

**Supplemental Table S1.** Comparison of clinical features between IIM-ILD patients with and without pleural effusion.

Characteristics	Total (N=125)	Without pleural effusion (N=99)	With pleural effusion (N=26)	p-value
<i>Demographics</i>				
Follow-up time, (median [IQR])	24.00 [12.00, 48.00]	25.00 [17.00, 50.00]	15.50 [4.25, 40.00]	<b>0.031</b>
Age, years, mean (S.D.)	48.90 (12.54)	47.98 (12.51)	52.42 (12.23)	0.108
Female gender, n (%)	81 (64.8)	64 (64.6)	17 (65.4)	1.000
Smoking, n (%)	21 (16.8)	17 (17.2)	4 (15.4)	1.000
<i>Diagnosis</i>				
DM, n (%)	41 (32.8)	32 (32.3)	9 (34.6)	0.825
PM, n (%)	5 (4.0)	3 (3.0)	2 (7.7)	0.278
CADM, n (%)	44 (35.2)	34 (34.3)	10 (38.5)	1.000
ASS, n (%)	23 (18.4)	18 (18.2)	5 (19.2)	0.696
IMNM, n (%)	12 (9.6)	12 (12.1)	0 (0.0)	0.135
<i>Clinical manifestations</i>				
Fever at presentation, n (%)	33 (26.4)	20 (20.2)	13 (50.0)	<b>0.005</b>
Rash, n (%)	88 (70.4)	70 (70.7)	18 (69.2)	1.000
Heliotrope rash, n (%)	54 (43.2)	42 (42.4)	12 (46.2)	0.825
Gotttron papule/sign, n (%)	64 (51.2)	55 (55.6)	9 (34.6)	0.078
V-neck sign, n (%)	41 (32.8)	32 (32.3)	9 (34.6)	0.818
Periungual erythema, n (%)	20 (16.0)	18 (18.2)	2 (7.7)	0.243
Skin ulcers, n (%)	32 (25.6)	29 (29.3)	3 (11.5)	0.079
Mechanic's hands, n (%)	43 (34.4)	36 (36.4)	7 (26.9)	0.488
Raynaud phenomenon, n (%)	22 (17.6)	17 (17.2)	5 (19.2)	0.778
Dysphagia, n (%)	25 (20.0)	17 (17.2)	8 (30.8)	0.166
Hoarseness, n (%)	15 (12.0)	12 (12.1)	3 (11.5)	1.000
Peripheral oedema, n (%)	26 (20.8)	19 (19.2)	7 (26.9)	0.420
Articular symptom, n (%)	74 (59.2)	62 (62.6)	12 (46.2)	0.178
Cardiovascular involvement, n (%)	13 (10.4)	8 (8.1)	5 (19.2)	0.142
Pulmonary hypertension, n (%)	19 (15.2)	13 (13.1)	6 (23.1)	0.226
Muscle involvement, n (%)	88 (70.4)	70 (70.7)	18 (69.2)	1.000
Pericardial effusion, n (%)	21 (16.8)	10 (10.1)	11 (42.3)	<b>&lt;0.001</b>
<i>ILD domain</i>				
HRCT score, (median [IQR])	112.99 [104.34, 136.97]	108.87 [104.06, 126.41]	145.57 [125.22, 185.17]	<b>&lt;0.001</b>
RP-ILD, n (%)	18 (14.4)	10 (10.1)	8 (30.8)	<b>0.013</b>
Lower lung zone consolidation, n (%)	42 (33.6)	29 (29.3)	13 (50.0)	<b>0.047</b>
HRCT pattern, n (%)		0.400		
NSIP	36 (28.8)	31 (31.3)	5 (19.2)	0.226
OP	48 (38.4)	38 (38.4)	10 (38.5)	0.994
NSIP combine OP	41 (32.8)	30 (30.3)	11 (42.3)	0.246
<i>Laboratory features</i>				
WBC, mean (S.D.)	6.96 (3.31)	6.58 (3.15)	8.38 (3.55)	<b>0.013</b>
HB, mean (S.D.)	123.89 (16.66)	125.57 (14.22)	117.50 (23.04)	<b>0.027</b>
LY%, mean (S.D.)	20.39 (11.09)	21.53 (11.14)	16.04 (9.96)	<b>0.024</b>
NLR, (median [IQR])	3.95 [2.53, 5.65]	3.55 [2.46, 5.09]	4.92 [3.78, 8.97]	<b>0.008</b>
ESR, mm/h, (median [IQR])	26.00 [15.00, 41.00]	24.00 [13.00, 35.44]	39.09 [19.00, 68.10]	<b>0.025</b>
CRP, mg/L, (median [IQR])	4.97 [2.14, 15.87]	4.73 [1.92, 8.52]	21.67 [11.20, 41.18]	<b>&lt;0.001</b>
CK level, U/L, (median [IQR])	183.00 [67.00, 2143.00]	208.00 [64.50, 2263.22]	136.00 [89.25, 1873.50]	0.773
LDH, U/L, (median [IQR])	369.60 [255.00, 569.00]	350.00 [241.00, 555.50]	426.00 [284.00, 676.65]	0.243
ALT, U/L, (median [IQR])	47.00 [19.00, 103.00]	48.00 [19.00, 100.74]	46.50 [22.25, 102.25]	0.745
AST, U/L, (median [IQR])	57.00 [23.00, 128.00]	52.00 [23.00, 117.50]	63.50 [27.00, 161.25]	0.245
Albumin, g/L, mean (S.D.)	34.78 (5.16)	35.81 (4.71)	30.87 (4.98)	<b>&lt;0.001</b>
ANA (≥1:80), n (%)	75 (60.0)	57 (57.6)	18 (69.2)	0.369
MSAs positive, n (%)	118 (94.4)	96 (97.0)	22 (84.6)	0.034
<i>Subtypes of MSAs, n (%)</i>				
Anti-MDA5	49 (39.2)	41 (41.4)	8 (30.8)	0.373
Anti-Mi2	15 (12.0)	11 (11.1)	4 (15.4)	0.513
Anti-TIF-γ	6 (4.8)	4 (4.0)	2 (7.7)	0.604
Anti-NXP2	10 (8.0)	9 (9.1)	1 (3.8)	0.686
Anti-Jo1	24 (19.2)	18 (18.2)	6 (23.1)	0.582
Anti-PL7	13 (10.4)	11 (11.1)	2 (7.7)	1.000
Anti-PL12	6 (4.8)	5 (5.1)	1 (3.8)	1.000
Anti-EJ	9 (7.2)	8 (8.1)	1 (3.8)	0.684
Anti-OJ	6 (4.8)	5 (5.1)	1 (3.8)	1.000
Anti-SAE	2 (1.6)	2 (2.0)	0 (0.0)	1.000
Anti-SRP	13 (10.4)	12 (12.1)	1 (3.8)	0.299
Anti-HMGCR	8 (6.4)	8 (8.1)	0 (0.0)	0.203
<i>Therapies</i>				
Exposure to high-dose glucocorticoid (>80 mg), n (%)	53 (42.4)	39 (39.4)	14 (53.8)	0.265
<i>No. of immunosuppressants, on top of steroid, n (%)</i>				
0	23 (18.4)	14 (14.1)	9 (34.6)	<b>0.018</b>
1	65 (52.0)	51 (51.5)	14 (53.8)	0.832
>=2	37 (29.6)	34 (34.3)	3 (11.5)	<b>0.023</b>
IVIg, n (%)	34 (27.2)	21 (21.2)	13 (50.0)	<b>0.006</b>
Exposure to pirfenidone, n (%)	24 (19.2)	18 (18.2)	6 (23.1)	0.582

Bold indicates statistical significance.

DM: dermatomyositis; IMNM: immune-mediated necrotising myopathy; PM: polymyositis; CADM: clinically amyopathic dermatomyositis; ASS: antisynthetase syndrome; MSAs: myositis-specific autoantibodies; MDA5: melanoma differentiation-associated gene 5; TIF1-γ: transcriptional intermediary factor 1 gamma; SAE: small ubiquitin-like modifier activating enzyme; NXP2: nuclear matrix protein 2; Jo-1: histidyl-tRNA-synthetase; PL-12: alanyl-tRNA synthetase; PL-7: threonyl-tRNA synthetase; EJ: glycyl-tRNA synthetase; OJ: isoleucyl-tRNA synthetase; HMGCR: 3-hydroxy-3-methylglutaryl-coenzyme A reductase; SRP: signal recognition particle; ILD: interstitial lung disease; RP-ILD: rapidly progressive interstitial lung disease; HRCT: high-resolution computed tomography. NSIP: nonspecific interstitial pneumonia; OP: organising pneumonia; WBC: white blood cell count; HB: haemoglobin; LY%: percentage of lymphocyte; NLR: neutrophil/lymphocyte ratio; CK: creatine kinase; ALT: alanine aminotransferase; AST: aspartate aminotransferase; LDH: lactate dehydrogenase; CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; ANA: anti-nuclear antibody; IVIG: intravenous immunoglobulin.

Supplemental Table S2. Clinical features of IIM-ILD patients with RP-ILD and predictors of RP-ILD.

Characteristics	Total (N=125)	Without RP-ILD (N=107)	With RP-ILD (N=18)	p-value	Univariate Cox regression analysis HR (95% CI)	p-value
<i>Demographics</i>						
Follow-up time, (median [IQR])	24.00 [12.00, 48.00]	27.00 [17.00, 51.00]	3.00 [1.00, 8.65]	<b>&lt;0.001</b>		
Age, years, mean (S.D.)	48.90 (12.54)	48.46 (12.77)	51.56 (10.99)	0.334		
Female gender, n (%)	81 (64.8)	71 (66.4)	10 (55.6)	0.428		
Smoking, n (%)	21 (16.8)	17 (15.9)	4 (22.2)	0.503		
<i>Diagnosis</i>						
DM, n (%)	41 (32.8)	34 (31.8)	7 (38.9)	0.552		
PM, n (%)	5 (4.0)	5 (4.7)	0 (0.0)	1.000		
CADM, n (%)	44 (35.2)	41 (38.3)	3 (16.7)	<b>0.006</b>	4.376 (1.723-11.115)	<b>0.002</b>
ASS, n (%)	23 (18.4)	15 (14.0)	8 (44.4)	0.288		
IMNM, n (%)	12 (9.6)	12 (11.2)	0 (0.0)	0.075		
<i>Clinical manifestations</i>						
Fever at presentation, n (%)	33 (26.4)	23 (21.5)	10 (55.6)	<b>0.007</b>	4.083 (1.61-10.355)	<b>0.003</b>
Rash, n (%)	88 (70.4)	71 (66.4)	17 (94.4)	<b>0.023</b>	8.403 (1.116-63.269)	<b>0.039</b>
Heliotrope rash, n (%)	54 (43.2)	41 (38.3)	13 (72.2)	<b>0.010</b>	3.918 (1.392-11.027)	<b>0.010</b>
Gotttron papule/sign, n (%)	64 (51.2)	53 (49.5)	11 (61.1)	0.448		
V-neck sign, n (%)	41 (32.8)	31 (29.0)	10 (55.6)	<b>0.033</b>	2.878 (1.132-7.315)	<b>0.026</b>
Periungual erythema, n (%)	20 (16.0)	16 (15.0)	4 (22.2)	0.487		
Skin ulcers, n (%)	32 (25.6)	24 (22.4)	8 (44.4)	0.076		
Mechanic's hands, n (%)	43 (34.4)	41 (38.3)	2 (11.1)	<b>0.031</b>	0.223 (0.051-0.972)	<b>0.046</b>
Raynaud phenomenon, n (%)	22 (17.6)	21 (19.6)	1 (5.6)	0.194		
Dysphagia, n (%)	25 (20.0)	22 (20.6)	3 (16.7)	1.000		
Hoarseness, n (%)	15 (12.0)	11 (10.3)	4 (22.2)	0.230		
Peripheral oedema, n (%)	26 (20.8)	22 (20.6)	4 (22.2)	1.000		
Articular symptom, n (%)	74 (59.2)	64 (59.8)	10 (55.6)	0.798		
Cardiovascular involvement, n (%)	13 (10.4)	10 (9.3)	3 (16.7)	0.399		
Pulmonary hypertension, n (%)	19 (15.2)	15 (14.0)	4 (22.2)	0.475		
Muscle involvement, n (%)	88 (70.4)	80 (74.8)	8 (44.4)	<b>0.013</b>	0.297 (0.117-0.753)	<b>0.011</b>
Pleural effusion, n (%)	26 (20.8)	18 (16.8)	8 (44.4)	<b>0.013</b>	3.669 (1.446-9.309)	<b>0.006</b>
Pericardial effusion, n (%)	21 (16.8)	17 (15.9)	4 (22.2)	0.503		
<i>ILD domain</i>						
HRCT score (median [IQR])	112.99 [104.34, 136.97]	110.26 [104.00, 128.50]	145.52 [115.22, 173.75]	<b>&lt;0.001</b>	1.01 ( 1.002-1.018)	<b>0.015</b>
Lower lung zone consolidation, n (%)	42 (33.6)	32 (29.9)	10 (55.6)	<b>0.033</b>	2.846 (1.122-7.223)	<b>0.028</b>
HRCT patterns, n (%)				<b>0.019</b>		
NSIP	36 (28.8)	35 (32.7)	1 (5.6)	<b>0.019</b>	0.126 (0.017-0.951)	<b>0.045</b>
OP	48 (38.4)	41 (38.3)	7 (38.9)	0.963		
NSIP combine OP	41 (32.8)	31 (29.0)	10 (55.6)	<b>0.044</b>	2.606 (1.028-6.606)	<b>0.044</b>
<i>Laboratory features</i>						
WBC, mean (S.D.)	6.96 (3.31)	7.07 (3.29)	6.31 (3.41)	0.370		
HB, mean (S.D.)	123.89 (16.66)	125.13 (15.52)	116.50 (21.34)	<b>0.042</b>	0.973 (0.95-0.996)	<b>0.021</b>
LY%, mean (S.D.)	20.39 (11.09)	20.67 (11.62)	18.69 (7.17)	0.485		
NLR, (median [IQR])	3.95 [2.53, 5.65]	3.95 [2.52, 5.80]	4.27 [2.58, 5.25]	0.741		
CK level, U/L, (median [IQR])	183.00 [67.00, 2143.00]	208.00 [68.00, 2609.50]	102.00 [50.75, 526.00]	0.082		
ESR, mm/h, (median [IQR])	26.00 [15.00, 41.00]	24.00 [13.00, 40.00]	30.63 [22.00, 45.75]	0.079		
CRP, mg/L, (median [IQR])	4.97 [2.14, 15.87]	4.75 [2.00, 12.71]	15.66 [5.48, 46.79]	<b>0.001</b>	1.019 (1.008-1.031)	<b>0.001</b>
LDH, U/L, (median [IQR])	369.60 [255.00, 569.00]	366.00 [241.50, 566.50]	387.50 [304.50, 559.00]	0.473		
ALT, U/L, (median [IQR])	47.00 [19.00, 103.00]	48.00 [19.00, 105.50]	46.50 [32.50, 77.25]	0.803		
AST, U/L, (median [IQR])	57.00 [23.00, 128.00]	52.00 [23.00, 121.00]	76.00 [43.75, 161.25]	0.249		
Albumin, g/L, mean (S.D.)	34.78 (5.16)	35.53 (4.89)	30.35 (4.56)	<b>&lt;0.001</b>	0.839 (0.772-0.912)	<b>&lt;0.001</b>
ANA (≥1:80), n (%)	75 (60.0)	68 (63.6)	7 (38.9)	0.068		
MSAs positive, n (%)	118 (94.4)	100 (93.5)	18 (100.0)	0.592		
<i>Subtypes of MSAs, n (%)</i>						
Anti-MDA5	49 (39.2)	34 (31.8)	15 (83.3)	<b>&lt;0.001</b>	9.425 (2.722-32.638)	<b>&lt;0.001</b>
Anti-Mi2	15 (12.0)	15 (14.0)	0 (0.0)	0.125		
Anti-TIF-γ	6 (4.8)	5 (4.7)	1 (5.6)	1.000		
Anti-NXP2	10 (8.0)	10 (9.3)	0 (0.0)	0.355		
Anti-Jo1	24 (19.2)	23 (21.5)	1 (5.6)	0.193		
Anti-PL7	13 (10.4)	12 (11.2)	1 (5.6)	0.690		
Anti-PL12	6 (4.8)	6 (5.6)	0 (0.0)	0.592		
Anti-EJ	9 (7.2)	8 (7.5)	1 (5.6)	1.000		
Anti-OJ	6 (4.8)	6 (5.6)	0 (0.0)	0.592		
Anti-SAE	2 (1.6)	2 (1.9)	0 (0.0)	1.000		
Anti-SRP	13 (10.4)	13 (12.1)	0 (0.0)	0.212		
Anti-HMGCR	8 (6.4)	8 (7.5)	0 (0.0)	0.601		
<i>Therapies</i>						
Exposure to high-dose glucocorticoid (>80 mg), n (%)	53 (42.4)	41 (38.3)	12 (66.7)	<b>0.037</b>	2.911 (1.092-7.759)	<b>0.033</b>
<i>No. of immunosuppressants, on top of steroid, n (%)</i>						
0	23 (18.4)	18 (16.8)	5 (27.8)	0.516		
1	65 (52.0)	56 (52.3)	9 (50.0)			
≥2	37 (29.6)	33 (30.8)	4 (22.2)			
IVIg, n (%)	34 (27.2)	27 (25.2)	7 (38.9)	0.257		
Exposure to pirfenidone, n (%)	24 (19.2)	22 (20.6)	2 (11.1)	0.521		

Bold indicates statistical significance.

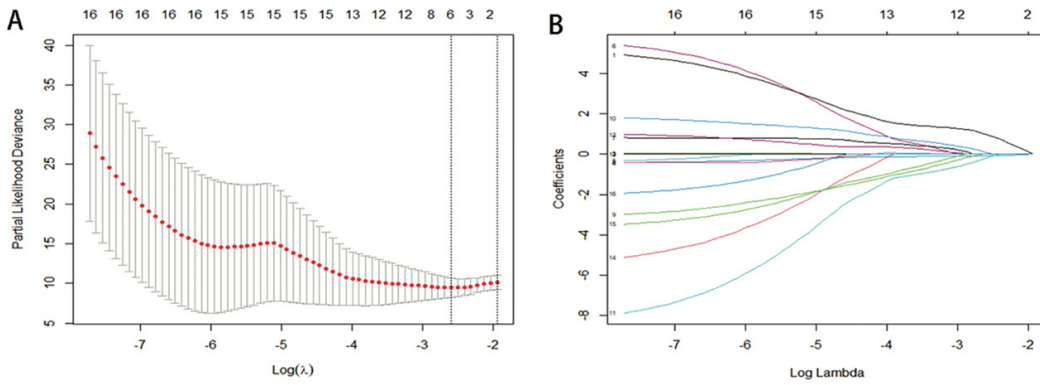
DM: dermatomyositis; IMNM: immune-mediated necrotising myopathy; PM: polymyositis; CADM: clinically amyopathic dermatomyositis; ASS: antisynthetase syndrome; MSAs: myositis-specific autoantibodies; MDA5: melanoma differentiation-associated gene 5; TIF1-γ: transcriptional intermediary factor 1 gamma; SAE: small ubiquitin-like modifier activating enzyme; NXP2: nuclear matrix protein 2; Jo-1: histidyl-tRNA-synthetase; PL-12: alanyl-tRNA synthetase; PL-7: threonyl-tRNA synthetase; EJ: glycyl-tRNA synthetase; OJ: isoleucyl-tRNA synthetase; HMGCR:3-hydroxy-3-methylglutaryl-coenzyme A reductase; SRP: signal recognition particle; ILD: interstitial lung disease; RP-ILD: rapidly progressive interstitial lung disease; HRCT: high-resolution computed tomography. NSIP: nonspecific interstitial pneumonia; OP: organising pneumonia; WBC: white blood cell count; HB: haemoglobin; LY%: percentage of lymphocyte; NLR: neutrophil/lymphocyte ratio; CK: creatine kinase; ALT: alanine aminotransferase; AST: aspartate aminotransferase; LDH: lactate dehydrogenase; CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; ANA: anti-nuclear antibody; IVIG: intravenous immunoglobulin.

**Supplemental Table S3.** Clinical features of IIM patients with non-survivors and predictors of mortality.

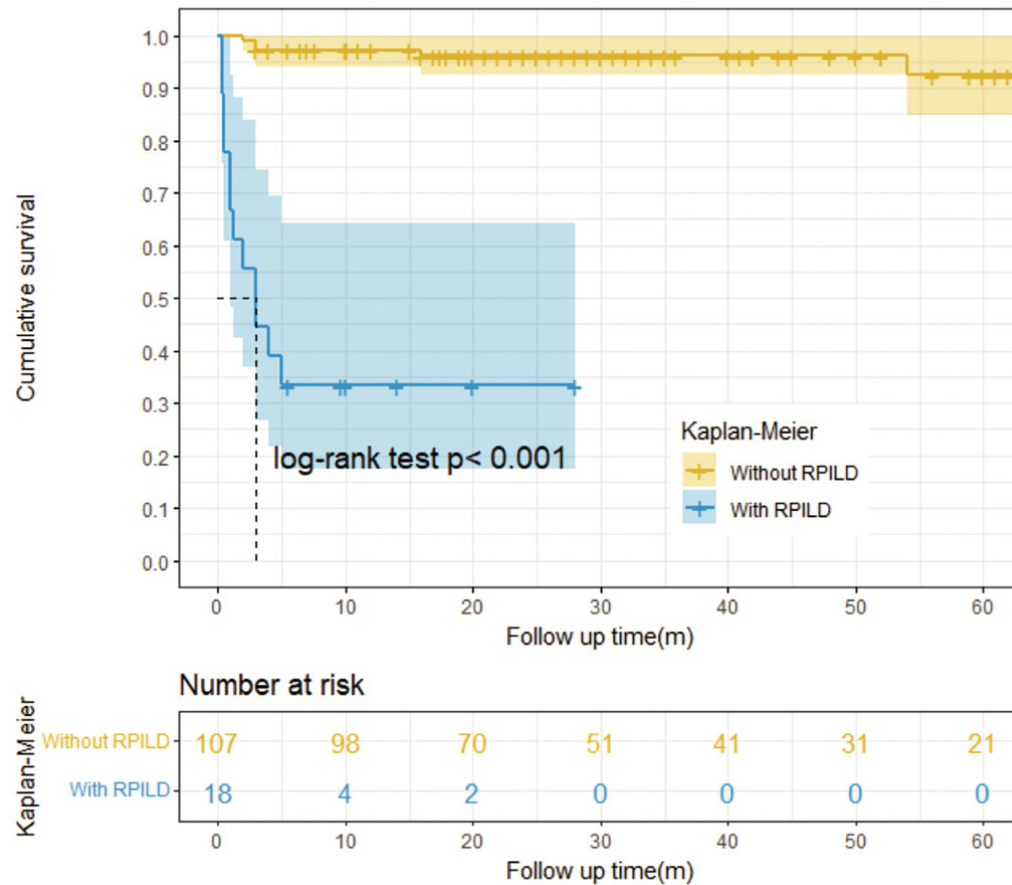
Characteristics	Total (N=158)	Survivors (N=140)	Non-survivors (N=18)	p-value	Univariate Cox regression analysis HR (95% CI)	p-value
<i>Demographics</i>						
Follow-up time, (median [IQR])	24.50 [14.25, 48.00]	26.00 [17.00, 48.00]	2.50 [1.00, 3.75]	<0.001		
Age, years, mean (S.D.)	47.04 (13.00)	46.48 (13.09)	51.44 (11.71)	0.128		
Female gender, n (%)	97 (61.4)	86 (61.4)	11 (61.1)	1.000		
Smoking, n (%)	28 (17.7)	24 (17.1)	4 (22.2)	0.529		
<i>Diagnosis</i>						
DM, n (%)	49 (31.0)	42 (30.0)	7 (38.9)	0.443		
PM, n (%)	7 (4.4)	6 (4.3)	1 (5.6)	0.579		
CADM, n (%)	26 (16.5)	20 (14.3)	6 (33.3)	0.087		
ASS, n (%)	48 (30.4)	44 (31.4)	4 (22.2)	0.424		
IMNM, n (%)	28 (17.7)	28 (20.0)	0 (0.0)	0.078		
<i>Clinical manifestations</i>						
Fever at presentation, n (%)	37 (23.4)	28 (20.0)	9 (50.0)	<b>0.014</b>	4.053 (1.594-10.306)	<b>0.003</b>
Rash, n (%)	103 (65.2)	86 (61.4)	17 (94.4)	<b>0.007</b>	11.151 (1.475-84.331)	<b>0.020</b>
Heliotrope rash, n (%)	66 (41.8)	55 (39.3)	11 (61.1)	0.126		
Gottron papule/sign, n (%)	73 (46.2)	61 (43.6)	12 (66.7)	0.080		
V-neck sign, n (%)	50 (31.6)	42 (30.0)	8 (44.4)	0.281		
Periungual erythema, n (%)	23 (14.6)	20 (14.3)	3 (16.7)	0.729		
Skin ulcers, n (%)	38 (24.1)	29 (20.7)	9 (50.0)	<b>0.015</b>	4.235 (1.617-11.091)	<b>0.003</b>
Mechanic's hands, n (%)	49 (31.0)	46 (32.9)	3 (16.7)	0.188		
Raynaud phenomenon, n (%)	26 (16.5)	23 (16.4)	3 (16.7)	1.000		
Dysphagia, n (%)	35 (22.2)	31 (22.1)	4 (22.2)	1.000		
Hoarseness, n (%)	18 (11.4)	16 (11.4)	2 (11.1)	1.000		
Peripheral oedema, n (%)	29 (18.4)	24 (17.1)	5 (27.8)	0.329		
Articular symptom, n (%)	86 (54.4)	78 (55.7)	8 (44.4)	0.453		
Cardiovascular involvement, n (%)	17 (10.8)	12 (8.6)	5 (27.8)	<b>0.028</b>	3.412 (1.215-9.578)	<b>0.020</b>
Pulmonary hypertension, n (%)	19 (15.2)	16 (15.0)	3 (16.7)	0.737		
Muscle involvement, n (%)	115 (72.8)	105 (75.0)	10 (55.6)	0.095		
Pleural effusion, n (%)	28 (17.7)	18 (12.9)	10 (55.6)	<0.001	6.697 (2.638-16.997)	<0.001
Pericardial effusion, n (%)	22 (13.9)	17 (12.1)	5 (27.8)	0.138		
<i>ILD domain</i>						
ILD, n (%)	125 (79.1)	107 (76.4)	18 (100.0)	<b>0.014</b>	29.138 (0.212-4000.214)	0.179
HRCT score (median [IQR])	112.99 [104.34, 136.97]	110.26 [104.06, 133.20]	127.32 [112.31, 165.45]	<b>0.018</b>	1.01 (1.001-1.019)	<b>0.022</b>
RP-ILD, n (%)	18 (11.4)	6 (4.3)	12 (66.7)	<0.001	41.882 (13.122-133.681)	<0.001
Lower lung zone consolidation, n (%)	42 (26.6)	33 (23.6)	9 (50.0)	<b>0.024</b>	3.23 (1.277-8.171)	<b>0.013</b>
HRCT patterns, n (%)				0.381		
NSIP	36 (28.8)	33 (23.6)	3 (16.7)			
OP	48 (38.4)	41 (29.3)	7 (38.9)			
NSIP combine OP	41 (32.8)	33 (23.6)	8 (44.4)	0.106		
<i>Laboratory features</i>						
WBC, mean (S.D.)	7.06 (3.19)	7.10 (3.11)	6.71 (3.88)	0.625		
HB, mean (S.D.)	125.93 (17.28)	127.50 (15.99)	113.72 (22.12)	<b>0.001</b>	0.959 (0.938-0.981)	<0.001
LY%, mean (S.D.)	21.64 (11.47)	22.00 (11.36)	18.89 (12.27)	0.281		
NLR, (median [IQR])	3.63 [2.31, 5.57]	3.47 [2.27, 5.64]	4.66 [3.45, 5.25]	0.178		
CK level, U/L, (median [IQR])	253.50 [72.25, 2447.61]	253.50 [71.25, 2537.50]	210.50 [81.50, 1215.75]	0.730		
ESR, mm/h, (median [IQR])	24.00 [12.00, 40.75]	23.00 [11.00, 37.43]	37.50 [23.00, 65.25]	<b>0.010</b>	1.013 (1.001-1.025)	<b>0.038</b>
CRP, mg/L, (median [IQR])	4.75 [1.91, 13.24]	4.58 [1.43, 10.86]	15.66 [5.34, 35.08]	<b>0.001</b>	1.012 (1.001-1.022)	<b>0.031</b>
LDH, U/L, (median [IQR])	360.50 [243.50, 575.75]	351.00 [241.00, 571.00]	459.14 [319.75, 672.24]	0.176		
ALT, U/L, (median [IQR])	49.00 [20.00, 105.75]	49.00 [19.75, 105.25]	48.00 [28.75, 103.50]	0.553		
AST, U/L, (median [IQR])	56.00 [24.25, 129.50]	51.00 [23.00, 124.75]	97.50 [46.75, 187.75]	0.093		
Albumin, g/L, mean (S.D.)	35.44 (5.17)	36.18 (4.82)	29.68 (4.15)	<0.001	0.796 (0.73-0.867)	<0.001
ANA (≥1:80), n (%)	67 (42.4)	59 (42.1)	8 (44.4)	1.000		
MSAs positive, n (%)	147 (93.0)	130 (92.9)	17 (94.4)	1.000		
<i>Subtypes of MSAs, n (%)</i>						
Anti-MDA5	50 (31.6)	39 (27.9)	11 (61.1)	<b>0.007</b>	4.254 (1.637-11.057)	<b>0.003</b>
Anti-Mi2	17 (10.8)	14 (10.0)	3 (16.7)	0.415		
Anti-TIF-γ	8 (5.1)	8 (5.7)	0 (0.0)	0.598		
Anti-NXP2	13 (8.2)	13 (9.3)	0 (0.0)	0.365		
Anti-Jo-1	27 (17.1)	25 (17.9)	2 (11.1)	0.740		
Anti-PL7	15 (9.5)	14 (10.0)	1 (5.6)	1.000		
Anti-PL12	151 (95.6)	134 (95.7)	17 (94.4)	0.579		
Anti-EJ	149 (94.3)	131 (93.6)	18 (100.0)	0.599		
Anti-OJ	151 (95.6)	133 (95.0)	18 (100.0)	1.000		
Anti-SAE	156 (98.7)	138 (98.6)	18 (100.0)	1.000		
Anti-SRP	135 (85.4)	117 (83.6)	18 (100.0)	0.077		
Anti-HMGCR	143 (90.5)	125 (89.3)	18 (100.0)	0.221		
<i>Therapies</i>						
Exposure to high-dose glucocorticoid (>80 mg), n (%)	68 (43.0)	58 (41.4)	10 (55.6)	0.314		
<i>No. of immunosuppressants, on top of steroid, n (%)</i>						
0	32 (20.3)	25 (17.9)	7 (38.9)	0.075		
1	84 (53.2)	76 (54.3)	8 (44.4)			
>=2	42 (26.6)	39 (27.9)	3 (16.7)			
IVIg, n (%)	43 (27.2)	37 (26.4)	6 (33.3)	0.577		
Exposure to pirfenidone, n (%)	24 (15.2)	24 (17.1)	0 (0.0)	0.077		

Bold indicates statistical significance.

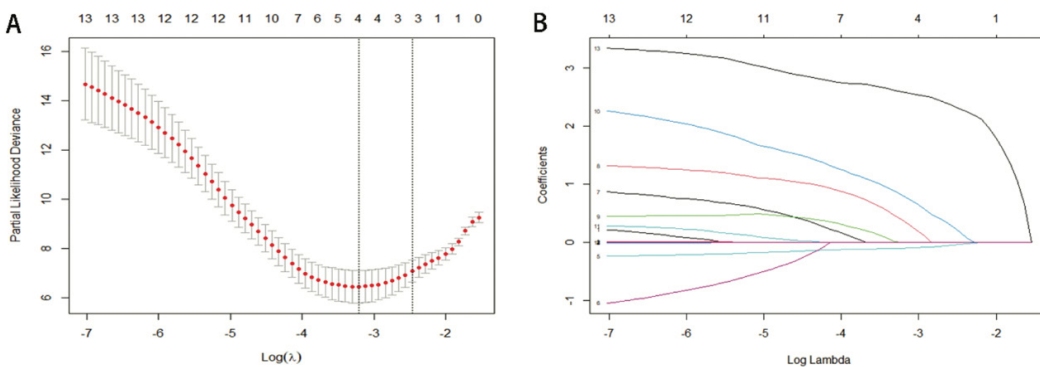
DM: dermatomyositis; IMNM: immune-mediated necrotising myopathy; PM: polymyositis; CADM: clinically amyopathic dermatomyositis; ASS: antisynthetase syndrome; MSAs: myositis-specific autoantibodies; MDA5: melanoma differentiation-associated gene 5; TIF1-γ: transcriptional intermediary factor 1 gamma; SAE: small ubiquitin-like modifier activating enzyme; NXP2: nuclear matrix protein 2; Jo-1: histidyl-tRNA-synthetase; PL-12: alanyl-tRNA synthetase; PL-7: threonyl-tRNA synthetase; EJ: glycyl-tRNA synthetase; OJ: isoleucyl-tRNA synthetase; HMGCR: 3-hydroxy-3-methylglutaryl-coenzyme A reductase; SRP: signal recognition particle; ILD: interstitial lung disease; RP-ILD: rapidly progressive interstitial lung disease; HRCT: high-resolution computed tomography. NSIP: nonspecific interstitial pneumonia; OP: organising pneumonia; WBC: white blood cell count; HB: haemoglobin; LY%: percentage of lymphocyte; NLR: neutrophil/lymphocyte ratio; CK: creatine kinase; ALT: alanine aminotransferase; AST: aspartate aminotransferase; LDH: lactate dehydrogenase; CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; ANA: anti-nuclear antibody; IVIG: intravenous immunoglobulin.



**Supplementary Fig. S1.** The most valuable predictors of RP-ILD in IIM-ILD patients were selected using LASSO Cox regression. **(A)** Selection of the tuning parameter ( $\lambda$ ) in the LASSO Cox regression analysis using 10-fold cross-validation via minimum criteria in IIM-ILD patients. **(B)** LASSO coefficient profiles of 16 candidate predictors. RP-ILD: rapidly progressive interstitial lung disease.



**Supplementary Fig. S2.** Kaplan-Meier survival curves for IIM patients with and without RP-ILD. The cumulative survival rate was significantly lower in IIM patients with RP-ILD than those without RP-ILD (33.3% vs. 94.4%;  $p < 0.001$ ). RP-ILD: rapidly progressive interstitial lung disease.



**Supplementary Fig. S3.** The most valuable predictors of mortality in IIM patients were selected using LASSO Cox regression. **(A)** Selection of the tuning parameter ( $\lambda$ ) in the LASSO Cox regression analysis using 10-fold cross-validation via minimum criteria in IIM patients. **(B)** LASSO coefficient profiles of 13 candidate predictors.