

Supplementary Table S1. Extended baseline clinical and serological characteristics of the study population by sex.

Variable	Overall (n=228)	Females (n=157)	Males (n=71)	p-value
<i>Ethnicity</i>				
Caucasian, N (%)	223 (97.8)	155 (98.7)	68 (95.8)	0.207
Hispanic, N (%)	3 (1.3)	1 (0.6)	2 (2.8)	0.207
Other, N (%)	2 (0.9)	1 (0.6)	1 (1.4)	0.207
<i>MAA</i>				
SSB, N	4 (1.8)	3 (2)	1 (1.4)	1.000
Ro60, N (%)	6 (2.6)	5 (3.2)	1 (1.4)	0.668
ACA, N (%)	2 (0.9)	2 (1.3)	0 (0)	1.000
ATA, N (%)	0 (0)	-	-	-
SmD, N (%)	1 (0.4)	1 (0.6)	0 (0)	1.000
Ku, N (%)	6 (2.7)	5 (3.3)	1 (1.4)	0.667
Pm/Scl, N (%)	9 (3.9)	7 (4.5)	2 (2.8)	0.724
RNP, N (%)	1 (0.4)	1 (0.7)	0 (0)	1.000
ANCA, N (%)	3 (1.3)	1 (0.6)	2 (2.8)	0.229
<i>MSA</i>				
Mi2 α , N (%)	16 (7.1)	9 (5.9)	7 (9.9)	0.282
Mi2 β , N (%)	13 (5.8)	8 (5.2)	5 (7)	0.556
TIF1 γ , N (%)	11 (4.9)	7 (4.6)	4 (5.6)	0.746
NXP2, N (%)	8 (3.6)	4 (2.6)	4 (5.6)	0.267
SAE, N (%)	6 (2.7)	5 (3.3)	1 (1.4)	0.667
PL-7, N (%)	9 (4)	6 (3.9)	3 (4.2)	1.000
PL-12, N (%)	9 (4)	8 (5.2)	1 (1.4)	0.278
EJ, N (%)	4 (1.8)	3 (2)	1 (1.4)	1.000
OJ, N (%)	2 (0.9)	2 (1.3)	0 (0)	1.000
Cn1a, N (%)	2 (0.9)	2 (1.3)	0 (0)	1.000
SRP, N (%)	4 (1.8)	3 (2)	1 (1.4)	1.000
Ks, N (%)	1 (0.4)	1 (0.6)	0 (0)	1.000
Zo, N (%)	1 (0.4)	1 (0.6)	0 (0)	1.000
<i>Muscular involvement</i>				
Abnormal muscular MRI, N (%)	53 (24.4)	35 (23.3)	18 (26.9)	0.812
Dysphagia, N (%)	49 (21.6)	30 (19.2)	19 (26.8)	0.201
<i>Lung involvement</i>				
NSIP, N (%)	84 (58.7)	59 (59)	25 (58.1)	0.924
UIP, N (%)	23 (16.1)	16 (16)	7 (16.3)	0.967
OP, N (%)	15 (10.8)	9 (9.4)	6 (14)	0.555
LID, N (%)	1 (0.7)	0 (0)	1 (2.3)	0.301
NSIP/OP, N (%)	11 (7.7)	8 (8)	3 (7)	1.000
Emphysema, N (%)	12 (5.3)	7 (4.5)	5 (7)	0.523
Pneumomediastinum, N (%)	0 (0)	-	-	-
Diaphragm involvement, N (%)	1 (0.5)	0 (0)	1 (1.5)	0.319
<i>Articular involvement</i>				
Joint deformity, N (%)	11 (4.9)	8 (5.3)	3 (4.2)	1.000
RF, N (%)	30 (13.6)	22 (14.7)	8 (11.4)	0.514
ACPA, N (%)	11 (5)	6 (4)	5 (7)	0.339
<i>Cutaneous involvement</i>				
Gottron's sign, N (%)	19 (8.3)	11 (7)	8 (11.3)	0.281
Shawl sign, N (%)	14 (6.1)	11 (7)	3 (4.2)	0.558
V sign, N (%)	35 (15.4)	26 (16.6)	9 (12.7)	0.451
Holster's sign, N (%)	9 (3.9)	8 (5.1)	1 (1.4)	0.280
Cutaneous ulcers, N (%)	15 (6.6)	12 (7.6)	3 (4.1)	0.052
Calcinosis cutis, N (%)	2 (0.9)	2 (1.3)	0 (0)	1.000
Periungual erythema, N (%)	13 (5.7)	9 (5.7)	4 (5.6)	1.000
RP, N (%)	45 (20.1)	31 (20.1)	14 (20)	0.982

ANA: antinuclear antibodies; ATS: anti-topoisomerase I, ENA: extractable nuclear antigen antibodies; Ro/SSA: Sjögren's-syndrome-related antigen A; La/SSB: Sjögren's-syndrome-related antigen B; ACA: anti-centromere antibodies; Sm: anti-Smith antibodies; RN: anti-ribonucleoprotein antibodies; Mi2A: anti-Mi-2 alpha; Mi2B: anti-Mi-2 beta; TIF1- γ : anti-transcription intermediary factor 1 gamma; MDA5: anti-melanoma differentiation-associated gene 5; NXP: anti-nuclear matrix protein 2; SAE: anti-small ubiquitin-like modifier activating enzyme (SAE1/2); Ku: anti-Ku antibodies; PL7: anti-threonyl-tRNA synthetase; PL12: anti-alanyl-tRNA synthetase; JO1: anti-histidyl-tRNA synthetase (Jo-1); EJ: anti-glycyl-tRNA synthetase; OJ: anti-isoleucyl-tRNA synthetase; Ro52: anti-Ro52/TRIM21; cN1A: anti-cytosolic 5'-nucleotidase 1; SRP: anti-signal recognition particle; HMGCR: anti-3-hydroxy-3-methylglutaryl-CoA reductase; PM/Scl: anti-PM/Scl complex (PM/Scl-75 and/or PM/Scl-100); ANCA: anti-neutrophil cytoplasmic antibodies; Ro60: anti-Ro60; KS: anti-asparaginyl-tRNA synthetase; ; Zo: anti-phenylalanyl-tRNA synthetase; ILD: interstitial lung disease; NSIP: non-specific interstitial pneumonia; UIP: usual interstitial pneumonia; OP: organising pneumonia; LID: lymphocytic interstitial pneumonia; NSIP/OP: mixed NSIP and organizing pneumonia pattern; ICU: intensive care unit; TLC: total lung capacity; RF: rheumatoid factor; ACPA: anti-citrullinated antibodies; MTX: methotrexate; MMF: mycophenolate mofetil; AZA: azathioprine; IVIG: intravenous immunoglobulin; RTX: rituximab; CYS A: cyclosporine A; Jaki: Janus kinase inhibitor; HCQ: hydroxychloroquine; CYC: cyclophosphamide; MITAX: myositis intention to treat activity index; MYOAC: myositis disease activity assessment tool; MMT: manual muscle testing; PhGA: physician's global assessment.

Supplementary Table S2. Current immunosuppressive and immunomodulatory treatments at the time of assessment.

MTX, N (%)	51 (22.4)	35 (22.3)	16 (22.5)	0.968
MMF, N (%)	41 (18)	28 (17.8)	13 (18.3)	0.931
AZA, N (%)	25 (11)	21 (13.4)	4 (5.6)	0.083
IVIG, N (%)	7 (3.1)	4 (2.5)	3 (4.2)	0.680
RTX, N (%)	14 (6.1)	11 (7)	3 (4.2)	0.558
CYS A, N (%)	35 (15.4)	17 (10.8)	18 (25.4)	0.005
GC daily, median (IQR)	1 (5)	1 (5)	5 (12.5)	0.060
Tacrolimus, N (%)	1 (0.4)	1 (0.6)	0 (0)	1.000
Jaki, N (%)	5 (2.2)	5 (3.2)	0 (0)	0.328
HCQ, N (%)	44 (19.3)	32 (20.4)	12 (16.9)	0.537
CYC, N (%)	1 (0.4)	1 (0.6)	0 (0)	1.000

MTX: methotrexate; MMF: mycophenolate mofetil; AZA: azathioprine; IVIG: intravenous immunoglobulin; RTX: rituximab; CYS A: cyclosporine A; Jaki: Janus kinase inhibitor; HCQ: hydroxychloroquine; CYC: cyclophosphamide; MITAX: myositis intention to treat activity index; MYOACT: Myositis Disease Activity Assessment Tool; MMT: Manual Muscle Testing; PhGA: physician's global assessment.

Supplementary Table S3. Full univariable linear regression analyses for MYOACT.

Variables	Univariable analyses			Multivariable analyses		
	Coeff. β	95% CI	<i>p</i> -value	Coeff. β	95% CI	<i>p</i> -value
Gender (male)	0.046	0.013 to 0.078	0.006	0.037	0.006 to 0.056	0.014
Age at diagnosis	0.002	0.001 to 0.003	0.000	0.001	0.000 to 0.002	0.060
Disease duration	0.015	-0.002 to 0.002	0.821	0.001	-0.001 to 0.003	0.225
<i>MAA</i>						
ANA	0.016	-0.018 to 0.049	0.358			
ENA	0.003	-0.028 to 0.034	0.843			
<i>MAA</i>						
SSA/Ro	-0.007	-0.041 to 0.027	0.680			
SSB/La	0.045	-0.071 to 0.160	0.448			
Ro52	0.007	-0.028 to 0.042	0.699			
Ro60	0.052	-0.042 to 0.146	0.278			
ACA	-0.064	-0.227 to 0.098	0.436			
ATA	-	-	-			
SMD	-0.098	-0.327 to 0.131	0.399			
Ku	-0.093	-0.187 to 0.001	0.053			
Pm/ScI	-0.014	-0.092 to 0.064	0.728			
RNP	-0.109	-0.338 to 0.120	0.349			
ANCA	0.016	-0.117 to 0.149	0.236			
<i>MSA</i>						
Mi2 α	0.006	-0.053 to 0.066	0.840			
Mi2 β	-0.023	-0.089 to 0.042	0.486			
TIF1g	-0.005	-0.076 to 0.066	0.882			
MDA5	0.068	0.011 to 0.125	0.019	0.091	0.044 to 0.138	0.0001
NXP2	-0.003	-0.085 to 0.080	0.951			
SAE	0.003	-0.092 to 0.098	0.950			
PL7	0.075	-0.002 to 0.153	0.056			
PL12	-0.003	-0.081 to 0.075	0.933			
Jo1	0.009	-0.022 to 0.041	0.569			
EJ	-0.006	-0.122 to 0.109	0.914			
OJ	-0.044	-0.207 to 0.119	0.594			
eN1A	-0.084	-0.247 to 0.078	0.306			
SRP	0.001	-0.114 to 0.117	0.982			
HMGCR	0.252	0.025 to 0.480	0.030			
KS	-0.108	-0.337 to 0.121	0.353			
Zo	-0.038	-0.267 to 0.191	0.745			
<i>IIM subgroup</i>						
DM	-0.016	-0.049 to 0.017	0.352	-0.063	-0.117 to -0.009	0.022
PM	-0.030	-0.075 to 0.015	0.195	-0.079	-0.138 to -0.020	0.009
ASyS	0.017	-0.014 to 0.048	0.284	-0.032	-0.084 to 0.021	0.233
IMNM	0.023	-0.004 to 0.050	0.096			
OL	0.059	-1.000 to 0.131	0.796			

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Variables	Univariable analyses			Multivariable analyses		
	Coeff. β	95% CI	p-value	Coeff. β	95% CI	p-value
<i>Muscular involvement</i>						
Abnormal muscular MRI	0.007	-0.014 to 0.028	0.507			
Endomysial infiltrate	-0.019	-0.039 to 0.002	0.078			
Perimysial infiltrate	-0.005	-0.027 to 0.017	0.672			
Atrophy	-0.003	-0.023 to 0.017	0.766			
Vacuoles	-0.005	-0.024 to 0.014	0.588			
Myositis, N (%)	0.032	-0.009 to 0.073	0.130			
Elevated CPK, N (%)	0.041	0.010 to 0.071	0.010	0.023	-0.001 to 0.048	0.062
Elevated aldolase, N (%)	0.27	-0.006 to 0.061	0.111			
Dysphagia	0.014	-0.023 to 0.051	0.454			
<i>Lung involvement</i>						
ILD (HRCT)	0.027	-0.004-0.58	0.089			
NSIP	0.026	-0.013 to 0.065	0.190			
UIP	-0.008	-0.060 to 0.044	0.763			
OP	0.033	-0.062 to 0.070	0.910			
LID	-0.106	-0.332 to 0.121	0.358			
NSIP/OP	-0.004	-0.075 to 0.067	0.905			
ICU admission	0.125	0.010 to 0.239	0.033	0.059	-0.028 to 0.14	0.182
Emphysema	-0.14	-0.088 to 0.060	0.704			
Pneumomediastinum	-	-	-			
Diaphragm involvement	-	-	-			
<i>Articular involvement</i>						
Arthritis	0.003	-0.027 to 0.034	0.219			
Joint deformity	-0.008	-0.079 to 0.063	0.824			
RF	-0.011	-0.58 to 0.035	0.629			
ACPA	-0.007	-0.078 to 0.065	0.856			
<i>Cutaneous involvement</i>						
Mechanic's hands	0.019	-0.017 to 0.054	0.301			
Gottron's papules	0.014	-0.021 to 0.050	0.428			
Gottron's sign	-0.009	-0.064 to 0.046	0.749			
Shawl sign	0.022	-0.041 to 0.085	0.501			
V sign	0.014	-0.028 to 0.056	0.502			
Holster sign	0.019	-0.059 to 0.096	0.636			
Heliotrope rash	0.008	-0.032 to 0.047	0.702			
Cutaneous ulcers	0.018	-0.043 to 0.079	0.556			
Calcinosis cutis	-0.088	-0.250 to 0.074	0.283			
Periungual erythema	0.047	-0.018 to 0.113	0.152			
RP	-0.021	-0.060 to -0.017	0.274			
<i>Other</i>						
Malignancy	0.016	-0.072 to 0.104	0.724			
Fever	0.047	0.002 to 0.092	0.041	0.016	-0.019 to 0.051	0.367
<i>Damage assessment</i>						
MDI extent, median (IQR)	0.870	0.717 to 1.022	0.0001	0.834	0.684 to 0.981	0.0001
MDI severity, median (IQR)	0.020	-0.016 to 0.056	0.284			

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