6 Clinical Features of Myositis: Skin Manifestations

Peter B. Chansky and Victoria P. Werth

Key Points to Remember

- Dermatomyositis (DM) is a unique autoimmune disease within the family of idiopathic inflammatory myopathies that presents with characteristic cutaneous findings.
- Skin disease can present as activity (potentially reversible) with erythema, scale, erosions, or ulcerations or evolve into damage (irreversible chronic lesions) with poikiloderma or calcinosis cutis.
- Classic cutaneous manifestations of DM include the "heliotrope" rash on the eyelids, "Gottron papules or sign" on the hands/extensor surfaces, psoriasiform-like plaques on the scalp, "V sign" on the upper chest, "shawl sign" on the upper back or posterior neck/shoulders, "mechanic's hands" on the lateral or palmar sides of the fingers, "holster sign" on the lateral thighs, and nailfold changes.
- Calcinosis cutis is highly prevalent in children with juvenile DM and is associated with the anti-NXP-2 antibody in both adults and children.
- The anti-MDA5 antibody, seen in a subtype of clinically amyopathic DM, can present with palmar papules, severe cutaneous ulceration, ischemic digits, and a rapidly progressive and potentially fatal interstitial lung disease.
- Mechanic's hands are seen with antisynthetase and anti-PM-Scl autoantibodies, while anti-Mi-2 is associated with the classic rashes of DM.
- DM patients with the anti-TIF1-γ antibody often present with hyperkeratotic palmar papules, psoriasiform lesions, and telangiectatic and hypopigmented patches ("red on white").
- When considering a diagnosis of DM, it is critical to consider a broad differential because of the potential for overlapping symptoms with other connective tissue diseases.

P. B. Chansky
Corporal Michael J. Crescenz Veterans Affairs Medical Center (Philadelphia), Philadelphia, PA, USA

Department of Dermatology, Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA, USA

V. P. Werth (✉)
Corporal Michael J. Crescenz Veterans Affairs Medical Center (Philadelphia), Philadelphia, PA, USA

Department of Dermatology, Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA, USA

Department of Dermatology, Perelman Center for Advanced Medicine, Philadelphia, PA, USA
e-mail: werth@mail.med.upenn.edu

R. Aggarwal, C. V. Oddis (eds.), *Managing Myositis*, https://doi.org/10.1007/978-3-030-15820-0_6

Introduction

Dermatomyositis (DM), unique among the idiopathic inflammatory myopathies (IIM), exhibits a set of distinctive and bothersome cutaneous findings. The classic skin manifestations of DM are not seen in patients with polymyositis or necrotizing myopathy and are broadly described as being active or resulting from disease damage causing scarring. Activity reflects a potentially reversible process and is represented by varying degrees of erythema, scale, and erosion or ulceration. Damage, an irreversible finding, describes chronic skin lesions that result from scarring of active skin lesions, often represented by the presence or absence of poikiloderma and calcinosis cutis. Erythema, a sign of skin inflammation and irritation, ranges in severity from pink to red to dark red/violet and is usually the first cutaneous finding in DM. Scale (visible hyperkeratosis of the stratum corneum) is also a marker of elevated disease activity that can occur with or without erythema. Lichenification (thickening of the epidermis) indicates worsening disease activity that results from chronic and excessive rubbing due to pruritus. Poikiloderma, a characteristic dermatologic finding in both DM and cutaneous lupus, describes the stereotypical features of hypopigmentation, hyperpigmentation, telangiectasia, and epidermal atrophy, often occurring in a photo-distributed pattern. Calcinosis cutis refers to calcium deposits within the skin and is another sign of damage from cutaneous DM. Overall, the skin lesions of DM are irritating and extremely pruritic—leading to a significantly impaired quality of life [1–3]. It may help differentiate DM rashes from CLE, as they are generally less pruritic.

The cutaneous signs of DM readily occur in specific anatomic locations that are almost pathognomonic for the disease. The two most common and pathognomonic rashes of DM are the "heliotrope" rash on the eyelids (30–60% of cases) and "Gottron papules or sign" on the hands or other extensor surfaces (60–80% of cases). These rashes are included in both Bohan and Peter's classification criteria as well as the newer EULAR/ACR classification criteria for IIM [4, 5]. Most DM rashes are symmetric, helping to differentiate them from local skin reactions or infections.

Description of Individual DM Rashes

An overview of the most common cutaneous manifestations in DM is illustrated in Fig. 6.1. The face is often affected with generalized facial erythema and the hallmark "heliotrope" rash (Fig. 6.2). The facial erythema is a photosensitive phenomenon that resembles the "malar rash" seen in lupus but is distinguished in patients with DM by the involvement of the nasolabial fold (Fig. 6.2). The textbook "heliotrope" rash refers to a localized pink-to-dark red/violet eruption or erythema on the eyelids, with the upper eyelid mostly involved, and can be associated with significant periorbital edema (Fig. 6.2). This finding is particularly bothersome to patients, as it is easily visible and can be quite pronounced. Moreover, the scalp is another site of involvement and usually presents with widespread erythematous scaly psoriasiform-like plaques that can be very pruritic (Fig. 6.3). These plaques are easily mistaken for seborrheic dermatitis or psoriasis and may contribute to a misdiagnosis or delayed diagnosis of DM. Furthermore, nonscarring alopecia on the scalp is also a common manifestation in patients with DM.

The upper chest and neck/shoulder region are also common areas of DM involvement. Photodistributed erythema and poikiloderma on the upper chest are referred to as the "V sign" (Fig. 6.4), while a similar finding on the upper back and posterior neck/shoulders is referred to as the "shawl sign" (Fig. 6.5). These sun-exposed areas can initially present as activity with erythema and pruritus that later develops into damage or poikiloderma.

The hands and extensor surfaces of the upper and lower extremities can be additional sites of cutaneous involvement. Raised, erythematous papules or plaques with or without scaling of the knuckles of the dorsum of the hand describes the textbook presentation known as "Gottron pap-

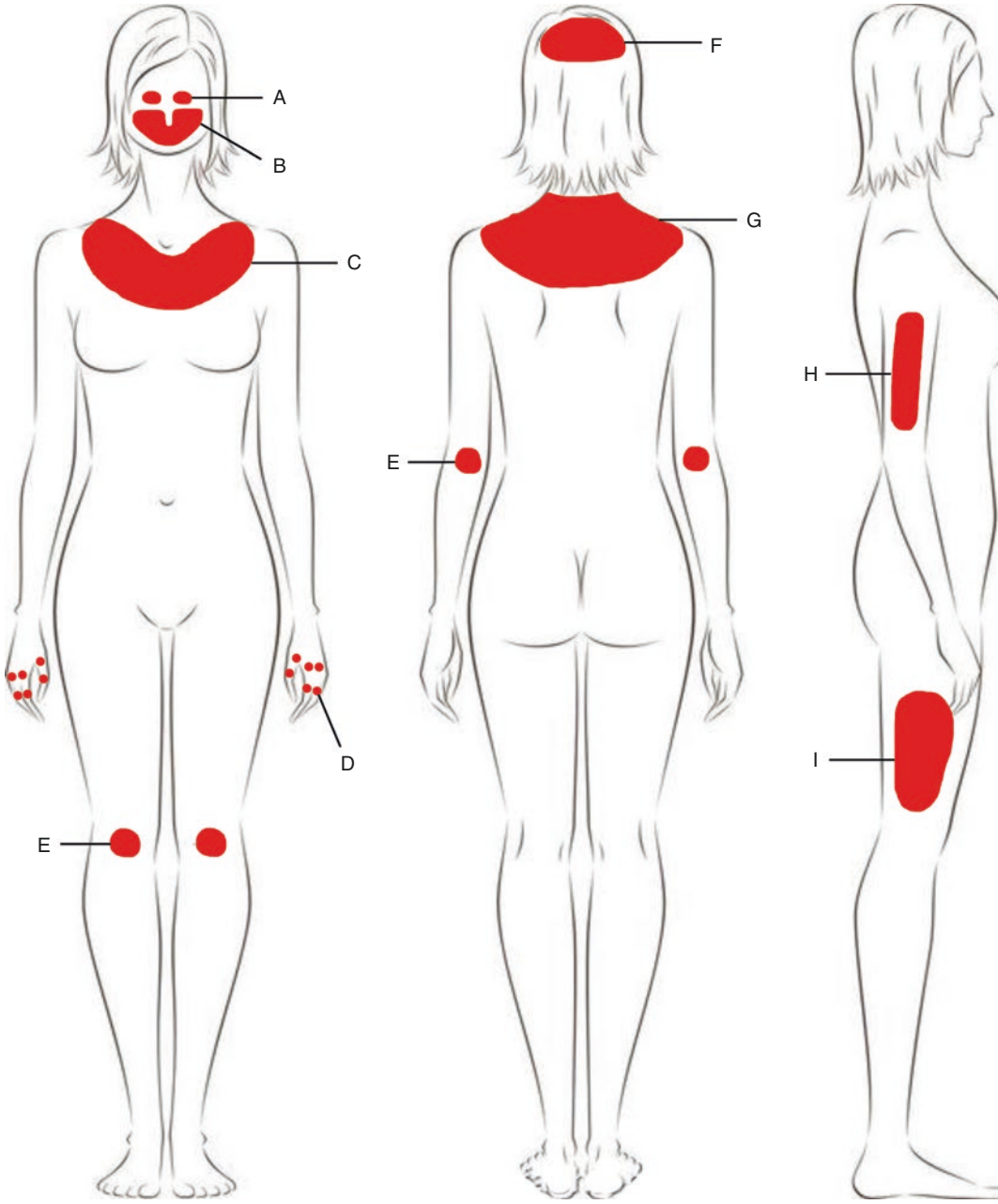

Fig. 6.1 Distribution of involvement in cutaneous DM ((**a**) heliotrope rash, (**b**) facial erythema involving nasolabial folds, (**c**) "V sign," (**d**) "Gottron papules," (**e**) "Gottron sign" on the knees and elbows, (**f**) scalp involvement, (**g**) "Shawl sign," (**h**) extensor erythema on lateral upper extremity, (**i**) "Holster sign" on lateral thigh)

ules" (Fig. 6.6). This eruption commonly involves the metacarpophalangeal joint, proximal interphalangeal joint, and distal interphalangeal joint, running linearly over the joints and tendons of the hand. In some patients, the erythema can occur on the dorsum of the hand and between the joints on the fingers. In particularly severe cases, patients may present with erosions, ulcerations, and scale. When erythematous, scaly macules are found in the same distribution over the joints of

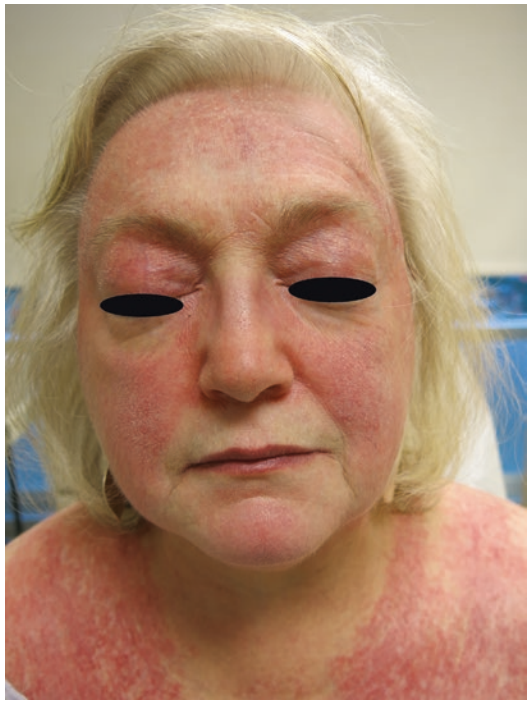

Fig. 6.2 Facial erythema with "heliotrope rash" and eyelid edema

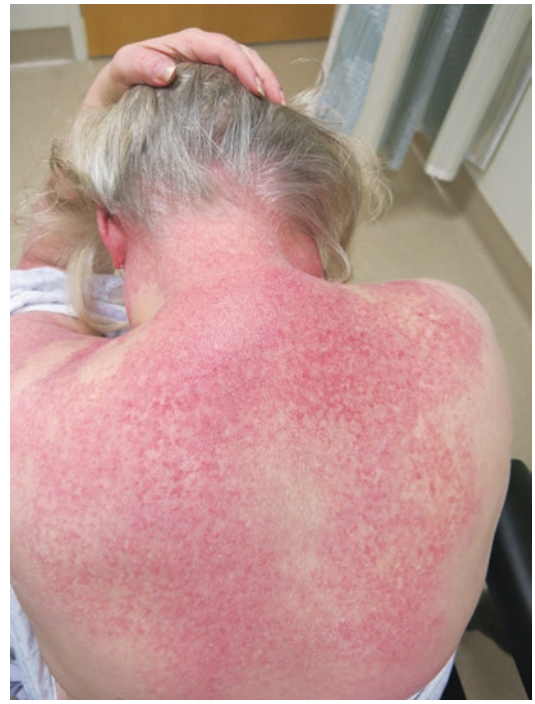

Fig. 6.5 Erythema and poikiloderma on the upper back ("Shawl sign")

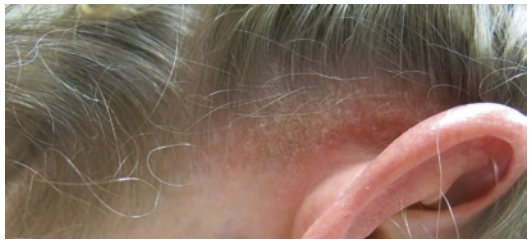

Fig. 6.3 Scaly, psoriasiform-like plaques on the scalp

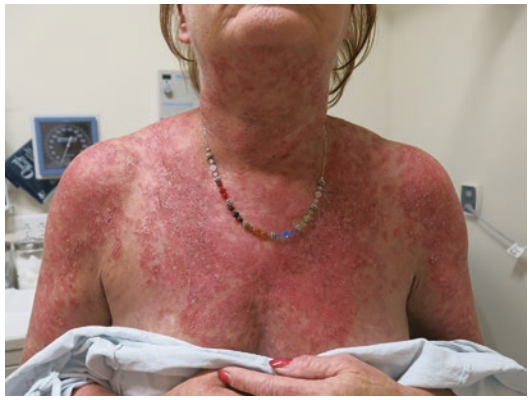

Fig. 6.4 Photodistributed erythema and poikiloderma on the chest ("V sign")

the hands or over the extensor surface of the elbows, knees, and ankles, it is known as "Gottron sign" (Fig. 6.7).

Finally, the lateral and palmar sides of the fingers may exhibit hyperkeratosis, lichenification, erythema, and scale known as "mechanic's hands" (Fig. 6.8). These features combine to produce horizontal fissuring, cracking, and lines on the skin that resemble the coarse hands of someone working in an industrial job or labor-intensive industry and occur most commonly on the radial aspect of the index and middle fingers and the ulnar aspect of the thumb. "Mechanic's hands" are frequently observed in patients with anti-synthetase autoantibodies and are reported in up to 70% of these patients [6] but have also been seen in patients possessing anti-PM-Scl autoantibodies as well as patients with classic and amyopathic DM without lung involvement.

The lower extremities can also be affected in DM. Erythema and scale on the lateral thighs is known as the "holster sign" (Fig. 6.9). These areas can exhibit poikiloderma. A similar, less

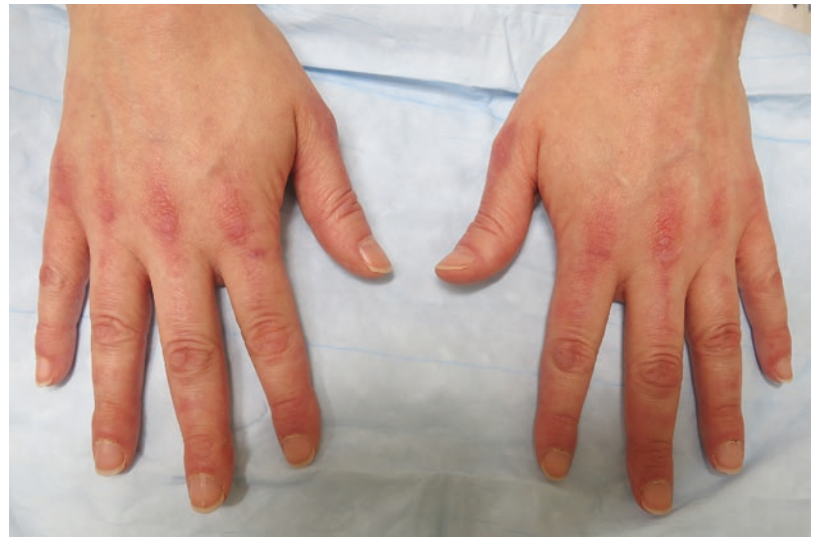

Fig. 6.6 Erythematous papules on dorsal knuckles ("Gottron papules")

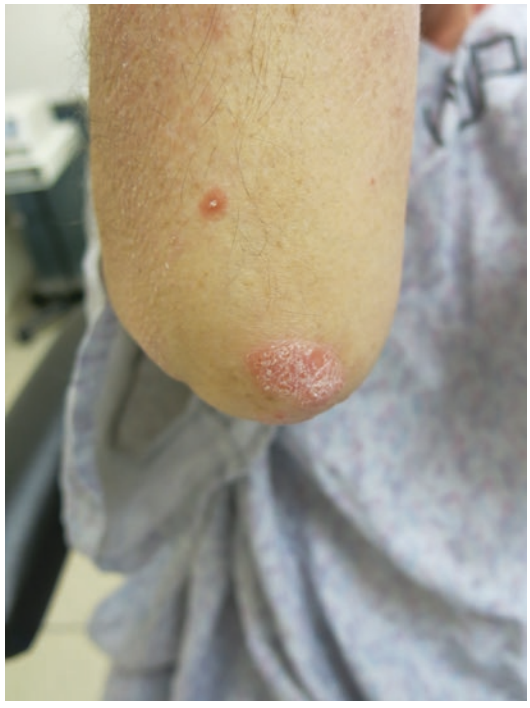

Fig. 6.7 Erythematous scaly plaque on the elbow ("Gottron sign")

common rash can be seen on the upper arms (Fig. 6.10). It is not well understood why this dermatologic finding presents in a traditionally sun-protected area of the body.

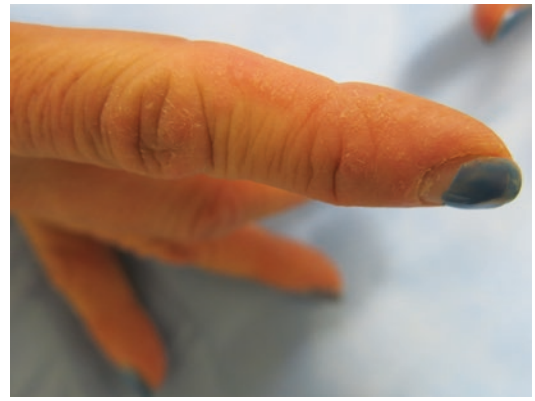

Fig. 6.8 Hyperkeratosis, lichenification, erythema, and scale on the sides of the finger ("mechanic's hands")

The presence of nailfold changes such as cuticular dystrophy, visible telangiectasias, and nailfold capillary dilation and dropout is also characteristic of DM (Fig. 6.11). Overgrowth of the nail beds can give the cuticles a classic "ragged" appearance. The nailbed capillary network may become dilated and visible with either the naked eye or a dermatoscope. Consequently, the enlarged vessels produce erythema around the cuticles termed periungual erythema, which, together with overgrowth, can be quite bothersome for

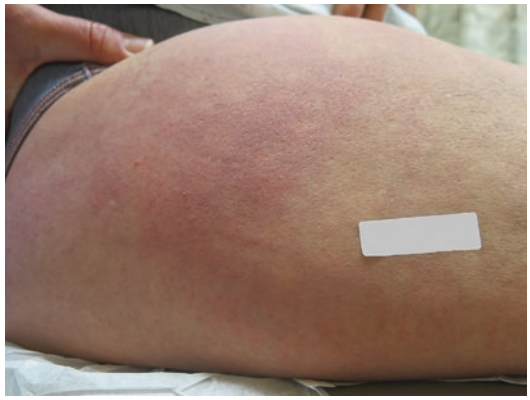

Fig. 6.9 Erythema and scale on the lateral thigh ("Holster sign")

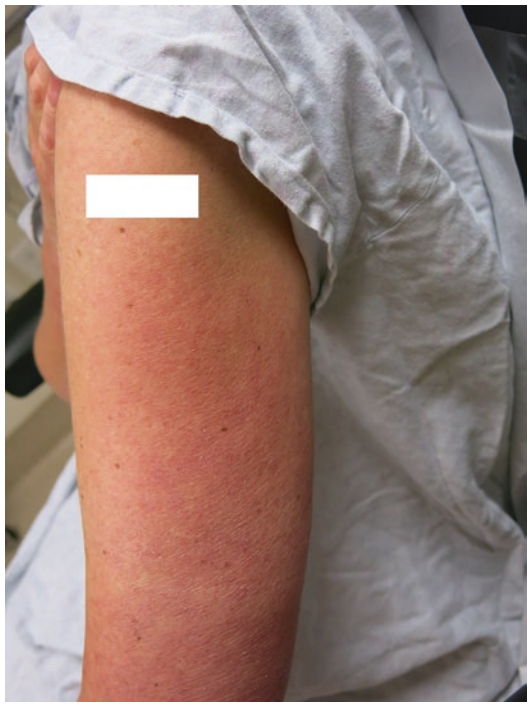

Fig. 6.10 Extensor erythema on the lateral upper arm

patients. The severity of the nailfold changes reflects disease activity, particularly in juvenile DM [7].

In several small studies, cutaneous ulceration as the early presentation of DM has been reported to reflect more severe disease or an underlying malignancy [8–10]; however, many DM patients with ulcers do not have cancer. Ulcerations can be associated with cutaneous vasculitis, calcinosis, panniculitis, or local microtrauma.

Fig. 6.11 Cuticular dystrophy ("ragged" cuticles) and visible telangiectasias on distal fingers

Calcinosis Cutis and Other Uncommon Rashes

Calcinosis cutis—the accumulation of calcium into hard nodules beneath the skin—occurs in intracutaneous, subcutaneous, fascial, or intramuscular locations, with a predilection for sites subjected to repeated microtrauma (the elbows, knees, flexor surfaces of the fingers, and buttocks). It is reported in up to 70% of children with juvenile DM (JDM) but is far less prevalent (approximately 20%) in adult DM patients [11–13]. Calcinosis usually develops in the upper extremities, such as the shoulders, arms, and hands, and is particularly resistant to treatment. It is linked to the duration of untreated disease as well as disease severity [14] and an increased risk for malignancy when associated with the anti-NXP-2 antibody [15]. Calcinosis leads to pain and functional compromise, particularly if the deposits are large and adjacent to a joint. Subsequent complications include extrusion of calcium deposits, ulceration, and infection of the overlying skin. Calcinosis cutis is also seen in other systemic autoimmune rheumatic disorders such as the limited form of systemic sclerosis (CREST). Thus, this physical exam finding requires a broad differential diagnosis. In JDM and DM, calcinosis is associated with anti-NXP-2 antibody.

Beyond the classic cutaneous findings discussed above, DM can also present with other less frequent skin manifestations. These include flagellate erythema, vesicular and bullous lesions, panniculitis, small vessel vasculitis, ichthyosis, widespread erythroderma, subcutaneous edema, and lipoatrophy. Flagellate erythema describes a specifically linear and streak-like distribution on the skin.

Rashes Associated with Clinically Amyopathic Dermatomyositis

A unique subtype of DM is clinically amyopathic DM (CADM), seen in approximately 20% of all DM cases in the USA [16]. These patients may have subtle signs of muscle involvement such as mildly elevated muscle enzymes and/or mild myopathic EMG or muscle biopsy abnormalities. Some CADM patients possess a unique autoantibody, termed anti-MDA5 antibody. This autoantibody has a characteristic cutaneous phenotype that includes palmar papules (Gottron papules but on the palmar side of the hand) (Fig. 6.12) with severe cutaneous ulcerations (Fig. 6.13) and ischemic digits sometimes leading to gangrene. It is very important to recognize these rashes, as up to 50% of such patients may present with or develop severe, rapidly progressive ILD, which portends a poor prognosis [17].

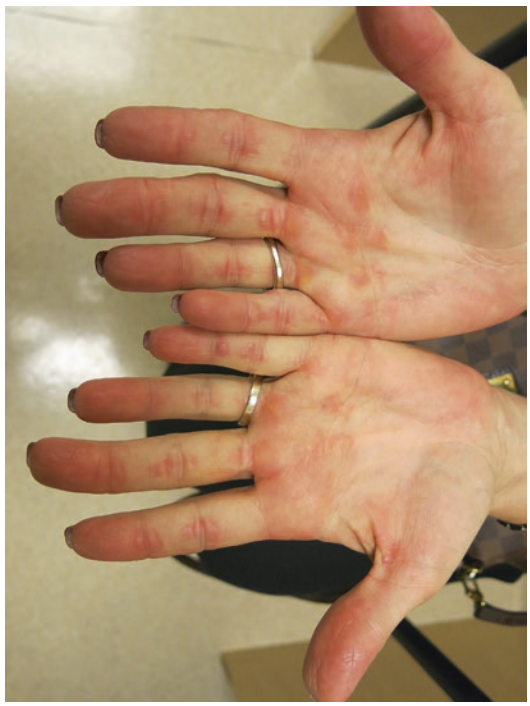

Fig. 6.12 Palmar papules associated with anti-MDA5 subtype of DM

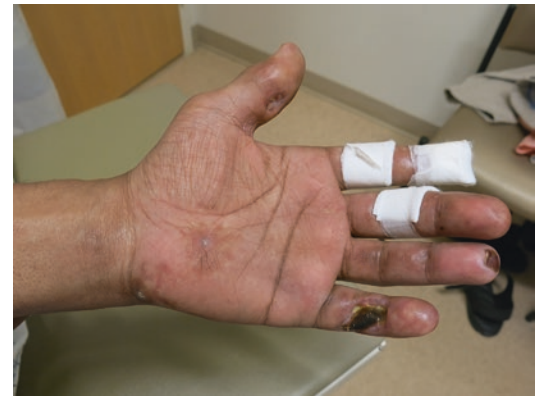

Fig. 6.13 Cutaneous ulceration associated with anti-MDA5 subtype of DM

Autoantibody Association of the Dermatomyositis Rashes

DM is associated with specific rashes and certain autoantibodies as seen with MDA-5 as described above. Similarly, mechanic's hands are seen with anti-synthetase and anti-PM-Scl autoantibodies, while anti-Mi-2 is associated with the classic rashes of DM including the heliotrope rash, Gottron's changes, the "shawl" and "V-neck" sign, cuticular overgrowth, and photosensitivity. Furthermore, DM patients with anti-TIF1-γ antibodies are more likely to demonstrate certain DM-specific cutaneous rashes including hyperkeratotic palmar papules, psoriasiform lesions, and the unique finding of telangiectatic and hypopigmented patches ("red on white") [18].

Differential Diagnosis

Ultimately, establishing a diagnosis of DM requires an astute dermatologist or a rheumatologist or neurologist with training or experience in DM, who can recognize many of the subtle features and characteristic distributions of the cutaneous manifestations of this disease. Overlapping

symptoms of other systemic autoimmune rheumatic disorders, such as rheumatoid arthritis, mixed connective tissue disease, Sjogren syndrome, systemic sclerosis, and subacute cutaneous lupus erythematosus, can contribute to a confusing clinical picture and an incorrect diagnosis.

A broad differential is important whenever considering a diagnosis of dermatomyositis. The appearance of a heliotrope rash must be evaluated for an allergic contact dermatitis or periorbital eczema. The facial erythema and malar rash seen in DM could be a sign of systemic lupus erythematosus (SLE). The differential for periungual erythema and visible nailfold telangiectasias includes scleroderma and less commonly SLE. The finding of photodistributed poikiloderma could also be seen in SLE, scleroderma, and rarely cutaneous T-cell lymphoma. The finding of erythematous scaly plaques on the extensor surface of the elbows, knees, and scalp could also present as psoriasis. Lastly, one must include the diagnosis of multicentric reticulohistiocytosis (MRH) and knuckle pads whenever considering the finding of Gottron papules on the joints of the dorsal hand.

The role of skin biopsy and its interpretation is discussed separately and may not be required in a typical DM case with classic rashes and confirmed muscle involvement. However, given the broad differential presented by the DM rashes discussed above, and in cases of less typical DM rashes, a skin biopsy may confirm one's clinical suspicion (Table 6.1).

Table 6.1 Characteristics of typical DM rashes

DM rash	Typical location	Frequency	Clinical association	Common differential	Figure
Gottron papules	Dorsum of the hands over MCP, PIP, and DIP joints, B/L	Common, ~70% [19]	Pathognomonic rash of DM	MRH, knuckle pads	Figure 6.6
Palmar papules	Palms, B/L	Rare	Associated with anti-MDA5 antibody	Callus	Figure 6.12
"V sign"	Upper chest, B/L	Common, ~83% [20]	All types of DM	Photodistributed drug eruption, SLE	Figure 6.4
"Shawl sign"	Upper back, posterior neck, and shoulders B/L	Common, ~63% [20]	All types of DM	Photodistributed drug eruption, SLE	Figure 6.5
"Holster sign"	Lateral thigh, B/L	Less common, ~28% [20]	All types of DM	Bruise, SLE	Figure 6.9
Nailfold capillary changes with cuticular overgrowth	Cuticles of fingernails, B/L	Common, capillary changes ~70%, cuticular overgrowth ~35% [20]	All types of DM	Scleroderma, SLE	Figure 6.11
Calcinosis	Shoulder girdle, elbows, hands, B/L	20–70% [11–13]	Associated with JDM and anti-NXP-2 autoantibodies	Scleroderma (CREST)	N/A
Mechanic's hands	Lateral and palmar side of the digits of the hand, B/L	Common, ~48% [20]	Associated with anti-synthetase and anti-PM-scl autoantibodies	Hand dermatitis, allergic contact dermatitis	Figure 6.8
Ulceration	Dorsal and/or ventral side of the hand and digits, B/L	Rare	Associated with anti-MDA5 antibody	Scleroderma, diabetes, chronic infection	Figure 6.13
Scalp	Scalp, all quadrants	Common, ~70% [20]	All types of DM	Psoriasis, seborrheic dermatitis	Figure 6.3
Heliotrope	Eyelids, B/L	30–60% [19, 20]	All types of DM	Contact dermatitis, eczema	Figure 6.2

References

1. Armadans-Tremolosa I, Selva-O'Callaghan A, Visauta-Vinacua B, Guilera G, Pinal-Fernandez I, Vilardell-Tarres M. Health-related quality of life and Well-being in adults with idiopathic inflammatory myopathy. Clin Rheumatol. 2014;33(8):1119–25.
2. Hundley JL, Carroll CL, Lang W, Snively B, Yosipovitch G, Feldman SR, et al. Cutaneous symptoms of dermatomyositis significantly impact patients' quality of life. J Am Acad Dermatol. 2006;54(2):217–20.
3. Goreshi R, Chock M, Foering K, Feng R, Okawa J, Rose M, et al. Quality of life in dermatomyositis. J Am Acad Dermatol. 2011;65(6):1107–16.
4. Bohan A, Peter JB. Polymyositis and dermatomyositis (first of two parts). N Engl J Med. 1975;292(7):344–7.
5. Bohan A, Peter JB. Polymyositis and dermatomyositis (second of two parts). N Engl J Med. 1975;292(8):403–7.
6. Uribe L, Ronderos DM, Diaz MC, Gutierrez JM, Mallarino C, Fernandez-Avila DG. Antisynthetase antibody syndrome: case report and review of the literature. Clin Rheumatol. 2013;32(5):715–9.
7. Christen-Zaech S, Seshadri R, Sundberg J, Paller AS, Pachman LM. Persistent association of nailfold capillaroscopy changes and skin involvement over thirty-six months with duration of untreated disease in patients with juvenile dermatomyositis. Arthritis Rheum. 2008;58(2):571–6.
8. Ponyi A, Constantin T, Garami M, Andras C, Tallai B, Vancsa A, et al. Cancer-associated myositis: clinical features and prognostic signs. Ann N Y Acad Sci. 2005;1051:64–71.
9. Mautner GH, Grossman ME, Silvers DN, Rabinowitz A, Mowad CM, Johnson BL Jr. Epidermal necrosis as a predictive sign of malignancy in adult dermatomyositis. Cutis. 1998;61(4):190–4.
10. Mahe E, Descamps V, Burnouf M, Crickx B. A helpful clinical sign predictive of cancer in adult dermatomyositis: cutaneous necrosis. Arch Dermatol. 2003;139(4):539.
11. Shimizu M, Ueno K, Ishikawa S, Yokoyama T, Kasahara Y, Yachie A. Cutaneous calcinosis in juvenile dermatomyositis. J Pediatr. 2013;163(3):921.
12. Muller SA, Winkelmann RK, Brunsting LA. Calcinosis in dermatomyositis; observations on course of disease in children and adults. AMA Arch Derm. 1959;79(6):669–73.
13. Weinel S, Callen JP. Calcinosis cutis complicating adult-onset dermatomyositis. Arch Dermatol. 2004;140(3):365–6.
14. Fisler RE, Liang MG, Fuhlbrigge RC, Yalcindag A, Sundel RP. Aggressive management of juvenile dermatomyositis results in improved outcome and decreased incidence of calcinosis. J Am Acad Dermatol. 2002;47(4):505–11.
15. Rogers A, Chung L, Li S, Casciola-Rosen L, Fiorentino DF. The cutaneous and systemic findings associated with nuclear matrix protein-2 antibodies in adult dermatomyositis patients. Arthritis Care Res (Hoboken). 2017;69:1909.
16. Bendewald MJ, Wetter DA, Li X, Davis MD. Incidence of dermatomyositis and clinically amyopathic dermatomyositis: a population-based study in Olmsted County, Minnesota. Arch Dermatol. 2010;146(1):26–30.
17. Moghadam-Kia S, Oddis CV, Sato S, Kuwana M, Aggarwal R. Anti-melanoma differentiation-associated gene 5 is associated with rapidly progressive lung disease and poor survival in US patients with amyopathic and myopathic dermatomyositis. Arthritis Care Res (Hoboken). 2016;68(5):689–94.
18. Fiorentino DF, Kuo K, Chung L, Zaba L, Li S, Casciola-Rosen L. Distinctive cutaneous and systemic features associated with antitranscriptional intermediary factor-1gamma antibodies in adults with dermatomyositis. J Am Acad Dermatol. 2015;72(3):449–55.
19. Kovacs SO, Kovacs SC. Dermatomyositis. J Am Acad Dermatol. 1998;39(6):899–920. quiz 1–2.
20. Chansky PB, Olazagasti JM, Feng R, Werth VP. Cutaneous dermatomyositis disease course followed over time using the cutaneous dermatomyositis disease area and severity index (CDASI). J Am Acad Dermatol. 2018;79:464–9.