Supplemental Table 1. Characteristics of dead patients

	Dead patients without SPNM (n=7)	Dead patients with SPNM (n=8)	P-value
Diagnosis			
Dermatomyositis, n (%)	2 (28.6)	2 (25.0)	1.000
Polymyositis, n (%)	3 (42.9)	0 (0)	0.155
CADM, n (%)	2 (28.6)	6 (75.0)	0.201
RP-ILD, n (%)	2 (28.6)	7 (87.5)	0.073
Age, years, median (IQR)	71 (67, 72)	59 (56, 65)	0.023
Female, n (%)	3 (32.9)	3 (27.5)	2.000
Smoking habits, n (%)	4 (57.1)	4 (50.0)	1.000
Myositis specific autoantibodies status			
Anti-MDA5 antibody-positive, n (%)	1 (14.3)	8 (100)	0.004
Anti-ARS antibody-positive, n (%)	2 (28.6)	0 (0)	0.388
Anti-TIF1-γ antibody-positive, n (%)	0 (0)	0 (0)	None
Anti-Mi-2 antibody-positive, n (%)	2 (28.6)	0 (0)	0.388
Antibodies-negative (※1), n (%)	2 (28.6)	0 (0)	
Anti-SSA (Ro52) antibody-positive, n (%)	0 (0)	0 (0)	None
Treatments			
Maximum prednisolone dose, mg/day, median (IQR) ((※2)	50 (40, 55)	60 (56, 60)	0.096
Methylprednisolone pulse therapy, n (%)	5 (71.4)	7 (87.5)	0.897
Intravenous cyclophosphamide	3 (42.9)	7 (87.5)	0.200
Tacrolimus, n (%)	3 (42.9)	7 (87.5)	0.200
Cyclosporin, n (%)	0 (0)	3 (37.5)	0.244
Tofacitinib, n (%)	0 (0)	6 (75.0)	0.015

IVIg, n (%)	2 (28.6)	0 (0)	0.388
Plasma exchange, n (%)	1 (14.3)	6 (75.0)	0.067
Treatments, types			
GC monotherapy, n (%)	2 (28.6)	0 (0)	0.388
GC with one immunosuppressant, n (%)	2 (28.6)	0 (0)	0.388
GC with two immunosuppressants, n (%)	3 (42.9)	3 (37.5)	1.000
GC with three or more immunosuppressants, n (%)	0 (0)	5 (62.5)	0.044
Laboratory test result			
LDH	333 (283, 585)	392 (325, 449)	0.908
CK	141 (37, 221)	72 (54, 97)	0.643
KL-6	1183 (822, 1532)	983 (760, 1576)	0.817
CRP	0.40 (0.12, 1,12)	0.31 (0.17, 0.55)	0.727
ferritin	343.0 (243.3, 746.5)	902.7 (456.3, 1411.7)	0.121
Use of a ventilator, n (%)	3 (42.9)	6 (75)	0.460
Cause of death			
ILD worsening, n (%)	4 (57.1)	6 (75)	
Cancer, n (%)	3 (42.9)	1 (12.5)	
Tension pneumothorax, n (%)	0	1 (12.5)	

SPNM: spontaneous pneumomediastinum, CADM: clinically amyopathic dermatomyositis, RP-ILD: rapidly progressive interstitial lung disease, IVIg: intravenous immunoglobulin, LDH: lactate dehydrogenase, CK: creatine kinase, KL-6: Krebs von Lungren-6, CRP: C-reactive protein, ILD: interstitial lung disease

※1 "Antibodies-negative" means that anti-MDA5, ARS, TIF1-γ, and Mi-2 antibodies were negative. Data regarding other myositis-specific antibodies was not collected in this study, because only anti-MDA5, ARS, TIF1-γ, and Mi-2 antibodies were approved tests in a daily practice setting in Japan.

※2 Maximum prednisolone dose means the highest amount of prednisolone dose or equivalent after excluding glucocorticoids dose of methylprednisolone pulse therapy.

Supplemental Table 2. Hazard ratio of SPNM occurrence for death adjusted by age and rapidly progressive interstitial lung disease (RP-ILD)

	Hazard ratio (95% confidential interval)	P-value
SPNM occurrence	3.13 (1.03 - 9.52)	0.044
RP-ILD	10.94 (3.03 - 39.47)	< 0.001
Age	1.09 (1.02 - 1.16)	0.010

Supplemental Table 3. Characteristics of anti-MDA5 antibody-positive patients and anti-ARS antibody-positive patients

	Anti-ARS antibody positive (n = 53)	Anti-MDA5 antibody positive (n = 40)	P-value
Diagnosis			
Dermatomyositis, n (%)	23 (43.4)	7 (17.5)	0.015
Polymyositis, n (%)	10 (18.9)	0 (0)	0.010
CADM, n (%)	20 (37.7)	33 (82.5)	< 0.001
RP-ILD, n (%)	3 (5.7)	19 (47.5)	< 0.001
SPNM occurrence, n (%)	6 (11.3)	15 (37.5)	0.006
Age, years, median (IQR)	66 (53, 70)	58 (48, 65)	0.014
Female, n (%)	26 (49.1)	21 (52.5)	0.905
Smoking habits, n (%)	25 (47.2)	16 (40.0)	0.632
Treatments			
Maximum prednisolone dose, mg/day, mean ± SD	40 (40, 50)	50 (50, 60)	< 0.001
Methylprednisolone pulse therapy, n (%)	3 (5.7)	22 (55.0)	< 0.001
Intravenous cyclophosphamide, n (%)	18 (34.0)	37 (92.5)	< 0.001
Tacrolimus, n (%)	34 (64.2)	37 (92.5)	0.003
Cyclosporin, n (%)	6 (11.3)	7 (17.5)	0.583
Tofacitinib, n (%)	1 (1.9)	10 (25.0)	0.002

IVIg, n (%)	2 (3.8)	2 (5.0)	1.000
Plasma exchange, n (%)	0 (0)	10 (25.0)	< 0.001
Treatments, types			
GC monotherapy, n (%)	8 (15.1)	0 (0)	< 0.001
GC with one immunosuppressant, n (%)	32 (60.4)	2 (5.0)	< 0.001
GC with two immunosuppressants, n (%)	12 (22.6)	27 (67.5)	< 0.001
GC with three or more immunosuppressants, n (%)	1 (1.9)	11 (27.5)	0.001
Laboratory test result			
LDH, median (IQR)	314 (254, 467)	340 (259, 412)	0.978
CK, median (IQR)	218 (93, 1093)	92 (58, 198)	0.002
KL-6, median (IQR)	782 (560, 1429)	890 (558, 1348)	0.994
CRP, median (IQR)	0.43 (0.23, 2.00)	0.43 (0.37, 1.51)	0.989
ferritin, median (IQR)	259.35 (88.4, 764.3)	572.51 (318.8, 868.9)	0.009
Use of a ventilator, n (%)	2 (3.8)	7 (17.5)	0.063
Death within one year, n (%)	2 (3.8)	9 (22.5)	0.015

CADM: clinically amyopathic dermatomyositis, SPNM: spontaneous pneumomediastinum, RP-ILD: rapidly progressive interstitial lung disease, IVIg: intravenous immunoglobulin, GC: glucocorticoids, LDH: lactate dehydrogenase, CK: creatine kinase, KL-6: Krebs von Lungren-6, CRP: C-reactive protein, ILD: interstitial lung disease