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Characteristics of patients with dermatomyositis from 2011-2021 at a tertiary care center

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To the Editor:

Herein, we present a retrospective chart review study assessing all adult (>18 years of age) patients diagnosed with dermatomyositis at the University of California, Davis (UC Davis) in Sacramento, California between 2011-2021. This study is designed to better characterize the variable manifestations and associations of this protean disease.

Dermatomyositis (DM) is the most common type of idiopathic inflammatory myopathy. It is defined by both highly reproducible and distinct skin manifestations along with varying systemic manifestations [1]. Owing to its heterogeneity, the true prevalence is difficult to determine, but some estimates show annual incidence of 1.9-7.7/1,000,000 persons and prevalence between 1/10,000-50,000 persons [2].

This autoimmune disease can impact physiologic systems throughout the body and may even be associated with internal malignancy. The cutaneous manifestations of dermatomyositis can be defined as pathognomonic, characteristic, compatible, less common, rare, recently-described, and nonspecific [1]. Gottron papules, Gottron sign, and heliotrope rash are considered pathognomonic, whereas nail fold changes, shawl sign, V-sign, Holster sign, and scalp involvement are characteristic. Compatible manifestations include poikiloderma, periorbital edema, and facial swelling. Less common findings include vesiculobullous, necrotic or ulcerative lesions, cutaneous vasculitis, and calcinosis cutis. Rare manifestations are mechanic's hands, flagellate erythema, deck chair sign, follicular hyperkeratosis, panniculitis, mucinosis, erythroderma, and oral mucosal changes. Raynaud phenomenon is considered a nonspecific finding [1]. Systemic manifestations are variable, including myopathy, which is found in 80% of diagnosed patients, cardiopulmonary involvement, gastrointestinal disease, and malignancy [1]. Autoantibodies can aid in the clinical diagnosis and can help predict phenotype, though testing remains variable [2]. Characterization of diverse patient populations, like that of UC Davis, can help define the variable presentations of dermatomyositis to inform clinicians and researchers and to improve patient care.

Methods

This study was approved by the UC Davis Institutional Review Board. Patients aged 18 or greater with an ICD 10 (M33.10) diagnosis of dermatomyositis from May 1st, 2011 to May 1st, 2021 were selected from electronic medical records. A total of 185 charts resulted and 21 were excluded based on diagnosis of juvenile dermatomyositis prior to age 18. Eight individuals were excluded due to physician-documented rule-out of dermatomyositis. The remaining 156 individuals were included in

statistical analysis. Chi-square, fisher exact and two sample t-tests compared variables such as age, pulmonary involvement, and frequency of malignancy.

Results

Demographics

The mean age of our study cohort was 59.4 years (SD 17.5). Two-thirds were female (68%, 106/156), consistent with prior reports [3] and the majority (55.8%, 87/156) were White, followed by 13.5% (21/156) Hispanic or Latino, 11.5% (18/156) African American or Black, 5% (8/156) Asian, and 14% Unknown (**Table 1**).

Table 1. *Demographics of patients with dermatomyositis.*

Demographic	Value (%) N=156
Age (years)	Mean 59.4
Legal sex:	
Female	106 (68.0)
Male	50 (32.1)
Race/ethnicity:	
African American or Black	18 (11.5)
Asian	8 (5.13)
Hispanic or Latino	21 (13.5)
White	87 (55.8)
Unknown	22 (14.1)

Care setting

Most diagnoses were made in the outpatient setting (85.3%, 133/156). Strikingly, only half of patients received care from a dermatologist throughout the course of their disease (53.2%, 83/156). In our center, the diagnosis of dermatomyositis was most often made by a rheumatologist (43.6%), followed by a dermatologist (30.8%), and an internist (11.5%).

Past medical history

Concomitant autoimmune disease was common in our population (44%, 69/156) and one in five had a family history of autoimmune disease (21.8%, 34/156) (**Table 2**). In our population, 13.5% (21/156) had metabolic syndrome defined by having diabetes, hypertension, and dyslipidemia prior to the diagnosis of DM. When tested and documented in the electronic medical record, 62% (46/74) of patients had a vitamin D deficiency prior to their DM diagnosis. As anticipated [4], a significant proportion also had a history of tobacco use prior to DM diagnosis.

Table 2. *Past medical history.*

Pre-existing medical conditions	Number (%) N=156
Type 2 diabetes mellitus	37 (23.7)
Hypertension	84 (53.8)
Dyslipidemia	56 (35.9)
GERD	57 (36.5)
Depression	39 (25)
History of tobacco use	53 (34.0)
Other autoimmune disease	69 (44.2)
Family history of autoimmune disease	34 (21.8)

Cutaneous findings

As noted above, cutaneous findings in DM can be pathognomonic, characteristic, compatible, less common, rare, recently-described, and nonspecific [1]. In our study, the most common DM findings documented were Gottron papules (pathognomonic), (50.6% of patients), followed by nail fold changes (characteristic), (29.4%) including capillary abnormalities, erythema, and ragged cuticles. These changes were followed by the characteristic V-sign (26.3%) and shawl signs (23.1%). Less commonly reported were heliotrope rash (19.9%), Gottron sign (16.7%), holster sign (7.7%), and calcinosis cutis (2%), (**Table 3**). Of note, when complement levels were tested, 94.1 % (16/17) of patients with low complement C3 or C4 levels had evidence of skin disease.

Table 3. *Cutaneous findings.*

Cutaneous finding	Number (%) N=156
Gottron's papules	79 (50.6)
Nail fold changes	46 (29.5)
Heliotrope rash	45 (28.8)
V-sign	42 (26.9)
Shawl sign	36 (23.1)
Gottron sign	26 (16.7)
Scalp changes	20 (12.8)
Holster sign	12 (7.7)
Mechanic's hands	12 (7.7)
Calcinosis cutis	3 (1.9)

Evidence of myositis

DM can be classified as myopathic or clinically/laboratory amyopathic based on the presence of both weakness and evidence of myopathy in laboratory and imaging data [5]. Two-thirds of patients demonstrated weakness on examination (64.1%, 100/156) and 88.2% (15/17) of patients with low complement C3 or C4 levels were myopathic with clinical or laboratory evidence of muscle destruction, consistent with hypomyopathic DM [5]. The remaining two patients were not tested. The other 35.9% (56/156) of our population was clinically amyopathic. When tested, 37.5% (21/56) of this group had evidence of muscle destruction by elevated muscle enzymes, electromyography, and/or muscle magnetic resonance imaging (MRI) consistent with dermatomyositis. Clinically amyopathic patients warrant further investigation to assess these subtle signs of disease activity.

Myopathy can be assessed in both non-invasive and invasive ways. Non-invasive testing includes laboratory evaluation of creatinine kinase, aldolase, aspartate aminotransferase (AST), alanine transaminase (ALT), lactate dehydrogenase, and imaging such as MRI. Invasive testing includes both electromyography and muscle biopsy. In our population, elevated creatinine kinase levels (50.4% of those tested, 66/131), averaged 2503. For elevated aldolase (37.7% of those tested, 46/122), the mean was 21.3. One in four patients tested had elevated AST (25.2%, 34/135) with a mean of 142, and one in five had elevated ALT (19.3%, 26/135) with a mean of 130.

Muscle MRI was utilized in 16.7% (26/156) patients, 22 of whom reported muscle edema. Furthermore, 26.3% (41/156) of patients underwent electromyography, 32 of whom had abnormal results. A muscle biopsy was performed in 21.1% of subjects. Those whose dermatomyositis was diagnosed by rheumatologists or internists were 2.5 times more likely to get a muscle biopsy than those diagnosed by dermatologists (26.4% of patients versus 1.3%); however, this association did not reach significance (OR=2.5, 95% CI: 0.94 to 6.70, P=0.065).

Malignancy

DM is known to be associated with malignancy, which can precede DM diagnosis, be identified concurrently with DM diagnosis, or follow DM diagnosis. In our population, 22.4% (35/156) of patients had malignancy-associated dermatomyositis. The mean age of those with a malignancy was 67.6 years compared to 57 years in those without a malignancy, a significant difference (P<0.001).

Antibody positivity

Two-thirds of those tested had positive antinuclear antibodies (ANA), (67.7%, 69/102). The most common specific autoantibodies were anti-SSA/Ro (9.6% of overall cohort) followed by rheumatoid factor (7.1%), anti-smooth muscle (4.5%), and anti-Jo one (4.5%). Few patients tested positive in our system for anti-MDA5, anti-TIF1ɣ, anti-NXP2, and anti-Mi2. As expected, all three patients who were MDA-5+ had pulmonary involvement [2]. All three patients with positive anti-SAE autoantibodies had severe skin findings and dysphagia, as in previous reports [2,5]. We were unable to clearly identify many disease associations with particular antibodies in our cohort owing to small sample size and a lack of cohesive antibody testing given the ten-year period that was studied.

Dermatopathology findings

Only 41.7% of patients had a skin biopsy performed and, as expected, interface dermatitis was reported in nearly all, (96.9%, 63/65) [1]. In one third of available biopsy reports, increased mucin was reported, (30.8%, 20/65). Few reported thickened basement membrane, (7.7%, 5/65) and several described perivascular dermatitis as well, (23.1%, 15/65) (**Table 4**).

Table 4. *Dermatopathology findings.*

Skin biopsy finding	Number (%) N=65
Interface dermatitis	63 (96.9)
Perivascular dermatitis	15 (23.1)
Increased mucin	20 (30.8)
Thickened basement membrane	5 (7.7)

Systemic findings

The most common systemic manifestation of dermatomyositis was pulmonary involvement (25%), typically interstitial lung disease. We identified an association between pulmonary involvement and race/ethnicity (P=0.012). Compared to White patients, African American or Black patients were nearly five times more likely to have pulmonary involvement (OR=4.79, 95% CI: 1.65 to 13.9, P=0.0039). Asians were nearly four times as likely to have pulmonary involvement (OR=3.83, 95% CI: 0.87 to 16.8, P=0.075). Hispanic or Latino patients did not differ significantly from White patients (P=0.51). Dysphagia (15.4%) and cardiac involvement (3.9%) were less common. Pulmonary involvement was followed by dysphagia and cardiac involvement in our population (**Table 5**).

Table 5. *Systemic findings in patients with dermatomyositis.*

Systemic finding	Number (%) N=156
Pulmonary involvement	39 (25)
Presence of dysphagia	24 (15.4)
Cardiac involvement	6 (3.9)

Discussion

This study describes a diverse population [7] of individuals with dermatomyositis and varied disease presentations. We identify trends in past medical history, presenting signs and symptoms, and clinical care. In our study, the most common cutaneous manifestation was Gottron papules, and the most common cutaneous histopathologic finding was, as expected, interface dermatitis. The most common systemic finding was interstitial lung disease, which correlated with race/ethnicity in our population.

One of the most striking findings of this study was that only 53.2% of patients received care from a dermatologist during the course of their disease. As such, documented cutaneous features and cutaneous histopathology results were limited in our study. Additionally, 25% of our patients had clinically documented depression (39/156). Taken together, this study highlights the need for more multidisciplinary care of this population.

Conclusion

Dermatomyositis is a variable disease that can be difficult to diagnose. Multidisciplinary management of patient care can promote rapid diagnosis and appropriate treatment for best outcomes. Further studies with larger cohorts are needed to refine knowledge on DM etiology, disease presentation, and prognostic indicators.

Potential conflicts of interest

The authors declare no conflicts of interest.

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