HHS Public Access

Author manuscript

J Am Acad Dermatol. Author manuscript; available in PMC 2024 April 14.

Published in final edited form as:

J Am Acad Dermatol. 2007 December ; 57(6): 937–943. doi:10.1016/j.jaad.2007.08.024.

Number, characteristics, and classification of patients with dermatomyositis seen by dermatology and rheumatology departments at a large tertiary medical center

Rhonda Q. Klein, MD, MPH[a], **Valerie Teal, MS**[b], **Lynne Taylor, PhD, MS**[b], **Andrea B. Troxel, ScD**[a,b], **Victoria P. Werth, MD**[a,c]

[a]University of Pennsylvania School of Medicine, Philadelphia, Pennsylvania

[b]Center for Clinical Epidemiology and Biostatistics, University of Pennsylvania, Philadelphia, Pennsylvania

[c]Philadelphia Department of Veterans Affairs Medical Center, Philadelphia, Pennsylvania

Abstract

Background—The current diagnostic criteria for dermatomyositis (DM) exclude patients without muscle involvement. As a result there is a paucity of research related to the complete spectrum of the disease.

Objective—The goal of this study was to evaluate differences in the clinical manifestations of DM seen by dermatology relative to rheumatology. We hypothesized that patients with minimal (hypomyopathic) or no (amyopathic) muscle disease would more likely be seen in dermatology, whereas those with more severe (classic) muscle disease would be seen in rheumatology.

Methods—We performed a retrospective chart review of patients with DM seen by our dermatology and rheumatology departments to classify spectrum, presentation, and complications. Patients seen between July 1, 2003, and June 30, 2006, were identified by *Current Procedural Terminology billing* code 710.3. Patients with mixed connective tissue diseases or miscoded DM were excluded.

Results—In all, 131 (65%) patients seen in dermatology, 58 (29%) in rheumatology, and 13 (6%) in both departments were identified. In all, 83 (69%) patients seen in dermatology, 27 (23%) in rheumatology, and 10 (8%) in both departments met criteria for inclusion in the study. The number of patients seen in rheumatology given the classification of classic DM (CDM) (24 of 27 [89%]), hypomyopathic DM (2 of 27 [7%]), and amyopathic DM (ADM) (1 of 27 [4%]) differed significantly from dermatology, where CDM comprised 27 of 83 (33%), hypomyopathic DM comprised 23 of 83 (28%), and ADM comprised 33 of 83 (40%) of the population, respectively ($P < .001$). Sex, ethnicity, and rates of interstitial lung disease differed between departments. There was no difference in the rates of interstitial lung disease between CDM and ADM ($P = .30$). The

Reprint requests: Victoria P. Werth, MD, Department of Dermatology, Hospital of the University of Pennsylvania, 3600 Spruce St, 2 Maloney Bldg, Philadelphia, PA 19104. werth@mail.med.upenn.edu.

Conflicts of interest: None declared.

A previous oral and poster presentation of this material was given at the 68th Annual Society for Investigative Dermatology Conference & the American DermatoEpidemiology Network Meeting, Los Angeles, Calif, May 9-12, 2007.

degree of muscle involvement did not correlate with the rates of DM-associated malignancy (*P* = .57). Few patients with ADM had muscle biopsy (n = 1) or electromyography (n = 7) testing. Positive anti-Jo-1 was seen in 2 of 96 patients (2%; one CDM and one ADM, both with interstitial lung disease), reflecting an overall low prevalence of this autoantibody, or a potential problem with the laboratory assay.

Limitations—Patients reflect the population in only one institution and, thus, the results may not be generalizable to other settings or referral centers. Because this is a retrospective chart review, results are limited by missing data and nonstandardized physical examinations and laboratory data across patients and physicians.

Conclusions—There is a clear difference in DM presentation to dermatology and rheumatology by degree of myositis-complicated disease. (J Am Acad Dermatol 2007;57:937-43.)

The current diagnostic criteria for dermatomyositis (DM) requires the presence of muscle disease. As a result, many physicians have difficulty diagnosing DM with minimal muscle disease (hypomyopathic DM [HDM]) or without muscle disease (amyopathic DM [ADM] or DM sine myositis). This contributes to delays in the diagnosis, treatment, and identification of complications related to the disease.[1] It is hypothesized that there are differences in the disease manifestations of patients who are seen in dermatology versus other medical disciplines, potentially contributing to different perceptions of the prevalence of HDM and ADM.

Classic DM (CDM) is characterized by proximal weakness, with laboratory and diagnostic testing revealing an inflammatory myopathy. Dermatologic manifestations of DM include a heliotrope rash, Gottron's papules, linear extensor erythema, cuticle and nail bed hyperemia, periorbital edema, facial swelling, a malar rash involving the nasolabial folds, V- or shawl-shaped macular erythema over the chest and back, hyperkeratosis of the palmar and lateral surfaces of the fingers, periungual telangiectasias, poikiloderma, and scaly alopecia.[2] Patients with HDM have clinically amyopathic disease for 6 months or longer with minimal clinical or muscle testing abnormalities (muscle enzyme levels, electromyography [EMG], muscle biopsy, muscle magnetic resonance imaging [MRI]).[1] Abnormalities in liver function tests, lactic acid dehydrogenase, creatine phosphokinase (CPK), and aldolase may be seen in CDM and HDM. Patients with ADM have classic cutaneous manifestations of DM without objective abnormalities on radiologic or laboratory screening.[3] With all types of DM, it is important to note that cutaneous disease does not always parallel muscle disease in its onset, activity, or response to therapy.

Because the Bohan and Peter[4,5] diagnostic criteria for DM have been used in most studies, and muscle disease is a required criterion, patients with HDM and ADM are often not included in research. Although the rates of malignancy and interstitial lung disease (ILD) are believed to be the same in HDM and ADM, there is a general lack of clinical research into the skin-predominant forms of the disease.[1]

Using the above definitions, patients with ADM are estimated to comprise 10% to 20% of all patients with DM seen in dermatology referral clinics of US academic health centers; however, the accuracy of this estimate and its correlation with the population seen by

rheumatologists is unknown.[6] The goal of this study was to evaluate differences in the clinical manifestations of DM seen by dermatology relative to rheumatology to better understand the differences in the spectrum of DM disease presentation and how it may affect further classification and outcome-oriented research for DM.

METHODS

To classify spectrum, presentation, and complications from DM seen by department, we performed a retrospective chart review at our hospital, a 625-bed tertiary medical center. Patients seen in the rheumatology and dermatology clinics between July 1, 2003, and June 30, 2006, and designated with a diagnosis of DM were identified by *Current Procedural Terminology* billing code 710.3. A total of 202 patients with this billing code repository were identified, and all charts were reviewed by the investigators. Patients with mixed connective tissue diseases or overlap syndromes (ie, rheumatoid arthritis, scleroderma, lupus erythematosus, Sjögren's syndrome, polyarteritis nodosa, sarcoidosis) were excluded. Patients miscoded or given a misdiagnosis of DM were also excluded. Patients were identified as adult- or juvenile-onset DM based on their age of symptom onset (Tables I, II, and III). This research was approved by our institutional review board (protocol #804873).

Classification

Clinic notes and objective results were used to classify patients with CDM, HDM, or ADM.[1,7,8] Patients were classified as having CDM if they had both clinically significant muscle weakness and laboratory evidence of muscle inflammation or had been treated with systemic immunosuppressive therapy for greater than or equal to consecutive 2 months during the first 6 months of cutaneous manifestations. Patients were designated as having HDM if there was some evidence of myositis on objective testing, with DM skin disease present for longer than 6 months without clinically significant muscle weakness. The classification of ADM was made for patients with hallmark cutaneous manifestations of DM without any history of muscle weakness or abnormalities on objective testing, and no history of immunosuppressive therapy for longer than 2 months during the first 6 months of diagnosis or use of hydroxyurea.[1,3] Many patients were classified as having ADM based on clinical findings and normal muscle enzymes.

Study design

Each chart was reviewed and a data worksheet that included epidemiologic information, symptom onset, age of diagnosis, subjective clinical information, objective physical examination findings, and laboratory data was completed for each patient. Classification of patients into disease subtypes was made after the review of the patient's records and completion of the worksheet, based on both clinician impression and supporting laboratory evidence.

Data analysis

Summary tables illustrating various clinical and laboratory characteristics by department and of various subgroups are included. In comparing patients seen by dermatology and rheumatology, Fisher's exact test and analysis of variance were used for categorical and

continuous variables, respectively. All *P* values are two-sided and a significance level of .05 was applied throughout.

RESULTS

There were 131 (65%) patients seen in dermatology, 58 (29%) in rheumatology, and 13 (6%) in both departments identified by Current Procedural *Terminology* billing code 710.3. After exclusion criteria were applied, 83 (69%) patients seen in dermatology, 27 (23%) in rheumatology, and 10 (8%) in both departments were included, with 58 (48%) cases of CDM, 35 (29%) ADM, and 27 (23%) HDM identified. The number of patients seen by rheumatology and classified as having CDM (24 of 27 [89%]), HDM (2 of 27 [7%]), and ADM (1 of 27 [4%]) differed significantly from dermatology, where CDM comprised 27 of 83 (33%), HDM 23 of 83 (28%), and ADM 33 of 83 (40%) of the population, respectively ($P < .001$). Sex and ethnicity differed between departments, with more male ($P = .003$) and African American ($P = .002$) patients seen in rheumatology.

The degree of muscle involvement by classification (CDM, HDM, or ADM) did not correlate with rates of DM-associated malignancy ($P = .57$). Of patients with CDM and muscle biopsy, EMG, and MRI testing available, 33 of 35 (94%), 33 of 41 (81%), and 17 of 27 (63%), respectively, had results consistent with myositis (Table IV). Objective invasive diagnostic testing was performed much less often in patients with ADM than CDM, with few patients with ADM having muscle biopsy (n = 1), EMG (n = 7), or MRI (n = 6) testing.

By department, rates of muscle biopsy, EMG, and MRI testing were quite varied (Table IV). In dermatology, 83.3% (15 of 18) of patients, 52.6% (20 of 38), and 48.3% (14 of 29) with muscle biopsy, EMG, and MRI testing, respectively, had results consistent with muscle inflammation. Of the patients seen by dermatology, muscle biopsy was performed in 15 cases of CDM, two of HDM, and one of ADM; EMG was performed in 19 cases of CDM, 12 of HDM, and 7 of ADM; and MRI testing was performed in 11 cases of CDM, 12 of HDM, and 6 of ADM. Half (50%) of the patients with HDM seen by dermatology had a positive muscle biopsy result with 41.7% (5 of 12) and 50% (6 of 12) of patients with HDM having positive EMG and MRI testing, respectively. By definition, none of the patients with ADM had positive objective testing. In rheumatology, 88.2% (15 of 17) of patients, 76.5% (13 of 17), and 54.5% (6 of 11) had muscle biopsy, EMG, and MRI testing indicative of myositis, respectively. Of the patients seen by both rheumatology and dermatology with objective testing, 100% (all of 4), 71.4% (5 of 7), and 42.9% (3 of 7) had positive muscle biopsy, EMG, and MRI results consistent with DM, respectively.

When examining records of patients with HDM to try and detect trends predictive of further diagnostic testing, CPK values did not correlate with further diagnostic and invasive testing. Further, as represented by Table IV, not many more patients with HDM received EMG, MRI, or muscle biopsies than patients with ADM. Only 7 of the 27 patients with HDM in this population had CPKs more than 200 U/L, with only two of these 7 having grossly elevated CPK values (elevated CPK values for these 7 patients with HDM were: 204 U/L, 242 U/L, 253 U/L, 259 U/L, 380 U/L, 2018 U/L, 3840 U/L). Of these 7 patients, 5 had diagnostic testing–all MRI–with only 3 of the 5 MRIs indicating active muscle inflammation.

The rheumatology population had greater rates of ILD (51.9%) than the patients seen by dermatology (27.2%) ($P = .032$). Although ILD differed between departments, there was no difference in rates of ILD between CDM and ADM ($P = .2697$). Interestingly, there were only 9 (7.5%) patients (7 CDM, 2 ADM) with both mechanic's hands and ILD, 12 (10%) patients with mechanic's hands without ILD (8 CDM, 2 HDM, 2 ADM), and 34 (28.3%) patients with ILD without mechanic's hands (16 CDM, 11 HDM, 7 ADM). Positive anti-Jo-1 was seen in 2 of 96 patients (2.1%; one CDM and one ADM, both with ILD), reflecting either low prevalence of this auto-antibody or a potential problem with the laboratory assay.

DISCUSSION

It is evident from these results that there is a clear difference in the prevalence of myositis in patients with DM seen in dermatology versus rheumatology. The treatment of patients with primary cutaneous disease versus primary muscle symptoms is very different and, thus, dermatologists and rheumatologists must consider these variants when designing a therapeutic strategy. The implications for the HDM subset, however, are less clear. Gerami et al[1] have proposed the term “clinically” ADM to reflect patients who present with primary skin disease and are later given a diagnosis of either HDM or ADM. Although our analyses separate HDM and ADM, in reality, it may make sense in further analyses to consider these groups together to better reflect clinical reality, particularly with no clear pattern distinguishing which patients receive diagnostic and invasive testing.

Even when the diagnosis of DM is suggested, many practitioners do not perform invasive testing for HDM or ADM unless muscle symptoms are present and/or muscle enzymes are elevated, as supported by our data set.[9] Some believe the definition of ADM to include no signs of muscle involvement after thorough clinical examination, laboratory studies, and imaging, although many practitioners would agree that EMG and muscle biopsy are less sensitive than muscle enzyme studies in detecting myositis.[10] It is well known that muscle enzymes can be normal even in active disease with myositis and, thus, there are no perfect parameters for documenting muscle inflammation.[11] Given sampling errors and low specificity of muscle biopsy and EMG, some practitioners defer more invasive diagnostic testing and imaging if no muscle symptoms are present, and proceed with sequential objective physical examinations, documenting muscle strength and function, because skin findings may precede muscle disease or be the only manifestations of DM.[12]

The overwhelming majority of patients in our data set had adult-onset disease (Tables I and III). This is likely a characteristic of the adult-focused tertiary medical center where the study was performed, coupled with the fact that juvenile-onset DM, particularly amyopathic, is less prevalent than adult-onset DM.[13] The prevalence of DM seen at our institution may reflect the highly specialized nature of our tertiary care center. In terms of patient demographics, patients with ADM in our data set were more likely to be Caucasian and female. This observation has been made previously, but the underlying explanation for this trend is unclear.[9] Whether the prevalence of ADM seen in our patient population reflects the referral nature of our medical center, the increasing prevalence of this disease subset, or the recognition of this disease is debatable.[14]

Considering that tertiary medical centers are largely referral based or inpatient consult based, it can be deduced that the demographics of our patient population are generalizable to the disease process itself; however, access to outpatient medical care is often difficult at large medical institutions. Both dermatology and rheumatology departments at our facility have months-long waiting lists and, thus, many patients are seen for the first time as inpatients when they present with very severe ILD, malignancy, or myositis. The higher proportion of African Americans seen by rheumatology in our data set may reflect the large west Philadelphia urban population that surrounds our medical center; these patients tend to be admitted with advanced disease. Although insurance status and methods of referral were variables not collected in this data set, we presume that the differences in race seen by dermatology and rheumatology may be accounted for by the combination of inpatient rheumatology consults and larger ADM Caucasian population seen by dermatology, and fewer private patients seen by rheumatology than dermatology.

Because this is a retrospective chart review study, there are several limitations to our data. First and foremost, the conclusions of this research are based on one institution. By using a sample from a large referral center, we cannot determine whether unusual circumstances or referral patterns affected our study population and, thus, the generalizability of our results. Secondly, by performing a retrospective study, not all subjective and objective physical and cutaneous findings are recorded in patient charts. Thus, missing data are more common with retrospective reviews than prospective research. Another limitation of this study was in the analysis of ILD, mechanic's hands, and Jo-1 antibody laboratory results. Associations between these variables have been demonstrated in numerous studies; however, because of missing data and only two positive anti-Jo-1 results, no statistical inferences could be made about these parameters.[15,16]

Anti-Jo-1 antibodies have been shown to be associated with ILD and arthritis, but the relationship is stronger in polymyositis than DM. Anti-Jo-1 auto-antibodies have been reported in 30% of patients with polymyositis and 13% of patients with DM, with lung specificity reported to be as high as 95%.[17,18] Because ILD is a common early manifestation of both polymyositis and DM that does not necessarily parallel disease course, most practitioners advise obtaining chest radiograph examinations, chest computed tomography, pulmonary function tests with diffusion capacity, and anti-Jo-1 antibody assays in the initial investigation regardless of the presence of respiratory symptoms.[19] The low incidence of positive anti-Jo-1 antibodies in our data set is surprising. Because numerous laboratories were used to process specimens from our patients, the relationship between Jo-1 and DM should be revisited. Nonetheless, with no difference in rates of ILD by degree of myositis, it is important to emphasize and encourage early pulmonary testing by both dermatologists and rheumatologists.

With different patients presenting to dermatology and rheumatology, it is essential that both disciplines work together to understand the pathogenesis and triggers of this complex autoimmune disorder, along with treatment strategies.

Acknowledgments

Supported in part by grants from the National Institutes of Health (NIH K24-AR 02207) and a Veterans Affairs Merit Review Grant (Dr Werth).

Abbreviations used

ADM	amyopathic dermatomyositis
CDM	classic dermatomyositis
CPK	creatine phosphokinase
DM	dermatomyositis
EMG	electromyography
HDM	hypomyopathic dermatomyositis
IL	interstitial lung disease
MRI	magnetic resonance imaging

REFERENCES

1. Gerami P, Schope JM, McDonald L, Walling HW, Sontheimer RD. A systematic review of adult-onset clinically amyopathic dermatomyositis (dermatomyositis sine myositis): a missing link within the spectrum of the idiopathic inflammatory myopathies. J Am Acad Dermatol. 2006; 54: 597–613. [PubMed: 16546580]
2. Sontheimer RD. Cutaneous features of classic dermatomyositis and amyopathic dermatomyositis. Curr Opin Rheumatol. 1999; 11: 475–82. [PubMed: 10551671]
3. Callen JP, Jorizzo JL. Amyopathic dermatomyositis (dermatomyositis sine myositis). J Am Acad Dermatol. 1992; 26: 505–6.
4. Bohan A, Peter JB. Polymyositis and dermatomyositis (first of two parts). N Engl J Med. 1975; 292: 344–7. [PubMed: 1090839]
5. Bohan A, Peter JB. Polymyositis and dermatomyositis (second of two parts). N Engl J Med. 1975; 292: 403–7. [PubMed: 1089199]
6. Jorizzo J. Dermatomyositis: practical aspects. Arch Dermatol. 2002; 138: 114–6. [PubMed: 11790176]
7. Sontheimer RD. The management of dermatomyositis: current treatment options. Expert Opin Pharmacother. 2004; 5: 1083–99. [PubMed: 15155110]
8. Euwer RL, Sontheimer RD. Amyopathic dermatomyositis (dermatomyositis siné myositis) [reply]. J Am Acad Dermatol. 1992; 26: 506–8.
9. Caproni M, Cardinali C, Parodi A, Giomi B, Papini M, Vaccaro M, et al. Amyopathic dermatomyositis: a review by the Italian group of immunodermatology. Arch Dermatol. 2002; 138: 23–7. [PubMed: 11790163]
10. Schmid MH, Trueb RM. Juvenile amyopathic dermatomyositis. Br J Dermatol. 1997; 136: 431–3. [PubMed: 9115933]
11. Takken T, Elst E, Spermon N, Helders PJM, Prakken ABJ, van der Net J. The physiological and physical determinants of functional ability measures in children with juvenile dermatomyositis. Rheumatology (Oxford). 2003; 42: 158–62.
12. Reed AM, Lopez M. Juvenile dermatomyositis: recognition and treatment. Pediatr Drugs. 2002; 4: 315–21.
13. Reed AM. Myositis in children. Curr Opin Rheumatol. 2001; 13: 428–33. [PubMed: 11604600]

14. Callen JP. Dermatomyositis. Lancet. 2000; 355: 53–7. [PubMed: 10615903]
15. Miller FW. Myositis-specific autoantibodies; touchstones for understanding the inflammatory myopathies. JAMA. 1993; 270: 1846–9. [PubMed: 8411528]
16. Hochberg MC, Feldman D, Stevens MB, Arnett FC, Reichlin M. Antibody to Jo-1 in polymyositis/dermatomyositis: association with interstitial pulmonary disease. J Rheumatol. 1984; 11: 663–5. [PubMed: 6334746]
17. Targoff IN, Reichlin M. Measurement of antibody to Jo-1 by ELISA and comparison to enzymatic inhibitory activity. J Immunol. 1987; 138: 2874–82. [PubMed: 3553328]
18. Targoff IN. Idiopathic inflammatory myopathy: autoantibody update. Curr Rheumatol Rep. 2002; 4: 434–41. [PubMed: 12217249]
19. Fathi M, Dastmalchi M, Rasmussen E, Lundberg IE, Tornling G. Interstitial lung disease, a common manifestation of newly diagnosed polymyositis and dermatomyositis. Ann Rheum Dis. 2004; 63: 297–301. [PubMed: 14962966]

Table I

Characteristics by department

	Dermatology	Rheumatology	Both
Charts reviewed	131	58	13
Total after exclusion	83 (69%)	27 (23%)	10 (8%)
Sex ($P = .003$)			
Female	76 (92%)	18 (67%)	8 (80%)
Male	7 (8%)	9 (33%)	2 (20%)
Ethnicity ($P = .002$)			
Caucasian	72 (87%)	15 (56%)	8 (80%)
African American	7 (8%)	9 (33%)	1 (10%)
Other	4 (5%)	3 (11%)	1 (10%)
DM category ($P < .001$)			
Classic	27 (33%)	24 (89%)	7 (70%)
Amyopathic	33 (40%)	1 (4%)	1 (10%)
Hypomyopathic	23 (28)	2 (7%)	2 (20%)
Symptom onset ($P = .26$)			
Adult	77 (93%)	23 (85%)	9 (90%)
Juvenile	6 (7%)	4 (15%)	1 (10%)
ILD ($P = .032$)			
Present	22 (27%)	14 (52%)	7 (70%)
Absent	59 (73%)	13 (48%)	3 (30%)
Unknown	2 (2%)	0 (0%)	0 (0%)

DM, Dermatomyositis; *ILD*, interstitial lung disease.

P values reflect comparisons between dermatology and rheumatology.

Table II

Characteristics of patients with classic dermatomyositis

	Classic DM			
Department	**Dermatology**	**Rheumatology**	**Both**	***P* value**
No. of patients	27	24	7	
Sex				
Female	25 (93%)	15 (63%)	5 (71%)	.015*
Male	2 (7%)	9 (38%)	2 (29%)	
Ethnicity				
Caucasian	23 (85%)	12 (50%)	6 (86%)	.014*
African American	2 (7%)	9 (38%)		
Other	2 (7%)	3 (13%)	1 (14%)	
Age at DM diagnosis, y	45.4 (19)	45.0 (20)	40.4 (19)	.93
Age at first DM symptoms, y	43.3 (20)	42.3 (20)	38.9 (19)	.85
Symptom onset to DM diagnosis, y	2.1 (8)	2.7 (7)	1.6 (2)	.78
Weakness				
None	1 (4%)	2 (8%)		.18
2/4	3 (11%)			
3/4		2 (8%)		
4/4	23 (85%)	20 (83%)	7 (100%)	
Neuropathy				
Present	7 (26%)	6 (25%)	2 (29%)	1.0
Absent	20 (74%)	18 (75%)	5 (71%)	
Dysphagia				
Present	14 (52%)	17 (71%)	6 (86%)	.25
Absent	13 (48%)	7 (29%)	1 (14%)	
Malignancy				
Yes	7 (26%)	2 (8%)		.15
No	20 (74%)	22 (92%)	7 (100%)	
ILD				
Present	6 (22%)	13 (54%)	4 (57%)	.023*
Absent	21 (78%)	11 (46%)	3 (43%)	
CPK†	1798 (4532)	3616 (6611)	352 (331)	.27
Aldolase†	15 (23)	27 (47)	12 (4)	.27
Muscle biopsy				
c/w DM	14 (93%)	15 (94%)	4 (100%)	1.0
Not c/w DM	1 (7%)	1 (6%)		

CPK, Creatine phosphokinase; *c/w*, consistent with; *DM*, dermatomyositis; *ILD*, interstitial lung disease.

** P* values reflect comparisons between dermatology and rheumatology.

† Values are not normally distributed.

Table III

Characteristics by dermatomyositis-specific muscle disease

	Classic DM	Amyopathic DM	Hypomyopathic DM	*P* value
Total after exclusion	58	35	27	
Sex				
Female	45 (78%)	31 (89%)	26 (96%)	.064
Male	13 (22%)	4 (11%)	1 (4%)	
Ethnicity				
Caucasian	41 (71%)	29 (83%)	25 (93%)	.18
African American	11 (19%)	5 (14%)	1 (4%)	
Other	6 (10%)	1 (3%)	1 (4%)	
Disease onset				
Adult	51 (88%)	34 (97%)	24 (89%)	.32
Juvenile	7 (12%)	1 (3%)	3 (11%)	
Age at first DM symptoms, y	42.3 (19)	45.0 (17)	38.0 (16)	.32
Age at DM diagnosis, y	44.6 (19)	48.4 (16)	43.1 (17)	.46
Symptom onset to DM diagnosis, y	2 (7)	3.4 (7)	5.1 (7)	.21
Malignancy				
Malignancy	9 (16%)	3 (9%)	2 (7%)	.57
Cutaneous manifestations				
Heliotrope	36 (62%)	18 (51%)	13 (48%)	.42
Gottron's papules	50 (86%)	31 (89%)	24 (89%)	1.0
Gottron's sign	50 (86%)	30 (86%)	24 (89%)	1.0
V sign	41 (71%)	23 (66%)	23 (85%)	.20
Shawl sign	40 (69%)	20 (57%)	23 (85%)	.056
Periorbital edema	43 (74%)	22 (63%)	17 (63%)	.42
Linear extensor erythema	43 (74%)	33 (94%)	24 (89%)	.029 *
Calcinosis	10 (17%)	1 (3%)	4 (15%)	.10
Periungual erythema	43 (74%)	25 (71%)	17 (63%)	.55
Cuticular changes	29 (50%)	11 (31%)	7 (26%)	.065
Mechanic's hands	15 (26%)	4 (11%)	2 (7%)	.072
Facial erythema	44 (76%)	25 (71%)	21 (78%)	.86
Livedo reticularis	10 (17%)	1 (3%)	1 (4%)	.054
Hair thinning	13 (22%)	2 (6%)	5 (19%)	.096
ILD				
Present	23 (40%)	9 (26%)	11 (44%)	.27
Absent	35 (60%)	26 (74%)	14 (56%)	
CPK †				
Average	2407 (5361)	91 (51)	353 (788)	.0092 *
Aldolase †				
Average	19 (34)	6 (2)	10 (15)	.078

	Classic DM	Amyopathic DM	Hypomyopathic DM	*P* value
ESR[†]	48 (101)	20 (25)	24 (24)	.21
EMG				
c/w DM	33 (81%)	0 (0%)	5 (36%)	<.001[*]
Not c/w DM	8 (20%)	7 (100%)	9 (64%)	
MRI				
c/w DM	17 (63%)	0 (0%)	6 (43%)	.012[*]
Not c/w DM	10 (37%)	6 (100%)	8 (57%)	
Muscle biopsy				
c/w DM	33 (94%)	0 (0%)	1 (33%)	.0042[*]
Not c/w DM	2 (6%)	1 (100%)	2 (67%)	
CT chest				
ILD	14 (74%)	6 (86%)	2 (50%)	.35
Normal	1 (5%)	1 (14%)	1 (25%)	
Other	4 (21%)	0 (0%)	1 (25%)	

CPK, Creatine phosphokinase; *CT*, computed tomography; *c/w*, consistent with; *DM*, dermatomyositis; *EMG*, electromyography; *ESR*, erythrocyte sedimentation rate; *ILD*, interstitial lung disease; *MRI*, magnetic resonance imaging.

[*] *P* values reflect comparisons between classic DM and amyopathic DM.

[†] Values are not normally distributed.

Table IV

Characteristics of patients with objective diagnostic testing (muscle biopsy, electromyography, or magnetic resonance imaging results) by department and by dermatomyositis subtype

	Dermatology	Rheumatology	Both departments	CDM	HDM	ADM
Muscle biopsy	15/18 *	15/17 *	4/4 *	33/35 *	1/3 *	0/1 *
EMG	20/38 *	13/17 *	5/7 *	33/41 *	5/14 *	0/7 *
MRI	14/29 *	6/11 *	3/7 *	17/27 *	6/14 *	0/6 *
Muscle biopsy + EMG	5	8	1	13	1	0
Muscle biopsy + MRI	4	3	0	6	0	1
EMG + MRI	13	4	4	11	7	3

ADM, Amyopathic dermatomyositis (DM); *CDM*, classic DM; *EMG*, electromyography; *HDM*, hypomyopathic DM; *MRI*, magnetic resonance imaging.

* Fraction is the number of patients with results consistent with DM over the total number of patients with test performed.