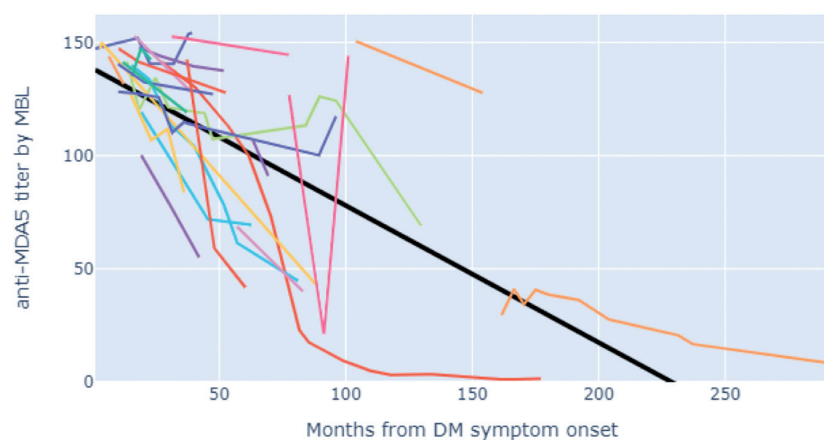
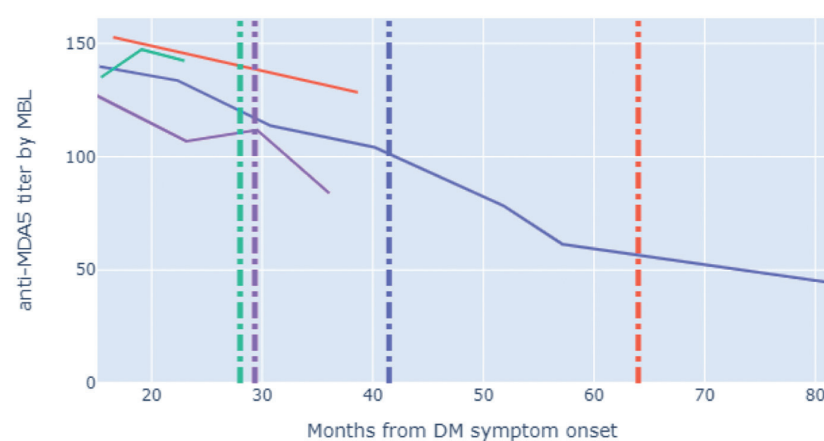


**Supplementary Table S1.** Demographic and disease characteristics of the anti-MDA5-positive and anti-Jo1-positive cohorts.

| Patient characteristics          | anti-Jo1 patients<br>(n=51)<br>Median (IQR) | anti-MDA5 patients<br>(n=51)<br>Median (IQR) | p-value  |
|----------------------------------|---|--|----------|
| Patient age at IIM symptom onset | 47.2 (33.7-55.5)                            | 47.3 (39.7-54.3)                             | 0.401    |
| Patient age at JH cohort entry   | 48.9 (34.4-58.0)                            | 48.8 (41.1-55.3)                             | 0.824    |
| Male sex                         | 29%   | 27%  | 0.779    |
| Race                             |   |  | 0.510    |
| White                            | 63%   | 47%  |          |
| Black                            | 28%   | 31%  |          |
| Asian                            | 4%  | 6%   |          |
| Other                            | 6%  | 10%  |          |
| Declined                         | 0%  | 2%   |          |
| Unknown                          | 0%  | 4%   |          |
| Ethnicity                        |   |  | 0.692    |
| Hispanic                         | 0%  | 2%   |          |
| Non Hispanic                     | 88%   | 86%  |          |
| Unknown                          | 12%   | 12%  |          |
| Duration of follow-up            | 5.6 (3.0-7.6)                               | 3.7 (1.4-5.9)                                | 0.019    |
| Ro52 positive                    | 75%   | 42%  | 0.001    |
| Gottrons sign or papules         | 28%   | 98%  | <0.00001 |
| Heliotrope sign                  | 28%   | 75%  | <0.00001 |
| Synovitis                        | 59%   | 57%  | 0.907    |
| Calcinosis                       | 4%  | 35%  | <0.00001 |
| Elevated CPK                     | 77%   | 15%  | <0.00001 |
| Maximum CPK                      | 720.0 (221.5-2162.0)                        | 101.0 (52.0-160.0)                           | <0.0001  |
| Maximum aldolase                 | 17.6 (10.0-32.3)                            | 8.8 (6.9-10.8)                               | <0.0001  |
| History of malignancy            | 8%  | 12%  | 0.527    |
| Interstitial lung disease        | 80%   | 80%  | 0.961    |
| Proximal muscle weakness         | 94%   | 73%  | 0.004    |
| Death                            | 14%   | 10%  | 0.516    |

**Supplementary Fig. S1.** Longitudinal MDA5 antibody titers (assayed by ELISA) over follow-up time in clinic. The majority of patients have decreasing titers over time, as indicated by the black line, which is fit to a regression of all individual anti-MDA5 data points.**Supplementary Fig. S2.** Longitudinal MDA5 antibody titers were available on four of the nine patients who experienced clinical remission. Of note, two patients (purple and blue lines) achieved clinical remission while maintaining anti-MDA5 positivity >32 units. Vertical lines indicate the date clinical remission was achieved. Most patients who went into clinical remission did not have available sera at the time of remission.