



AUTOIMMUNE CONNECTIVE TISSUE DISEASES

## DIFFUSE ERYTHEMA WITH 'ANGEL WINGS' SIGN IN JAPANESE PATIENTS WITH ANTI-SMALL UBIQUITIN-LIKE MODIFIER ACTIVATING ENZYME ANTIBODY-ASSOCIATED DERMATOMYOSITIS

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**Background:** Myositis-specific autoantibodies (MSAs) are closely associated with distinct clinical subsets of dermatomyositis (DM). In 2007, a novel MSA-reacting small ubiquitin-like modifier activating enzyme (SAE) was identified in some patients with DM. Owing to the low proportion of patients with DM having the anti-SAE antibody (1.5%–3% in Asia and 6.7%–8.4% in Europe among all patients with DM), their clinical features have not been sufficiently analysed. We here demonstrate the clinical features in six Japanese cases of anti-SAE antibody-associated DM, and review the current published works.

**Observation:** Previous reports described the rashes in patients with anti-SAE antibody-associated DM as widespread skin involvement and dark-red/pigment-like skin rashes. We found diffuse erythema that sometimes resulted in erythroderma in our 6 patients. Erythema was found over the shoulder and lumbar regions, except in the inferior border of the scapular regions, which characteristically appeared like 'angel wings'. In addition, we retrospectively evaluated the clinical characteristics of our 7 patients and 46 previously reported patients with anti-SAE antibody-associated DM. The average age was 63 years, and the ratio between men and women was about 1: 2. All the patients had typical cutaneous manifestations of DM (Heliotrope rash, Gottron's papules and Gottron's signs, shawl sign and V-neck signs). Forty-six (87%) of the 53 patients also developed muscle weakness, including dysphagia (26 cases). Thirty-one (67%) of the 46 patients developed rashes before muscle involvement. Interstitial lung disease (ILD) was found in 47% patients. Malignancy was found in 21% patients.

**Key message:** 'Angel wings' sign may be characteristic of anti-SAE antibody-associated DM because none of the 60 DM cases without anti-SAE antibody at our department in the same period showed the pattern of erythema on their backs. Dysphagia would be an





important clinical issue in patients of anti-SAE antibody-associated DM.

