Supplementary Table S1. The prevalence of ILD and RP-ILD in the Brazilian and Japanese patients with anti-MDA5(+) DM/ADM stratified by the time intervals between the disease diagnosis and symptoms onset.

	Brazil (n=34)	Japan (n=65)	*p*
ILD			
Time interval*, n (%)			
≤ 1 month	4/8 (50.0)	20/24 (83.3)	0.152
1 - 3 months	1/2 (50.0)	14/16 (87.5)	0.314
≥ 3 months	7/20 (35.0)	17/25 (68.0)	0.038
All	12/30 (40.0)	61/65 (93.9)	<0.001
RP-ILD			
Time interval*, n (%)			
≤ 1 month	1/8 (12.5)	13/24 (54.2)	0.053
1 - 3 months	0/2 (0)	11/16 (68.8)	<0.001
≥ 3 months	0/20 (0)	8/25 (32.0)	<0.001
All	1/30 (3.3)	32/65 (49.3)	<0.001

Data are presented as the frequency (%).
ADM: amyopathic dermatomyositis; DM: dermatomyositis; ILD: interstitial lung disease; RP-ILD: rapidly progressive interstitial lung disease.
* +Time interval between the onset of any symptoms related to DM/ADM or ILD and the diagnosis of the disease.

Supplementary Table S2. Clinical characteristics and outcomes of Brazilian and Japanese patients with anti-MDA5(+) dermatomyositis.

Variables	Brazil without RP-ILD (n=33)	Japan without RP-ILD (n=31)	Japan with RP-ILD (n=34)	*p* [a]	*p* [b]
Demographics					
Median age at diagnosis (years)	42 (35-49)	48 (39-66)	53 (46-64)	0.026	<0.001
Female, n (%)	20 (60.6)	22 (71.0)	22 (64.7)	0.438	0.803
Ethnicity, n (%)					
Asia	0	31 (100)	34 (100)	<0.001	<0.001
Africa	0	0	0		
Europe	0	0	0		
North-America	0	0	0		
South-America	33 (100)	0	0		
Oceania	0	0	0		
Clinical diagnosis, n (%)					
DM	19 (57.6)	6 (19.4)	6 (17.6)	0.001	0.001
ADM	14 (42.4)	25 (80.6)	28 (82.4)		
Dt: symptom onset and disease diagnosis (mo.)	4 (2-13)	3 (1-4)	2 (1-3)	0.027	0.005
Clinical manifestations, n (%)					
Fever	10 (30.3)	6 (19.4)	20 (58.8)	0.392	0.027
Weight loss	20 (60.6)	12 (38.7)	11 (32.4)	0.133	0.028
Gottron's papules	32 (97.0)	25 (80.6)	29 (85.3)	0.051	0.197
Gottron's sign	32 (97.0)	30 (96.8)	32 (94.1)	>0.999	>0.999
Heliotrope rash	30 (90.9)	10 (32.3)	9 (26.5)	<0.001	<0.001
Facial erythema	20 (60.6)	21 (67.7)	22 (64.7)	0.061	0.800
V-sign	15 (45.5)	7 (22.6)	8 (23.5)	0.069	0.750
Shawl sign	10 (30.3)	8 (25.8)	7 (20.6)	0.784	0.410
Scratch dermatitis	2 (6.1)	8 (25.8)	4 (11.8)	0.041	0.673
Raynaud's phenomenon	16 (48.5)	0	1 (2.9)	-	<0.001
Digital ulcers	8 (24.2)	1 (3.2)	1 (2.9)	0.027	0.013
Skin ulcers	7 (21.2)	3 (9.7)	6 (17.6)	0.305	0.765
Holster's sign	0	3 (9.7)	1 (2.9)	-	-
Calcinosis	9 (27.3)	0	0	-	-
Mechanic's hands	7 (21.2)	19 (61.3)	26 (76.5)	0.002	<0.001
Joint involvement					
Isolated arthralgia	10 (30.3)	4 (12.9)	16 (47.1)	0.132	0.212
Arthritis	14 (42.4)	14 (45.2)	5 (14.7)	0.999	0.015
Myalgia	17 (51.5)	11 (35.5)	7 (20.6)	0.218	0.011
Muscle weakness					
Upper limbs					
Grade V	8 (24.2)	21 (67.8)	23 (67.7)	0.001	0.001
Grade IV	23 (69.7)	9 (29.0)	10 (29.4)		
Grade III	2 (6.1)	1 (3.2)	0		
Grade II	0	0	1 (2.9)		
Grade I	0	0	0		

Variables	Brazil without RP-ILD (n=33)	Japan without RP-ILD (n=31)	Japan with RP-ILD (n=34)	p^a	p^b
Lower limbs					
Grade V	9 (27.3)	20 (64.6)	21 (61.8)	0.007	0.008
Grade IV	21 (63.6)	9 (29.0)	11 (32.4)		
Grade III	3 (9.1)	1 (3.2)	2 (5.8)		
Grade II	0	1 (3.2)	0		
Grade I	0	0	0		
Dysphagia, n (%)	10 (30.3)	1 (3.2)	3 (8.8)	0.006	0.033
Lung involvement, n (%)					
Dyspnoea	12 (36.4)	7 (22.6)	23 (67.6)	0.280	0.015
ILD confirmed by HRCT	14 (42.4)	30 (96.8)	34 (100)	<0.001	<0.001
Onset of ILD					
Acute onset (< 1 months)	4 (12.1)	10 (32.3)	20 (58.8)	<0.001	<0.001
Subacute onset (1–3 months)	4 (12.1)	6 (19.4)	10 (29.4)		
Chronic onset (> 3 months)	3 (9.1)	3 (9.7)	4 (11.8)		
Asymptomatic	3 (9.1)	11 (35.5)	0		
RP-ILD	0	0	34 (100)	-	-
HRCT findings of ILD					
Lower lobe consolidation	3 (9.1)	17 (54.8)	24 (70.6)	0.404	<0.001
Lower lobe reticulation	5 (15.2)	17 (54.8)	21 (61.8)	<0.001	0.001
Random GGO	8 (24.2)	10 (32.3)	16 (47.1)	0.003	0.128
Laboratory data					
CPK (U/L)	79 (48-555)	107 (69-153)	151 (75-240)	0.672	0.429
AST (U/L)	48 (26-86)	45 (34-81)	52 (38-82)	0.820	0.459
ALT (U/L)	39 (24-65)	38 (24-76)	41 (23-67)	0.863	0.890
LDH (U/L)	437 (270-765)	313 (263-415)	335 (293-490)	0.016	0.136
CRP (mg/dL)	4.1 (1.0-9.6)	0.3 (0.1-0.5)	0.6 (0.3-1.4)	<0.001	<0.001
Treatment, n (%)					
MP pulse therapy	17 (51.5)	17 (54.8)	22 (64.7)	0.625	0.327
IVIG	13 (39.4))	7 (22.6)	16 (47.1)	0.183	0.624
Drugs used up to 6 months after diagnosis					
(Hydroxy)chloroquine	18 (54.6)	0	0	-	-
Methotrexate	12 (36.4)	1 (3.2)	0	0.001	
Azathioprine	19 (57.6)	0	0	-	-
Cyclosporine	8 (24.2)	1 (3.2)	4 (11.8)	0.027	0.217
Leflunomide	3 (9.1)	0	0	-	-
Tacrolimus	0	29 (93.5)	30 (88.2)	-	-
Cyclophosphamide	1 (3.0)	26 (83.9)	30 (88.2)	<0.001	<0.001
Mycophenolate mofetil	4 (12.1)	0	1 (2.9)	-	-
Rituximab	0	0	0	-	-
JAK inhibitors	0	1 (3.2)	3 (8.8)	-	-
Follow-up duration (months)	58 (31-82)	57 (27-98)	46 (20-88)	0.968	0.404
Outcomes, n (%)					
Relapse	11 (33.3)	6 (19.4)	7 (20.6)	0.263	0.280
Death	3 (9.1)	0	9 (26.5)	-	0.109
Complications, n (%)					
Severe infection	4 (12.1)	2 (6.5)	3 (8.8)	0.673	0.709
Malignancy	3 (9.1)	4 (12.9)	1 (2.9)	0.704	0.356
Current disease status, n (%)					
Active disease	3/33 (9.1)	2/31 (6.5)	2/26 (7.7)		
Complete clinical response	17/33 (51.5)	21/31 (67.7)	17/26 (65.4)		
Remission	13/33 (39.4)	8/31 (25.8)	7/26 (26.9)		
Current medications, n (%)					
Systemic glucocorticoids					
In use	6/33 (18.2)	26/31 (83.9)	22/26 (84.6)	<0.001	<0.001
Dose (mg/day)	0 (0-0)	3 (1-5)	5 (2.8-9.3)	<0.001	<0.001
IVIG	0	1/31 (3.2)	1/26 (3.8)	-	-
(Hydroxy)chloroquine	1/33 (3.0)	0	0	-	-
Methotrexate	6/33 (18.2)	1/31 (3.2)	0	<0.001	-
Azathioprine	6/33 (18.2)	0	0	-	-
Cyclosporine	4/33 (12.1)	2/31 (6.5)	2/26 (7.7)	0.673	0.685
Leflunomide	4/33 (12.1)	0	0	-	-
Tacrolimus	0	23/31 (74.2)	21/26 (80.8)	-	-
Cyclophosphamide	0	0	0	-	-
Mycophenolate mofetil	5/33 (15.2)	3/31 (9.7)	4/26 (15.4)	0.709	>0.999
Rituximab	3/33 (9.1)	0	0	-	-
JAK inhibitors	0	1/31 (3.2)	2/26 (7.7)	-	-

Data are presented as mean ± standard deviation, median (25%-75%), or frequency (%).
[a] Comparison between Brazilian patients without RP-ILD and Japanese patients without RP-ILD
[b] Comparison between Brazilian patients without RP-ILD and Japanese patients with RP-ILD
ADM: amyopathic dermatomyositis; ALT: alanine aminotransferase; AST: aspartate aminotransferase; CPK: creatine phosphokinase; CRP: C-reactive protein; DM: dermatomyositis; HRCT: high-resolution computed tomography; GGOs: ground-glass opacities; ILD: interstitial lung disease; IVIG: intravenous immunoglobulin; JAK: Janus kinase; mo: months; MP: methylprednisolone; RP-ILD: rapidly progressive interstitial lung disease; D: time intervals.

Supplementary Table S3. Stratification of Brazilian and Japanese patients with anti-MDA5(+) dermatomyositis according to the year of diagnosis.

Variables	Brazil Before 2018 (n=22)	Brazil After 2019 (n=12)	Japan Before 2018 (n=33)	Japan After 2019 (n=32)	p^a	p^b
Demographics						
Median age at diagnosis (years)	42 (37-51)	41 (25-49)	52 (45-64)	49 (41-65)	0.005	0.033
Female, n (%)	13 (59.1)	7 (58.3)	22 (66.7)	22 (68.7)	0.580	0.720
Ethnicity, n (%)						
Asia	0	1 (8.3)	33 (100)	32 (100)	<0.001	<0.001
Africa	0	0	0	0		
Europe	0	0	0	0		
North-America	0	0	0	0		
South-America	22 (100)	11 (91.7)	0	0		
Oceania	0	0	0	0		
Clinical diagnosis, n (%)						
DM	15 (68.2)	4 (33.3)	2 (6.1)	10 (31.3)	<0.001	>0.999
ADM	7 (31.8)	8 (66.7)	31 (93.9)	22 (68.8)		
Dt: symptom onset and disease diagnosis (mo)	3.7 (0.9-10.1)	9.8 (2.4-13.5)	1.9 (1.3-3.1)	2.3 (1.2-3.5)	0.186	0.004
Clinical manifestations, n (%)						
Fever	4 (18.2)	6 (50.0)	13 (39.4)	13 (40.6)	0.138	0.735
Weight loss	11 (50.0)	9 (75.0)	10 (30.3)	13 (40.6)	0.166	0.088
Gottron's papules	22 (100)	10 (83.3)	26 (78.8)	28 (87.5)	0.034	0.658
Gottron's sign	22 (100)	10 (83.3)	31 (93.9)	31 (96.9)	0.511	0.176
Heliotrope rash	21 (95.1)	9 (75.0)	11 (33.3)	8 (25.0)	<0.001	0.005
Facial erythema	13 (59.1)	7 (58.3)	22 (66.7)	21 (65.6)	0.582	0.740
V-sign	8 (36.4)	7 (58.3)	6 (18.2)	9 (28.1)	0.206	0.090
Shawl sign	4 (18.2)	6 (50.0)	6 (18.2)	9 (28.1)	>0.999	0.284
Scratch dermatitis	2 (9.1)	0	4 (12.1)	8 (25.0)	>0.999	-
Raynaud's phenomenon	11 (50.0)	5 (41.7)	1 (3.0)	0	0.001	-
Digital ulcers	4 (18.2)	4 (33.3)	2 (6.1)	0	0.204	-
Skin ulcers	2 (9.1)	5 (41.7)	6 (18.2)	3 (9.4)	0.454	0.025
Holster's sign	0	0	1 (3.0)	3 (9.4)	-	-
Calcinosis	5 (22.7)	4 (33.3)	0	0	-	-
Mechanic's hands	2 (9.1)	6 (50.0)	23 (69.7)	22 (68.8)	<0.001	0.303
Joint involvement						
Isolated arthralgia	7 (31.8)	3 (25.0)	12 (36.4)	8 (25.0)	0.779	>0.999
Arthritis	9 (40.9)	5 (41.7)	5 (15.2)	14 (43.8)	0.056	>0.999
Myalgia	10 (45.5)	8 (66.7)	6 (18.2)	12 (37.5)	0.038	0.102
Muscle weakness						
Upper limbs						
Grade V	3 (13.6)	5 (41.7)	24 (72.3)	20 (62.5)	<0.001	0.214
Grade IV	19 (86.4)	5 (41.7)	8 (24.2)	11 (34.4)		
Grade III	0	2 (16.6)	1 (3.0)	0		
Grade II	0	0	0	1 (3.1)		
Grade I	0	0	0	0		
Lower limbs						
Grade V	3 (13.6)	6 (50.1)	22 (66.6)	19 (59.4)	<0.001	0.541
Grade IV	18 (81.8)	4 (33.3)	9 (27.3)	11 (34.4)		
Grade III	1 (4.6)	2 (16.6)	2 (6.1)	1 (3.1)		
Grade II	0	0	0	1 (3.1)		
Grade I	0	0	0	0		
Dysphagia, n (%)	7 (31.8)	4 (33.3)	1 (3.0)	3 (9.4)	0.005	0.075
Lung involvement, n (%)						
Dyspnoea	6 (27.3)	7 (58.3)	14 (42.4)	16 (50.0)	0.391	0.740
ILD confirmed by HRCT	7 (31.8)	8 (66.7)	33 (100)	31 (96.9)	>0.999	0.015
Onset of ILD						
Acute onset (< 1 months)	1 (4.5)	3 (25.0)	19 (57.6)	11 (34.4)	0.001	0.032
Subacute onset (1–3 months)	4 (18.2)	1 (8.3)	5 (15.2)	11 (34.4)		
Chronic onset (> 3 months)	1 (4.5)	2 (16.6)	5 (15.2)	2 (6.3)		
Asymptomatic	1 (4.5)	2 (16.6)	4 (12.1)	7 (21.9)		
RP-ILD	0	1 (8.3)	19 (57.6)	15 (46.9)	-	0.032
HRCT findings of ILD						
Lower lobe consolidation	2 (9.1)	1 (8.3)	20 (60.6)	21 (65.6)	0.001	0.002
Lower lobe reticulation	3 (13.6)	2 (16.6)	19 (57.6)	19 (59.4)	0.007	0.017
Random GGO	5 (22.7)	4 (33.3)	13 (39.4)	13 (40.6)	0.543	0.739

Variables	Brazil Before 2018 (n=22)	Brazil After 2019 (n=12)	Japan Before 2018 (n=33)	Japan After 2019 (n=32)	p^a	p^b
Laboratory data						
CPK (U/L)	100 (70-877)	59 (35-92)	150 (66-233)	115 (74-167)	0.536	0.012
AST (U/L)	51 (27-86)	42 (22-115)	40 (34-66)	62 (38-97)	0.812	0.207
ALT (U/L)	42 (27-63)	35 (22-73)	34 (23-56)	58 (30-92)	0.291	0.302
LDH (U/L)	511 (369-821)	267 (233-455)	318 (269-394)	335 (295-461)	0.002	0.328
CRP (mg/L)	4.5 (1.1-9.1)	1.3 (0.8-14.8)	0.4 (0.2-0.8)	0.5 (0.2-0.9)	<0.001	0.001
Treatment, n (%)						
MP pulse therapy	9 (40.9)	9 (75.0)	14 (42.4)	26 (81.3)	>0.999	>0.687
IVIG	5 (22.7)	9 (75.0)	16 (48.5)	7 (21.9)	0.088	<0.003
Drugs used up to 6 months after diagnosis						
(Hydroxy)chloroquine	13 (59.1)	5 (41.7)	0	0	-	-
Methotrexate	6 (27.3)	6 (50.0)	0	1 (3.1)	-	<0.001
Azathioprine	14 (63.6)	5 (41.7)	0	0	-	-
Cyclosporine	3 (13.6)	5 (41.7)	4 (12.1)	1 (3.1)	>0.999	<0.001
Leflunomide	1 (4.5)	2 (16.6)	0	0	-	-
Tacrolimus	0	0	28 (84.9)	31 (96.9)	-	-
Cyclophosphamide	0	2 (16.6)	28 (84.9)	28 (90.3)	-	<0.001
Mycophenolate mofetil	1 (4.5)	3 (25.0)	0	1 (3.1)	-	0.056
Rituximab	0	0	0	0	-	-
JAK inhibitors	0	0	0	4 (12.5)	-	-
Follow-up duration (months)	71 (44-117)	32 (16-58)	91 (70-119)	30 (22-48)	0.303	0.706
Outcomes, n (%)						
Relapse	9 (40.9)	2 (16.6)	9 (27.3)	4 (12.5)	0.382	0.658
Death	2 (9.1)	1 (8.3)	5 (15.2)	4 (12.5)	0.689	>0.999
Complications, n (%)						
Severe infection	2 (9.1)	2 (16.6)	2 (6.1)	3 (9.4)	>0.999	0.603
Malignancy	3 (13.6)	0	4 (12.1)	1 (3.1)	>0.999	0.999
Current disease status, n (%)						
Active disease	1/22 (4.6)	2/12 (16.7)	1/29 (3.5)	3/28 (10.7)	0.594	0.839
Complete clinical response	10/22 (45.4)	8/12 (66.7)	17/29 (58.6)	21/28 (75.0)		
Remission	11/22 (50.0)	2/12 (16.6)	11/29 (37.9)	4/28 (14.3)		
Current medications, n (%)						
Systemic glucocorticoids						
In use	3/22 (13.6)	4/12 (33.3)	24/29 (82.8)	24/28 (85.7)	<0.001	0.002
Dose (mg/day)	0 (0.0)	0 (0-5)	3 (1-7)	5 (2-5)	<0.001	0.021
IVIG	0	0	2/29 (6.9)	0	-	-
(Hydroxy) chloroquine	1/22 (4.6)	0	0	0	-	-
Methotrexate	3/22 (13.6)	3/12 (25.0)	0	1/28 (3.6)	-	<0.001
Azathioprine	2/22 (9.1)	4/12 (33.3)	0	0	-	-
Cyclosporine	1/22 (4.6)	3/12 (25.0)	3/29 (10.3)	1/28 (3.6)	0.642	<0.001
Leflunomide	3/22 (13.6)	1/12 (8.3)	0	0	-	-
Tacrolimus	0	0	21/29 (72.4)	23/28 (82.1)	-	-
Cyclophosphamide	0	0	0	0	-	-
Mycophenolate mofetil	1/22 (4.6)	5/12 (41.7)	2/29 (6.9)	5/28 (17.9)	>0.999	<0.001
Rituximab	3/22 (13.6)	1/12 (8.3)	0	0	-	-
JAK inhibitors	0	0	1/29 (3.5)	2/28 (7.1)	-	-

Data are presented as mean ± standard deviation, median (25%-75%), or frequency (%).

[a] Comparison between Brazilian and Japanese patients who were diagnosed before 2018.

[b] Comparison between Brazilian and Japanese patients who were diagnosed after 2019.

ADM: amyopathic dermatomyositis; ALT: alanine aminotransferase; AST: aspartate aminotransferase; CPK: creatine phosphokinase; CRP: C-reactive protein; DM: dermatomyositis; HRCT: high-resolution computed tomography; GGOs: ground-glass opacities; ILD: interstitial lung disease; IVIG: intravenous immunoglobulin; JAK: Janus kinase; mo: months; MP: methylprednisolone; RP-ILD: rapidly progressive interstitial lung disease; D: time intervals.