**Supplementary figure 1.** Detection of AIRE with flow-cytometric analysis.

(a) Detection of human AIRE from splenocytes in huAIRE-Tg with flowcytometric analysis. Wild-type non-Tg mice (WT) served as a negative control. Percentages of AIRE\(^+\)CD19\(^-\) cells surrounded by purple squares are indicated. One representative result from a total of more than three experiments is shown.

(b) Flow-cytometric analysis of AIRE expression from peripheral blood. Peripheral blood mononuclear cells from Patient 4 (before the treatment: Before Tx) but not from the other five patients showed AIRE\(^+\) cells. Peripheral blood from two healthy donors served as control.
**Supplementary table 1**: List of the patients

| Patient | Age | Sex | Diagnosis       | Interstitial pneumonia | Autoantibody       |
|---------|-----|-----|-----------------|-------------------------|--------------------|
| 1       | 85  | F   | Dermatomyositis | —                       | —                  |
| 2       | 60  | F   | Dermatomyositis | —                       | —                  |
| 3       | 64  | F   | Myositis        | —                       | Anti-SRP*          |
| 4       | 61  | M   | ADM             | Severe                  | Anti-ARS**         |
| 5       | 60  | F   | Myositis        | —                       | —                  |
| 6       | 50  | F   | Dermatomyositis | Mild                    | —                  |

* anti-signal recognition particle antibody

** anti-aminoacyl tRNA synthetase antibody
## Supplementary table 2: Primers and probes

| Gene        | Primer                                      |
|-------------|---------------------------------------------|
| **OVA**     | Forward: 5’-CTGTCTGGCATCTCCTCAGC-3’         |
|             | Reverse: 5’-TGACCCCTACCACCTCTCTGC-3’        |
| **AIRE**    | Forward: 5’-CGGTGGAGGTGAGGCTAGG-3’          |
|             | Reverse: 5’-CAGCCGTCACAGCAGATGAG-3’         |
| **Mouse Hprt** | Forward: 5’-TTCCCTGGTTAAGCAGTACAGC-3’     |
|             | Reverse: 5’-CCAAACAAAGTCTGGCCTGTAC-3’       |
| **Human Hprt** | Forward: 5’-GGTCAGGCAGTATAATCCAAAGATG-3’   |
|             | Reverse: 5’-AACAAAGTCTGGCCTATATCCAACAC-3’  |

| Gene        | Dual-Labeled Probe (FAM/TAMRA)               |
|-------------|---------------------------------------------|
| **OVA**     | 5’-TCTCAAGCTGTCCATGCACATGCAGC-3’             |
| **AIRE**    | 5’-ACGGCACAACCTCGTCCCTCATTCTTCTCCT-3’       |
| **Mouse Hprt** | 5’-TTCCAGGAGGTCTCCTTCACCAGCAAGCT-3’       |
| **Human Hprt** | 5’-TCGTTGGGTCCCTTCACCAGCAAGC-3’            |