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Clinical Characteristics and Outcomes of Juvenile and Adult Dermatomyositis

Dermatomyositis (DM) is an idiopathic inflammatory myopathy with bimodal onset age distribution. The age of onset is between 5-18 yr in juvenile DM and 45-64 yr in adult DM. DM has a distinct clinical manifestation characterized by proximal muscle weakness, skin rash, extramuscular manifestations (joint contracture, dysphagia, cardiac disturbances, pulmonary symptoms, subcutaneous calcifications), and associated disorders (connective tissue disease, systemic autoimmune diseases, malignancy). The pathogenesis of juvenile and adult DM is presumably similar but there are important differences in some of the clinical manifestations, associated disorders, and outcomes. In this study, we investigated the clinical characteristics and outcomes of 16 patients with juvenile DM and 48 with adult DM. This study recognizes distinctive characteristics of juvenile DM such as higher frequency of neck muscle involvement, subcutaneous calcifications, and better outcomes.

Key Words : *Juvenile Dermatomyositis; Adult Dermatomyositis; Clinical Characteristics; Outcomes*

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INTRODUCTION

Dermatomyositis (DM) is one of the idiopathic inflammatory myopathies. It is clinically characterized by progressive symmetrical proximal muscle weakness and a specific skin manifestation. It has extramuscular manifestations such as joint contractures, dysphagia, cardiac disturbance, pulmonary symptoms, and subcutaneous calcifications. DM also has an association with malignant disease, and various autoimmune and connective tissue diseases. The average age at diagnosis is 40 yr, and almost twice as many women are affected as men (1). DM has bimodal onset age distribution. Most cases of juvenile DM present between 5-14 yr of age and most adult DM in the fifth and sixth decades of life (2).

The incidence of juvenile DM is 2-3 cases per one million per year (2-5), and adult DM is 5-10 cases per one million per year (6). Major differences between juvenile and adult DM include the greater potential for calcinosis, the presence of vascular inflammation, and the potential for lipodystrophy accompanied by insulin resistance in juvenile DM (7). DM is still associated with morbidity and mortality rates as high as 4-50%, principally related to life-threatening muscle weakness and cardiac and lung complications.

Although there have been many reports on DM, there is a few study that compares juvenile to adult DM for clinical characteristics and outcomes. The aim of this study is to clarify the differences in clinical characteristics and outcomes between juvenile and adult DM.

MATERIALS AND METHODS

A total of 64 patients with juvenile DM (n=16) and adult DM (n=48) were enrolled in this study. All patients were treated at Severance Hospital (Yongdong and Shinchon) or Konyang University Hospital between January 1996 and December 2006. Diagnosis of DM in all cases was based on the criteria defined by Bohan and Peter (8, 9): symmetric muscle weakness, increased muscle enzymes, myopathic changes on electromyography, typical histological findings on muscle biopsy, and characteristic dermatologic signs. In this study, we excluded patients with amyopathic dermatomyositis.

We defined juvenile DM as onset of symptoms before 18 yr of age and adult DM as onset of symptoms after 18 yr of age. We analyzed the following data from hospital records: the age at diagnosis; current age; sex; duration of disease; distribution and degree of weakness; electrophysiological study findings; presence of extramuscular manifestations such as joint contractures, dysphagia, cardiac disturbance, pulmonary symptoms, and subcutaneous calcifications; and associated disorders such as mixed connective tissue disease, rheumatoid arthritis, scleroderma, systemic lupus erythematosus, and malignancy.

We analyzed the laboratory findings such as creatine kinase (CK), alanine and aspartate aminotransferase (ALT, AST), antinuclear antibodies (ANA), anti- Ro/SSA, anti-SSB/La, anti-histidyl-tRNA synthesase (anti-Jo-1), anti-double-stranded DNA, and rheumatoid factor (RF) at the time of presence of

muscle weakness.

Principles of treatment for both juvenile DM and adult DM were as follows: initial treatment was started with prednisolone in a daily dose of 0.5-1 mg/kg. Dose reduction was usually initiated after 4-6 weeks of full-dose therapy depending on the degree of clinical improvement and steroid side effects. In general, the dose of prednisolone was reduced by 5 mg/day per week, reaching a level of 25-35 mg/day by the end of the second month with a subsequent slower rate of reduction or conversion to an alternate-day regimen. Oral methotrexate (7.5-15 mg/week) or azathioprine (2-3 mg/kg/day) was used in combination with prednisolone from the start in certain patients, namely those with very severe myositis and those with a long interval before commencement of treatment. They were also added as second-line therapy in patients who responded slowly to steroid therapy, developed prominent steroid side effects, or suffered relapse with steroid therapy alone.

Patients were followed up on a regular basis and were examined at 1-3 month intervals in an outpatient setting. During each visit, patients underwent detailed physical and neurological examinations. The minimal follow-up duration was 34 months. Patients who died before 34 months were included in the study.

Evaluation of outcomes was performed on all patients between September and December 2006. The outcomes of juvenile and adult DM patients during the evaluation period were defined as 1) remission: disappearance of skin manifestations, normalization of muscle strength, normal function of major organs, normalization of both serum muscle enzyme levels and electromyographic abnormalities-all persisting after therapy discontinuation; 2) improvement: improvement of muscle strength and skin manifestations, functions of involved major organs, and biochemical data with therapy; 3) deterioration: worsening of muscle strength and skin manifestations, functions of involved major organs, and biochemical findings despite therapy; and 4) death: death of patient despite therapy during the follow-up period.

Statistical analysis was performed using SPSS version 11.0 (SPSS Inc. Chicago, IL, U.S.A.). For group comparisons involving binary data, we used chi-square test and Fisher's exact test. Cumulative survival rates were calculated, using Kaplan-Meier method for the whole series. The significance of the survival difference was examined by the log-rank test, and $P<0.05$ was considered statistically significant.

RESULTS

Clinical and laboratory characteristics of juvenile DM

Details of each patient are given in Table 1. There were 16 patients (patient Nos. 1-16), 6 males and 10 females, who had typical muscle pathology and electrophysiological findings consistent with DM. Onset age varied from 4-12 yr. The mean duration of disease was 52 months (range, 34-110 months). Onset symptoms were skin rash in 6 patients and limb weakness in 10. Most showed symmetrical proximal muscle weakness, and none showed facial weakness. Eight out of 16 patients showed neck muscle weakness. Six patients showed dysphagia. Six patients showed right bundle branch block or ST-T wave abnormality on electrocardiography (EKG). None of the patients showed interstitial lung disease on chest radiography. Six patients showed subcutaneous calcifications. One patient showed systemic lupus erythematosus, scleroderma, and joint contracture. None of the patients showed mixed connective tissue disease, rheumatoid arthritis, or malignancy.

CK was increased in 12 patients. ALT was increased in 8 patients. AST was increased in 12 patients. ANA with a speckled pattern were present in 5/12 (41.7%) patients. Anti-Ro/SSA antibodies and anti-ds DNA antibodies were detected in 1/12 (8.3%) patients. Anti-La/SSB antibodies, anti-Jo-1 antibodies, RF were negative in all patients.

Clinical and laboratory characteristics of adult DM

Details of each patient are given in Table 1. There were 48 patients (patient Nos.17-64), 17 males and 31 females, who had typical muscle pathology and electrophysiological findings consistent with DM. Onset age varied from 20-73 yr. The mean duration of disease was 41 months (range, 3-110 months). Onset symptoms were skin rash in 31 patients and limbs weakness in 17. Most patients showed symmetrical proximal muscle weakness and few showed facial weakness. Five out of 48 patients showed neck muscle weakness. Nine patients showed dysphagia. Eleven out of 48 patients showed right bundle branch block, ST-T wave abnormality, or tachyarrhythmia on EKG. Six patients showed interstitial lung disease on chest radiography. Six patients showed nasopharyngeal cancer, ovarian cancer, thyroid cancer, or cervical cancer. Five patients showed mixed connective tissue disease and three patients showed scleroderma. Two patients showed systemic lupus erythematosus and one patient showed subcutaneous calcifications, joint contracture, and rheumatoid arthritis.

CK was increased in 46 patients. ALT was increased in 36 patients. AST was increased in 40 patients. ANA with a speckled pattern were present in 17/36 (44.7%) patients. Anti-Ro/SSA antibodies were detected in 9/36 (25.0%) patients. Anti-Jo-1 antibodies were detected in 6/36 (16.7%) patients. Anti-ds DNA antibodies were detected in 2/36 (5.6%) patients. Anti-La/SSB antibodies and RF were detected in patient respectively out of 36.

Clinical and laboratory comparison between juvenile and adult DM patients

The frequency of neck muscle weakness and subcutaneous calcification in patients with juvenile DM was significantly higher than that of patients with adult DM ($P<0.05$, Table

Table 1. Characteristics of juvenile and adult dermatomyositis (DM) patients

Patient No.	Sex/ Age (yr)	Age of onset (yr)	Follow up duration (months)	Onset Sx	Distribution of muscle weakness						Joint contracture	Dysphagia	Cardiac disturbance (EKG)	Pulmonary symptom (Chest radiography)	Subcutaneous calcification	Associated disorders	Malignancy
					Face	Neck	U/Ex		L/Ex								
							Prox	Distal	Prox	Distal							
Juvenile DM																	
1	M/14	11	36	R	5	4+	4+	5	4+	5	–	–	RBBB	–	+	–	–
2	F/10	5	58	L	5	5	4+	5-	4	5-	–	+	ND	–	–	–	–
3	F/11	7	40	A	5	4+	4	5-	4	5-	–	–	RBBB	–	–	–	–
4	M/14	10	39	U	5	5	4	5	4+	5	–	–	ST	–	+	–	–
5	F/13	9	42	R	5	5	4+	5	4+	4+	–	+	NL	–	–	–	–
6	F/15	11	45	L	5	4+	4+	5-	4	5-	–	–	NL	–	+	–	–
7	F/14	4	110	A	5	4	4+	4+	4	5-	–	–	ND	–	–	–	–
8	F/14	11	36	R	5	5	4	5-	4+	5-	–	–	ST	–	–	SLE	–
9	F/13	10	35	L	5	4	4-	5-	4	4+	–	+	NL	–	–	–	–
10	F/15	12	34	A	5	4	3	4+	3+	4+	+	+	NL	–	+	–	–
11	M/11	7	38	R	5	5	4-	5-	4-	5-	–	–	NL	–	–	–	–
12	F/10	6	42	R	5	5	4+	5	4+	5-		–	NL	–	–	–	–
13	M/13	9	45	U	5	5	4+	5	5-	5		–	ST	–	+	–	–
14	M/15	6	100	R	5	4+	4+	5	4	5-		+	NL	–	+	SCL	–
15	F/12	8	48	A	5	5	5-	5	4+	5		–	NL	–	–	–	–
16	M/16	9	80	L	5	4	4+	5-	4+	5-		+	RBBB	–	–	–	–
Adult DM																	
17	F/55	51	38	R	5	5	4+	5-	4-	4+	–	–	ST	–	–	–	–
18	M/24	20	46	L	5	5	5	5	4-	4+	–	–	–	–	–	–	–
19	F/57	54	36	A	5	4+	4-	5	4	5	–	–	–	–	–	–	–
20	F/62	59	36	L	5	5	4-	5	4	4+	+	–	RBBB	ILD	–	SCL	–
21	F/54	50	48	R	5	5	3+	4+	3+	4+	–	–	–	–	–	–	–
22	M/59	55	40	U	5	5	4-	4+	4-	4+	–	+	RBBB	–	–	–	–
23	F/56	52	42	R	5	5	4	4	3	4	–	+	–	–	–	RA	–
24	M/67	65	13	R	5	5	4	5	4-	5	–	–	–	ILD	–	–	–
25	M/57	56	11	R	5	5	4	4+	4	4+	–	–	–	–	–	–	Naso Ca
26	M/53	43	110	R	5	5	5	5	5	5	–	+	ST	–	+	MCTD	–
27	F/45	41	40	R	5	3	4	5	4	5	–	–	–	–	–	SCL	–
28	F/69	69	6	A	5	5	3	5-	4-	5	–	+	–	–	–	–	Ovarian Ca
29	M/70	66	45	R	4	5	4	5	4+	5	–	–	ST	–	–	–	–
30	F/46	45	3	L	5	5	4+	5	4+	5	–	–	–	ILD	–	MCTD	–
31	F/55	52	36	R	5	5	3	5	3	5	–	+	–	–	–	–	–
32	M/64	60	44	L	5	5	5	5	4+	5	–	–	ST	–	–	–	–
33	F/34	30	42	R	5	5	4+	5	5	5	–	–	–	–	–	–	–
34	F/47	42	60	R	5	5	5-	5-	5-	5-	–	–	–	–	–	–	Cervical Ca
35	F/32	27	50	A	5	4+	4	4+	4	5	–	–	–	–	–	–	–
36	F/54	50	48	R	5	5	5-	5	5-	5	–	–	–	–	–	–	Thyroid Ca
37	F/50	46	38	L	5	5	5	5	4-	5-	–	–	–	–	–	–	–
38	F/32	29	36	R	5	5	4+	5-	4-	5-	–	–	–	–	–	–	–
39	M/76	73	36	R	5	5	4	4+	4	4+	–	–	–	–	–	–	–
40	M/37	37	5	A	5	3	3	4+	4	4+	–	–	TA	ILD	–	MCTD	–
41	F/34	30	40	R	5	5	4-	5	4-	5-	–	–	–	–	–	–	–
42	F/30	26	38	R	5	5	4-	4+	4-	4+	–	–	–	–	–	–	–
43	M/56	52	42	R	5	5	4-	4+	4-	4+	–	+	–	–	–	–	Naso Ca
44	F/24	20	44	A	5	4-	4-	5	4-	5	–	–	ST	–	–	–	–
45	F/25	21	40	L	5	5	3	4+	4-	4+	–	–	–	–	–	SLE	–
46	M/24	21	36	R	5	4+	4-	4+	4-	4+	–	–	–	–	–	–	–
47	M/32	26	72	U	5	5	4+	5	5	5	–	–	–	–	–	–	–
48	F/27	21	68	R	5	5	4	5-	4+	5-	–	–	–	–	–	–	–
49	M/28	24	47	A	4+	5	4+	5-	4+	5-		–	–		–	–	–
50	F/30	27	34	R	5	5	4+	5	4-	5-		-	RBBB	ILD	-	SLE	-
51	F/37	31	70	L	5	5	5-	5	4+	5		+	-		-	-	-

(Continued next page)

Table 1. *(Continued from the previous page)* Characteristics of juvenile and adult dermatomyositis (DM) patients

Patient No.	Sex/ Age (yr)	Age of onset (yr)	Follow up duration (months)	Onset Sx	Distribution of muscle weakness						Joint contracture	Dysphagia	Cardiac disturbance (EKG)	Pulmonary symptom (Chest radiography)	Subcutaneous calcification	Associated disorders	Malignancy
					Face	Neck	U/Ex		L/Ex								
							Prox	Distal	Prox	Distal							
52	M/39	33	65	A	5	4+	4+	5	5-	5-		–	–		–	–	–
53	F/32	27	58	R	5	5	4+	5	4+	5		–	–		–	MCTD	–
54	F/33	29	47	L	5	5	5	5	4	4+		–	–		–	–	–
55	M/54	51	35	A	5	4+	4+	4+	4+	4+		+	–		–	SCL	–
56	F/45	42	36	U	5	5	4+	5	5-	5		–	–		–	–	Ovarian Ca
57	F/35	31	45	U	5	5	4+	5	5-	5		–	TA		–	–	–
58	F/32	28	46	R	5	5	5-	5	5-	5		–	–		–	–	–
59	M/58	57	5	U	5	5	4+	5	5-	5		–	–	ILD	–	–	–
60	M/40	37	36	A	5	4+	4+	5-	4+	5-		+	–		–	–	–
61	F/26	22	44	L	5	5	5-	5	4+	5		–	–		–	–	–
62	F/38	35	36	R	5	5	4+	5-	5-	5		–	ST		–	MCTD	–
63	F/40	36	47	L	5	5	5-	5	4+	5		–	–		–	–	–
64	F/45	41	45	A	5	4+	4+	4+	4+	4+		–	–	–	–	–	–

R, rash; A, all limds weakness; U, upper limb weakness; L, lower limb weakness; ST, ST-T wave abnormality; RBBB, right bundle branch block; TA, tachyarrythmia; NL, normal; ND, not done; ILD, interstitial lung disease; SCL, scleroderma; SLE, systemic lupus erythematosus; MCTD, mixed connective tissue disease; Naso Ca, nasopharyngeal cancer; Ovarian Ca, ovarian cancer; Cervical Ca, cervical cancer; Thyroid Ca, thyroid cancer; Sx, symptom; prox, moximal.

Table 2. Comparison of clinical and laboratory characteristics between juvenile and adult DM patients

	Juvenile DM (n=16)	Adult DM (n=48)	*P* value
Neck muscle weakness	8 (50%)	10 (20.8%)	0.03
Joint contracture	1 (6%)	1 (2.1%)	NS
Dysphagia	6 (37.5%)	9 (18.8%)	NS
EKG abnormality	6 (37.5%)	11 (22.9%)	NS
Interstitial lung disease	0	6 (12.5%)	NS
Subcutaneous calcification	6 (37.5%)	1 (2.1%)	0.01
Overlap syndrome	2 (12.5%)	11 (22.9%)	NS
Malignancy	0	6 (12.5%)	NS
Raised CK	12 (75.0%)	46 (95.8%)	NS
Raised ALT	8 (50.0%)	36 (78.3%)	NS
Raised AST	12 (75.0%)	40 (87.0%)	NS
Positive ANA	5 (41.7%)*	17 (44.7%)†	NS
Positive anti-Ro/SSA	1 (8.3%)*	9 (25.0%)†	NS
Positive anti-La/SSB	0*	1 (2.8%)†	NS
Positive anti-Jo-1	0*	6 (16.7%)†	NS
Positive anti-ds DNA	1 (8.3%)*	2 (5.6%)†	NS
Positive RF	0*	1 (2.8%)†	NS

*Data available for twelve patients; †data available for 36 patients.
DM, dermatomyositis; CK, creatine kinase; ALT, alanine aminotransferase; AST, aspartate aminotrnasferase; ANA, antinuclear antibodies; Jo-1, histidyl-tRNA synthetase; RF, rheumatoid factor; ds DNA, double-stranded DNA; NS, not significant.

2). Comparing the frequency of dysphagia, EKG abnormality, interstitial lung disease, joint contracture, overlap syndrome, and malignancy, there was no statistically significant difference between juvenile and adult DM (Table 2).

Comparing the frequency of raised CK, raised ALT, raised AST, positive ANA, positive anti-Ro/SSA, positive anti-La/SSB, positive anti-Jo-1, positive anti-ds DNA, and positive RF, there was no statistically significant difference between juvenile and adult DM (Table 2).

Table 3. Outcomes of juvenile and adult DM patients

	Juvenile DM (n=16)	Adult DM (n=48)	*P* value by Fisher's exact test
Remission	9 (56.2%)	18 (37.5%)	NS
Improvement	6 (37.5%)	17 (35.4%)	NS
Deterioration	1 (6.3%)	3 (6.3%)	NS
Death	0 (0%)	10 (20.8%)	<0.001

DM, dermatomyositis; NS, not significant.

The median follow-up duration of juvenile and adult DM patients was 52 months (range, 34-110 months) and 41 months (range, 3-110 months), and there was no statistically significant difference between two groups. The outcomes of patients with juvenile and adult DM are summarized in Table 3. In the 16 patients with juvenile DM, 9 patients (56.2%) had remission. Six patients (37.5%) had improvement of juvenile DM. The 1 remaining patient (6.3%) had a deteriorating course. None of the 11 patients with juvenile DM died during follow-up. Eighteen out of 48 adult DM patients (37.5%) had remission and 17 (35.4%) had improvement. Three patients (6.3%) had deterioration of adult DM. Ten out of the total 48 patients with adult DM died during the follow up. The mortality rate of adult DM was 20.8% and significantly higher than juvenile DM (0%) (P<0.001). The survival difference between juvenile and adult DM by log-rank test was statistically not significant (P=0.0544) (Fig. 1).

The characteristics of patients with adult DM who died

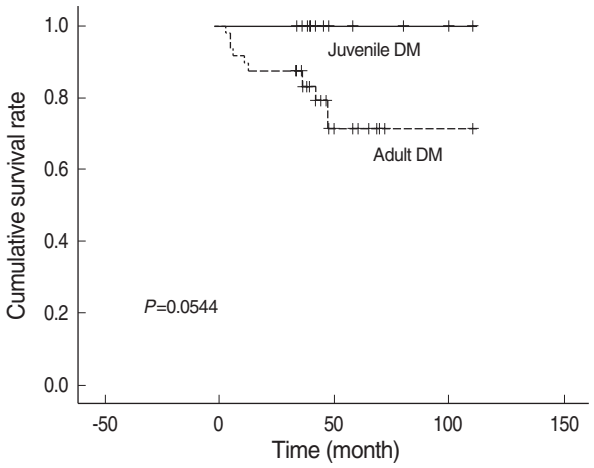

Fig. 1. Survival curve of 16 patients with juvenile DM and 48 with adult DM. Number of available adult DM patients: 42 at 1-yr follow up, 40 at 3-yr follow up, and 38 at 9-yr follow up.

Table 4. Cause of death in 10 patients with adult DM

Patient No.	Sex/age (yr)	Duration of disease (month)	Cause of death
20	F/62	36	Pulmonary hypertension/ILD
24	F/67	13	Sepsis
25	M/57	11	Cancer
28	F/69	6	Cancer
30	F/46	3	Pneumonia
36	F/54	48	Cancer
40	M/37	5	Pneumonia
43	M/56	42	Cancer
56	F/45	36	Cancer
59	M/58	5	Pneumonia

DM, dermatomyositis; ILD, interstitial lung disease.

are shown in Table 4. Death was caused by cancer in 5 patients and by pulmonary complications in other 4 patients that included pulmonary hypertension due to interstitial lung disease in 1 and aspiration pneumonia in 3. The 3 patients with aspiration pneumonia concomitantly had moderate ventilatory insufficiency. One patient died from sepsis.

DISCUSSIONS

DM is characterized by subacute (over several weeks to months), progressive, and proximal muscle weakness. In our study, most patients with juvenile and adult DM had subacute, progressive, and proximal muscle weakness.

It is known that DM has a bimodal age distribution: one peak occurs in children between 5-14 yr of age and a second, larger peak occurs between 45-64 yr of age (10, 11). In our study, onset age distribution was similar to that of previous reports on juvenile DM (2, 12), but there was a wider variation of onset age distribution in adult DM. It is reported that females outnumber males by 2: 1 (10, 11), and our study also showed female predominance in both juvenile and adult DM.

In our study, the difference of neck muscle weakness frequency between juvenile and adult DM was statistically significant (P=0.03). The reason of this result has been not explained until now and further study is needed. Neck flexors muscle weakness is common in DM, but in some cases, there is also severe weakness of neck extensor muscles. This has been encountered particularly in patients with DM associated with scleroderma and mixed connective tissue disease (13). In our study, neck flexor muscle weakness was present in 8 out of 16 patients with juvenile DM (50.0%) but neck extensor muscle weakness was not. Neck flexor muscle weakness was present in 10 out of 48 patients with adult DM (20.8%) and in these cases, 3 patients with mixed connective tissue disease or scleroderma also showed neck extensor muscle weakness.

Skin rash often precedes the onset of weakness by weeks to months. Early in its course, rash and muscle enzyme elevations may be the sole manifestations of DM (14). In our study, most patients with juvenile and adult DM had proximal muscle weakness. The rash preceded muscle weakness in 6 out of 16 patients with juvenile DM (37.5%) and 23 out of 48 patients with adult DM (47.9%).

EKG abnormalities such as nonspecific ST-T wave change, varying degrees of heart block and bundle branch block, and tachyarrhythmia occur in up to 40% of DM patients (15). In our study, EKG abnormalities were found in 6 out of 16 patients with juvenile DM (37.5%) and 11 out of 48 with adult DM (22.9%).

Calcinosis of the skin or muscle is unusual in adults, but may occur in up to 40% of children or adolescents with DM (16). In our study, subcutaneous calcifications were found in 6 out of 16 patients with juvenile DM (37.5%) and 1 out of 48 with adult DM (2.1%). The mechanism of calcinosis development remains uncertain and there is still no standard treatment for the condition. Juvenile DM with pathologic calcinosis is strongly associated with the tumor necrosis factor alpha (TNF-α) 308 allele and increased production of TNF-α (17).

Interstitial lung disease (ILD) is often associated with considerable morbidity and mortality. The frequency of ILD in DM varies, ranging from 5-47% in the different series and tending to increase with the duration of the muscle disease; the overall frequency is probably close to 9%. In our study, ILD was found in 12.5% of adult DM patients and in none of the juvenile DM patients. DM is an uncommon etiology of ILD and most patients with ILD associated with DM present between the ages of 20-40 yr (18). Therefore, we postulate that in the course of time, ILD may develop in juvenile DM patients-the risk of ILD increasing with increased duration of DM.

The relationship of DM to malignancy has been recently clarified (19). The reported frequency of malignancy in DM

has varied from 6-60% in most large population-based cohort studies, revealing a frequency of about 20-25%. Malignancy is more common in older patients (>50 yr) and rare in childhood (20, 21). In our study, malignancy was found in 6 out of 48 patients with adult DM (12.5%) and in none with juvenile DM. There have been reports explaining the association of DM with malignancy. Some authors have postulated that tumor antigens produce autoantibodies that cross-react with various components of muscle in a small percentage of patients (22, 23). Less likely, the tumor releases myotoxic substances that instigate muscle fiber necrosis and inflammation. A third possibility is that both the neoplasm and myositis have a common pathogenesis.

A wide variety of malignancies have been reported in patients with DM. Gynecologic malignancy, particularly ovarian carcinoma, may be overrepresented in DM (19, 24). In our study, gynecologic malignancy such as ovarian cancer and cervical cancer were found in 3 out of 48 patients with adult DM. Asians with DM are often found to have nasopharyngeal cancer (25). In our study, nasopharyngeal cancer was found in 2 out of 48 patients with adult DM.

The inflammatory myopathies in approximately 20% of patients are associated with connective tissue diseases such as scleroderma, mixed connective tissue disease, Sjögren syndrome, systemic lupus erythematosus, and rheumatoid arthritis (10, 11), and is called the "overlap syndrome". In our study, overlap syndrome was found in 12.5% of juvenile DM patients and 22.9% of adult DM patients.

A several studies have evaluated the short- and long-term outcomes of DM (26-31). We observed that 56.2% of juvenile DM patients achieved remission whereas 37.5% improved and 6.3% worsened in clinical status. In contrast, we observed that 37.5% of adult DM patients achieved remission whereas 35.4% improved and 27.1% worsened in clinical status or died. Our findings confirm the results from other studies showing DM remission rates varying from 25-70% (32-35). We observed that the outcomes of juvenile DM were than adult DM.

In previous studies on the survival of DM patients, survival ranged from 72-84%, 3-73%, and 42-85% at 2, 5, and 10 yr, respectively (2, 26, 32, 34, 36). In our study, we found that 88%, 83%, and 72% of adult DM patients were still alive at 1, 3, and 4 yr at follow up, respectively (Fig. 1). The survival difference between juvenile and adult DM by log-rank test was statistically not significant (P=0.0544). This suggests that if more patients were enrolled in the study, a statistically significant difference may have been seen between juvenile and adult DM.

Kang et al. (37) have demonstrated that ILD occurs in patients with adult DM, and in this subset of idiopathic inflammatory myopathy, survival of patients is poor. In our study, 5 adult DM patients with ILD died.

DM has further been reported to be associated with increased mortality with a total mortality rate ranging from 4-50% (32, 34, 38, 39). Cancer, lung, and cardiovascular complications are generally cited as the most common causes of death in DM patients (26, 27, 35, 36, 40). In our series, the mortality rate of juvenile and adult DM patients was 0% and 20.8%. Among the 10 adult DM patients who died, 5 died within 1 yr of DM diagnosis. The leading cause of death was cancer in 50% of cases.

The second leading cause of death in our patients was aspiration pneumonia (33.3% of cases), which occurred during the first 5 months of adult DM diagnosis. Aspiration pneumonia was more often due to DM-related moderate ventilatory insufficiency (with decreased cough reflex and inability to take maximum inspiration) in these patients.

In conclusion, the clinical characteristics of juvenile DM reveal higher frequency of neck muscle involvement and subcutaneous calcifications than those of adult DM. The outcomes of juvenile DM are better than those of adult DM, and more aggressive therapy for juvenile DM should be advocated. We suggest that ILD and malignancy are strongly associated with mortality of DM. Further studies are needed on a population basis to determine whether the adequate treatment of ILD or malignancy reduces the mortality of DM patients and why the frequency of ILD and malignancy is higher in adult DM patients.

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