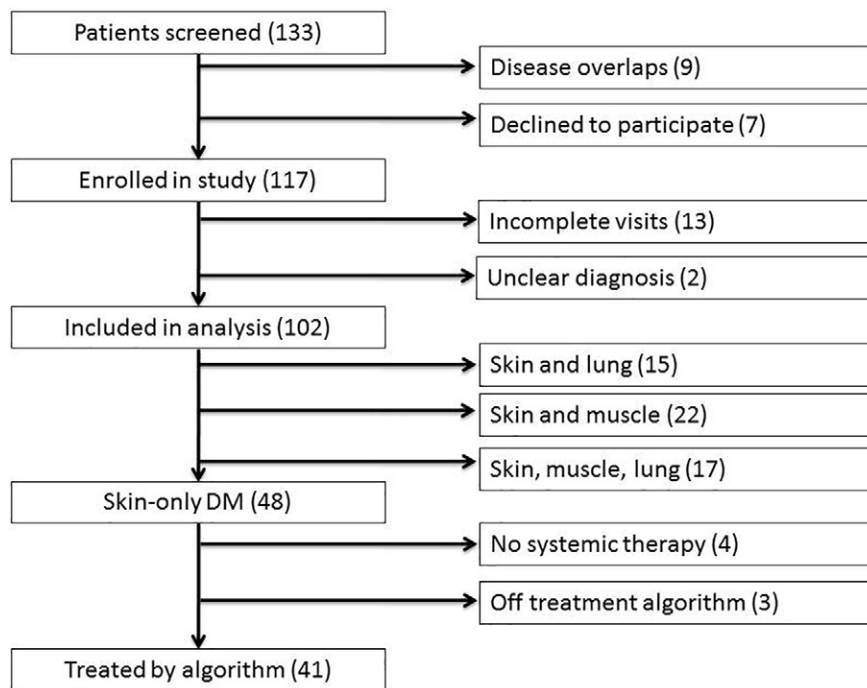


Fig. 2. Selection of study participants.

inhibitors were an option. Systemic corticosteroid medications were not routinely used in the treatment of patients with cutaneous DM but a course of medium-to-high dose corticosteroid medications was used to provide relief in cases of severe disease. Lastly, topical agents such as corticosteroid or immunomodulator medications were commonly used by participants but not included in the algorithm because they are not considered a systemic treatment.

Cutaneous dermatomyositis disease area and severity index

The CDASI is a validated tool for the assessment of cutaneous DM that provides a disease activity score with a range from 0 to 100 (Goreshi et al. 2012; Hornung et al. 2012; Klein et al. 2008; Yassaee et al. 2010). A recent study showed that at the University of Pennsylvania, mild DM is defined as CDASI activity scores of 19 or less but scores of 20 or more are representative of moderate-to-severe cutaneous DM (Anyanwu et al. 2013). A clinically relevant improvement in disease was measured by a 4-point change in CDASI scores (Anyanwu et al. 2015). In this current study, the CDASI is used prospectively to capture disease severity at each visit.

Classification of patients

Patients were classified by the presence of clinically evident skin, lung, and/or muscle disease at any time throughout their disease course. Four disease categories were defined: skin-only, skin and lung, skin and

muscle, and skin, muscle, and lung (Fig. 2). No patients had cardiac involvement. The presence of lung disease was determined by a low-diffusing capacity for carbon monoxide (i.e., <80% predicted) on pulmonary function tests in the absence of anemia or pulmonary hypertension, and compatible findings on high resolution computed tomography scans of the lung. Objective muscle weakness, muscle enzyme elevations, and abnormal electromyograph study results were indicative of muscle involvement. Patients who required systemic therapy for lung or muscle disease were excluded (Fig. 2). Patients who were included in the current study had none of these abnormalities including muscle weakness or muscle enzyme elevations. The aforementioned treatment algorithm was applied to patients with skin-only DM.

Patients were stratified by the most aggressive treatment required. Each patient was included in one of four groups (Table 1). Patients who received other agents in the past and discontinued their use before enrollment in the study were included in the group that best described their therapeutic regimen while in the study. The use of prednisone was not a reason for exclusion from any treatment group but the dose and duration of use were noted. The goal was to use adjunctive therapies to minimize and eventually discontinue the use of prednisone.

Statistical methods

Data was exported from the Research Electronic Data Capture database and analyzed with Microsoft Excel and GraphPad. Frequency

Table 1
Treatment of DM in 41 patients with skin-only disease

Treatment Group	Skin-only DM n (%)	Received Prednisone	Duration of Current Treatment Months, IQR	Median CDASI Score at Last Visit
Hydroxychloroquine	10 (24.4)	3	17.5 (6.5-54.8)	17.5 (10.3-22.8)
Other antimalarial medications	13 (31.7)	2	21.0 (14.0-52.0)	13.0 (4.5-18.0)
Antimalarial medications with cytotoxic drugs	16 (39.0)	5	23.0 (5.0-45.5)	15.0 (9.0-31.0)
IVIg	2 (4.9)	1	49.5 (22.0-77.0)	6.5 (6.0-7.0)
Summary	41	11	24.0 (7.0-50.0)	13.5 (6.5-26.0)

CDASI = Cutaneous Dermatomyositis Disease Area and Severity Index; DM, dermatomyositis; IQR, interquartile range; IVIg, intravenous immunoglobulin.

and percentage for categorical variables and median and interquartile range (IQR) for continuous variables were computed.

Results

Part 1–102 participants

Of the 133 patients who were screened in the dermatology clinic between July 2008 and April 2013, 9 patients had an overlap with another autoimmune disease or the diagnosis of DM was not clear, 7 patients declined to participate, and 117 patients were eventually enrolled in the study (Fig. 2). One hundred and four of these participants completed at least one visit and two participants were excluded from the analysis because they were found to have diagnoses other than DM. A total of 102 patients were included in the analysis (Table 2). Of all participants, 26 patients (25.5%) presented to their first clinic visit without a diagnosis, 46 patients (45.1%) with a diagnosis of DM, and 28 patients (27.5%) had been previously diagnosed with other diseases including cutaneous lupus erythematosus (CLE), psoriasis, undifferentiated connective tissue disease, and rosacea. Thirty-three patients completed one visit and 69 patients completed more than one visit. For all patients, the median duration of follow-up was 13 months (IQR, 0–31).

Seven of 102 patients (6.9%) did not receive any systemic therapy during the study period. Four patients had skin-only disease, two patients had skin and lung disease, and one patient had skin and muscle disease. The median final CDASI score for all patients in this group was 12.0 (IQR, 6.0–26.0). The patients with skin-only disease had a median initial CDASI score of 19.5 (IQR, 12.0–27.3) and a median final CDASI score of 13.5 (IQR, 7.5–24.0). Four of the seven patients were treated in the past but did not require systemic therapy while enrolled in the study. Three patients did not require treatment throughout their disease course.

A total of 27 adverse reactions (Table 3) to medications that were administered for DM treatment were noted in 20 of 102 patients (19.6%). Of the 84 patients who received antimalarial medications at any point during the course of their disease, 11 patients (13.1%) developed a drug eruption with the use of antimalarial medications. Eight of these reactions were to HCQ. Six of 11 patients who

Table 2
Patient characteristics

Patient Characteristic	n (%)
Sex	
Female	86 (84.3)
Male	16 (15.7)
Race/Ethnicity	
Caucasian	91 (89.2)
African American	5 (4.9)
Hispanic/Latino	4 (3.9)
Asian	2 (1.9)
Tobacco Use	
Never	60 (58.8)
Past	38 (37.2)
Current	4 (3.9)
Diagnosis	
Clinically Amyopathic DM	63 (61.7)
Classic DM	39 (38.2)
Disease severity	
Mild	57 (55.9)
Moderate-to-Severe	45 (44.1)
	Median (IQR)
Age at onset, y	48 (38–57)
Disease duration, y	4 (2–10)

DM, dermatomyositis; IQR, interquartile range.

Table 3
Adverse reactions

Medication	Adverse Reaction Observed Incidences (n)
Hydroxychloroquine	Rash (8) Gastrointestinal disturbance (2) Transaminitis (1) Hair loss (1) Mood disturbance (1) Headaches (1)
Quinacrine	Rash (1) Body aches (1) Fatigue (1)
Chloroquine	Rash (1) Visual disturbance (1)
Methotrexate	Transaminitis (2) Hair loss (1)
Azathioprine	Gastrointestinal disturbance (2) Transaminitis (1)
Intravenous immunoglobulin	Cerebral vascular accident (1)

experienced a drug eruption subsequently received and tolerated other antimalarial medications. In the other five cases, another antimalarial medication was not prescribed. Prednisone was used to treat skin, muscle, or lung disease in 47 of 102 patients (46.1%).

Part 2–41 participants with skin-only disease

Eleven patients (26.8%) with skin-only disease received prednisone (Table 1). The median period of prednisone use throughout their treatment course was 15 months (IQR, 6.5–24). The median maximum dose was 20 mg/d (IQR, 10–55) and the median current dose was 2 mg/d (IQR, 0–5). Four of 11 patients received doses that were greater than 5 mg/d at any time during their treatment course and only 2 patients were administered doses that were greater than 5 mg/d for more than 2 months.

Hydroxychloroquine

Ten patients with skin-only disease were managed with HCQ alone (Table 1). HCQ was insufficient for four patients and thus, quinacrine was prescribed at the time of their last study visit. The four patients for whom treatment with HCQ was insufficient had a median treatment duration of 27.5 months (IQR, 6.0–61.5) and median final CDASI score of 20 (IQR, 11.5–24) before the addition of quinacrine. Since 4 of the 10 patients in this group required an escalation in therapy, only 6 of 41 patients with skin-only DM (14.6%) who required systemic therapy were controlled with HCQ alone.

Other antimalarial medications

Of the 41 patients with skin-only DM, 13 patients were managed with other antimalarial medications, 12 of whom received these agents for more than 8 weeks. The median final CDASI score for the 12 patients with skin-only disease who received adequate trials of antimalarial medications was 11.5 (IQR, 4.5–16.5). Nine of the 13 patients with skin-only disease (69.2%) in this group were managed with HCQ and quinacrine for 29 months (IQR, 18.0–52.0). The last recorded CDASI score for patients administered this combination was 13 (IQR, 4.5–18.0). The remaining four patients with skin-only disease in this treatment group were treated with chloroquine (2 patients), chloroquine with quinacrine (1 patients), and quinacrine alone (1 patient). Approximately 31.7% of patients with skin-only disease who required systemic therapy were maintained at this level of the treatment algorithm.

Antimalarial medications with cytotoxic agents

Of the 41 patients with skin-only DM who required systemic therapy, 16 patients (39.0%) were managed with a combination of antimalarial medications and cytotoxic agents. This included seven patients on MTX, eight patients on MMF, and one patient on AZA. Five of the eight patients on MMF had tried and failed therapy with MTX before initiating treatment with MMF. The patient treated with AZA had previously used MMF 1 g twice daily for 3 months without any improvement. The seven patients on MTX and three other patients on MMF had not used any other cytotoxic drug before their current regimen.

Intravenous immunoglobulin treatment

DM was treated with IVIg in two patients with skin-only disease. One patient did not tolerate treatment with AZA, HCQ, or quinacrine and tried treatment with MTX, MMF, and chloroquine for a year each before initiating IVIg treatment. The patient was on IVIg treatment alone for 29 months prior to enrollment and continued for 48 months while information was recorded in the database. To date, this patient has received more than 6 years of treatment with IVIg and the CDASI score at the time of the last visit was 6. The other patient with skin-only DM was treated with IVIg in combination with HCQ, quinacrine, and MTX. This patient was treated with this combination for 22 months before IVIg because there was minimal skin and muscle disease. The last recorded CDASI score for this patient while on the combination treatment was 7.

Discussion

The use of antimalarial medications in the management of cutaneous DM is supported by several retrospective case series that report on the clinical response in up to 75% of cases (Cosnes et al. 1995; Lam and Vleugels 2012). In a retrospective case series of 17 adult patients with DM, nine patients experienced some improvement when treated with antimalarial medications (Ang and Werth 2005). In four of those patients, the addition of quinacrine to HCQ was required to achieve a response and two patients benefited from substituting chloroquine for HCQ (Ang and Werth 2005).

In this analysis of adult patients with DM, 56.1% of patients were managed with antimalarial medications and 43.9% required the use of cytotoxic drugs or IVIg treatment in combination with antimalarial medications. HCQ was used for 24.4% of patients, HCQ and quinacrine for 22.0%, chloroquine for 4.9%, and chloroquine and quinacrine for 2.4%. Although 24.4% of patients with skin-only disease were managed with HCQ alone during the study period, this overestimates the efficacy of HCQ monotherapy because four patients in this group required the addition of quinacrine. Thus, just 14.6% of patients with skin-only disease were managed with HCQ alone. Our results suggest that HCQ is less effective in the treatment of patients with cutaneous DM than patients with CLE (Chang et al. 2011; Wahie et al. 2011). After a median duration of treatment of 24 months, the median final CDASI score for all patients with skin-only disease was 13.5. Most patients did not experience complete resolution of inflammatory skin activity and most patients had at least mild disease activity at the time of their final visit.

In our patient population, a cutaneous drug eruption was noted in 13% of patients who were exposed to antimalarial medications. We suggest that this adverse effect is not a class effect because half of the patients who experienced an initial reaction went on to receive other antimalarial agents and no subsequent adverse cutaneous reactions were observed. Pelle and Callen (2002) reported the development of a cutaneous eruption in 31% of patients with DM and 3% of patients with CLE. Interestingly, two of four patients who subsequently received chloroquine also had a drug reaction with

chloroquine (Pelle and Callen 2002). Both studies show a relatively high incidence of cutaneous reactions to antimalarial medications in patients with DM.

Prospective trials report a significant improvement in cutaneous DM symptoms with the use of IVIg treatment (Gottfried et al. 2000; Saito et al. 2008). Resolutions of cutaneous and muscle disease are also achieved when IVIg treatment was used in addition to other systemic therapies (Danieli et al. 2002, 2009). The only randomized controlled trial to date on the use of IVIg treatment to manage cutaneous DM evaluated the effect of IVIg on muscle and cutaneous involvement in patients with refractory DM (Dalakas et al. 1993). Significant improvements in cutaneous disease were noted in eight patients. An appreciable clinical response began a mean of 15 days after the first infusion and was definitive after the second but most patients required repeated treatments to maintain the response. Our results highlight the clinical response that is possible with a prolonged use of IVIg treatment for cutaneous DM. Although there are few patients in this group, the beneficial effect of IVIg treatment is especially notable given that patients who were treated with IVIg were the most refractory.

Although this study provides useful information on the treatment of patients with DM, there are several limitations. The database was designed to include patients with DM at any point in their disease or treatment course. As a result, many patients were enrolled after a trial of one or more therapeutic agents, which impaired our ability to assess skin activity at baseline for all patients and limited our ability to draw conclusions about a treatment effect. Secondly, treatment of cutaneous DM requires an algorithmic approach that depends on the additive effects of several agents. Since patients with a more severe disease status received escalating doses of therapy, it is difficult to compare final CDASI scores across treatment groups. Treatment decisions are often driven by which organs are involved, which results in several possible treatment regimens and few patients in each group. Clinical trials are necessary to elucidate the efficacy of each therapeutic agent alone or in combination with others.

In addition, the use of prednisone in a small number of patients may have contributed to clinical improvements and changes in CDASI activity scores for these groups, which again limited our ability to draw conclusions about treatment effect. Lastly, participants were recruited from a referral-based clinic at a tertiary medical center. A referral bias must be considered because patients were likely to have more refractory disease. Nonetheless, this study of a large cohort of adult patients with DM contributes to the understanding of DM management. Our data highlight the outcomes of the CDASI treatment algorithm and underscores the need for more effective therapies.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at <http://dx.doi.org/10.1016/j.ijwd.2017.05.001>.

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