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MANIFESTATIONS OF THE SKIN IN DERMATOMYOSITIS AND THEIR DIAGNOSTIC SIGNIFICANCE: A LITERATURE REVIEW

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ABSTRACT

Background. Dermatomyositis (DM) is an idiopathic inflammatory myopathy in which characteristic cutaneous manifestations often precede or occur independently of muscle involvement. Skin findings are essential for diagnosis, phenotype classification and prognostic assessment, reflecting underlying immunopathologic processes.
Aim of the study. To review and summarize current evidence on the spectrum of cutaneous manifestations in dermatomyositis, their associations with myositis-specific autoantibodies, and implications for prognosis, monitoring and management.
Materials and methods. A narrative review of peer-reviewed literature was performed. Searches were conducted in PubMed and in the Medical University of Gdańsk (GUMed) online library catalog. Search terms included "dermatomyositis", "cutaneous manifestations," "autoantibodies" and related phenotype and outcome terms. Priority was given to recent cohort studies, key reviews and illustrative case series; articles in English and Polish were considered. Selected sources were synthesized qualitatively to describe clinical morphology, serologic correlations, validated skin assessment tools and therapeutic approaches.
Conclusions. Classic cutaneous signs (heliotrope rash, Gottron papules/sign, shawl and V-neck distributions, poikiloderma) remain central to DM recognition. Distinct skin phenotypes correlate with specific autoantibodies (e.g., anti-MDA5 with ulcerations and high ILD risk; anti-TIF1-γ with extensive eruptions and malignancy association; anti-NXP2 with calcinosis). Standardized scoring (CDASI) aids objective assessment. Integrating detailed dermatologic evaluation with antibody profiling improves early diagnosis, risk stratification and individualized management. Prospective, phenotype-driven studies and refinement of classification criteria-particularly to better capture amyopathic variants-are needed.

KEYWORDS

Dermatomyositis; Cutaneous Manifestations; Myositis-Specific Autoantibodies; Anti-MDA5; Interstitial Lung Disease; Shawl Sing; CDASI; anti-TIF1-γ; anti-NXP2, JAK Inhibitors; Clinically Amyopathic Dermatomyositis; Heliotrope Rash; Calcinosis Cutis; Poikiloderma

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1. Introduction

Definition of dermatomyositis.

Dermatomyositis (DM) is a rare, acquired autoimmune disease within the group of idiopathic inflammatory myopathies (IIM), characterized by muscle weakness and the presence of typical cutaneous lesions (1,3). It belongs to a clinically heterogeneous group of disorders which - despite a common picture of myopathy with predominant involvement of proximal muscles - differ in affected muscle groups, clinical profile and histopathological features (1-3). In dermatomyositis, characteristic cutaneous signs constitute a key element of the disease phenotype and often coexist with subacute muscle inflammation (1-3).

Epidemiology

Dermatomyositis occurs significantly more often in women, as confirmed by population data from different regions. In a cohort from Olmsted County (USA), as many as 90% of patients were women and the mean age at onset was 57 years, indicating predominance in middle and older adult age groups (4). A similar demographic structure was reported in a study from northern Spain, where over two-thirds of cases involved women and the mean age of patients with idiopathic inflammatory myopathies - with dermatomyositis predominating - was around 53 years (5). In both populations DM was the most frequently diagnosed subtype of inflammatory myopathies - accounting for about half of all cases - with some patients presenting a clinically amyopathic form. Incidence rates remained at similar levels (0.9–1.1 per 100,000 person-years), confirming that dermatomyositis remains a rare but well-defined disease entity, most often affecting women in their fifth and sixth decades of life (4,5).

Pathophysiology and pathogenesis of dermatomyositis

Dermatomyositis is an autoimmune disease in which muscle and skin damage results from the interaction of humoral and cellular mechanisms together with genetic and environmental factors. A key initiating process is deposition of the complement complex C5b–C9 (MAC) in the walls of muscle capillaries. Complement activation - preceded by formation of C3b and C4b - leads to endothelial cell injury, ischemia and secondary atrophic degeneration of muscle fibers, especially in perifascicular zones that are furthest from blood supply (perifascicular atrophy). Chronic injury results in reduced capillary density and progressive degeneration of muscle fibers (6).

The histopathological picture of DM is characterized by predominance of perivascular and perimysial inflammatory infiltrates composed mainly of CD4+ T cells, macrophages, dendritic cells and B lymphocytes (6,7). The presence of MAC deposits and a reduced number of vessels relative to muscle fibers confirm ischemic microangiopathy as a central element of pathogenesis (8). In contrast to polymyositis, cytotoxic damage of fibers by CD8+ cells is less frequently observed in DM (9,10).

Autoantigen–autoantibody reactions also play a significant role, especially myopathy-specific autoantibodies (e.g., anti–Mi-2, Anti-TIF1-γ, anti-MDA5, anti–SAE) (6,8). Some autoantigens are overexposed or altered in regenerating muscle fibers, which favors activation of T and B lymphocytes (6,8). Concurrently, activation of the type I interferon pathway - characteristic for DM - occurs mainly via plasmacytoid dendritic cells and regenerating muscle cells (11).

Genetic factors also influence disease development, particularly the presence of class II HLA alleles (e.g., DRB1*0301, DQA1*0501) that increase susceptibility to autoimmune responses (6,8). Genetic predisposition may be modulated by environmental factors including viral infections, UV radiation, vitamin D deficiency or exposure to certain drugs (6,8). Interaction of these elements leads to establishment of chronic inflammation, microcirculatory dysfunction and progressive muscle damage (8).

Dermatologic significance in dermatomyositis

Cutaneous lesions play a key role in diagnosis and assessment of dermatomyositis because they frequently precede muscle symptoms or appear concurrently with them (6). Patients may present a wide spectrum of manifestations including various types of erythema, photosensitivity, pigmentary disorders, pruritus, and adnexal involvement such as periungual changes or alopecia (6). In the amyopathic form, where clinical and laboratory features of muscle involvement are absent for at least six months, the characteristic dermatologic picture forms the basis of diagnosis (12). Contemporary classification systems emphasize the role of the skin, allowing diagnosis of ADM (amyopathic dermatomyositis) without the necessity of muscle biopsy if typical skin lesions are present without symmetric muscle weakness (13). Importantly, the cutaneous phenotype can provide information about the autoantibody profile and predict disease course. For example, anti–Mi-2 antibodies are associated with classic features such as heliotrope rash, Gottron papules, V-neck or shawl sign, while Anti-TIF1-γ antibodies more often accompany widespread photoerythematous changes. Conversely, patients with anti-MDA5 antibodies typically display skin ulcerations, palmar papules, diffuse hair loss, inflammatory subcutaneous changes or mucosal symptoms (14). Such broad and varied dermatologic expression makes the skin not only an early disease signal but also a valuable tool in assessing subtype, activity and potential prognosis.

2. Classic cutaneous manifestations of DM

2.1. Heliotrope rash

Clinical picture

The heliotrope rash is a hallmark cutaneous manifestation of dermatomyositis and typically appears as violaceous periorbital erythema accompanied by edema, most prominently affecting the upper eyelids, although the cheeks and nasal bridge may also be involved (15). Its clinical presentation varies depending on disease duration, skin phototype and overall inflammatory activity. In more advanced stages, the rash may develop a characteristic reticulated, hypovascular pattern surrounding conspicuous telangiectatic vessels, reflecting chronic microvascular injury (16). Rarely, a bullous variant can occur and has been associated with more severe systemic involvement, particularly an increased risk of interstitial lung disease (16).

2.2. Gottron papules and Gottron sign

Gottron papules and the Gottron sign are two classic, highly characteristic cutaneous elements in dermatomyositis. Gottron papules are elevated, violaceous-erythematous papules or small plaques, often with mild scaling, located directly over the hand joints - most commonly the MCP, PIP and DIP joints (3,14). They may leave pigmentary changes, skin thinning or minor scarring over time. The Gottron sign is their “flattened

counterpart" - erythematous or pink-violet patches on extensor surfaces, not only of the hands but also elbows, knees or - less commonly - ankles. They are usually not accompanied by marked purpura, and any scaling tends to be subtle. Both signs reflect the same pathology - inflammatory-cutaneous involvement over bony prominences - and together constitute one of the most typical clinical signals of DM. (3,14,17,18)

Inverse Gottron's sign refers to keratotic, firm papules appearing on the flexor sides of the fingers - i.e., in locations opposite to classical Gottron papules (19). This atypical distribution is strongly associated with the presence of anti-MDA5 antibodies, which occur in the clinically amyopathic form of dermatomyositis and frequently coexist with rapidly progressive interstitial lung disease (RP-ILD) (20,21). The appearance of inverse Gottron's sign in a patient should therefore suggest this specific subtype and prompt urgent antibody testing and pulmonary assessment.

2.3.Shawl sign

The shawl sign is a symmetric violaceous erythema involving the nape, posterior aspects of the shoulders and the upper back, resembling a cloak draped over the shoulders (6, 22). Poikiloderma - a mixture of epidermal atrophy, telangiectasia and pigmentary disturbance- may coexist in the same area (18). The shawl sign is sufficiently characteristic that it is described as one of the typical cutaneous manifestations of dermatomyositis, including paraneoplastic forms, where a shawl-pattern erythema may be the first indication of an underlying malignancy (23).

2.4. Poikiloderma

Poikiloderma in dermatomyositis represents a complex skin change that includes concurrent telangiectasia, areas of hypo- and hyperpigmentation and superficial skin atrophy (14). It is most commonly located on the nape, posterior shoulders, upper back, arms, buttocks and within the V-neck area (14). Although poikiloderma in DM usually coexists with photodistributed lesions - which is particularly characteristic for this disorder (24) - it may also affect photoprotected sites (24). In some cases - especially in children - poikiloderma may present atypically with diffuse, scaly skin changes that significantly complicate diagnosis and may delay recognition (24.) It is worth emphasizing that poikiloderma occurring in inflammatory myopathy is referred to as poikilodermatomyositis, distinct from poikiloderma vasculare atrophicans, which represents a variant of cutaneous T-cell lymphoma such as mycosis fungoides (25).

2.5. V-sign

The V-neck sign is a characteristic cutaneous manifestation of dermatomyositis, presenting as confluent macular, sometimes atrophic erythema located on the anterior neck and the upper chest (18). The V-neck sign is also associated with the presence of anti-Mi-2 antibodies, which classically correlate with the typical cutaneous DM phenotype (26).

3. Cutaneous changes of prognostic significance

3.1. Skin ulcerations

Cutaneous ulcerations in dermatomyositis (DM) are an important marker of severe disease and show a strong association with specific serologic profiles. They are most characteristic for the anti-MDA5-positive phenotype, where digital, periungual and periarticular ulcerations often result from pronounced vascular injury induced by the type I interferon pathway (27,28.) Cohort studies have shown that the presence of ulcerations was one of the strongest predictors of anti-MDA5 positivity (OR ~10), particularly when lesions were localized to the fingertips (28). In patients with this antibody profile the risk of interstitial lung disease (ILD), especially the rapidly progressive variant, is significantly increased - however, this association was present only in those with concurrent anti-MDA5 antibodies (28). Case reports confirm that cutaneous ulcerations may be an early sign of severe course in both children and adults - for example, in patients with anti-NXP-2 antibodies (29) or in DM associated with malignancy and intense endothelial injury via interferon mechanisms (27). Importantly, ulcerations may also occur in a subset of patients with Anti-TIF1-γ antibodies, although they are rarer and present with greater heterogeneity in that group (27). Clinical descriptions indicate that early recognition of this cutaneous phenotype allows implementation of intensive immunosuppressive therapy and pulmonary monitoring even if respiratory symptoms are absent. (30) Taken together, these data suggest that skin ulcerations - especially when combined with anti-MDA5 seropositivity - are a significant marker of risk for systemic complications and require urgent, in-depth pulmonary diagnostics (27-30).

3.2. Mechanic's hands

Mechanic's hands are a characteristic cutaneous manifestation of dermatomyositis, particularly frequent in patients with anti-ARS antibodies, notably anti-Jo-1 (31-33). They present as symmetric thickening, hyperkeratosis and cracking of the skin on the ulnar side of the thumbs and the radial sides of the index and

middle fingers, which may mimic chronic hand eczema (31,32). Histopathologically the lesions show acanthosis, hyperkeratosis, a lymphohistiocytic infiltrate with interface dermatitis features and the presence of Civatte bodies; a well-described pattern is the so-called pseudocheckerboard pattern - alternating vertical bands of ortho- and parakeratosis interrupted by horizontal zones of ortho-keratosis (31,32). Mechanic's hands are strongly associated with presence of ILD - studies show that most patients with this feature develop interstitial lung disease - making them an important marker of a high-risk phenotype in dermatomyositis (31,33). Because they can be misdiagnosed as eczema and are linked to severe pulmonary involvement, recognition of mechanic's hands should prompt urgent respiratory evaluation and testing for anti-ARS antibodies (31–33).

3.3. Subcutaneous calcifications

Calcinosis in dermatomyositis is dystrophic, meaning deposition of insoluble calcium salts - most often hydroxyapatite or carbonate apatite - in the skin and soft tissues despite normal serum calcium and phosphate concentrations (34). Its frequency is markedly higher in children, affecting 20–75% of patients with juvenile DM, whereas in adult dermatomyositis it occurs in up to 20% of patients (35). The clinical picture is diverse: from superficial or deep nodules, through tumorous forms, to extensive plate-like calcinosis termed calcinosis universalis (36). Newer imaging techniques, such as whole-body computed tomography, have allowed distinction of five precise distribution patterns of deposits: clustered, disjoint, interfascial, confluent and fluid-filled (37). Calcinosis significantly impairs patient functioning, and its pathogenesis includes neutrophil activation, mitochondrial dysfunction and vascular abnormalities (35).

3.4. Periungual telangiectasias

Periungual telangiectasias are a manifestation of dermatomyositis (6) and reflect the underlying microangiopathic process of the disease. Clinically they present as erythema, petechiae and tenderness of the nail folds, described as Keining's sign (38). Capillaroscopic studies demonstrate dilated and tortuous capillary loops, avascular areas, disorganization of vascular architecture and periungual hemorrhages, and their presence correlates both with disease activity and immunologic phenotype (39–41). These changes are present in the majority of patients already at diagnosis and can be monitored noninvasively using dermoscopy or videocapillaroscopy. Recent observations also indicate associations between specific capillary patterns and myopathy autoantibodies - for example, more frequent periungual bleeding and vessel dilatation among patients with anti-TIF1 antibodies (42), Some studies suggest that capillary patterns may reverse with treatment, highlighting their dynamism and dependence on current inflammatory activity (42,43).

4. Cutaneous phenotypes and autoantibodies

4.1. TIF1-γ

Dermatomyositis with anti-TIF1-γ antibodies is characterized by extensive and often severe cutaneous changes, which are one of the most important elements of this disease phenotype. In most studies patients present classic DM signs such as heliotrope rash, Gottron sign and Gottron papules, although their frequency may differ between groups with and without associated malignancy (44). Some patients also exhibit typical DM erythematous changes of the nape and chest such as V-sign or shawl sign, while features such as mechanic's hands or skin ulcerations are less common (<10%) (44).

A characteristic feature of the anti-TIF1-γ variant is that skin lesions can be particularly extensive; the literature has also described the rare but highly specific "red-on-white" sign and a variant with "islands of sparing" resulting from intense inflammation. (45) These changes can be a valuable diagnostic clue, particularly since the skin phenotype often predominates clinically over muscle involvement (45,46).

In studies comparing patients with and without malignancy it was observed that Gottron papules occurred more frequently in patients without cancer (63% vs 20%), whereas other typical changes - heliotrope rash, Gottron sign or photodistributed erythema - occurred with similar frequency (44). Another study did not find significant differences in the frequency of cutaneous changes between patients with and without cancer, except for an increased frequency of dysphagia suggesting a link to malignancy but not being a direct skin manifestation. (47).

Some studies emphasize that anti-TIF1-γ-positive patients are prone to severe erythematous-papular eruptions, which may stem from the role of TIF1-γ as an inhibitor of TNF and mechanisms related to antitumor responses (46,48).

4.2. NXP2

Dermatomyositis with anti-NXP2 antibodies forms a characteristic phenotype in which cutaneous symptoms are often less pronounced than in classic DM, and some patients present dermatomyositis sine dermatitis (i.e., without typical skin changes) (49). Compared with other DM subtypes, anti-NXP2-positive patients show a reduced frequency of heliotrope rash and Gottron lesions, and their skin inflammation is usually milder (50). Simultaneously, anti-NXP2 is associated with a high risk of subcutaneous changes - especially calcinosis, which may be extensive, early and aggressive (51,52). In adults peripheral edema and marked muscle involvement with slight or atypical skin manifestations are more frequent (49,50). Recent data confirm regional differences in this phenotype: in adults from India nonspecific skin changes predominate, whereas in children typical DM features remain frequent (53). Overall, anti-NXP2 defines a DM subtype characterized by milder cutaneous inflammation but pronounced subcutaneous tissue involvement and frequent calcinosis.

4.3. MDA5

MDA5-positive dermatomyositis (MDA5 DM) is a subtype characterized by severe course and high risk of pulmonary complications. The most characteristic cutaneous changes include deep, painful ulcerations, palmar papules and features such as “hikers’ feet” and inverse Gottron sign, which often coexist with rapidly progressive interstitial lung disease (RP-ILD) and marked proximal muscle weakness (54-56). Ulcerations typically localize on the digits, nail folds or within Gottron’s papules and over joints (knees, elbows), and their pathomechanism involves vasculopathy and intravascular thromboses, which in extreme cases can lead to digital gangrene (57). Additionally, in MDA5 DM palmar hyperkeratotic papules, heliotrope rash, shawl sign, V-sign and other less specific changes such as oral ulcerations, panniculitis, alopecia or flagellate erythema are observed and may indicate worse prognosis (57). Presence of anti-MDA5 antibodies is associated with poorer response to standard glucocorticoid and immunosuppressive therapy, increased risk of complications and higher mortality, underscoring the need for early diagnosis, intensive monitoring and aggressive treatment (54,58).

5. Amyopathic dermatomyositis (ADM) and skin-limited variants of DM

Amyopathic dermatomyositis (ADM) includes patients who present with typical, pathognomonic cutaneous changes of dermatomyositis but who show no clinical or laboratory evidence of muscle involvement for at least six months (12). Despite the introduction of EULAR/ACR criteria, ADM remains diagnostically challenging, leading to frequent misdiagnoses - up to 41% of patients are initially diagnosed as lupus or undifferentiated connective tissue disease (59). Importantly, some patients with ADM do not meet the minimal point threshold in EULAR/ACR criteria, because those criteria include only three skin features (heliotrope rash, Gottron papules and Gottron sign), while patients often present a broader spectrum of signs such as facial, neck, scalp or limb erythema and scaling (60). Distinguishing ADM from classic DM requires emphasizing that typical skin manifestations persist in the total absence of clinical myopathy - this is the foundation of diagnosis and classification of this variant.

5.1 Autoantibody frequencies

The autoantibody profile in dermatomyositis (DM) and oligosymptomatic variants such as clinically amyopathic DM (CADM/ADM) clearly reflects clinical differences between these entities. In CADM/ADM anti-MDA5 antibodies are detected much more frequently, reaching about 36–37% of cases, which is several times higher than in classic DM where the prevalence is 4.8–11.5% (61,62). Presence of anti-MDA5 in this group correlates with the typical skin phenotype and increased risk of interstitial lung disease, particularly the acute course, shown in both Western and Asian populations (62,12). Anti-TIF1-γ antibodies are also observed more often in CADM (up to 31%), however - unlike in classic DM - they do not show clear clinical correlations; in DM they are associated with more severe skin changes and increased cancer risk (61,62). Classic DM is characterized by higher frequency of synthetase autoantibodies, particularly anti-Jo-1, which are a major myositis-specificity in that form (62). At the same time ADM/CADM is more often seronegative, which together with omission of many characteristic skin features from EULAR/ACR criteria complicates classification (60). In both groups the most common accompanying autoantibody remains anti-Ro52, although its clinical significance differs: in CADM it correlates with chronic ILD and vasomotor symptoms, while in DM it more often associates with mechanic’s hands, fever and concomitant CTD (62). All these observations emphasize that CADM/ADM is immunologically and clinically distinct, in which autoantibody profile - especially anti-MDA5 presence - has crucial prognostic significance and should determine the extent of organ monitoring, particularly of the respiratory system.

6. Scale of cutaneous disease activity

Assessment of skin activity in dermatomyositis relies on several standardized tools, most frequently the Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI). This scale has been partially validated and enables detailed measurement of inflammatory activity and irreversible skin damage across 15 anatomical locations. CDASI separates activity features - such as erythema, scaling or ulcerations - from permanent damage elements including poikiloderma and calcinosis, producing two independent scores that reflect current inflammatory state and cumulative damage (63,64). The scale shows high interobserver reproducibility and good sensitivity to clinical changes, making it a standard tool in clinical research on DM (63,64).

The second instrument developed for precise characterization of skin lesions is the Cutaneous Assessment Tool (CAT), which includes 21 items divided into activity, damage and mixed domains. CAT allows classification of lesions by type (e.g., pathognomonic, vascular, acral) and their current features, taking into account erythema intensity, secondary damage or healing traces. CAT provides a wide scoring range (activity 0–175), enabling capture of subtle clinical changes, although its interobserver reliability may be lower than that of CDASI, particularly in damage components (65).

A broader systemic view in assessing dermatomyositis - including skin changes - is provided by the Myositis Intention to Treat Activity Index (MITAX), which evaluates disease activity in seven systems: cutaneous, muscular, pulmonary, cardiac, articular, gastrointestinal and general. Each manifestation receives a score from 0 to 4, and the global score reflects total disease activity (66). MITAX, as a multi-organ tool, is especially useful in studies of disease course and treatment response, since MITAX values correlate with disease severity, presence of ILD and need for intensified immunosuppression (66).

Comparing these three tools, CDASI remains the most precise and best-validated index for assessing cutaneous changes in DM, particularly useful in clinical trials (63-65). CAT enables broader classification of lesion types and subtle features but has less stability between raters (65). MITAX is a systemic activity measure in which skin is only one domain; its clinical value increases in monitoring overall disease activity and predicting long-term prognosis (66). In practice these scales complement each other - CDASI and CAT serve precise skin evaluation, while MITAX places these findings in the wider picture of systemic DM activity.

7. Treatment focused on cutaneous manifestations

8. Glucocorticosteroids (GCS)

Glucocorticosteroids remain the foundation of dermatomyositis treatment despite a lack of recent randomized trials conclusively proving their efficacy (67). Prednisone or prednisolone at approximately 1 mg/kg/day is typically used for induction of remission, often combined with immunosuppressants such as methotrexate or azathioprine to increase treatment efficacy, reduce relapses and enable faster steroid tapering (67,69). Although pulse methylprednisolone may rapidly improve features such as mechanic's hands, vascular lesions or Gottron papules, cutaneous changes - particularly heliotrope erythema and pigmentary alterations - may persist for months, suggesting poorer steroid responsiveness and that chronic skin changes do not always reflect active inflammation (68). Data also indicate frequent discordant responses of muscle and skin to therapy and variable steroid efficacy for cutaneous lesions, while steroids remain necessary for muscle or lung involvement (70–72).

Hydroxychloroquine (HCQ)

Hydroxychloroquine is widely used as a first-line drug for cutaneous manifestations of dermatomyositis due to its broad use in dermatology and rheumatology (73,74). However, available data indicate limited efficacy of HCQ as monotherapy or combined only with topical treatment, with better effects observed when used together with glucocorticosteroids (75). Notably, HCQ is associated with a substantial rate of acute cutaneous reactions - observed in up to 31% of DM patients - as confirmed in contemporary and earlier clinical observations (76,77). Cases of exacerbation of DM skin lesions after HCQ initiation have been reported, with worsening rash from several weeks to months after starting treatment and resolution after drug discontinuation; these reactions more commonly affect patients with Anti-TIF1-γ or anti-NXP2 antibodies (78). Given limited efficacy, frequent adverse effects and risk of exacerbating skin lesions, current evidence does not support HCQ as first-line therapy for cutaneous DM, underscoring the need for high-quality prospective trials (75).

Methotrexate, mycophenolate

Methotrexate (MTX) is one of the most commonly used systemic first-line agents for cutaneous dermatomyositis, especially in patients intolerant or unresponsive to antimalarials. (79) The drug demonstrates efficacy for both skin lesions and muscle involvement, making it an important steroid-sparing therapy in CDM (79). In a retrospective study by Hornung et al. of steroid-refractory patients, MTX induced a clinical response

in 73% of patients, resulting in significant improvement in CDASI scores (mean reduction from 14.1 to 5.5) (80). In clinical practice many centers prefer MTX as the first systemic agent for CDM patients and for CADM patients who are unresponsive or intolerant to antimalarials or who present with severe skin disease at diagnosis. Before starting therapy a full laboratory work-up is recommended, including CBC with differential, renal and liver function tests and screening for viral hepatitis (81). Treatment typically begins at 15 mg/week, increased by 2.5 mg every 2–4 weeks up to a maximum of 25 mg/week if no clinical response is achieved (82).

Mycophenolate mofetil (MMF) is an important option for patients with dermatomyositis refractory to conventional therapy. In a retrospective series of 12 patients who did not respond to standard therapies or did not tolerate them, MMF at doses of 500 mg–1 g/day improved skin status and muscle strength in 10 patients, with good tolerability and initial effects seen after 4–8 weeks (83). A small observational comparison of MTX and MMF found no significant differences in speed of improvement, though the proportion of patients achieving ≥40% reduction in CDASI-A was higher in the MMF group both at first assessment (54% vs 27%) and at last visit (77% vs 55%) (84). Authors note small sample sizes and lack of randomization limit definitive conclusions and emphasize need for large prospective trials. Additional clinical observations suggest MMF may be effective as maintenance therapy in severe DM, including cases with anti-MDA5 antibodies and rapidly progressive ILD, where single cases reported improvement in respiratory function and reduction of autoantibody titers (85).

JAK inhibitors (tofacitinib, ruxolitinib)

Tofacitinib, a selective JAK1/JAK3 inhibitor, is a promising option in refractory dermatomyositis, though caution is warranted due to possible long-term risks of malignancy and major adverse cardiovascular events (MACE). Early clinical reports showed improvement in pruritus and skin lesions as early as 4 weeks in patients with difficult-to-treat cutaneous DM (86). In an open 12-week study all patients with refractory DM achieved a response according to ACR/EULAR criteria after tofacitinib monotherapy, with significant reductions in skin activity measured by CDASI and a favorable safety profile (87). Additionally, in amyopathic DM patients with anti-MDA5 antibodies and concomitant ILD, tofacitinib combined with glucocorticoids improved survival and pulmonary parameters (88).

Baricitinib, a JAK1/JAK2 inhibitor, shows promising efficacy for cutaneous lesions in dermatomyositis. In a prospective study involving 12 patients clinical improvement was observed after 4 weeks, with marked reduction of skin severity by week 12 (89). In an open trial of 16 patients treated with baricitinib or ruxolitinib improvement of skin lesions was seen after 3 months, though without significant effect on muscle strength (90). Similarly, a case series of 12 patients showed reduction of mean CDASI score from 31 to 16 over 12 weeks, and 11 of 12 patients met criteria for clinically meaningful improvement (91).

Ruxolitinib demonstrates promising efficacy in dermatomyositis. Ladislau et al. reported improvement of skin lesions, muscle strength and reduction of type I interferon levels after JAK inhibitor monotherapy (92). Efficacy is also supported by case reports - e.g., rapid skin improvement and steroid dose reduction in a patient with Anti-TIF1-γ antibodies (93). Positive effects were also observed with topical ruxolitinib, with significant skin improvement and reduction of interferon markers (94). Concurrently, the FDA has warned about the risk of serious cardiovascular events associated with JAK inhibitors, requiring caution (95).

Photoprotection

Photoprotection is a fundamental element in the treatment of cutaneous manifestations of dermatomyositis. UV radiation exacerbates skin lesions, and DM patients demonstrate increased photosensitivity due in part to a greater number of apoptotic cells in skin and genetic predispositions that enhance apoptosis after UV exposure and delay clearance of apoptotic debris (96,97). Therefore strict sun protection - including sun avoidance, use of high-SPF sunscreens and protective clothing - is recommended as a first step in therapy. It forms the basis of management before initiating topical or systemic treatments such as glucocorticoids, antimalarials or immunomodulatory therapies (70).

8. Differential diagnosis of DM skin lesions

Dermatomyositis vs Subacute Cutaneous Lupus Erythematosus (SCLE)

Subacute cutaneous lupus erythematosus (SCLE) and dermatomyositis (DM) exhibit photosensitive, erythematous-scaly lesions, but their morphology and distribution differentiate the two entities. In SCLE annular lesions with central clearing or papulosquamous foci resembling psoriasis predominate, typically limited to UV-exposed areas - lateral face, "V"-shaped neck, upper trunk and dorsolateral forearms. SCLE lesions do not scar and often resolve with hypopigmentation (98-100). A characteristic facial feature of SCLE is predominantly lateral face involvement, with sparing of the eyelids and without edema. In DM, pathognomonic features such as violaceous eyelid edema (heliotrope eruption) and Gottron papules on extensor surfaces of the hands are present (103). Facial erythema in DM more often involves the nasolabial folds, allowing differentiation from the malar rash of SLE. Additionally, DM commonly shows photodistributed poikiloderma (shawl sign, V-sign) involving the upper chest and back, often with associated pruritus, distinguishing it from SCLE where poikiloderma is absent and lesions remain more confined to photoexposed skin with annular or psoriasiform morphology (99,100).

Dermatomyositis vs systemic sclerosis (scleroderma)

Scleroderma and DM can coexist, but their skin features are distinct. Systemic sclerosis is dominated by skin thickening, sclerodactyly, telangiectasias and Raynaud phenomenon resulting from fibrosis and microvascular dysfunction (99, 102, 103). DM does not cause skin hardening to the degree characteristic of scleroderma; its changes are more inflammatory, erythematous and often photodistributed. Furthermore, presence of Gottron papules, heliotrope rash or poikiloderma is unique to DM and not seen in scleroderma. Calcinosis cutis may occur in both diseases, but is distinctly more common in juvenile DM (14,103).

Dermatomyositis vs photodermatoses

Photodermatoses, like DM, manifest exacerbation after UV exposure. However, in DM UV exposure is a clear exacerbating factor linked to increased apoptotic cell counts in skin - associated with genetic predisposition and impaired clearance of apoptotic cells. In classic photodermatoses the skin reaction is directly induced by UV, but lacks DM-specific signs such as heliotrope rash or Gottron lesions. Moreover DM involves changes in partially shielded areas and characteristic poikiloderma (shawl sign, holster sign), which is not typical of pure phototoxic or photoallergic reactions (99,105,106,107).

Dermatomyositis vs SINAM (statin-induced necrotizing autoimmune myopathy)

Although DM and SINAM belong to the group of inflammatory myopathies, they differ in clinical and cutaneous profile. SINAM features severe, progressive myopathy with very high CK values (often 2,000–20,000 IU/L), but typically does not cause characteristic DM skin changes (108-110). Series of cases describe rashes in a subset of patients with immune-mediated necrotizing myopathy, but these do not take the form of DM pathognomonic lesions like heliotrope rash or Gottron changes (111). Biopsy in SINAM shows muscle fiber necrosis with regeneration and scant inflammatory infiltrate, whereas DM is dominated by perivascular inflammation and epidermal damage. Moreover DM often presents with cutaneous features, photosensitivity and poikiloderma, which are not observed in SINAM (111).

9. Conclusions

Cutaneous manifestations are a central element of the clinical picture of dermatomyositis and are crucial for diagnosis, assessment of activity, immunologic phenotype and prognosis. In many cases skin lesions precede development of myopathy or remain the only detectable manifestation, as in amyopathic dermatomyositis, highlighting the role of the dermatologist in early recognition (6,12,13).

Classic cutaneous signs of dermatomyositis - such as heliotrope rash, Gottron papules and sign, V-sign, shawl sign and poikiloderma - remain the most characteristic and best-established markers of the disease, enabling its distinction from other photosensitive dermatoses and connective tissue diseases (14,15,18,24). Their presence, distribution and morphology not only support diagnosis but also provide information about inflammatory activity and potential internal organ involvement.

Increasing evidence indicates that specific cutaneous phenotypes closely correlate with myopathy-specific autoantibody profiles. Cutaneous ulcerations, inverse Gottron sign and palmar papules are particularly prognostically important, showing strong association with anti-MDA5 antibodies and high risk of rapidly progressive interstitial lung disease (27,28,54–58). Conversely, extensive, atypical and severe skin lesions in patients with Anti-TIF1-γ antibodies may be an important warning sign for concomitant malignancy (44–48).

Amyopathic dermatomyositis and skin-limited variants pose a significant diagnostic challenge, especially given current classification criteria which do not encompass the full spectrum of cutaneous

manifestations (59,60). In this group autoantibody profile - particularly anti-MDA5 - is of key prognostic importance and should determine the scope of organ monitoring, especially of the respiratory system (61,62).

Assessment of skin activity using standardized instruments such as CDASI, CAT and MITAX enables objective clinical evaluation, monitoring of treatment response and comparison of clinical study outcomes (63–66). Of these, CDASI remains the best-validated and most useful tool for assessing cutaneous activity in dermatomyositis.

Therapeutic management of dermatomyositis requires a multi-faceted approach that considers both systemic treatment and photoprotection as well as phenotype-targeted therapy. Limited efficacy of antimalarials, variable response of cutaneous lesions to glucocorticoids and promising results with JAK inhibitors emphasize the need for further research on targeted treatments, particularly for patients with refractory and severe cutaneous disease (75–77,86–94).

In summary, the skin in dermatomyositis is not merely an affected organ but serves as a crucial "diagnostic window" reflecting complex immunologic processes, disease phenotype and potential systemic complications. Thorough assessment of cutaneous manifestations should be an integral part of diagnosis, classification and monitoring of patients with dermatomyositis, and further research into their prognostic and therapeutic significance remains an important direction for modern dermatology and rheumatology.

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REFERENCES

1. Dourmishev, L. A., & Dourmishev, A. L. (Eds.). (2009). *Dermatomyositis: Advances in recognition, understanding and management*. Springer.
2. James, W. D. (2020). *Andrews' diseases of the skin: Clinical dermatology* (13th ed., pp. 157–183). Elsevier.
3. DeWane, M. E., Waldman, R., & Lu, J. (2020). Dermatomyositis: Clinical features and pathogenesis. *Journal of the American Academy of Dermatology, 82*(2), 267–281.
4. Kronzer, V. L., Kimbrough, B. A., Crowson, C. S., Davis, J. M., III, Holmqvist, M., & Ernste, F. C. (2023). Incidence, prevalence, and mortality of dermatomyositis: A population-based cohort study. *Arthritis Care & Research, 75*(2), 348–355.
5. Corrales-Selaya, C., Prieto-Peña, D., Martínez-López, D., Benavides-Villanueva, F., & Blanco, R. (2025). Epidemiology of dermatomyositis and other idiopathic inflammatory myopathies in Northern Spain. *Biomedicines, 13*(10), 2537.
6. Qudsiya, Z., & Waseem, M. (2025). Dermatomyositis. In *StatPearls*. StatPearls Publishing. (Updated August 7, 2023).
7. Engel, A. G., & Arahata, K. (1986). Mononuclear cells in myopathies: Quantitation of functionally distinct subsets, recognition of antigen-specific cell-mediated cytotoxicity in some diseases, and implications for the pathogenesis of the different inflammatory myopathies. *Human Pathology, 17*(7), 704–721. https://doi.org/10.1016/S0046-8177(86)80180-0
8. Thompson, C., Piguet, V., & Choy, E. (2018). The pathogenesis of dermatomyositis. *British Journal of Dermatology, 179*(6), 1256–1262.

9. Emslie-Smith, A. M., & Engel, A. G. (1990). Microvascular changes in early and advanced dermatomyositis: A quantitative study. *Annals of Neurology, 27*(4), 343–356.
10. Vattemi, G., Mirabella, M., Guglielmi, V., Lucchini, M., Tomelleri, G., Ghirardello, A., & Doria, A. (2014). Muscle biopsy features of idiopathic inflammatory myopathies and differential diagnosis. *Autoimmunity Highlights, 5*(3), 77–85.
11. Haq, S. A., & Tournadre, A. (2015). Idiopathic inflammatory myopathies: From immunopathogenesis to new therapeutic targets. *International Journal of Rheumatic Diseases, 18*(8), 818–825. https://doi.org/10.1111/1756-185X.12736
12. Gerami, P., Schope, J. M., McDonald, L., Walling, H. W., & Sontheimer, R. D. (2006). A systematic review of adult-onset clinically amyopathic dermatomyositis (dermatomyositis sine myositis): A missing link within the spectrum of the idiopathic inflammatory myopathies. *Journal of the American Academy of Dermatology, 54*, 597–613. https://doi.org/10.1016/j.jaad.2005.10.041
13. Lundberg, I. E., Tjärnlund, A., Bottai, M., et al. (2017). 2017 EULAR/ACR classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups. *Annals of the Rheumatic Diseases, 76*, 1955–1964. https://doi.org/10.1136/annrheumdis-2017-211468
14. Muro, Y., Sugiura, K., & Akiyama, M. (2016). Cutaneous manifestations in dermatomyositis: Key clinical and serological features—a comprehensive review. *Clinical Reviews in Allergy & Immunology, 51*, 293–302. https://doi.org/10.1007/s12016-015-8496-5
15. Mainetti, C., Terziroli Beretta-Piccoli, B., & Selmi, C. (2017). Cutaneous manifestations of dermatomyositis: A comprehensive review. *Clinical Reviews in Allergy & Immunology, 53*, 337–356.
16. Sagut, P., & Elston, D. M. (2024). Images in dermatology: Dermatomyositis. *Journal of the American Academy of Dermatology, 91*(3), 606–611. https://doi.org/10.1016/j.jaad.2024.04.066
17. Auriemma, M., Capo, A., Meogrossi, G., & Amerio, P. (2014). Cutaneous signs of classical dermatomyositis. *Giornale Italiano di Dermatologia e Venereologia, 149*, 505–517.
18. Bogdanov, I., Kazandjieva, J., Darlenski, R., & Tsankov, N. (2018). Dermatomyositis: Current concepts. *Clinics in Dermatology, 36*, 450–458.
19. Jain, S., & Sharma, A. (2020). Inverse Gottron's sign in anti-MDA5 antibody-associated dermatomyositis. *Rheumatology (Oxford)*. https://doi.org/10.1093/rheumatology/keaa108
20. Selva-O'Callaghan, A., Pinal-Fernandez, I., Trallero-Araguás, E., Milisenda, J. C., Grau-Junyent, J. M., & Mammen, A. L. (2018). Classification and management of adult inflammatory myopathies. *Lancet Neurology, 17*, 816–828.
21. Irie, K., Matsumura, N., Hoshi, M., & Yamamoto, T. (2021). Inverse Gottron's papules in patients with dermatomyositis: An underrecognized but important sign for interstitial lung disease. *International Journal of Dermatology, 60*, e62–e65.
22. Ho, J. D., Burton, A. T. M., McKenzie, T., Best, C., Clare-Lyn Shue, A., Smith-Matthews, S., Fraser, K., & Anderson, A. (2024). Case report: "Fur stole and turtleneck" and "halter-back" signs: An expanded wardrobe for dermatomyositis. *Frontiers in Immunology, 15*, 1400575. https://doi.org/10.3389/fimmu.2024.1400575
23. Ranjit, E., Sapra, A., Etherton, A., Wasey, W., & Bhandari, P. (2020). 'Shawl sign' as a paraneoplastic dermatosis. *Cureus, 12*(4), e7498. https://doi.org/10.7759/cureus.7498
24. Alaboud, S., Al Hawsawi, W., Alqahtani, N., Aldosari, M., Alhawsawi, W. K., & Al Hawsawi, K. (2023). Juvenile dermatomyositis presenting as generalized poikiloderma: A case report. *Cureus, 15*(12), e50573. https://doi.org/10.7759/cureus.50573
25. Herath, H. M. M. T. B., Keragala, B. S. D. P., Pahalagamage, S. P., Janappriya, G. H. C. C., Kulatunga, A., & Gunasekera, C. N. (2018). Erythroderma and extensive poikiloderma—a rare initial presentation of dermatomyositis: A case report. *Journal of Medical Case Reports, 12*(1), 83. https://doi.org/10.1186/s13256-018-1618-y
26. Mondal, S., Roongta, R., & Sinha, D. (2021). V sign in dermatomyositis. *Archives of Medicine and Health Sciences, 9*(2), 355–356. https://doi.org/10.4103/amhs.amhs_207_21
27. Yamamoto, R., Kokubu, H., Ikuno, Y., Kato, M., Koike, T., Takahashi, T., & Fujimoto, N. (2025). Anti-transcription intermediate factor 1-γ antibody-positive dermatomyositis with refractory skin ulcers and rapid cancer progression. *The Journal of Dermatology, 52*(3), e229–e230. https://doi.org/10.1111/1346-8138.17482
28. Narang, N. S., Casciola-Rosen, L., Li, S., Chung, L., & Fiorentino, D. F. (2015). Cutaneous ulceration in dermatomyositis: Association with anti-melanoma differentiation-associated gene 5 antibodies and interstitial lung disease. *Arthritis Care & Research, 67*(5), 667–672. https://doi.org/10.1002/acr.22498
29. Wang, Y. T., Zhang, Y., Tang, T., Luo, C., Liu, M. Y., Xu, L., Wang, L., & Tang, X. M. (2022). Anti-nuclear matrix protein 2+ juvenile dermatomyositis with severe skin ulcer and infection: A case report and literature review. *World Journal of Clinical Cases, 10*(11), 3579–3586. https://doi.org/10.12998/wjcc.v10.i11.3579
30. Martínez-Díaz, L., Jiménez-de la Cruz, L., Moret-Rueda, M., & Calero-Paniagua, I. (2023). Skin ulcers associated with dermatomyositis MDA5. Úlceras cutáneas asociadas a dermatomiositis MDA5. *Medicina Clinica, 160*(3), 143. https://doi.org/10.1016/j.medcli.2022.09.021

31. Dandelooy, J., van Hal, P. T., Even, P., Lechkar, B., Siozopoulou, V., Lambert, J., & Aerts, O. (2016). Not just ordinary hand dermatitis: Mechanic's hands revealing dermatomyositis. *Journal of the European Academy of Dermatology and Venereology, 30*(7), 1223–1224. https://doi.org/10.1111/jdv.13149
32. Kanno, K., Kishibe, M., Honma, M., & Ishida-Yamamoto, A. (2025). Pathogenesis of pseudocheckerboard pattern in mechanic's hands. *The Journal of Dermatology, 52*(9), 1418–1423. https://doi.org/10.1111/1346-8138.17826
33. Mori, T., Ohashi, T., Kato, Y., Ohtsuka, M., & Yamamoto, T. (2016). Clinicopathological analysis of mechanic's hand associated with dermatomyositis. *The Australasian Journal of Dermatology, 57*(4), e123–e126. https://doi.org/10.1111/ajd.12381
34. Mormile, I., Mosella, F., Turco, P., et al. (2023). Calcinosis cutis and calciphylaxis in autoimmune connective tissue diseases. *Vaccines (Basel), 11*.
35. Davuluri, S., Chung, L., & Lood, C. (2024). Calcinosis in dermatomyositis. *Current Opinion in Rheumatology, 36*(6), 453–458. https://doi.org/10.1097/BOR.0000000000001036
36. Liu, X., & Yao, H. H. (2024). Diffuse subcutaneous calcinosis in dermatomyositis. *QJM*. https://doi.org/10.1093/qjmed/hcae045
37. Cervantes, B. A., Gowda, P., Rider, L. G., et al. (2024). Development of a computed tomography calcium scoring technique for assessing calcinosis distribution, pattern and burden in dermatomyositis. *Rheumatology (Oxford), 63*, 58–63.
38. Przekazińska-Boager, K., & Brzezicka-Ciach, U. (2014). Dermatomyositis as a manifestation of cancer. *Dermatology Review/Przegląd Dermatologiczny, 101*(1), 40–45. https://doi.org/10.5114/dr.2014.41070
39. Argobi, Y., & Smith, G. P. (2019). Tracking changes in nailfold capillaries during dermatomyositis treatment. *Journal of the American Academy of Dermatology, 81*, 275–276.
40. Xu, H., & Qian, J. (2025). The role of nailfold video-capillaroscopy in the assessment of dermatomyositis. *Rheumatology, 64*, 2987–2994.
41. Dan, J., Sprow, G., Afarideh, M., Concha, J., Kodali, N., Vazquez, T., Diaz, D., & Werth, V. (2022). Evaluation of nailfold capillaries in dermatomyositis using a dermatoscope [Abstract]. *Journal of Investigative Dermatology, 142*, S38.
42. Salgueiro, C., Poblete, M. J., Robles-Silva, C., Abarzúa, Á., & Vera-Kellet, C. (2023). Trichoscopic, oral, and periungual fold findings as activity and damage markers in dermatomyositis patients and their correlation with myositis antibodies. *Archives of Dermatological Research, 315*(6), 1603–1613. https://doi.org/10.1007/s00403-023-02554-0
43. Hamaguchi, Y., Mugii, N., Matsushita, T., & Takehara, K. (2021). Long-term changes in nail fold capillary abnormalities and serum fibroblast growth factor 23 levels in dermatomyositis patients with anti-MDA5 antibody. *Journal of Dermatology, 48*(1), 106–109. https://doi.org/10.1111/1346-8138.15589
44. Tang, K. Y., Zhang, H. L., Zhang, X. Y., & Jin, H. Z. (2024). Clinical and laboratory features between anti-TIF1γ dermatomyositis with and without malignancy: 37 case series and a review. *The Journal of Dermatology, 51*(12), 1646–1657. https://doi.org/10.1111/1346-8138.17426
45. Li, C. L., Ma, S. H., & Wu, C. Y. (2023). Anti-TIF1-γ antibody-positive dermatomyositis. *International Journal of Rheumatic Diseases, 26*(12), 2583–2584. https://doi.org/10.1111/1756-185X.14895
46. Fiorentino, D. F., Kuo, K., Chung, L., Zaba, L., Li, S., & Casciola-Rosen, L. (2015). Distinctive cutaneous and systemic features associated with antitranscriptional intermediary factor-1γ antibodies in adults with dermatomyositis. *Journal of the American Academy of Dermatology, 72*, 449–455.
47. Motegi, S. I., Sekiguchi, A., Ikeuchi, H., Sakairi, T., Ogawa, H., Fujii, T., . . . Ishikawa, O. (2020). Clinical features of anti-transcription intermediary factor 1γ (TIF1γ)-positive dermatomyositis with internal malignancy and investigation of the involvement of TIF1γ expression in tumors in the pathogenesis of cancer-associated dermatomyositis. *The Journal of Dermatology, 47*(12), 1395–1402. https://doi.org/10.1111/1346-8138.15526
48. Xu, S., Hu, X., Wang, J., et al. (2023). Polymyositis and dermatomyositis biomarkers. *Clinica Chimica Acta, 547*, 117443.
49. Ichimura, Y., Konishi, R., Shobo, M., Inoue, S., Okune, M., Maeda, A., . . . Nishino, I. (2022). Anti-nuclear matrix protein 2 antibody-positive inflammatory myopathies represent extensive myositis without dermatomyositis-specific rash. *Rheumatology, 61*(3), 1222–1227.
50. Rogers, A., Chung, L., Li, S., Casciola-Rosen, L., & Fiorentino, D. F. (2017). Cutaneous and systemic findings associated with nuclear matrix protein 2 antibodies in adult dermatomyositis patients. *Arthritis Care & Research, 69*(12), 1909–1914. https://doi.org/10.1002/acr.23210
51. Valenzuela, A., Chung, L., Casciola-Rosen, L., & Fiorentino, D. (2014). Identification of clinical features and autoantibodies associated with calcinosis in dermatomyositis. *JAMA Dermatology, 150*(7), 724–729. https://doi.org/10.1001/jamadermatol.2013.10416
52. Fredi, M., Bartoli, F., Cavazzana, I., Ceribelli, A., Carabellese, N., Tincani, A., Satoh, M., & Franceschini, F. (2017). Calcinosis in poly-dermatomyositis: Clinical and laboratory predictors and treatment options. *Clinical and Experimental Rheumatology, 35*(2), 303–308.

53. Manwatkar, A., Padiyar, S., Nair, A., Jha, A., Kumar, S., Yadav, B., Prakash, J. A. J., Das, J. K., & Mathew, J. (2023). Clinical profile of anti-NXP-2 antibody-positive inflammatory myositis and outcome in an Indian population. *Clinical Rheumatology, 42*(12), 3289–3297. https://doi.org/10.1007/s10067-023-06751-0
54. Lu, X., Peng, Q., & Wang, G. (2024). Anti-MDA5 antibody-positive dermatomyositis: Pathogenesis and clinical progress. *Nature Reviews Rheumatology, 20*(1), 48–62. https://doi.org/10.1038/s41584-023-01054-9
55. Goel, N., Sankar, J., Kumar, A., et al. (2024). MDA5 dermatomyositis—Skin says it all. *QJM, 117*(12), 868–869.
56. Wang, G., Yang, R., Yan, D., et al. (2024). Cutaneous ulcers in anti-MDA5-positive dermatomyositis with rapidly progressive interstitial lung disease: A multicentre retrospective cohort study. *Archives of Medical Science*. https://doi.org/10.5114/aoms/182885
57. MDA5 DM Consortium. (2023). *Mucocutaneous and articular manifestations of anti-MDA5 dermatomyositis* [Consortium report/abstract].
58. Xie, H., Zhang, D., Wang, Y., et al. (2023). Risk factors for mortality in patients with anti-MDA5 antibody-positive dermatomyositis: A meta-analysis and systematic review. *Seminars in Arthritis and Rheumatism, 62*, 152231. https://doi.org/10.1016/j.semarthrit.2023.152231
59. Ghazi, E., Sontheimer, R. D., & Werth, V. P. (2013). The importance of including amyopathic dermatomyositis in the idiopathic inflammatory myositis spectrum. *Clinical and Experimental Rheumatology, 31*(1), 128–134.
60. Patel, B., Khan, N., & Werth, V. P. (n.d.). *Applicability of EULAR/ACR classification criteria for dermatomyositis to amyopathic disease* [Article; publication details not provided].
61. Fornaro, M., Girolamo, F., Giannini, M., Coladonato, L., Capuano, A., Capodiferro, M., D'Abbicco, D., Ruggieri, M., Mastrapasqua, M., & Iannone, F. (2024). [Title not provided]. *Clinical and Experimental Rheumatology, 42*(2), 288–294.
62. Gan, Y. Z., Li, Y. H., Zhang, L. H., Ma, L., He, W. W., Jin, Y. B., An, Y., & Ye, H. (2020). [Comparison of clinical and immunological features between clinically amyopathic dermatomyositis and typical dermatomyositis]. *Beijing Da Xue Xue Bao Yi Xue Ban, 52*(6), 1001–1008. https://doi.org/10.19723/j.issn.1671-167X.2020.06.003
63. Tiao, J., Feng, R., Bird, S., Choi, J. K., Dunham, J., George, M., Gonzalez-Rivera, T. C., Kaufman, J. L., Khan, N., Luo, J. J., Micheletti, R., Payne, A. S., Price, R., Quinn, C., Rubin, A. I., Sreih, A. G., Thomas, P., Okawa, J., & Werth, V. P. (2017). The reliability of the Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI) among dermatologists, rheumatologists and neurologists. *British Journal of Dermatology, 176*(2), 423–430.
64. Ahmed, S., Chen, K. L., & Werth, V. P. (2020). The validity and utility of the Cutaneous Disease Area and Severity Index (CDASI) as a clinical outcome instrument in dermatomyositis: A comprehensive review. *Seminars in Arthritis and Rheumatism, 50*(3), 458–462. https://doi.org/10.1016/j.semarthrit.2020.01.002
65. Klein, R. Q., Bangert, C. A., Costner, M., Connolly, M. K., Tanikawa, A., Okawa, J., Rose, M., Fakharzadeh, S. S., Fiorentino, D., Lee, L. A., Sontheimer, R. D., Taylor, L., Troxel, A. B., & Werth, V. P. (2008). Comparison of the reliability and validity of outcome instruments for cutaneous dermatomyositis. *British Journal of Dermatology, 159*(4), 887–894. https://doi.org/10.1111/j.1365-2133.2008.08711.x
66. Danieli, M. G., Paladini, A., Longhi, E., Tonacci, A., & Gangemi, S. (2023). A machine learning analysis to evaluate the outcome measures in inflammatory myopathies. *Autoimmunity Reviews, 22*(7), 103353. https://doi.org/10.1016/j.autrev.2023.103353
67. Guo, J., Wang, W., Huang, A., & Mei, C. (2024). Pharmacological strategies in dermatomyositis: Current treatments and future directions. *Medical Science Monitor, 30*, e944564. https://doi.org/10.12659/MSM.944564
68. Sevim, E., Kobrin, D., Casal-Domínguez, M., & Pinal-Fernandez, I. (2024). A comprehensive review of dermatomyositis treatments—from rediscovered classics to promising horizons. *Expert Review of Clinical Immunology, 20*(2), 197–209. https://doi.org/10.1080/1744666X.2023.2270737
69. Bolosiu, H. D., Man, L., & Rednic, S. (1999). The effect of methylprednisolone pulse therapy in polymyositis/dermatomyositis. *Advances in Experimental Medicine and Biology, 455*, 349–357. https://doi.org/10.1007/978-1-4615-4857-7_54
70. Callen, J. P., & Wortmann, R. L. (2006). Dermatomyositis. *Clinics in Dermatology, 24*(5), 363–373. https://doi.org/10.1016/j.clindermatol.2006.07.001
71. Dawkins, M. A., Jorizzo, J. L., Walker, F. O., Albertson, D., Sinal, S. H., & Hinds, A. (1998). Dermatomyositis: A dermatology-based case series. *Journal of the American Academy of Dermatology, 38*(3), 397–404.
72. Femia, A. N., Vleugels, R. A., & Callen, J. P. (2013). Cutaneous dermatomyositis: An updated review of treatment options and internal associations. *American Journal of Clinical Dermatology, 14*(4), 291–313.
73. Lam, C., & Vleugels, R. A. (2012). Management of cutaneous dermatomyositis. *Dermatologic Therapy, 25*(2), 112–134.
74. Vleugels, R. A., & Callen, J. P. (2009). Dermatomyositis: Current and future treatments. *Expert Review of Dermatology, 4*(6), 581–594.
75. Le Clainche-Compagnie, A., Traineau, H., Teboul, A., Dupin, N., & Chasset, F. (2025). Efficacy and tolerability of hydroxychloroquine in dermatomyositis. *Journal of the European Academy of Dermatology and Venereology*. Advance online publication. https://doi.org/10.1111/jdv.70176

76. Wolstencroft, P. W., Casciola-Rosen, L., & Fiorentino, D. F. (2018). Association between autoantibody phenotype and cutaneous adverse reactions to hydroxychloroquine in dermatomyositis. *JAMA Dermatology, 154*(10), 1199–1203.
77. Pelle, M. T., & Callen, J. P. (2002). Adverse cutaneous reactions to hydroxychloroquine are more common in patients with dermatomyositis than in patients with cutaneous lupus erythematosus. *Archives of Dermatology, 138*(9), 1231–1233.
78. Rypka, K., Buonomo, M., Buechler, C., Benolken, M., Swigost, A., Konstantinov, N., Gaddis, K., & Goldfarb, N. (2023). A case series of hydroxychloroquine exacerbating the dermatomyositis rash. *Dermatology Online Journal, 29*(5). https://doi.org/10.5070/D329562407
79. Dalakas, M. C. (2011). Inflammatory myopathies: Management of steroid resistance. *Current Opinion in Neurology, 24*(5), 457–462.
80. Hornung, T., Ko, A., Tuting, T., Bieber, T., & Wenzel, J. (2012). Efficacy of low-dose methotrexate in the treatment of dermatomyositis skin lesions. *Clinical and Experimental Dermatology, 37*(2), 139–142.
81. Cobos, G. A., Femia, A., & Vleugels, R. A. (2020). Dermatomyositis: An update on diagnosis and treatment. *American Journal of Clinical Dermatology, 21*, 339–353.
82. Briemberg, H. R., & Amato, A. A. (2003). Dermatomyositis and polymyositis. *Current Treatment Options in Neurology, 5*(5), 349–356.
83. Edge, J. C., Outland, J. D., Dempsey, J. R., & Callen, J. P. (2006). Mycophenolate mofetil as an effective corticosteroid-sparing therapy for recalcitrant dermatomyositis. *Archives of Dermatology, 142*(1), 65–69. https://doi.org/10.1001/archderm.142.1.65
84. Grinnell, M., Keyes, E., Diaz, D., Vazquez, T., Feng, R., & Werth, V. P. (2022). Mycophenolate mofetil and methotrexate efficacy in dermatomyositis. *British Journal of Dermatology, 187*(3), 437–438. https://doi.org/10.1111/bjd.21235
85. Ling, G. C., Su, C., Guo, Y. A., Qiu, X., Liu, J. W., Xiao, M., Xiao, Y. Y., Yang, S., Zhang, J. Y., & Xie, J. J. (2024). Mycophenolate mofetil after tacrolimus for refractory clinically amyopathic dermatomyositis: A case report. *Frontiers in Pharmacology, 15*, 1472667. https://doi.org/10.3389/fphar.2024.1472667
86. Kurtzman, D. J. B., Wright, N. A., Lin, J., et al. (2016). Tofacitinib citrate for refractory cutaneous dermatomyositis: An alternative treatment. *JAMA Dermatology, 152*(8), 944–945. https://doi.org/10.1001/jamadermatol.2016.0866
87. Paik, J. J., Casciola-Rosen, L., Shin, J. Y., Albayda, J., Tiniakou, E., Leung, D. G., et al. (2021). Study of tofacitinib in refractory dermatomyositis: An open-label pilot study of ten patients. *Arthritis & Rheumatology, 73*(5), 858–865. https://doi.org/10.1002/art.41602
88. Chen, Z., Wang, X., & Ye, S. (2019). Tofacitinib in amyopathic dermatomyositis-associated interstitial lung disease. *The New England Journal of Medicine, 381*(3), 291–293. https://doi.org/10.1056/NEJMc1900045
89. Zhao, Q., Zhu, Z., Fu, Q., et al. (2022). Baricitinib for the treatment of cutaneous dermatomyositis: A prospective, open-label study. *Journal of the American Academy of Dermatology, 87*(6), 1374–1376. https://doi.org/10.1016/j.jaad.2022.08.025
90. Landon-Cardinal, O., Guillaume-Jugnot, P., Toquet, S., et al. (2023). JAK inhibitors for the treatment of adult dermatomyositis: A pilot study. *Journal of the American Academy of Dermatology, 88*(4), 924–926. https://doi.org/10.1016/j.jaad.2022.10.055
91. Landon-Cardinal, O., Guillaume-Jugnot, P., Bolko, L., et al. (2019). JAK inhibitors: A promising molecular-targeted therapy in dermatomyositis [Abstract]. *Arthritis & Rheumatology, 71*(Suppl. 10).
92. Ladislau, L., Suárez-Calvet, X., Toquet, S., et al. (2018). JAK inhibitor improves type I interferon induced damage: Proof of concept in dermatomyositis. *Brain, 141*(6), 1609–1621. https://doi.org/10.1093/brain/awy105
93. Jing, X., Min, W., Xia, L., & Shanshan, L. (2025). Ruxolitinib in adult dermatomyositis with anti-TIF1γ antibody: A case report and literature review. *Frontiers in Immunology, 16*, 1591631. https://doi.org/10.3389/fimmu.2025.1591631
94. Lanis, A., Kim, H., Lu, S., Tsai, W. L., Kaneshiro, A., Ehrlich, A., Martin, G., Schiffenbauer, A., & Shenoi, S. (2023). Efficacy of topical ruxolitinib for cutaneous dermatomyositis. *JAAD Case Reports, 45*, 24–26. https://doi.org/10.1016/j.jdcr.2023.09.043
95. Chung, M. P., & Paik, J. J. (2022). Past, present, and future in dermatomyositis therapeutics. *Current Treatment Options in Rheumatology, 8*(4), 71–90. https://doi.org/10.1007/s40674-022-00193-6
96. Dourmishev, L., Meffert, H., & Piazena, H. (2004). Dermatomyositis: Comparative studies of cutaneous photosensitivity in lupus erythematosus and normal subjects. *Photodermatology, Photoimmunology & Photomedicine, 20*(5), 230–234. https://doi.org/10.1111/j.1600-0781.2004.00115.x
97. Werth, V. P., et al. (n.d.). Photosensitivity in rheumatic diseases. *Journal of Investigative Dermatology Symposium Proceedings, 9*(1), 57–63.
98. Grönhagen, C. M., & Nyberg, F. (2014). Cutaneous lupus erythematosus: An update. *Indian Dermatology Online Journal, 5*(1), 7–13. https://doi.org/10.4103/2229-5178.126020
99. Habibullah, T., Habibullah, A., & Simsim, R. (2021). Skin manifestations of rheumatological diseases. In H. Almoallim & M. Cheikh (Eds.), *Skills in rheumatology* (Chapter 15). Springer.

100. Mazzoni, D., Kubler, P., & Muir, J. (2021). Recognising skin manifestations of rheumatological disease. *Australian Journal of General Practice, 50*(12), 873–878. https://doi.org/10.31128/AJGP-02-21-5863
101. Smith, R. L., Sundberg, J., Shamiyah, E., Dyer, A., & Pachman, L. M. (2004). Skin involvement in juvenile dermatomyositis is associated with loss of end row nailfold capillary loops. *The Journal of Rheumatology, 31*(8), 1644–1649.
102. LeRoy, E. C., Black, C., Fleischmajer, R., Jablonska, S., Krieg, T., Medsger, T. A., Jr., et al. (1988). Scleroderma (systemic sclerosis): Classification, subsets and pathogenesis. *The Journal of Rheumatology, 15*(2), 202–205.
103. Wigley, F. M. (2002). Clinical practice. Raynaud's phenomenon. *The New England Journal of Medicine, 347*(13), 1001–1008. https://doi.org/10.1056/NEJMcp013013
104. Ceribelli, A., Fredi, M., Taraborelli, M., Cavazzana, I., Franceschini, F., Quinzanini, M., et al. (2012). Anti-MJ/NXP-2 autoantibody specificity in a cohort of adult Italian patients with polymyositis/dermatomyositis. *Arthritis Research & Therapy, 14*(2), R97.
105. Ledo, E. (1993). Photodermatosis. Part I: Photobiology, photoimmunology, and idiopathic photodermatoses. *International Journal of Dermatology, 32*(6), 387–396.
106. Gozali, M. V., Zhou, B. R., & Luo, D. (2016). Update on treatment of photodermatosis. *Dermatology Online Journal, 22*(2), 13030/qt1rx7d228.
107. Grossberg, A. L. (2013). Update on pediatric photosensitivity disorders. *Current Opinion in Pediatrics, 25*, 474–479.
108. Nazir, S., Lohani, S., Tachamo, N., et al. (2017). Statin-associated autoimmune myopathy: A systematic review of 100 cases. *Journal of Clinical Rheumatology, 23*, 149–154. https://doi.org/10.1097/RHU.0000000000000497
109. Huda, S. A., Yadava, S., Kahlown, S., et al. (2020). Statin-induced necrotizing autoimmune myopathy. *Proceedings (Baylor University Medical Center), 34*, 185–186.
110. Selva-O'Callaghan, A., Alvarado-Cardenas, M., Pinal-Fernández, I., et al. (2018). Statin-induced myalgia and myositis: An update on pathogenesis and clinical recommendations. *Expert Review of Clinical Immunology, 14*, 215–224. https://doi.org/10.1080/1744666X.2018.1440206
111. Williams, B., Horn, M. P., Banz, Y., Feldmeyer, L., & Villiger, P. M. (2021). Cutaneous involvement in anti-HMGCR positive necrotizing myopathy. *Journal of Autoimmunity, 123*, 102691. https://doi.org/10.1016/j.jaut.2021.102691