A Single Center Retrospective Analysis of Inflammatory Dermatologic Diagnoses and Misdiagnoses Associated with Dermatomyositis

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Introduction

- Dermatomyositis (DM) is an idiopathic inflammatory myopathy with distinct skin symptoms.
- Accurate diagnosis of DM's skin manifestations is crucial for timely screening of comorbidities like interstitial lung disease and malignancy[1,2].
- The protean nature of DM's skin manifestations often leads to misdiagnosis.[3]
- DM patients may also experience increased rates of other skin conditions due to immunological disruptions from underlying autoimmune disease or systemic treatments.[4]

Objective

To determine the prevalence of dermatologic and SLE related diagnoses among individuals with DM and to characterize the misdiagnosis process, particularly concerning DM-mimicking diseases, including SLE, eczema, psoriasis, rosacea, among others.

Methods

Retrospective cohort study of 167 dermatomyositis (DM) from the Johns Hopkins Myositis Center registry. Epic™ diagnoses related to dermatologic or SLE-related ICD codes were identified. Each diagnosis was assigned to one of 40 relevant disease groupings, 20 of which were deemed inflammatory. Patient demographics and clinical misdiagnosis were determined through retrospective chart review. A diagnosis was classified as a potential misdiagnosis if it occurred before DM diagnosis and disappeared after DM diagnosis. Each misdiagnosis was confirmed through careful chart review.

Inclusion criteria:
- Adult-onset dermatomyositis
- > 1 Johns Hopkins dermatology visit
- 1 or more dermatological (LXXX or skin cancer C44.XX) or SLE-related (M32.X) ICD code.

Exclusion criteria:
- Juvenile DM and mixed connective tissue disorder.

Results

Table 1: Cohort demographics

Total cohort size	167
Age at diagnosis, years (mean, SD)	48.6 (13.9)
Sex (n, %)	
Male	32 (20%)
Female	135 (80%)
Race (n, %)	
White	115 (69%)
Black	32 (19%)
Asian	14 (8%)
Hispanic	7 (4%)
Cancer-associated DM[a] (n, %)	
Yes	23 (14%)
No	144 (86%)
Interstitial Lung Disease (n, %)	
Yes	70 (42%)
No	97 (58%)
Muscle involvement (n, %)	
Myopathic	106 (63%)
Amyopathic/hypomyopathic	61 (37%)

Table 2: Misdiagnosis of inflammatory or SLE diagnoses in patients with DM

Disease group	Prevalence (%)	Pre-DM diagnosis (%)	Misdiagnosis (% of pre-DM)	True Prevalence (% of cohort)	Misdiagnosed (% of cohort)	Misdiagnosis, days, average (SD)[b]	Misdiagnosis, days, median
Eczema	60 (36%)	23 (38%)	16 (70%)	44 (26%)	16 (10%)	463 (659)	239
other	29 (17%)	12 (41%)	9 (75%)	20 (12%)	9 (5%)	694 (798)	241
contact	18 (11%)	6 (33%)	5 (83%)	13 (8%)	5 (3%)	173 (137)	194
seborrheic	16 (10%)	2 (13%)	0 (0%)	14 (8%)	0 (0%)	N/A	N/A
atopic	13 (8%)	7 (54%)	2 (29%)	11 (7%)	2 (1%)	154 (116)	154
Dermatitis unspecified[c]	52 (31%)	15 (29%)	11 (73%)	41 (25%)	11 (7%)	182 (179)	109
Rosacea	17 (10%)	7 (41%)	3 (43%)	14 (8%)	3 (2%)	122 (75)	114
SLE	15 (9%)	10 (29%)	7 (70%)	8 (5%)	7 (4%)	417 (735)	71
Psoriasis	14 (8%)	4 (29%)	3 (75%)	11 (7%)	3 (2%)	84 (36)	98
Acneiform	13 (8%)	0 (0%)	0 (0%)	13 (8%)	0 (0%)	N/A	N/A
Urticaria	10 (6%)	3 (30%)	1 (33%)	9 (5%)	1 (1%)	31 (0)	31
CLE	7 (4%)	5 (71%)	4 (80%)	3 (2%)	4 (2%)	353 (324)	236
Lichenoid[d]	6 (4%)	3 (50%)	1 (33%)	5 (3%)	1 (1%)	461 (0)	461
Granulomatous	5 (3%)	2 (40%)	0 (0%)	5 (3%)	0 (0%)	N/A	N/A
SJS/TEN/EM	3 (2%)	1 (33%)	1 (100%)	2 (1%)	1 (1%)	11 (0)	11
Dermatitis, interface	3 (2%)	1 (33%)	0 (0%)	3 (2%)	0 (0%)	N/A	N/A
Vasculitis	2 (1%)	2 (100%)	0 (0%)	2 (1%)	0 (0%)	N/A	N/A
Panniculitis	1 (1%)	0 (0%)	0 (0%)	1 (1%)	0 (0%)	N/A	N/A
Pemphigoid	1 (1%)	1 (100%)	0 (0%)	1 (1%)	0 (0%)	N/A	N/A
Sclerotic	1 (1%)	0 (0%)	0 (0%)	1 (1%)	0 (0%)	N/A	N/A
Neutrophilic Dermatosis	1 (1%)	0 (0%)	0 (0%)	1 (1%)	0 (0%)	N/A	N/A

Table 3: Cohort outcomes

Mean dermatologic or SLE diagnoses (SD)	3.4 (2.6)
Misdiagnosis* (n, %)	
Yes	41/167 (25%)
Myopathic	21/106 (19.8%)
Amyopathic/hypomyopathic	20/61 (32.8%)
No	126 (75%)
Mean duration of misdiagnosis, days (SD)	344 (546)
Median duration of misdiagnosis, days	142

Table 4: Biopsy outcomes of patients with misdiagnosis, n = 48 (29%)

Disease	No biopsy	Spongiotic	Interface	Perivascular infiltrate	Total biopsies (% of disease group)
Eczema	5	1	9	1	11 (69%)
other	2	0	5	1	6 (75%)
atopic	1	0	2	0	2 (66%)
contact	2	1	2	0	3 (60%)
Dermatitis unspecified	1	0	9	1	10 (90%)
Rosacea	1	0	2	0	2 (66%)
SLE	1	0	6	0	6 (86%)
Psoriasis	1	0	1	1	2 (66%)
Urticaria	0	0	1	0	1 (100%)
CLE	0	0	4	0	4 (100%)
Lichenoid	0	0	1	0	1 (100%)
SJS/TEN/EM	1	0	0	0	0 (0%)

Discussion

Frequent diagnoses:
- Eczema (36% of patients), dermatitis unspecified (31%), rosacea (10%), SLE (8%), and psoriasis (8%) were the most frequently diagnosed conditions in DM patients.
- Eczema other (17%) was the most diagnosed form of eczema (eczema unspecified, nummular, eczematous dermatitis, etc).

Misdiagnosis among conditions diagnosed pre-DM :
- CLE (80% misdiagnosed), psoriasis (75%), dermatitis unspecified (73%), eczema (70%), and SLE (70%) had the highest rates of misdiagnosis among inflammatory conditions pre-DM diagnosis.
- Contact eczema was misdiagnosed most frequently (83%) among eczema subtypes, suggesting DM eruptions may clinically resemble this localized process.
- 6 patients had more than one misdiagnosis.
- CLE, eczema, and SLE had the longest durations of misdiagnosis, indicating diagnostic difficulties differentiating these diseases from DM.

Biopsies:
- Skin biopsies often led to diagnosis revision, though some showed unexpected findings like spongiotic dermatitis rather than classic DM interface dermatitis.
- Clinical-pathologic correlation aids diagnosis of complex skin diseases but remains imperfect.

Conclusions and Limitations

Conclusions:
- Our single center study reveals a high burden of inflammatory skin diseases among DM patients, with eczema, dermatitis, and rosacea most prevalent.
- Common inflammatory dermatologic diseases (eczema, psoriasis, rosacea) may be more prevalent in DM patients compared to the general population.
- CLE, SLE, psoriasis, contact eczema, and dermatitis were the most misdiagnosed conditions, with misdiagnosis lasting over a year in many cases.
- Skin biopsies often drove true diagnosis, though histopathologic exceptions exist.
- Provider education and tailoring the diagnostic differential may enable more timely and accurate diagnosis of DM's varied presentations.

Limitations:
- Single center retrospective design
- Diagnoses that came after DM diagnosis were not analyzed
- Not all misdiagnoses had skin biopsies

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[a] Cancer diagnosis +/- 5 years of DM diagnosis.
[b] Days between first instance of disease group diagnosis and confirmation of DM.
[c] Epic diagnoses: dermal hypersensitivity reaction, pruritic rash, intertrigo, chronic dermatitis, dermatitis, facial dermatitis, hand dermatitis, periorbital dermatitis, vulvar dermatitis, radiation dermatitis, perioral dermatitis, periorificial dermatitis
[d] Non-DM manifestations. Epic diagnoses: lichenoid dermatitis, lichen planus, oral lichen planus, lichen planopilaris, lichen sclerosus.

References:
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2. Olazagasti JM et al. PMID: 25721032
3. Yang SH et al. PMID: 32743506
4. Hornung T, Wenzel J. PMID: 24939511