

**Supplementary material**

Patients with IIM met the 2017 EULAR/ACR criteria and the 2017 European Neuromuscular Centre criteria for immune-mediated necrotising myopathy (IMNM). Exclusion criteria encompassed individuals under 18 years of age, pregnant or breast-

feeding women, those with other autoimmune diseases, tumours, specific infections, or other muscle disorders, as well as patients who had engaged in vigorous exercise within one week prior to recruitment. For IIM, the Myositis Disease Activity Assessment Visual Analogue Scale (MYO-CAT), Myositis Intention to Treat Activity

Index (MITAX) and the Cutaneous Assessment Tool (CAT) were utilised to evaluate disease activity and severity; Manual Muscle Testing 8 (MMT8) was used to evaluate muscle strength. Interstitial lung disease (ILD) was diagnosed through high-resolution CT scans.

**Supplementary Table S1.** Clinical data of healthy controls and patients with IIM.

Items	IIM (N=48)	LSM (N=6)	HC (N=35)	p value (HC vs. IIM)
<b>Females</b>	39/48 (81.3%)	1 (16.7%)	25/29 (71.4%)	0.3055
<b>Age (years)</b>	49 (46, 57)	34 (24, 55)	54 (46, 61)	0.0782
<b>Subtypes of IIM</b>				
DM	26 (54%)			
IMNM	11 (23%)			
ASS	8 (17%)			
Unclassified IIM (n=3)	3 (6%)			
<b>Myositis-specific antibodies</b>				
Anti-EJ	3 (6%)			
Anti-Jo-1	5 (10%)			
Anti-MDA5	10 (21%)			
Anti-TIF1- $\gamma$	10 (21%)			
Anti-Mi-2 $\beta$	3 (6%)			
Anti-SRP	6 (13%)			
MSA-negative	5 (10%)			
Anti-NXP2	1 (2%)			
Anti-PL-12(n=1)	1 (2%)			
Anti-TIF1- $\gamma$ /HMGCR	1 (2%)			
Anti-MDA5/EJ	1 (2%)			
Anti-MDA5/PL-12	1 (2%)			
Anti-MDA5/NXP2	1 (2%)			
<b>Laboratory data</b>				
CRP (mg/L)	6.030 (2.888, 8.903)			
ESR (mm/h)	49.00 (34.25, 67.50)			
C3 (g/L)	0.8630 (0.7730, 0.9915)			
C4 (g/L)	0.2255 (0.1765, 0.2630)			
Creatinine kinase (IU/L)	442.00 (87.25, 3015.00)			
AST (IU/L)	73.50 (38.75, 156.30)			
ALT (IU/L)	65.00 (28.75, 123.00)			
LDH (IU/L)	428.0 (314.5, 775.3)			
HLDH (IU/L)	318.0 (231.5, 570.5)			
CD3 cell Count	782.5 (412.5, 1126.0)			
CD4 cell Count (cell/ $\mu$ L)	372.0 (214.8, 735.0)			
CD8 cell Count (cell/ $\mu$ L)	235.5 (124.3, 390.8)			
CD4/CD8	1.655 (0.975, 2.583)			
B cell Count (cell/ $\mu$ L)	148.5 (62.75, 270.3)			
NK cell Count (cell/ $\mu$ L)	92 (46, 212)			
IgG (g/L)	13.00 (10.12, 16.85)			
IgA (g/L)	2200 (1545, 3130)			
IgM (g/L)	1330.0 (960.5, 1695.0)			
<b>Myositis activity score</b>				
MYOACT score of extra-muscular organ	2.167(1.667, 3.333)			
VAS score of muscle	2 (0, 3)			
VAS score of lung	4 (2, 6)			
MITAX score of extra-muscular organs	1.75 (1.00, 2.50)			
MITAX score of muscle	1 (0, 3)			
MITAX score of lung	2 (0, 7)			
CAT score of skin activity	3.50 (1.00, 5.75)			
CAT score of skin injury	2 (0, 4)			
MMT8 score	128.0 (105.3, 146.9)			

Data is represented as N (%) or median (25% Percentile, 75% Percentile).

IIM: idiopathic inflammatory myopathy; LSM: lipid storage myopathy; HC: healthy control; DM: dermatomyositis; IMNM: immune-mediated necrotising myopathy; ASS: anti-synthetase syndrome; EJ: glycyL-tRNA synthetase; Jo-1: histidyl-tRNA synthetase; MDA5: melanoma differentiation-associated gene 5; TIF1- $\gamma$ : transcription intermediary factor 1- $\gamma$ ; SRP: signal recognition particle; HMGCR: hydroxymethylglutaryl-CoA reductase; MSA: myositis-specific autoantibody; NXP2: nuclear matrix protein 2; PL-12: alanyl-tRNA synthetase; CK: creatine kinase; LDH: lactate dehydrogenase; HBDH: hydroxybutyrate dehydrogenase; ALT: alanine transaminase; AST: aspartate transaminase; MYOACT: myositis disease activity assessment visual analogue scale; VAS: visual analogue score; MITAX: myositis intention to treat activity index; CAT: cutaneous assessment tool; MMT8: manual muscle testing 8.

\* $p < 0.05$ .