

**Supplementary Table S1.** Demographic and clinical features of the 94 patients with anti-SRP immune-mediated necrotising myopathy.

Clinical characteristics	Patients, n/N (n%)
Females	64/94 (68.1%)
Age at disease onset ≤ 18 years	12/94 (12.7%)
Duration from the disease onset to diagnosis (mean ± SD, months)	13.0 ± 18.6
≤12 months	74/94 (78.7%)
>12 months	20/94 (21.3%)
Antecedent infection or vaccination	18/94 (19.1%)
Statin-exposure	8/94 (8.51%)
Onset form	
Acute-subacute onset (≤6 months)	36/94 (38.3%)
Chronic onset (>6 months)	58/94 (61.7%)
Initial symptoms	
Limb or trunk weakness	65/94 (69.1%)
Bulbar symptoms	7/94 (7.44%)
Hyper CKemia	12/94 (12.8%)
Myalgia	14/94 (14.9%)
Muscle weakness	
Legs predominantly than arms	46/94 (48.9%)
Proximal weakness dominant	88/94 (93.6%)
Laterality	
Neck weakness	19/94 (20.2%)
Dysphagia	74/94 (78.7%)
Weight loss	41/94 (43.6%)
Myalgia	42/94 (44.7%)
Dyspnoea	27/94 (28.7%)
Dyspnoea	27/94 (28.7%)
Extra-muscular symptoms	
Fever	10/94 (10.6%)
Skin rash	7/94 (7.45%)
Arthritis	14/94 (14.9%)
Raynaud's phenomenon	5/94 (5.32%)
Associated disorder	
Cancer	6/94 (6.38%)
Rheumatic disease	6/94 (6.38%)
Blood examination	
Peak CK (IU/L, mean ± SD)	6885.76 ± 6300.67
Antinuclear antibody positivity	45/68 (66.2%)
Thyroid dysfunction	26/60 (43.3%)
Elevated erythrocyte sedimentation rate	16/55 (29.1%)
Myositis antibody spectrum	
Isolated SRP	50 (53.2%)
Anti-Ro-52	40 (42.5%)
Anti-PM-Scl-75	6 (6.38%)
Anti-PM-Scl-100	2 (2.13%)
Anti-Mi-2	1 (1.06%)
Anti-Ku	4 (4.26%)
Abnormal electrocardiogram	21/68 (30.9%)
Decreased capacity of respiratory function	23/34 (67.6%)
Interstitial lung disease	28/73 (38.4%)
Regimens	
GC	67/72 (93.0%)
GC alone	3/72 (4.17%)
IVIg	48/72 (67.0%)
IVIg alone	4/72 (5.56%)
GC + IVIg	7/72 (11.1%)
GC + steroid-sparing agent	21/72 (29.2%)
GC + IVIg + steroid-sparing agent	36/72 (50.0%)
Plasma exchange	4/72 (5.56%)

Data are presented as number/total number (percentage) of patients.

CK: creatine kinase; GC: glucocorticoids; IVIg: intravenous immunoglobulin; SD: standard deviation; SRP: signal recognition particle.

**Supplementary Table S2.** Semi-quantitative MRI scores of lower limb skeletal muscle in patients with signal recognition particle myopathy.

Muscle	Oedema score (mean)	fatty infiltration score (mean)
Gluteus maximus	1.48	1.44
Sartorius	0.98	0.53
Rectus femoris	1.34	0.63
Vastus intermedius	1.16	0.67
Vastus medialis	1.02	0.63
Vastus lateralis	1.41	0.72
Adductor longus	1.06	0.67
Adductor magnus	1.85	1.11
Gracilis	0.89	0.44
Long head of biceps femoris	1.59	1.03
Semitendinosus	1.16	1.08
Semimembranosus	1.73	1.14
Front group	1.17	0.72
Medial group	1.24	0.73
Posterior group	1.47	1.07