

Supplementary materials

Table S1 : Results of immunodot assay, ELISA and indirect immunofluorescence on MDA5 transfected cells and HEp-2 cells for the 23 patients with anti-MDA5 positive dermatomyositis

| Patient ID | Immunodot | Immunodot intensity | ELISA (U/mL) | IIF on MDA5 transfected cells (inverse dilution) | IIF pattern on HEp-2 cells |
|----------------------|-----------|---------------------|--------------|--|----------------------------|
| MDA5 ⁺ 1 | positive | 85 | 108 | 1350 | MDA5 pattern |
| MDA5 ⁺ 2 | positive | 56 | 138 | 1350 | MDA5 pattern |
| MDA5 ⁺ 3 | Hpo | 103 | 94 | 1350 | Non-specific |
| MDA5 ⁺ 4 | Hpo | 114 | 143 | 12150 | MDA5 pattern |
| MDA5 ⁺ 5 | positive | 85 | 119 | 4050 | MDA5 pattern |
| MDA5 ⁺ 6 | Hpo | 103 | 105 | 1350 | MDA5 pattern |
| MDA5 ⁺ 7 | Hpo | 112 | 181 | 4050 | MDA5 pattern |
| MDA5 ⁺ 8 | Hpo | 179 | 153 | 12150 | MDA5 pattern |
| MDA5 ⁺ 9 | positive | 46 | 91 | 4050 | MDA5 pattern |
| MDA5 ⁺ 10 | positive | 44 | 159 | 12150 | MDA5 pattern |
| MDA5 ⁺ 11 | positive | 31 | 100 | 4050 | MDA5 pattern |
| MDA5 ⁺ 12 | Hpo | 175 | 142 | 12150 | MDA5 pattern |
| MDA5 ⁺ 13 | Hpo | 140 | 147 | 12150 | MDA5 pattern |
| MDA5 ⁺ 14 | positive | 39 | 109 | 1350 | MDA5 pattern |
| MDA5 ⁺ 15 | positive | 76 | 135 | 1350 | MDA5 pattern |
| MDA5 ⁺ 16 | positive | 38 | 155 | 4050 | Non-specific |
| MDA5 ⁺ 17 | positive | 60 | 120 | 12150 | MDA5 pattern |
| MDA5 ⁺ 18 | Hpo | 136 | 161 | 12150 | MDA5 pattern |
| MDA5 ⁺ 19 | positive | 42 | 154 | 12150 | Non-specific |
| MDA5 ⁺ 20 | Hpo | 127 | 140 | 12150 | MDA5 pattern |
| MDA5 ⁺ 21 | positive | 72 | 98 | 450 | MDA5 pattern |
| MDA5 ⁺ 22 | positive | 81 | 143 | 4050 | Non-specific |
| MDA5 ⁺ 23 | positive | 74 | 66 | 0 | MDA5 pattern |

The IIF pattern on HEp-2 cells was termed "MDA5 pattern" when the characteristic cytoplasmic staining in rare clustered cells was observed. Otherwise, it was termed "Non-specific".

Abbreviations: Hpo: high positive, IIF: indirect immunofluorescence.

Table S2 : Clinical features of the three clinical subgroups made depending on the frequency of RP-ILD, cutaneous manifestations, ulcerations, Raynaud phenomenon and arthritis

| | Group 1 (n=11) | Group 2 (n=3) | Group 3 (n=9) |
|---|-----------------------|----------------------|----------------------|
| Mean age \pm SD (years) | 61 \pm 13 | 50 \pm 13 | 46 \pm 15 |
| Women | 6/11 (55%) | 2/3 (67%) | 5/9 (56%) |
| Clinical manifestations | | | |
| RP-ILD | 11/11 (100%) | 0 (0) | 0 (0) |
| Cutaneous manifestations | 9/11 (82%) | 3/3 (100%) | 9/9 (100%) |
| Ulcerations | 4/11 (36%) | 3/3 (100%) | 0 (0) |
| Arthritis | 6/11 (55%) | 3/3 (100%) | 8/9 (89%) |
| Raynaud phenomenon | 2/11 (18%) | 3/3 (100%) | 0 (0) |
| Muscular damage | 6/11 (55%) | 3/3 (100%) | 6/9 (67%) |
| Mortality rate | 6/11 (55%) | 0 (0) | 1/9 (11%) |

Abbreviations: RP-ILD: rapidly progressive interstitial lung disease