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Incidence of Dermatomyositis and Clinically Amyopathic Dermatomyositis: A Population-Based Study in Olmsted County, Minnesota

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Abstract

Objective—To identify new and existing cases of dermatomyositis and its subtypes in Olmsted County, Minnesota, from 1976 through 2007, and to establish a population-based estimate of the incidence and prevalence of dermatomyositis and amyopathic dermatomyositis.

Design—Retrospective population-based study.

Setting—Community-based epidemiology project.

Patients—Using the Rochester Epidemiology Project, patients with a diagnosis of dermatomyositis were identified.

Main Outcome Measures—Incidence of dermatomyositis and clinically amyopathic dermatomyositis; risk of malignancy in clinically amyopathic dermatomyositis.

Results—Of the 29 patients identified, 6 (21%) of these had the clinically amyopathic subtype of dermatomyositis, and 22 (76%) were female. Overall age- and sex-adjusted incidence (95% confidence interval) of dermatomyositis including all subtypes was 9.63 (6.09-13.17) per 1,000,000 and was 2.08 (0.39-3.77) per 1,000,000 for clinically amyopathic dermatomyositis. Age- and sex-adjusted prevalence was 21.42 (13.07-29.77) per 100,000. Eight patients (28%) had a malignancy during the study period; risk of malignancy (odds ratio) for classic dermatomyositis compared with clinically amyopathic dermatomyositis was 4.61 but was not statistically significant (0.22-96.09) (*P*=.44).

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Conclusions—Dermatomyositis is a rare disease, and clinically amyopathic dermatomyositis represents an estimated 20% of all dermatomyositis cases. Larger population-based studies are needed to estimate the risk of malignancy associated with subtypes of dermatomyositis, particularly clinically amyopathic dermatomyositis.

Keywords

clinically amyopathic dermatomyositis; dermatomyositis; epidemiology; incidence; population-based study

Introduction

The term *amyopathic dermatomyositis* (ADM) was first coined by Pearson[1] in 1979 to describe a rare skin disease that has the same cutaneous symptoms as classic dermatomyositis (CDM) but lacks objective evidence of myopathy. ADM was not fully recognized as a distinct subset of dermatomyositis until the early 1990s.2 Initially thought to carry a favorable prognosis, ADM may in fact be linked with an increased risk of internal malignancy, as suggested by some case reports.3,4 Several studies also have indicated an increased risk of lung disease,[5,6] but the extent of this risk is unclear. It has been estimated that ADM represents about 10% of all cases of dermatomyositis; this percentage may be higher in Asian populations.[7] Multiple studies have reported on the incidence of cancer in patients with dermatomyositis or polymyositis.[8-11] The incidence of dermatomyositis itself, however, including the subtype ADM, is largely unknown.

Founded in 1966, the Rochester Epidemiology Project (REP) is a comprehensive medical record linking database for all residents of Olmsted County, Minnesota.[12] By using the REP, we aimed to ascertain the incidence and prevalence of dermatomyositis and its subtypes, along with the incidence of the subtypes ADM and clinically amyopathic dermatomyositis (CADM).

Methods

After obtaining Mayo Clinic Institutional Review Board approval, we retrieved from the REP and then reviewed patient records for all newly diagnosed cases of dermatomyositis in Olmsted County from 1976 through 2007. Patients with polymyositis or mixed connective tissue disease and patients who had denied research authorization were excluded from the study.

Definitions

Patients with a diagnosis of dermatomyositis were subclassified by disease subtype—CDM, ADM, CADM, CADM evolving into CDM, hypomyopathic dermatomyositis (HDM), and juvenile dermatomyositis—according to the definitions described in Table 1.7,13-15 Because patients with HDM and patients with ADM may have similar initial cutaneous symptoms and a subjective lack of muscle involvement, the term “clinically amyopathic dermatomyositis” (CADM) was designated to describe such patients. Also, because of the broad definitions used in our study, the designations of juvenile dermatomyositis and CADM were not mutually exclusive from the other dermatomyositis subtypes (Table 1).

Collected Data

Only patients who were residents of Olmsted County, Minnesota, at the time of disease diagnosis were included in the study results. Residents who received a previous diagnosis of dermatomyositis elsewhere but were living in Olmsted County as of the reference date were included only in the calculations of prevalence. Collected data included patient characteristics such as age, sex, race, place of diagnosis, residence at time of diagnosis, date and results of

skin and muscle biopsies, electromyography, magnetic resonance imaging, testing for autoantibodies and muscle enzymes, associated symptoms and illnesses, malignancy during the study period, and clinical follow-up.

Statistical Analysis

The incidence rates of CDM, ADM, and CADM were calculated by dividing the number of CDM events, ADM events, and CADM events, respectively, by the Olmsted County population for 1976 through 2007 determined by the US decennial census, with linear interpolation for intercensus years. Because of the low percentage of African Americans living in Olmsted County (3.8% in Olmsted County vs 12.8% nationally), the incidence rates were adjusted to the age and sex distribution of the 2000 US white population.[16] Overall prevalence of dermatomyositis in Olmsted County was also calculated using January 1, 2007, as a reference date.

Logistic regression was used to calculate the odds ratios for malignancy in all patients with CDM compared with patients with CADM. Survival was estimated using the Kaplan-Meier method. Models also evaluated the death events as expected survival probabilities based on total Minnesota population from 1970 to 2000. All tests were 2 sided with an α level of .05. Analyses were performed using SAS v9.1 software (SAS Institute, Inc, Cary, North Carolina).

Results

Patient Population

Between January 1, 1976, and December 31, 2007, 77 patients were identified from the REP as having a coded preliminary or final diagnosis of dermatomyositis; 4 patients (5.2%) who denied research authorization were excluded. Upon review of the charts, the clinical description and course of illness were typical of dermatomyositis for 40 patients. Diagnosis of dermatomyositis was excluded for the other 33 patients on the basis of the clinical records, including incorrect preliminary diagnosis of dermatomyositis and coding overlap with polymyositis. For incidence calculations, 11 of the 40 remaining patients were further excluded from the study group: 3 patients because the diagnosis was not made within the designated study dates and 8 patients because they were not residents of Olmsted County at the time of diagnosis. However, those 11 patients were included in prevalence calculations.

After all exclusions, 29 patients with a new diagnosis of dermatomyositis were included in the study analysis. We grouped patients into the following subclassifications: 20 patients with CDM (2 juvenile), 4 with ADM (1 juvenile), 2 with HDM, and 3 with CADM evolving into CDM (Table 2). Combining the patients with ADM and HDM gave 6 patients with CADM. Three patients had juvenile dermatomyositis (2 CDM, 1 ADM). Excluding the 3 juvenile cases (patients aged 2, 5, and 11 years), the mean age was 57.4 years (range, 30.5-95.0 years). Most patients were female (22/29, 76%), and the majority were white (24/29, 83%); 1 patient was Asian, 1 patient was Hispanic, and race was unknown for 3 patients. Clinical characteristics of the 6 patients with CADM are shown in Table 3 (published online only).

Epidemiology

The overall age- and sex-adjusted incidence of dermatomyositis including all subtypes was 9.63 per 1,000,000 (95% confidence interval [CI], 6.09-13.17); age-adjusted incidence was 13.98 per 1,000,000 (95% CI, 8.08-19.89) for females and 4.68 per 1,000,000 (95% CI, 1.15-8.20) for males. Overall age- and sex-adjusted incidence of ADM including the juvenile case was 1.31 per 1,000,000 (95% CI, 0.01-2.60); age-adjusted incidence was 2.07 per 1,000,000 (95% CI, 0-4.41) for females and 0.45 per 1,000,000 (95% CI, 0-1.32) for males. Overall age- and sex-adjusted incidence of CADM was 2.08 per 1,000,000 (95% CI,

0.39-3.77); age-adjusted incidence of CADM for females was 3.50 per 1,000,000 (95% CI, 0.43-6.58) and for males was 0.45 per 1,000,000 (95% CI, 0-1.32). Age- and sex-adjusted incidence of dermatomyositis per 1,000,000 persons by decade of diagnosis is shown in Table 4.

Prevalence was calculated using January 1, 2007, as a reference date; 26 cases were used to calculate prevalence. Age- and sex-adjusted overall prevalence of all subtypes of dermatomyositis was 21.42 per 100,000 (95% CI, 13.07-29.77). Age-adjusted prevalence was 33.08 per 100,000 (95% CI, 18.83-47.33) for women and 8.35 per 100,000 (95% CI, 0.81-15.90) for men.

Malignancy

Among the 29 patients with dermatomyositis, 8 (28%) had a diagnosis of cancer at some time during the study period. Cancer was diagnosed after dermatomyositis onset in 6 of the 29 patients (21%), including 1 patient with CADM evolving into CDM who received a diagnosis of esophageal adenocarcinoma 12 years after dermatomyositis was diagnosed. Endometrial cancer was diagnosed and treated in 1 patient 1.5 years before the onset of ADM but was no longer an active disease process at the time of dermatomyositis diagnosis. Additionally, dermatomyositis developed in 1 patient during chemotherapy for metastatic colon cancer. The odds ratio for concurrent or future development of malignancy in patients who have CDM compared with those who have CADM was 4.61 but was not statistically significant (95% CI, 0.22-96.09; P=.44).

Lung Disease

Of the 29 study patients, evidence of lung disease developed in 5 (17%). Results from computed tomography of the chest were available for 4 of these patients. Three of the 5 patients (2 with CDM, 1 with CADM evolving into CDM) had interstitial fibrosis, 1 patient (with CDM) had interstitial muscle weakness, and 1 patient (with CDM) received a diagnosis of bronchitis obliterans–organized pneumonia. No patients with CADM had evidence of lung disease.

Survival

Follow-up data were available for all 29 patients. Average duration of follow-up from the date of diagnosis was 6.85 years (range, 0.07-19.56 years). At the end of the study period, 8 of the 29 adult patients had died (28%). For 6 of these patients, the cause of death was attributed to metastatic disease (in the setting of dermatomyositis with associated malignancy). Kaplan-Meier estimates of overall 5- and 10-year survival for patients with a diagnosis of dermatomyositis was 0.80 (95% CI, 0.66-0.97) and 0.73 (95% CI, 0.56-0.96), respectively; expected 5- and 10-year survival for the age- and sex-matched general Minnesota population was 0.92 and 0.85, respectively.

Discussion

In 1990, Oddis et al[17] reported an incidence of 5.5 cases of dermatomyositis per million; of interest, that study showed a significant increase in incidence from the first to the second decade of the study: 2.5 per 1,000,000 in the first decade, increasing to 8.9 per 1,000,000 in the second. [17] We also sought to examine whether our study showed a similar increasing frequency of dermatomyositis over time (Table 4), but our patient population was too small to yield any significant conclusions. Our overall incidence of 9.63 per 1,000,000 from 1976 through 2007 is only slightly higher than the rate reported in the second decade by Oddis et al[17] and may more accurately reflect true incidence, because ours is a population-based study.

Approximately 20% of patients in our study (6 of 29) had CADM, which correlates with previously reported rates of 10% to 20%.[13] Of these 6 patients with CADM, 5 received their diagnosis since 2000 (the other patient, in 1981). It remains unclear whether the incidence of ADM is truly increasing or whether this represents increased awareness and clinician knowledge of this rare subtype of dermatomyositis.

In accordance with Sontheimer's classification scheme,[7,13-15] we defined CADM evolving into CDM as the presence of skin manifestations for more than 6 months before the onset of clinical or subclinical evidence of myopathy. Case studies suggest that, for some patients, ADM may progress to frank myopathy up to 10 years after the onset of cutaneous symptoms.[3,7] Our study identified 3 patients with CADM that progressed to CDM, but only 1 patient had CADM that progressed to myopathy (CDM) after a long period (14 years) with only cutaneous manifestations. This is consistent with previous reports indicating that progression of CADM to CDM after a long period of cutaneous-only disease is a relatively rare occurrence in the spectrum of CADM disease.[13] However, we acknowledge that true classification of disease in a patient perhaps may be known only after an entire lifetime.

Among our 29 patients with dermatomyositis, 8 (28%) had a diagnosis of cancer at some time during the study period. This rate is higher than that in previous reports: in 1992, Sigurgeirsson et al[10] reported a malignancy rate of 15% associated with dermatomyositis in a populated-based study in Sweden. Of note, our calculations included cancers diagnosed before the onset of dermatomyositis; if only those cancers diagnosed at or after the onset of dermatomyositis are included, our malignancy rate would be 21%, closer to the rate reported by Sigurgeirsson et al.10

We sought to examine the incidence of malignancy in patients with CADM compared with their CDM counterparts; however, with only 6 patients in our study population having CADM, our comparisons of malignancy incidence between the 2 groups were not statistically significant. Only 1 patient with CADM had a malignancy, and this cancer occurred 1.5 years before the diagnosis of dermatomyositis. In addition, of the 5 patients with lung disease, none had the CADM subtype. Case reports of a decreased risk of both malignancy and lung disease in CADM patients do exist in the medical literature.[13] However, a comparison of 120 patients with dermatomyositis by Klein et al[18] found no significant differences in risk of malignancy or lung disease between patients with CDM and CADM. The risk remains unclear, and screening for both malignancy and lung disease continues to be essential for all dermatomyositis patients regardless of disease subtype.

In our patients with CADM, 3 of 6 (50%) had elevated levels of anti-Jo-1 IgG antibodies. This antibody has been shown to be present in some patients with polymyositis and dermatomyositis. A review from Gerami et al[13] showed that 3 of 85 CADM patients (3.5%) and 1 of 21 patients with CADM and coexisting lung disease (4.8%) were shown to have positive anti-Jo-1 antibodies. With 3 of 6 patients testing positive for this antibody, our yield is higher than previously reported results and is at variance with the literature to date. Reasons for this discrepancy are unknown but may be related to our epidemiologic patient population in Olmsted County, Minnesota.

The strength of this report is that it is a population-based study performed using the REP, a well-validated medical record database for Olmsted County. However, it is possible that patients may have been excluded from our study because of lack of disease recognition by clinicians; even today, ADM continues to be a clinically difficult diagnosis. Given the small patient numbers in this population-based study, limited conclusions can be derived regarding associations of dermatomyositis and its subtypes with malignancy, lung disease, and prognosis. The retrospective nature of the study and reliance on the medical record precluded

comprehensive clinical data on all patients in our analysis. Dermatomyositis manifesting in nonwhite skin is an important clinical entity; our analysis is limited by the predominantly white population of Olmsted County, Minnesota.

Conclusion

CADM represents approximately 20% of all cases of dermatomyositis. Although our rate of malignancy associated with dermatomyositis is similar to that in previous reports in the literature, larger population-based studies are needed to estimate the risk of malignancy associated with subtypes of dermatomyositis, particularly with CADM.

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Ms. Reeder and Dr. Davis had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis.

Abbreviations

ADM	amyopathic dermatomyositis
CADM	clinically amyopathic dermatomyositis
CDM	classic dermatomyositis
CI	confidence interval
HDM	hypomyopathic dermatomyositis
REP	Rochester Epidemiology Project

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Table 1

Dermatomyositis Definitions

Type of Dermatomyositis	Definition
Classic (CDM)	Hallmark cutaneous manifestations with evidence of proximal muscle weakness occurring within 6 mo after onset of skin disease
Amyopathic (ADM)	Hallmark cutaneous manifestations of dermatomyositis for >6 mo without clinical or laboratory evidence of myopathy
	Exclusion criteria:
	1) Use of drugs capable of producing hallmark cutaneous changes characteristic of dermatomyositis
	2) Use of systemic immunosuppressive agents for >2 consecutive mo within the 6 mo after skin disease onset
Hypomyopathic (HDM)	Hallmark cutaneous manifestations without subjective muscle weakness for >6 mo but with objective evidence of subclinical muscle weakness on further testing (abnormal muscle enzymes, signs of myopathy on electromyography or muscle biopsy)
Clinically amyopathic (CADM)	A term used to describe both ADM and HDM
Clinically amyopathic evolving into classic (CADM→CDM)	Patients with cutaneous dermatomyositis with onset of muscle involvement >6 mo after onset of clinically significant skin disease
Juvenile (JDM)	Dermatomyositis of any subset occurring in a patient younger than 18 years

Adapted from Gerami et al.[13] Used with permission.

Table 2

Patient Characteristics by Subtype of Dermatomyositis

	Dermatomyositis Subtype[a]					
Characteristic	**CDM (n=20)**	**ADM (n=4)**	**HDM (n=2)**	**CADM (n=6)**	**CADM→CDM (n=3)**	**JDM (n=3)**
Percentage of the total group (N=29)	68.9%	13.8%	6.9%	20.7%	10.3%	10.3%
Mean age at diagnosis, y	51.9	48.5	65.9	54.3	49.3	6.8
Positive diagnostic studies						
Skin biopsy	9/10 (90%)	3/3 (100%)	2/2 (100%)	5/5 (100%)	2/2 (100%)	0/0 (0%)
EMG	18/19 (95%)	0/3 (0%)	2/2 (100%)	2/5 (40%)	3/3 (100%)	1/1 (100%)
Muscle biopsy	11/12 (92%)	0/0 (0%)	0/0 (0%)	0/0 (0%)	1/1 (100%)	2/2 (100%)
MRI	1/2 (50%)	0/1 (0%)	0/0 (0%)	0/1 (0%)	0/0 (0%)	0/1 (0%)
Malignancy	6/20 (30%)	1/4 (25%)	0/2 (0%)	1/6 (17%)	1/3 (33%)	0/3 (0%)

Abbreviations: ADM, amyopathic dermatomyositis; CADM→CDM, CADM evolving into CDM; CADM, clinically amyopathic dermatomyositis; CDM, classic dermatomyositis; EMG, electromyography; HDM, hypomyopathic dermatomyositis; JDM, juvenile dermatomyositis; MRI, magnetic resonance imaging.

[a]Values are no. of patients (%) unless otherwise stated. Subtypes are not mutually exclusive.

Table 3

Characteristics of Patients With CADM (published online only)

Patient/Sex	Classification	Age at Diagnosis, y	Year of Diagnosis	Biopsy Results Consistent With Dermatomyositis?[d]	Skin Findings	Muscle Enzymes[a]	Antibodies and Inflammatory Markers[b]	Duration of Follow-up, y
1/F	ADM	68.4	2002	Yes	GP, GS, heliotrope, shawl sign, extensor erythema, cuticle telangiectasias, photosensitivity, pruritus, scalp involvement	CK, 158 U/L; aldolase, 7.5 U/L	ANA, 1.7 U; ESR, 14 mm/h; SSA/Ro, 1 U; SSB/La, 1.4 U	5.67
2/F	ADM	57.0	2007	Yes	Heliotrope, shawl sign	CK, 77 U/L; aldolase, 3.2 U/L	ANA, 5.8 U; Jo-1, 2.4 U	.58
3/F	ADM	61.5	1981	Yes	GP, GS, heliotrope, shawl sign, scalp involvement, cuticle telangiectasias, extensor erythema	CK, 202 U/L	ANA, positive 1:64; ESR, 39 mm/h; SSB/La, negative[c]; RNP, negative[c]	19.50
4/M	ADM	6.98	2007	ND	GP, GS, heliotrope, extensor erythema, calcinosis	CK, 111 U/L; aldolase, 11.8 U/L	ANA, >1 U; ESR, 11 mm/h; anticentromere antibodies, negative[c]	.71
5/F	HDM	73.3	2000	Yes	GS, GP, heliotrope, shawl sign, pruritus, scalp involvement, extensor erythema, cuticle telangiectasias	CK, 75 U/L; aldolase, 5.3 U/L	ANA, 4.2 U; ESR, 23 mm/h; Jo-1, 2.9 U	7.96
6/F	HDM	58.5	2007	Yes	GS, GP, heliotrope, shawl sign, mechanic's hands, photosensitivity, extensor erythema, cuticle telangiectasias	CK, 99 U/L; aldolase, 5.8 U/L; LDH, 205 U/L	ANA >12 U; ESR, 3 mm/h; Jo-1, 3.4 U; SSA/Ro, 185.9 U; SSB/La, 22.4 U; RNP, 2.9 U	1.02

Abbreviations: ADM, amyopathic dermatomyositis; ANA, antinuclear antibody; CADM, clinically amyopathic dermatomyositis; CK, creatine kinase; ESR, erythrocyte sedimentation rate; F, female; GP, Gottron papules; GS, Gottron sign; HDM, hypomyopathic dermatomyositis; Jo-1, IgG anti-Jo–1 antibody; LDH, lactate dehydrogenase; M, male; ND, not done; RNP, IgG anti–ribonuclear protein antibody; SSA/Ro, IgG anti-SSA/Ro antibody; SSB/La, IgG anti-SSB/La antibody.

[a] Reference ranges. Aldolase: 0-16 years, <14.5 U/L; ≥17 years, <7.7 U/L. CK: 22-210 U/L. LDH: ≥18 years, 122-222 U/L.

[b] Reference ranges. ANA: negative, ≤1.0 U; positive, >1.0 U. ESR: 0-29 mm/h. Jo-1: negative, <1.0 U; positive, ≥1.0 U. SSA/La: negative, <1.0 U; positive, ≥1.0 U. SSA/Ro: negative, <1.0 U; positive, ≥1.0 U. RNP: negative, <1.0 U; positive, ≥1.0 U.

[c] Specific lab values unavailable.

[d] Biopsy results consistent with dermatomyositis included lichenoid interface dermatitis and dermal perivascular lymphocytic inflammation.

Table 4

Age- and Sex-Adjusted Incidence of Dermatomyositis per 1,000,000 Persons in Olmsted County, Minnesota, 1976-2007

Calendar Decade	No. of Cases	Incidence (95% CI)
1976-1985	3	4.4 (0-9.4)
1986-1995	9	9.4 (3.2-15.7)
1996-2007	17	12.5 (6.5-18.5)
Overall	29	9.6 (6.1-13.2)

Abbreviation: CI, confidence interval.