

Supplementary File

Supplementary Table S1. Demographic and clinical characteristics of IIM patients and HCs.

	IIM (n=44)	HC (n=11)
Age, years (mean $\pm$ SEM)	49.3 $\pm$ 1.8	45.8 $\pm$ 3.6
Female/male	31/13	8/3
Diagnosis DM/PM	33/11	/
Time since diagnosis, years	0.9 $\pm$ 0.2	/
CK, U/L	1305 $\pm$ 442.8	/
Leukocyte count, 10 ⁹ /L	7.8 $\pm$ 0.6	/
Lymphocyte count, 10 ⁹ /L	1.2 $\pm$ 0.1	/
Medication (%)		/
Prednisone or methylprednisolone, n (%)	34 (77)	/
Other immunosuppressive drugs, n (%)	31 (70)	/

All data are expressed as mean $\pm$ SEM.

IIM: idiopathic inflammatory myopathy; HC: healthy control; CK: creatine kinase.

Supplementary Table S2. Demographic and clinical characteristics of patients with or without anti-MDA5.

Demographic and clinical characteristics	Anti-MDA5 (+) (n=16)	Anti-MDA5 (-) (n=28)	<i>p</i> -value
Age, years (mean $\pm$ SEM)	48.4 $\pm$ 2	49.8 $\pm$ 2.6	0.66
Female/male, n	14/2 (88)	17/11 (61)	0.13
Diagnosis DM/PM, n	15/1 (94)	18/10 (64)	0.07
Time since diagnosis, years	0.5 $\pm$ 0.1	1.2 $\pm$ 0.4	0.1
Systemic features			
ILD, n (%)	16 (100)	17 (61)	<0.01
RP-ILD, n (%)	3 (19)	0 (0)	<0.05
Fever, n (%)	5 (31)	8 (29)	0.85
Arthritis/arthralgia, n (%)	11 (69)	2 (7)	<0.001
Dyspnea at presentation, n (%)	2 (13)	1 (4)	0.69
Dysphagia, n (%)	0 (0)	6 (21)	<0.05
Myagia, n (%)	3 (19)	8 (29)	0.93
Myasthenia, n (%)	7 (44)	18 (64)	0.35
ADM, n (%)	9 (56)	3 (11)	<0.01
Cardiomyopathy, n (%)	0 (0)	1 (4)	1
Mucocutaneous features			
Calcinosis, n (%)	0 (0)	0 (0)	1
Cutaneous ulceration, n (%)	8 (50)	1 (4)	<0.01
Heliotrope rash, n (%)	13 (81)	12 (43)	<0.05
Gottron papules/sign, n (%)	14 (88)	8 (29)	<0.001
Ro52 positivity, n (%)	14 (88)	15 (54)	0.05

IIM: idiopathic inflammatory myopathy; DM: dermatomyositis; PM: polymyositis; ILD: interstitial lung disease; RP-ILD: rapid progressive interstitial lung disease; ADM: amyopathic dermatomyositis.

Supplementary Table S3. Prevalence of myositis-specific antibodies in 44 IIM patients.

Myositis-specific antibodies	n	%
anti-MDA5	16	36
anti-PL-7	2	5
anti-SRP	4	9
anti-PL-12	2	5
anti-TIF1- γ	2	5
anti-Jo-1	2	5
anti-Mi-2 β	4	9
anti-PM Scl75	3	7
anti-Ku	2	5
anti-OJ	2	5
anti-EJ	3	7
anti-NXP2	3	7
negative MSA	8	18

IIM: idiopathic inflammatory myopathy; MDA5: melanoma differentiation-associated gene 5; PL-7: threonyl-tRNA synthetase; SRP: signal recognition particle; PL-12: alanyl-tRNA synthetase; TIF1- γ : transcription intermediary factor 1 γ ; Jo-1: histidyl-tRNA synthetase; Mi-2: complex nucleosome remodeling histone deacetylase; PM Scl: polymyositis/systemic sclerosis; OJ: isoleucyl-tRNA synthetase; EJ: glycyl tRNA synthetase; NXP2: nuclear matrix protein 2; MSA: myositis specific antibody.

Supplementary Table S4. Laboratory characteristics of anti-MDA5-positive and anti-MDA5-negative IIM patients.

Laboratory Parameters	Anti-MDA5 (+) (n=16)	Anti-MDA5 (-) (n=28)	<i>p</i> -value
CK, U/L	75.4 $\pm$ 26	2008 $\pm$ 663.4	<0.01
AST, U/L	83.4 $\pm$ 17.8	95.2 $\pm$ 19.7	0.66
ALT, U/L	75.5 $\pm$ 17.8	79.1 $\pm$ 14.2	0.87
LDH, U/L	311.6 $\pm$ 18.3	524.2 $\pm$ 76.3	<0.05
Cr, μ mol/L	50.4 $\pm$ 2.5	52.4 $\pm$ 2.9	0.63
ESR, mm/h	43 $\pm$ 5.7	39.8 $\pm$ 4.7	0.67
CRP, mg/L	10.4 $\pm$ 2.6	18.8 $\pm$ 4	0.09
ANA, U/ml	91.1 $\pm$ 31.4	110.6 $\pm$ 25.7	0.64
Ferritin, μ g/L	676 $\pm$ 135.9	473.5 $\pm$ 82	0.18
IgG, g/L	13.2 $\pm$ 0.9	13.6 $\pm$ 0.9	0.77
IgM, g/L	1.2 $\pm$ 0.2	1.4 $\pm$ 0.1	0.48
IgA, g/L	2.7 $\pm$ 0.2	2.4 $\pm$ 0.1	0.23
Leukocyte count, 10 ⁹ /L	5.4 $\pm$ 0.6	8 $\pm$ 0.5	<0.01
Lymphocyte count, 10 ⁹ /L	0.6 $\pm$ 0.1	1.5 $\pm$ 0.2	<0.001

All data are expressed as mean $\pm$ SEM.

IIM: idiopathic inflammatory myopathy; CK: creatine kinase; AST: aspartate aminotransferase; ALT: alanine transaminase; LDH: lactate dehydrogenase; Cr: creatinine; ESR: erythrocyte sedimentation rate; CRP: C-reactive protein; ANA: anti-nuclear antibody; Fer: ferritin.