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REVIEW ARTICLE

Adult and juvenile dermatomyositis treatment

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Abstract

Introduction: Dermatomyositis is a rare autoimmune inflammatory condition affecting skin and muscles. The disease can be seen in both adults and children. It can be associated with malignancy. Considering involvement of skin in the disease, many patients consult dermatologists for its treatment. Hence, knowledge about its presentation, complications, prognosis, and treatment is necessary.

Objective: The objective of this review article is to provide comprehensive information about treatment of dermatomyositis.

Methods: In this review article, we reviewed the published literature on adult and juvenile dermatomyositis to highlight the treatment. Articles published in peer-reviewed journals including reviews, clinical trials, case series, and case reports published in electronic database (MEDLINE/PubMed) through January 2021, cross references of respective articles and trials from clinicaltrials.gov were included for qualitative analysis of the literature.

Results: Treatment options for dermatomyositis include traditional immunosuppressive agents and immunomodulatory therapy. High-dose corticosteroids represent the first line of treatment while other immunosuppressive agents are also used, either in combination with or as an alternative to corticosteroids, after initial treatment failure. Some biological agents have been used for the treatment of dermatomyositis with variable responses.

Conclusion: Although several treatment options are available, several questions remain unanswered about the optimal treatment of dermatomyositis.

KEYWORDS
corticosteroids, dermatomyositis, immunosuppressants, malignancy, management

1 | INTRODUCTION

Idiopathic inflammatory myopathies are disorders affecting skeletal muscles. Its subtypes include polymyositis, dermatomyositis, inclusion body myositis, and immune-mediated necrotizing myopathy.[1] Dermatomyositis (DM) is a rare autoimmune inflammatory condition affecting skeletal muscle and skin.[2,3] It is characterized by inflammatory myopathy of unknown cause. Patients may present only with skin disease, with concomitant muscle disease, systemic involvement, or malignancy.[3] Muscle-related symptoms can range from

painless weakness to significant myalgia associated with muscle weakness. In addition to muscle and skin, involvement of other systems/organs including pulmonary system (lungs), joints, heart, and intestine may be seen in these patients (Table 1).[4]

Muscle weakness is progressive. Dermatomyositis can occur in adults and children. The reported mean age of disease onset in adult and children is 42 and 7.3 years.[5] In another study, mean age at disease onset for juvenile dermatomyositis was 81.97 months with a female predominance.[6] Many common features and differences exist in the presentation, associated conditions, and prognosis in adult versus juvenile dermatomyositis. For instance, vasculopathy is more common in juvenile dermatomyositis as compared to adult-onset dermatomyositis. Similarly, the risk of calcinosis and ulceration is higher in children but long-term prognosis is better.

In adults, myositis-specific antibodies are more likely to be present. Also, the disease is more likely to be associated with risk of interstitial lung disease, amyopathic disease, and malignancy in adults. Although findings of muscle biopsy and interferon gene signature are mostly similar, some differences exist in the pathology of dermatomyositis in adults and children.[7]

Clinically amyopathic dermatomyositis is a subgroup of dermatomyositis comprising 5–20% of total number of patients. In these patients, dermatological features are seen but clinical muscle weakness lasting ≥6 months is absent.[8]

Considering varied presentations, dermatomyositis poses diagnostic as well therapeutic challenge.[3] Muscle biopsy and plasma levels of miRNA are useful investigations in these patients.[9,10] A cross-sectional study comparing muscle biopsies in adults and children with dermatomyositis showed similar distribution of CD4 and CD8-positive cells. In children, there was predominance of CD4 in perimysium. Muscle biopsies in adults showed predominance of CD20 and CD68-positive cells. Distribution of CD20 in perivascular endomysium and CD68 in perimysium was similar in both groups.[5] Plasma levels of hsa-miR-4442 may be a useful biomarker for the diagnosis and disease activity of dermatomyositis.[10]

Dermatomyositis may present as paraneoplastic syndrome or with malignancies of different organs.[11] It has been suggested that in 26–70% patients, neoplasia occurs within 1 year of diagnosing dermatomyositis. However, the risk of neoplasia is high during 5 years after the diagnosis of myositis. Therefore, clinicians should perform annual screening for neoplasia especially during the initial years after the diagnosis of myositis.[12]

TABLE 1 Clinical features of dermatomyositis.

- Erythema (heliotrope erythema and Gottron's papules)
- Muscle weakness
- Myalgia
- Fever
- Malaise
- Weight loss
- Systemic manifestations involving lungs, joints, heart and intestine
- Calcinosis (more common in children)
- Ulcers (more common in children)

Considering the cutaneous involvement, dermatologists can play an important role in the diagnosis and management of dermatomyositis. Moreover, the disease is chronic and adversely affects the quality of life.

Useful tests for the diagnosis of dermatomyositis are listed in Table 2.[9,10,12]

In the following section, we briefly discuss the update on the treatment of adult and juvenile dermatomyositis. Articles published in peer-reviewed journals including reviews, clinical trials, case series, and case reports published in electronic database (MEDLINE/PubMed) through January 2021, cross references of respective articles and trials from clinicaltrials.gov were included for qualitative analysis of the literature.

2 | TREATMENT OF DERMATOMYOSITIS

The objectives of the treatment of dermatomyositis include improving the physical functions and avoiding development of disability. The treatment must be instituted early and requires a team approach involving physical therapist, dermatologist, and family physician. Involvement of other specialists may be required, depending on the particular manifestations of the disease.

Factors considered in the selection of treatment include severity of disease, systemic manifestations, and myositis-specific antibody profile.[13]

For cutaneous manifestation, photoprotection with protective cloths and sunscreens is the first line of management. Moisturizers and emollients may be useful for the control of pruritus and associated burning. Topical application of corticosteroids is also used for controlling local inflammation.

3 | CONVENTIONAL IMMUNOSUPPRESSANTS AND IMMUNOMODULATORS

Immunosuppressives and immunomodulatory agents are commonly used in the treatment of inflammatory myositis.[14] These agents include corticosteroids, methotrexate, hydroxychloroquine, tacrolimus, cyclophosphamide, cyclosporine, azathioprine, mycophenolate mofetil, and immunoglbulins.[4,15,16]

TABLE 2 Tests for the diagnosis of dermatomyositis.

- Myositis-specific antibodies (antisynthase antibodies, anti-signal recognition particle, anti-Mi-2, anti-CADM-140, anti-p155/p140, and anti-MJ, Anti-PMS1)
- Myositis associated antibodies (Anti-U1 ribonucleoprotein, anti-Ku, anti-PM-Scl)
- Muscle biopsy
- Plasma miRNA levels
- Serum creatine kinase or aldolase level
- Electromyography

4 | CORTICOSTEROIDS

Considering their anti-inflammatory and immunosuppressive action, corticosteroids are considered as first-line treatment for dermatomyositis. Both oral and parenteral therapies are used; selection of a particular agent being dependent on disease severity. The usual approach for the management includes an initial high oral dose of corticosteroids followed by a slow tapering. Oral glucocorticoids are used in the dose of 0.75–1 mg/kg/day.[4] Oral prednisolone is considered as the initial pharmacological treatment for dermatomyositis. If no satisfactory improvement is observed with initial therapy of prednisolone after 3 months, adjuvant therapy is often needed. After the initial therapy, dose of prednisolone should be tapered to either daily or alternate day treatment.[17] Treatment can be stopped within 1–2 years. With this treatment, patients can remain disease-free for several years. However, this is an empiric approach for the management of the disease.[2]

In patients with severe condition, intravenous methylprednisolone pulse doses of 1 g/day may be required.[4] Although corticosteroids are useful, adverse events are a common concern for regular use of corticosteroids. The adverse events associated with long-term use of corticosteroids include suppression of adrenal functions, immunosuppression, and osteoporosis. Avascular necrosis is another important concern with use of corticosteroids.

5 | OTHER IMMUNOSUPPRESSANTS AND IMMUNOMODULATORS

These agents are used as adjuvants with glucocorticoids for boosting the efficacy and decreasing the dose of corticosteroids.[4]

Methotrexate an antimetabolite is considered as first-line option for adjuvant therapy in patients not showing satisfactory response to oral corticosteroids. In adults, it is started in the lower dose (7.5–10 mg/week) and dose is slowly increased up to 25 mg/week.[4] With gradual increase in the dose of methotrexate, dose of corticosteroids is reduced. Gastrointestinal, hepatic, and hematological (neutropenia, thrombocytopenia) adverse events are reported with the use of methotrexate. Folic acid is used to reduce the incidence of adverse effects associated with methotrexate.

Intravenous immunoglobulin is an effective short-term option in refractory cases, but long-term efficacy is not known.[17] Intravenous immunoglobulins (2 g/kg) given as multiple dose is effective in patients with resistant disease and involvement of esophagus and throat muscles and lungs.[4] Limitations of intravenous immunoglobulins include cost and short duration of improvement. A case report suggested beneficial effect of combined therapy with corticosteroids, intravenous immunoglobulin, and mycophenolate mofetil in severe and rapidly progressive subtype, antimelanoma differentiation-associated gene 5 dermatomyositis.[18] A study reported higher mortality for treatment with glucocorticoids or immunosuppressants than with immunoglobulin.[19]

Hydroxychloroquine is a good choice for dermatological manifestations whereas cyclosporine may be useful for pulmonary involvement. Other immunosuppressants include azathioprine, mycophenolate mofetil, and tacrolimus.

Answers for many questions regarding optimal therapy for dermatomyositisare urgently warranted. A Cochrane review published in 2012 suggested absence of high-quality randomized clinical trials on usage of immunosuppressants in inflammatory myositis.[14] Even today, there is no consensus regarding the optimal dose, duration of treatment, frequency, and speed of dose tapering for immunosuppresants in DM. It is also not known when to add and the choice of adjuvant immunosuppressant in patients with steroid resistance. Evidence regarding efficacy and safety of traditional immunosuppressive agents is also not available, adding to the uncertainty about the most effective treatment for long-term remission.[2]

6 | RITUXIMAB

Many biologic drugs are currently used as off-label for the treatment of dermatomyositis. Among them, B-cell-depleting therapies such as rituximab seem to be promising options for the treatment of refractory dermatomyositis.[2,20] A randomized trial evaluating efficacy of rituximab in patients with refractory dermatomyositis reported significant improvement in the cutaneous disease activity. Cutaneous disease responded faster when treated earlier in the course of the disease than later.[21] Although a small study, the promising observations from the GRAID-2 registry, Germany suggest that rituximab may be a useful agent for the treatment of dermatomyositis. Rituximab is generally started after the disease has failed respond to multiple conventional immunosuppressives. Rituximab is available as intravenous infusions and administered at a dose of 1 g and the infusions are repeated after 2 weeks. The patients in the GRAID-2 registry received a mean of 3.09 infusions. There was no specific safety concern in these patients, and most of them demonstrated good tolerability. Infections represent the important adverse events in patients receiving rituximab.[22]

7 | ABATACEPT

In a small study, abatacept showed promising results in adult patients with refractory dermatomyositis in terms of lowering disease activity. Repeat muscle biopsies done after 6 months of treatment showed increased frequency of anti-inflammatory Foxp3$^+$ Tregs indicating muscle regeneration and better treatment responses.[23] According to the results of a small study by Tang and colleagues,[24] evaluating the in vivo effects of abatacept among patients with myositis from the ARTEMIS trial, circulatory CD4/CD8 ratio at the time of active disease may be a predictor of treatment efficacy. More evidence is required for evaluation of efficacy and safety of abatacept in dermatomyositis.

8 | TOFACITINIB

Considering the role of JAK/STAT signaling in the pathogenesis of calcinosis and interstitial lung disease in dermatomyositis, tofacitinib, may have a role in its treatment. Wendel and colleagues[25] have reported successful treatment of two such patients with tofacitinib. Over 28 weeks, there was impressive response with tofacitinib therapy. Both the patients did not develop new calcifications. Moreover, the preexisting calcinosis either remained stable or reduced in number, but no new lesions were seen. Importantly, in one patient with interstitial lung disease showed rapid clinical response to tofacitinib. Tofacitinib can also offer benefit of steroid-sparing agent. In both cases, the authors reported early tapering of corticosteroid with concomitant tofacitinib therapy. Treatment with tofacitinib was well tolerated without any major safety issue.

In another case report, an adult patient with anti-MDA-5 and anti-Ro52 antibody-positive hypomyopathic dermatomyositis with interstitial pulmonary involvement showed significant response in the form of improved physical performance, improvement in cutaneous condition and interstitial lung disease. The treatment was well tolerated.[15] In this case, the decision to start tofacitinib was based on interferon signature identified by flow cytometry.[15] Thus, tofacitinib may be further studied for patients with anti-MDA5 antibody-positive dermatomyositis.[26] It has also been suggested for the treatment of refractory anti-NXP2 and anti-TIF1γ dermatomyositis.[27] Tofacitinib citrate can thus be an option for refractory cutaneous dermatosis, but needs further evaluation through large and well-designed clinical trials.[28]

9 | OTHER THERAPIES

The results of a randomized, double-blind, placebo-controlled pilot study suggested that addition of infliximab injection to immunosuppressive and prednisone may be beneficial in some patients with refractory dermatomyositis.[29] Case series of three women suggested potential usefulness of apremilast, a phosphodiesterase four inhibitor as an adjuvant to other immunomodulating drugs in recalcitrant patients.[30] Apremilast may be also an option to evaluate in patients with refractory cutaneous dermatomyositis with severe pruritus.[31]

In a case series of three patients, oral apremilast 30 mg given twice daily with the treatment regimens resulted in significant improvement. It was useful in tapering of steroids and steroid-sparing agents. Interestingly, in two patients, other medications could be discontinued and apremilast was then used as a monotherapy.[30]

The exact mechanism of apremilast in the treatment of dermatomyositis is not known. It has been suggested to act by interference with Th1 and Th2 response.[30]

In a small retrospective study, repository corticotropin injection showed improvement as per physician's assessment.[32] Tocilizumab and ruxolitinib also needs to be studied in more details.[22,23] Ruxolitinib treatment of a patient with polycythemia vera with JAK2 V617F mutation and concomitant deramatomyositis resolved DM symptoms rapidly.[33] Physical dysfunction after treatment is not uncommon in patients with dermatomyositis, and it may be associated with age at disease onset, female gender, and CK levels prior to the treatment.[34] Persistence of CD244+ (CD28 null) T cells in muscles and relative loss of regulatory T cells are associated with poor clinical outcomes after treatment with traditional immunosuppressive therapy.[35] Physiotherapy and symptomatic treatment are also essential for improving general well-being and muscle strength of these patients.[4]

10 | CLINICALLY AMYOPATHIC DERMATOMYOSITIS

Amyopathic dermatomyositis, a subtype of dermatomyositis, is characterized by muscle weakness, Gottron papules, heliotrope rash, scalp and extremity erythema, pruritus, and fatigue. Differentiating features of amyopathic dermatomyositis from dermatomyositis are absence of characteristic muscle weakness and muscle enzyme elevation.[36] The treatment options include topical corticosteroids, systemic immunosuppressants, and immunomodulators.[36] Currently, no consensus exists on the choice of first-line treatment for these patients. It is also not known whether treatment should be the same as that of patients with classic dermatomyositis with muscle weakness. Systematic review of literature also did not yield any strong evidence. Evidence from cases series and case reports suggests use of more than one therapeutic agent in majority of patients either due to side effects or lack of efficacy. Antimalarial agents were most commonly used as the treatment option in these patients. Most promising treatment results in these patients were obtained with intravenous immunoglobulin, which resulted in improvement or remission.[8] In a case report, low-dose naltrexone was found to be effective in a patient with refractory amyopathic dermatomyositis.[36] Anakinra was effective in improving manifestations in a patient with severe and refractory clinically amyopathic dermatomyopathy.[37] Treatment options for patients with dermatomyositis are summarized in Tables 3 and 4.

TABLE 3 Suggested treatments in patients with dermatomyositis.

- For cutaneous symptoms: Photoprotection, topical emollients, moisturizers, topical corticosteroids
- Corticosteroids plus immunosuppressants/immunomodulators (methotrexate*/hydroxychloroquine**/azathioprine/cyclosporine***/mycophenolate mofetil/tacrolimus)
- Intravenous immunoglobulins
- Biologics (limited evidence)
- Supportive therapy
- Symptomatic treatment
- Physiotherapy

Note: *Commonly used **Good effect in skin related symptoms ***Preferred for patients with pulmonary manifestations.

TABLE 4 Summary of treatment options in dermatomyositis

Treatment option	Place in the therapy	Limitations
Corticosteroids	First-line treatment	Long-term use is associated with risk of suppression of adrenal functions, immunosuppression, and osteoporosis
Methotrexate	First-line option for adjuvant therapy in patients not showing satisfactory response to oral corticosteroids	Gastrointestinal, hepatic, and hematological adverse events
Intravenous immunoglobulin	Short-term option in refractory cases	Long-term efficacy is unknown, cost
Rituximab	Refractory dermatomyositis	Infusion related reactions, infections, cost
Abatacept and tofacitinib	Refractory dermatomyositis	Cost, limited evidence compared to other options mentioned above

11 | JUVENILE DERMATOMYOSITIS

Understanding of the pathogenesis and treatment response of juvenile dermatomyositis has been better than adult dermatomyositis. Gottron's papules and muscle weakness are the most common initial clinical presentations of juvenile dermatomyositis. Female gender, absence of Gowers' sign at disease onset, and photosensitivity at disease onset are predictors for better outcome. A 20-year retrospective study of 39 children reported no malignancy among children with juvenile dermatomyositis.[6] Calcinosis can occur in patients with severe and long-term active disease.

Treatment options for juvenile dermatomyositis include corticosteroids, intravenous immunoglobulin, hydroxychloroquine, mycophenolate mofetil, azathioprine, cyclophosphamide, cyclosporine, and methotrexate.[38–41]

Among these, high doses of corticosteroids are still the first line of treatment. Other immunosuppressants can be selected based on the severity of the condition.[40,42] Traditional therapies are effective in reducing mortality rates in the long run.

A recently published Childhood Arthritis and Rheumatology ResearchAlliance (CARRA) registry study (n = 39) involving patients with juvenile dermatomyositis evaluated methotrexate and prednisone; intravenous methylprednisolone, methotrexate, and prednisone; and intravenous methylprednisolone, methotrexate, prednisone, and intravenous immunoglobulin. All three regimens showed significant improvement in disease activity, but there were no significant differences between the groups.[43]

Cyclophosphamide has been shown to be effective and well tolerated in severe juvenile dermatomyositis in a study involving 56 patients. Treatment showed reduction in skin and muscle disease. Cyclophosphamide may be useful as a steroid-sparing agent because of its ability to reduce the dose of glucocorticoids. Short-term safety is good, but long-term side effects need to be studied.[44] Survey results of CARRA members supported the use of rituximab, abatacept, anti-TNFα drugs, and tocilizumab for the treatment of refractory juvenile dermatomyositis. Biologics are commonly used in combination with other agents.[45]

In a case report, ustekinumab was found to be effective in the treatment of amyopathic juvenile dermatomyositis associated with psoriasis.[46]

In patients with refractory disease, novel biological therapies are used, but evidence from well-designed clinical trials is required for strong recommendation.

A randomized trial in refractory cases of juvenile dermatomyositis reported significant improvement in the cutaneous disease activity with rituximab.[21] Rituximab especially for skin lesions and cyclophosphamide are options for patients with severe refractory disease.[47]

Infliximab, adalimumab, tofacitinib, and baricitinib (Janus kinase 1/2 inhibitor) are other options, which need further evaluation. No good evidence exists for tocilizumab in juvenile dermatomyositis.

Treatment of calcinosis is a challenge in juvenile dermatomyositis.[9] Early diagnosis and prompt therapy can prevent its development and also result in better outcome.[40] Different therapies including increased systemic immunosuppression, intravenous immunoglobulin, bisphosphonates, and topical sodium thiosulfate are the options, but there is no consensus.[48]

Clinical manifestation and myositis-specific antibodies may be useful for prediction of prognosis. Although there is significant improvement in the prognosis of juvenile myositis over recent years, long-term outcome vary considerably between different patients suggesting variability of clinical patterns among different patients.[42] Myositis-specific antibodies, muscle biopsy, magnetic resonance imaging, and biomarkers can be useful for the assessment of disease activity and prediction of outcome.[47] High levels of galectin-9, CXCL10, TNFRII, and galectin-1 are predictors of poor response to conventional therapy.[49] In these patients, aggressive therapy and monitoring may be required.

12 | FUTURE SCOPE FOR THE TREATMENT OF DERMATOMYOSITIS

Considering high expression of IL-18 in muscle tissue in inflammatory myopathies, it may represent a novel therapeutic target. Therapies against this target need further evaluation. Multicentric randomized clinical trials are required for establishing systematic plan for the treatment of dermatomyositis. In addition to the need of an effective treatment for dermatomyositis, especially in refractory cases, an effective and well-tolerated drug is also

necessary for the treatment of calcinosis among patients with dermatomyositis.

Blockers of the lymphotoxin signaling pathway, anti-B cell therapy, anti-interferon-alpha, anti-cytokine therapies, and anti-chemokine therapy are the future scopes for the treatment of dermatomyositis.[50] On-going clinical trials with apremilast, PF-06823859, KZR-616, ustekinumab, mesenchymal stromal cells, and subcutaneous immunoglobulin (IgPro20) will provide more insights about potential of therapies under investigation for the treatment of dermatomyositis. Other experimental therapies include adrenocorticotropic hormone gel for refractory cutaneous manifestations, pirfenidone in dermatomyositis interstitial lung disease, and sodium thiosulfate for the treatment of calcinosis associated with dermatomyositis.

13 | CONCLUSION

Dermatomyositis is a rare autoimmune, inflammatory disease affecting skin and muscles. Considering involvement of skin, patients may consult dermatologist for its management. Oral corticosteroids represent the first line of treatment of dermatomyositis. In patients not responding to it or those who are not the candidates for corticosteroids, other immunosuppressants and immunomodulatory agents are useful agents. Newer biologic agents are emerging as beneficial agents in dermatomyositis, but more evidence is required demonstrating their efficacy and safety.

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Anant Patil involved in writing and revising the manuscript. Jun Lu, Martin Kassir, and Mahsa Babaei involved in review and revising the manuscript. Mohamad Goldust involved in conception, writing, review and revising the manuscript.

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CONFLICT OF INTEREST

None.

DATA AVAILABILITY STATEMENT

Data sharing not applicable to this article as no datasets were generated or analysed during the current study.

DISCLAIMER

"We confirm that the manuscript has been read and approved by all the authors that the requirements for authorship as stated earlier in this document have been met and that each author believes that the manuscript represents honest work".

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