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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.

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

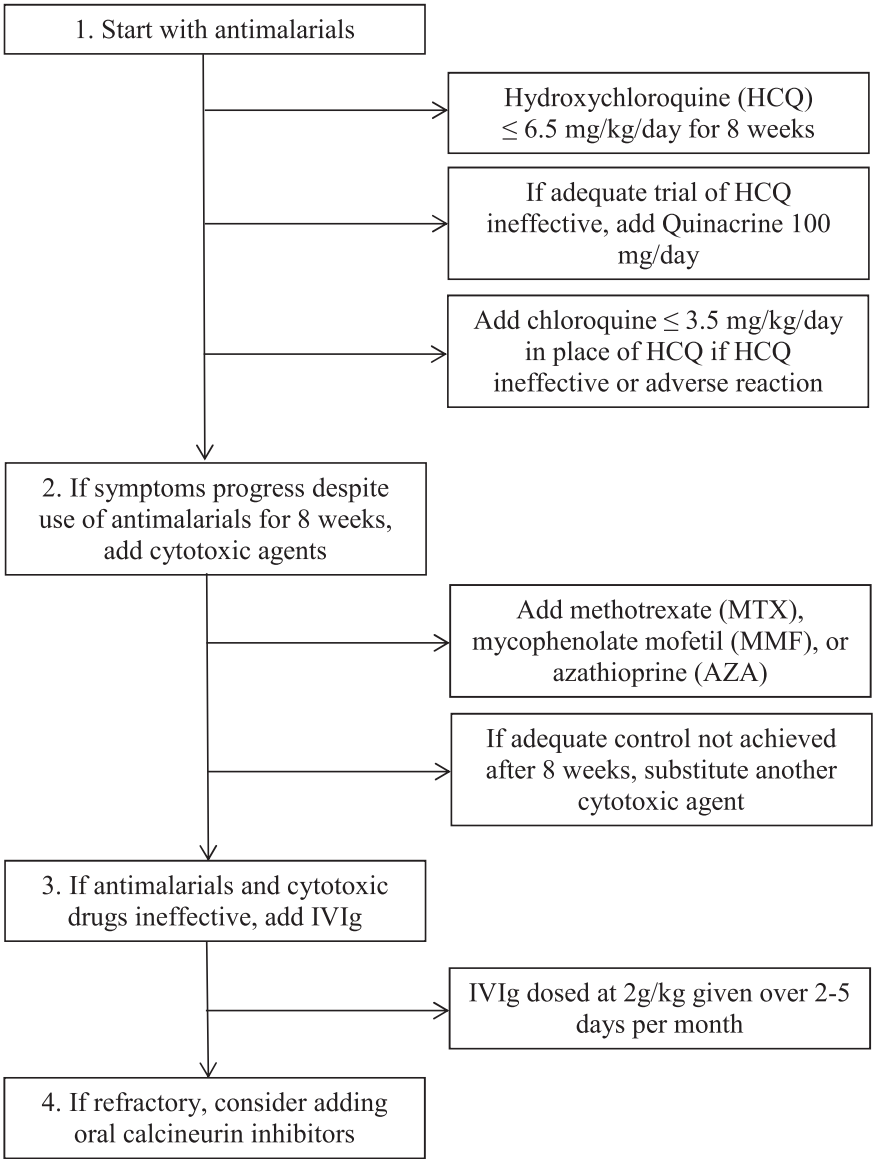

Fig. 1. Treatment algorithm.

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