

## **Abstract**

Patients with dermatomyositis positive for anti-aminoacyl tRNA synthetase (ARS) antibodies, also known as anti-synthetase syndrome (ASS), frequently present with mechanic's hand and interstitial lung disease (ILD). We first screened the antibody profiles of 59 patients with dermatomyositis, and then examined the cutaneous, muscular and pulmonary manifestations characteristic for patients with ASS. The anti-ARS antibodies Jo-1, PL-7, PL-12, EJ and KS, along with antibodies to TIF1- $\gamma$ , MDA5 and Mi-2 were examined. Among the 59 patients, 20, 21, 15 and 3 patients were classified into the ASS, non-ASS, myositis-specific antibody-negative, and unknown groups, respectively. Five of 16 patients (31%) with ASS had 6 relatives with a history of collagen diseases, within the second-degree of relationship, including 2 cases of dermatomyositis (vs. the non-ASS group,  $P=0.018$ ). Patients with ASS more frequently presented with fever and arthralgia, and had elevated levels of C-reactive protein. Nine of the 11 finger lesions (82%) clinically-diagnosed as mechanic's hands showed a psoriasiform tissue reaction. ILD was observed in 19 of 20 patients (95%) with ASS, and 8 of 21 patients (38%) in the non-ASS group, in which 6 patients possessed anti-MDA5 antibody. Patients with ASS showed higher serum levels of muscle enzymes, and 4 of 12 patients (33%) had fasciitis-dominant myopathy, while only 1 of 11 patients (9%) in the non-ASS group had fasciitis-dominant myopathy. Patients with ASS often present with a psoriasiform tissue reaction in the hand lesions and fasciitis-dominant myopathy, and the relatives of those with ASS are at high risk for collagen diseases.