The Arthritis of Inflammatory Childhood Myositis Syndromes

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ABSTRACT. ***Objective.*** Arthritis has been an associated finding in juvenile dermatomyositis (JDM), but its prevalence, course, and response to therapy has not been well described. We investigated the frequency, course, and clinical and radiographic features in a large cohort of patients with JDM.

Methods. The charts of 94 patients with idiopathic myositis (1984–99) were reviewed: 80 JDM, 3 juvenile polymyositis (JPM), 5 amyopathic JDM, and 6 overlap myositis syndromes. Compiled data included demographics, clinical features, a detailed description of the arthritis, investigations (radiographs, autoantibodies), course, and response to therapy. All radiographs were independently reviewed by a single radiologist.

Results. Sixty-one percent (95% CI 50–72%) of patients with JDM had arthritis. The arthritis was reported a median 4.5 mo (range –73.6 to 76.6 mo) after the JDM onset. When compared to patients with no arthritis, the occurrence of arthritis was not significantly related to sex, race, positive antinuclear antibody or rheumatoid factor, calcinosis, nodules, vasculitis, or Raynaud's phenomenon. The initial involvement was pauciarticular in 67% and polyarticular in 33%. In the pauci group, asymptomatic knee effusions were the predominant finding (n = 19, 58%), and in 18 patients may have been the result of steroid therapy. Two patients evolved from a pauci onset to a polyarticular course. All responded to therapy (corticosteroids; 47 were taking other medications) with remission of the arthritis within a median of 2.0 mo (range 0.1–64.5 mo). However, the arthritis recurred in 39% as the corticosteroids were tapered. Four patients with JDM eventually required corticosteroid wrist injections, with resolution of the arthritis. The arthritis was nonerosive in all cases. No patient with JPM had arthritis. Three of 5 patients with amyopathic JDM and 4 of 6 with overlap myositis syndrome had a nonerosive polyarthritis.

Conclusion. Nonerosive arthritis involving the knees, wrists, elbows, and fingers is a frequent manifestation of JDM and other idiopathic childhood myositis. The arthritis is seen early in the course of JDM and often responds to treatment. However, the arthritis may recur with tapering of corticosteroids despite remission of the JDM. In a significant proportion of JDM cases, arthritis is the major sequela and may warrant further medical therapy or intraarticular corticosteroid injections. (J Rheumatol 2001;28:192–7)

Key Indexing Terms:
JUVENILE DERMATOMYOSITIS NONEROSIVE ARTHRITIS IDIOPATHIC MYOSITIS

Juvenile dermatomyositis (JDM) is a rare childhood disease characterized by inflammation primarily affecting the skin and muscles. Arthritis is a common manifestation in patients with JDM. Arthritis has been reported in various clinical cohorts of JDM occurring 23–64% of the time[1-4], with a chronic polyarthritis persisting into adulthood described in 8 out of 62 (13%) patients with JDM[5]. However, the literature contains very little information describing the prevalence and clinical features of the arthritis in JDM.

Disability in JDM has been suggested to arise primarily from the proximal muscle weakness, contractures, and soft tissue calcinosis[6]. However, the extent to which the arthritis associated with JDM contributes to morbidity has not been determined. The potential pain and limitation conferred by the associated arthritis may become a clinically important problem, particularly when the other features of JDM are in remission.

We investigated the frequency, course, and clinical, immunological and radiographic features of arthritis in a large cohort of patients with JDM, as well as and the extent of disability conferred by the arthritis. We hypothesized that arthritis is a frequent occurrence in JDM, and in some patients may lead to significant, persistent morbidity.

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MATERIALS AND METHODS

All patients seen at the Hospital for Sick Children, a tertiary and quaternary referral center, between 1984 and 1999 with a primary diagnosis of idiopathic myositis were included. The referral center provides medical care to patients residing predominantly in Toronto but also includes a large patient population living in other parts of Ontario. Patients with mixed connective tissue disease (MCTD) were excluded. MCTD was defined by the Kasukawa criteria[7].

Patients were classified as having JDM, juvenile polymyositis (JPM), amyopathic JDM, and overlap myositis syndrome according to established criteria. The criteria of Bohan and Peter[8] were used to define patients as having definite or probable JDM and JPM. The criteria include symmetrical muscle weakness, muscle biopsy showing an inflammatory myopathy, elevated skeletal muscle enzymes, abnormal electromyographic and dermatologic features pathognomonic of JDM (heliotrope, Gottron's sign). Definite JDM is defined as rash plus at least 3 muscle criteria. Probable JDM consists of rash plus 2 muscle criteria. Amyopathic JDM is defined as patients with the characteristic JDM rash in the absence of any muscle criteria. Muscle involvement was additionally ruled out by the presence of a normal magnetic resonance image in our study patients. JPM is defined as 3 or 4 muscle criteria in the absence of rash. Overlap myositis syndrome includes patients with features of myositis who also fulfill some criteria for other connective tissue disorders such as systemic lupus erythematosus (SLE), progressive systemic sclerosis, rheumatoid arthritis (RA), polyarteritis, or Sjögren's syndrome.

In all, 80 patients with JDM were studied. Fourteen additional patients were studied: 5 with amyopathic JDM, 3 with JPM, and 6 with overlap myositis syndromes. All study patients were followed rigorously in a similar fashion, and since 1990 all patients were seen in a separate myositis clinic by certified pediatric rheumatologists with physiotherapy assessments at each visit.

Data, compiled from a retrospective chart review of all study patients, were extracted using specially prepared data forms. Two trained volunteer research assistants extracted and entered data into a computer database. Thirty out of a total of 94 charts were studied in duplicate as a validity check. The information extracted, where available, included demographics, clinical features of JDM, a detailed description of the arthritic manifestations, investigations (radiographic, autoantibody profile), course, and response to therapy, as well as the Childhood Health Assessment Questionnaire (CHAQ) score[9]. The CHAQ has been available and administered to our patients with JRA and JDM since 1990. The CHAQ is a 30 item scale that evaluates the patient's physical function in 8 domains (grooming and dressing, arising, eating, walking, hygiene, reach, grip, activity). The CHAQ yields a total score that estimates the amount of disability conferred by the JDM (0 = no disability, 3 = severe disability). The CHAQ was initially developed to measure physical function in children with arthritis, but has been validated in our patients with JDM[10]. Arthritis was defined as swelling or effusion of a joint or at least 2 of the following: heat, decreased range of motion, or tenderness/painful movement.

Radiographs of patients were taken when clinically indicated by the treating rheumatologist. All radiographs were independently reviewed by a single radiologist blinded to patient, type of myositis, and treatment. The radiographs were assessed for the presence or absence of osteopenia, calcification, erosions, joint space narrowing, and effusions.

Analysis of the data was done using the Data Desk 6.1 software program[11].

RESULTS

Demographics and clinical features. The number of patients in each group and their associated clinical manifestations at diagnosis are given in Table 1. There was a predominance of female patients in each category of inflammatory myositis, with the exception of JPM. Our study patients with JDM possessed the typical clinical features[1,2].

The median followup period in patients with JDM, amyopathic JDM, JPM, and overlap myositis syndrome was 3.8 (range 0.15–13.4 yrs), 4.9 (range 0.97–8.8 yrs), 6.9 (range 1.9–12.0 yrs), and 3.8 years (range 1.5–7.5 yrs), respectively.

Patients with JDM. From the 80 patients with JDM, 49 (61%; 95% CI 50–72%) were found to have an associated arthritis. The arthritis occurred a median 4.5 months (range –73.6 to 76.6) after the JDM diagnosis. The proportion of patients who presented with arthritis at the diagnosis of their JDM was 11% (9/80).

The initial articular involvement was pauciarticular (4 joints or less) in 67% (33/49) and polyarticular in 33% (16/49). In the pauciarticular group, asymptomatic knee effusions were the predominant finding (19/33, 58%), and in 18 patients followed initiation of corticosteroid therapy and therefore may have resulted from corticosteroid treatment. Two patients evolved from a pauciarticular onset to a polyarticular course. The distribution of the arthritis most commonly involved the knees, wrists, elbows, and fingers (Figure 1). However, as shown in Figure 1, both small and large joints of the upper and lower extremities were involved in some patients during the course of the arthritis.

The arthritis in most cases was painful but generally did not markedly limit function. The majority of patients denied having any difficulties with their activities of daily living as a result of their arthritis.

The CHAQ scores from the last rheumatology clinic visit were available in 65 patients with JDM (42 from patients with associated arthritis). The median CHAQ score was found to be 0 (range 0–2.75). Twelve JDM patients with arthritis had CHAQ scores greater than 0 (range 0.125–0.875). There were no significant differences in the median CHAQ scores between JDM patients with or without arthritis.

The antinuclear antibody (ANA) and rheumatoid factor (RF) were tested in 76 and 57 patients. ANA was assayed using indirect immunofluorescence (HEp-2 substrate). A positive ANA and RF found any time during the disease course was present in 79% and 5% of the screened patients, respectively. Only 2 JDM patients with arthritis had a positive RF. In both cases, the RF was tested only once at onset of JDM and before the development of arthritis. Compared to patients with no arthritis, the occurrence of arthritis was not significantly related to sex, race, positive ANA or RF, calcinosis, nodules, vasculitis, or Raynaud's phenomenon (Table 2). Forty patients were screened for myositis-specific autoantibodies (anti-synthetases, anti-SRP, anti-Mi-2, anti-KJ, anti-JP, anti-Fer, anti-Mas) and antibodies present in patients with overlap conditions (anti-PM-Scl, anti-Ku, anti-U1, U2, U3, and U5 RNP, and anti-Ro). We found that 2 patients tested positive to the myositis-specific autoantibody anti-Mi-2, but only one patient had a pauciarticular arthritis involving the knees. Two additional patients tested positive to anti-PM-Scl; one of these patients had polyarthritis of the large and small joints.

All patients with arthritis responded to therapy prescribed

Table 1. Clinical and demographic features of the study population at diagnosis.

	JDM		Amyopathic JDM		JPM		Overlap	
Median followup, yrs (range)	3.8 (0.15–13.4)		4.9 (0.97–8.8)		6.9 (1.9–12.0)		3.8 (1.5–7.5)	
Arthritis	Yes	No	Yes	No	Yes	No	Yes	No
Total number of patients	49	31	3	2	0	3	4	2
Sex (F:M)	35:14	20:11	2:1	1:1	—	1:2	3:1	0:2
Median age, yrs (range)	6.2 (1.7–15.8)	5.2 (1.8–11.7)	11.6 (5.7–12.2)	5.0 (0.5–9.5)	—	1.7 (1.1–15.4)	9.4 (6.4–15.7)	8.8 (8.3–9.2)
Race (% white)	37/45 (82)[†]	23/26 (88)	3/3 (100)	2/2 (100)	—	1/1 (100)	0/3 (0)	1/2 (50)
Muscle weakness (%)	47/49 (96)	27/31 (87)	0/3 (0)	0/2 (0)	—	3/3 (100)	4/4 (100)	2/2 (100)
Heliotrope rash (%)	39/49 (80)	19/31 (61)	1/3 (33)	1/2 (50)	—	0/3 (0)	1/4 (25)	1/2 (50)
Gottron's rash (%)	46/49 (94)	24/31 (77)	3/3 (100)	2/2 (100)	—	0/3 (0)	3/4 (75)	1/2 (50)
CPK > 300 U/l* (%)	18/45 (40)	13/29 (45)	0/2 (0)	0/2 (0)	—	3/3 (100)	2/4 (50)	0/2 (0)
AST > 40 U/l* (%)	39/47 (83)	18/26 (69)	0/1 (0)	1/2 (50)	—	3/3 (100)	3/4 (75)	1/2 (50)
ALT > 40 U/l* (%)	16/33 (48)	10/17 (59)	0/1 (0)	1/1 (100)	—	1/1 (100)	2/3 (67)	0/2 (0)
LDH > 900 U/l* (%)	4/10 (40)	3/9 (33)	—	0/1 (0)	—	1/1 (100)	—	—
Abnormal muscle biopsy (%)	35/38 (92)	19/20 (95)	0/1 (0)	0/1 (0)	—	3/3 (100)	4/4 (100)	1/1 (100)
Abnormal EMG (%)	33/47 (70)	20/21 (95)	1/2 (50)	0/1 (0)	—	2/2 (100)	4/4 (100)	1/2 (50)
Abnormal nailfolds (%)	38/49 (78)	19/31 (61)	2/3 (67)	1/2 (50)	—	1/3 (33)	2/4 (50)	1/2 (50)

[†]Ethnicity of the other 8 patients: aboriginal (n = 2), East Indian (n = 1), Black (n = 2), Oriental (n = 3).
*Muscle enzymes available at presentation of JDM.
CPK: creatine phosphokinase, AST: aspartate aminotransferase, ALT: alanine aminotransferase, LDH: lactate dehydrogenase, EMG: electromyogram, JDM: juvenile dermatomyositis, JPM: juvenile polymyositis.

Table 2. Relationship between arthritis and clinical features in patients with JDM. Reanalysis of data after reclassification of probable steroid knee effusions as "no arthritis" results in a significant relationship between arthritis and Raynaud's phenomenon (23% vs 2%; p = 0.005). No significant relationship occurs between arthritis and the other clinical features in JDM.

	Arthritis, n = 49	No Arthritis, n = 31	p
Sex (% female)	71	65	0.52
Race (% white, n = 71)	82	88	0.48
Positive ANA (%)	79	71	0.57
Positive RF (%)	4	3	0.85
Calcinosis (%)	24	26	0.89
Nodules (%)	12	10	0.72
Vasculitis (%)	24	10	0.10
Raynaud's phenomenon (%)	14	3	0.11

for myositis, with remission of the arthritis within a median of 2.0 months (range 0.1–64.5 mo). All received corticosteroid therapy, but 47 of 80 patients took additional medications including nonsteroidal antiinflammatory drugs (NSAID) (n = 18), methotrexate (MTX; 27), plaquenil (22), azathioprine (AZA; 1), cyclosporine (3), intravenous immunoglobulin (22), and cyclophosphamide (6). In 19 patients (39%), there was a recurrence of the arthritis when the corticosteroids were tapered. The arthritis flared despite the fact that these 19 patients were receiving other therapeutic agents including NSAID (9), MTX (6), plaquenil (4), AZA (1), and intravenous immunoglobulin (4). Four patients with JDM eventually required corticosteroid wrist injections, with resolution of the arthritis for many months. The 4 patients who received joint injections all presented with a stiff wrist that was clinically symptomatic but was unresponsive to NSAID and therapy for myositis.

Of the 49 patients with arthritis, 27 had radiographs that were available for review (9 had serial radiographs available). The areas studied included shoulders (4), elbows (8), wrists (7), hands (6), hips (3), knees (11), ankles (5), and feet (3). No erosions were seen but 2 patients had joint space narrowing that did suggest some degree of joint damage. The features of the radiographs are given in Table 3.

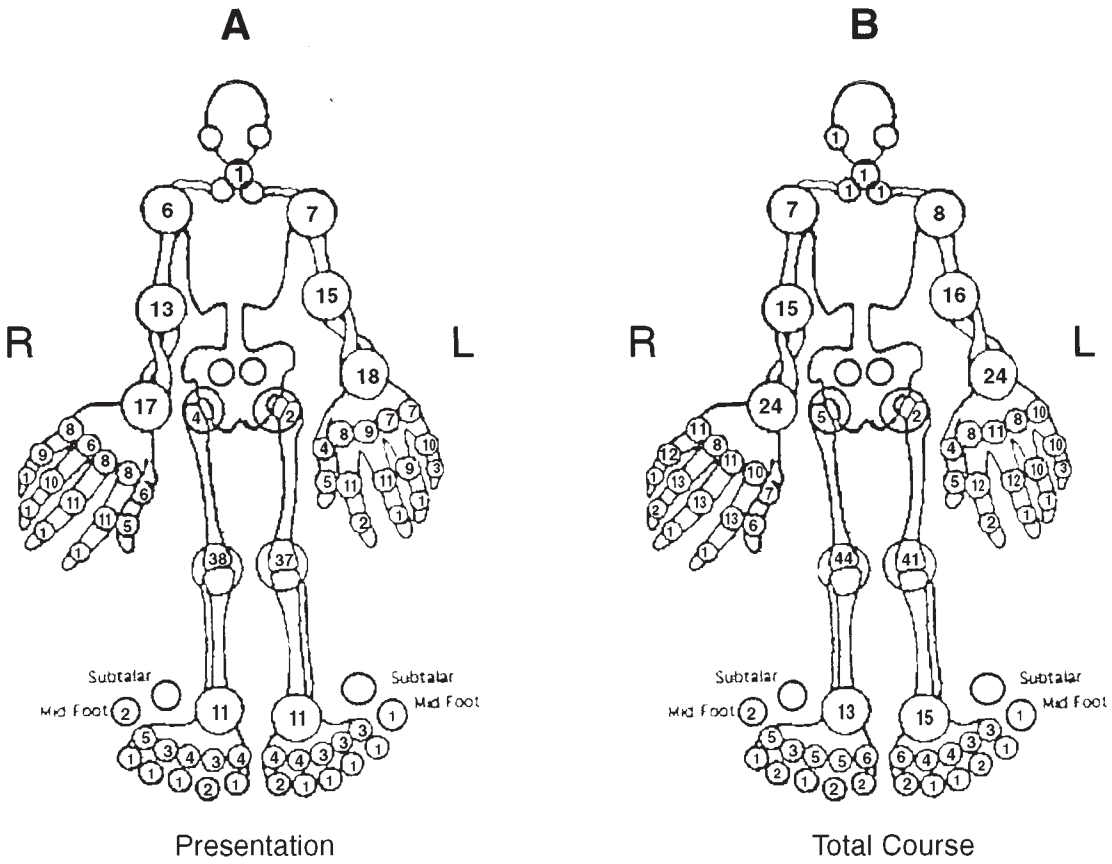

Figure 1. Distribution of joint involvement in 49 JDM patients with arthritis. The number represents the number of patients with arthritis in each designated joint at the onset of arthritis (presentation, A) and during entire course of study period (total course, B). Median followup period was 3.8 years (range 0.15–13.4).

Table 3. Radiographic features of JDM patients with arthritis.

	Osteopenia	Calcification	Erosions	Joint Space Narrowing	Effusions
JDM, n = 27 patients (%)	18 (67)	9 (33)	0 (0)	2 (7)	3 (11)

It is possible that some of our patients had asymptomatic knee effusions that were not true arthritis but rather were a complication of corticosteroid therapy. In these patients, we noted effusion(s) of the knee in the absence of joint line tenderness, limited range of movement, or increased warmth. We therefore reanalyzed our data by reclassifying the 18 patients with potentially steroid related knee effusions as having no arthritis. In this case, the prevalence rate of arthritis would be decreased to 39% (n = 31, 95% CI 28–50%), with a pauciarticular involvement in 48% and polyarticular involvement in 52%. With these 18 patients reclassified, the remaining patients with arthritis had a higher prevalence of Raynaud's phenomenon (23% vs 2%; p = 0.005). We found no relationship between arthritis and the remaining clinical features (Table 2) in our patients with JDM.

Other patients. The patients with JPM demonstrated no arthritis. In patients with amyopathic JDM, 3 of 5 patients had arthritis: nonerosive polyarthritis of small joints (n = 1) and small plus large joints (2). Four of 6 patients with overlap myositis syndromes had nonerosive polyarthritis affecting the small and large joints. Only 2 patients with arthritis had CHAQ scores greater than 0 (one with amyopathic JDM had a CHAQ score of 0.25, one with overlap myositis syndrome had a CHAQ score of 0.125).

No patient with amyopathic JDM was screened for myositis-specific autoantibodies. Some of the amyopathic JDM patients had autoantibody screening for ANA (3 positive, 2 negative), anti-dsDNA (4 negative), anti-extractable nuclear antigen (2 negative), and RF (5 negative).

DISCUSSION

We determined that arthritis is a frequent occurrence in JDM, manifesting in 61% of our patients. This falls within the upper range of previous prevalence rates of arthritis reported in JDM cohorts[1-4]. Even if we exclude the 18 patients with potential corticosteroid knee effusions, arthritis is still a common manifestation, with a prevalence of 39%.

The arthritis may occur before or after the onset of other

features of JDM, but the median presentation was within the first 5 months. This suggests that the arthritis is a true JDM associated clinical feature.

It is possible that in some patients, arthritis, especially asymptomatic knee effusions, may be a complication of therapy. The administration of high dose corticosteroids has been associated with the development of synovial effusions in up to 30% of treated patients[12]. In that study, Lally examined the relationship between high dose intravenous methylprednisolone (equivalent to ≥ 60 mg prednisone/day) in 35 patients with chronic obstructive pulmonary disease without a history of articular disease. Twelve patients developed small asymptomatic knee effusions during or shortly after the steroid pulse. In 9 cases, the knee joint was aspirated and analysis revealed a noninflammatory synovial fluid with normal viscosity and mucin clot formation. It was postulated that the mechanism of the effusion is related to fluid shifts across the synovial vasculature induced by corticosteroids.

There is controversy regarding the frequency and significance of corticosteroid related effusions. There is very little information in the literature to address this issue. Some people believe that this phenomenon only occurs in adults. However, in children receiving corticosteroid therapy, our practice has been to consider corticosteroid related effusions when examination reveals knee effusion in the absence of pain, limited range of movement, or increased warmth. No asymptomatic knee effusion in our patients was aspirated, and therefore treatment related effusions remain a possibility.

The arthritis in JDM is more similar to that seen in patients with SLE as opposed to juvenile rheumatoid arthritis. The articular involvement of fingers, wrists, knees, and elbows in our JDM patients is in a distribution commonly seen in SLE. As in SLE, the arthritis is very responsive to corticosteroid therapy and may flare when the corticosteroids are tapered[13,14]. Additionally, the nonerosive nature of the arthritis in JDM is similar to SLE, and contrasts with the erosive changes seen in some patients with JRA.

The severity of JDM is worse in patients who have clinical manifestations that include profound weakness and/or ischemic ulcers of the skin or gut. We examined for associations between JDM severity/clinical features and the development of arthritis; we found no relationship between the occurrence of arthritis and sex, race, ANA/RF positivity, calcinosis, nodules, vasculitis, or Raynaud's phenomenon. Hence, we could not find clinical, demographic, or severity features that predicted the presence or absence of arthritis in JDM. However, when we excluded patients with asymptomatic knee effusions in our analysis, the occurrence of arthritis became significantly related with the presence of Raynaud's phenomenon. The relevance of this potential relationship remains to be determined.

Myositis-specific autoantibodies (MSA) are autoantibodies that are directed against proteins or RNA found in patients with myositis. They are frequently found in adults with inflammatory myopathies, and much less commonly in children. In adults, MSA can be used to group patients who behave similarly in their clinical presentation, disease severity, response to therapy, and prognosis. Adults with anti-Mi-2 autoantibodies have classical manifestations of DM including proximal muscle weakness, myalgias, and the typical rash of DM including the V sign and shawl sign. These patients respond well to treatment with corticosteroids and have a good prognosis. The profile of adult patients with anti-synthetase autoantibodies (Jo-1, PL-7, PL-12) includes arthritis, "mechanic's" hands, fever, and interstitial lung disease. These patients exhibit only a moderate response to corticosteroid therapy and are susceptible to a flare of disease with tapering of therapy. Previously, we found MSA to be rare in JDM[15]. Two patients in that study who had anti-Mi-2 autoantibodies were included in this series; one had a pauciarticular arthritis involving the knees. None of our JDM study patients with arthritis had anti-synthetase autoantibodies. Thus, in our pediatric population MSA were not useful in the diagnosis of JDM or in prediction of which patients will develop arthritis.

The arthritis was persistent in 39% of our patients despite remission of the JDM. This rate is in contrast to the 13% rate of chronic polyarthritis reported by Hollister[5]. The arthritis may warrant therapy since patients often complain of pain despite the fact that the arthritis does not cause significant functional limitations in the majority (as confirmed by the mostly unremarkable CHAQ scores in our patients with JDM). Although the arthritis is very responsive to corticosteroids, some of our patients were treated with other antiarthritic agents because of our concerns about the safety of longterm corticosteroid use. When the other features of JDM in these patients are in remission (absence of rash with normal strength and muscle enzymes), we often managed the arthritis with other therapies including NSAID, MTX, or intraarticular steroid injections. In our experience, most cases of JDM associated arthritis respond well to these therapies, with minimal adverse effects compared with longterm use of corticosteroids.

Our patients with amyopathic JDM and overlap myositis syndrome also had a nonerosive polyarthritis affecting the small and large joints. No patient with JPM had arthritis. We determined that it was not possible to use the associated manifestation of arthritis to discriminate between the different types of idiopathic myositis syndromes. Additionally, arthritis is not simply an overlap feature but is truly a part of JDM.

There are some potential limitations to be considered in interpreting our results. JDM is a rare disease; however, our study included a fairly large cohort of patients. We believe that our reported prevalence of arthritis in JDM is an accurate value (for patients seen in a referral setting) and not limited by patient numbers, as shown by the fairly tight confidence intervals.

Variability can be encountered among radiologists in the interpretation of radiographs. As a result, all radiographs in

this study were reviewed in a standard fashion by a single radiologist blinded to the patient, type of myositis, and treatment. However, the number of radiographs that were examined was limited. In our study, not all patients had radiographs taken of their affected joint(s) and only 9 patients had serial radiographs available for review. Importantly, the most symptomatic or severely affected patients had radiographs taken, therefore our chance of missing any destructive changes is reduced.

Finally, we acknowledge that there was variable followup of our patients, as they were recruited at different times during the study period. This may limit our insight into the persistence and course of the arthritis encountered in JDM.

We conclude that a nonerosive arthritis similar to that seen in SLE is a frequent manifestation of JDM. The distribution of the arthritis primarily involves the fingers, wrists, elbows, and knees. The arthritis is seen early in the course of JDM and often responds to JDM treatment. However, the arthritis frequently recurs despite remission of the JDM, especially when the corticosteroids used for treatment of the myositis are tapered. More than one-third of our patients developed chronic polyarthritis requiring further treatment with medications or intraarticular steroid injections. The arthritis in JDM is a frequent and clinically important problem even when the myositis is in remission.

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