



## Anti-SUMO-activating enzyme E1 autoantibodies

<b>Acronyms</b>	SAE1, SAE2 (catalytic subunits of the <b>S</b> UMO- <b>a</b> ctivating <b>e</b> nzyme E1) SUMO ( <b>s</b> mall <b>u</b> biquitin related <b>m</b> odifier)
<b>Indications</b>	► Dermatomyositis  Differentiation of antigen specificities of antinuclear antibodies revealed by <u>ANA-IIFT</u> for academic purposes. Determination usually not indicated in ANA-IIFT negative sera. Diagnostic utility of antibodies at the moment unknown.
<b>see also</b>	<u>Autoantibodies in idiopathic inflammatory myopathies</u>
<b>Antigen</b>	The tight heterodimer SUMO activating enzyme E1 (SAE) consists of the subunits SAE1 (although known as Aos1; Mr 38,4 kDa) and SAE2 (Uba2; Mr 71,2 kDa), which are located within the nuclei, sparing the nucleoli, as revealed by IFT (Desterro et al. 1999; Gong et al. 1999) SUMO1-protein (101 amino acids; 11,6 kDa) is targeted to proteins during their posttranslational modification, not only for degradation within proteasomes but also for a variety of other cellular processes (nuclear transport, transcriptional regulation, apoptosis etc.). For ligation SUMO1 is activated by SAE2 resulting in the cleavage of four C-terminal amino acids, adenylation of the C-terminal carboxyl group and formation of a thioester bond followed by transfer to E2 and finally in attachment to the target protein (Wang et al. 2009).
<b>Autoantibodies</b>	By IIFT the antibodies show a coarse speckled nuclear staining pattern. Antibodies to both subunits SAE1 and SAE2 have been demonstrated in the same patient.
<b>Prevalence</b>	Anti-SAE autoantibodies have been demonstrated in two of twenty patients (10 %) suffering from dermatomyositis (Betteridge et al. 2007).
<b>Detection</b>	Immunoprecipitation of <sup>35</sup> S-methionine-labeled proteins extracted from K562-cells (Betteridge et al. 2007).
<b>Literature</b>	Betteridge Z, Gunawardena H, North J, Slinn J, McHugh N: Identification of a novel autoantibody directed against small ubiquitin-like modifier activating enzyme in dermatomyositis. <i>Arthritis Rheum</i> (2007); 56(9): 3.132 - 3.137 (PMID: <a href="#">17763420</a> ).  Desterro JM, Rodriguez MS, Kemp GD, Hay RT: Identification of the enzyme required for activation of the small ubiquitin-like protein SUMO-1. <i>J Biol Chem</i> (1999); 274:10.618 - 10.624 (PMID: <a href="#">10187858</a> ).  Gong L, Li B, Millas S, Yeh ET: Molecular cloning and characterization of human AOS1 and UBA2 components of the sentrinactivating enzyme complex. <i>FEBS J</i> (1999); 448:185 -189 (PMID: <a href="#">10217437</a> ).  Wang J, Lee B, Cai S, Fukui L, Hu W, Chen Y: Conformational transition associated with E1-E2 interaction in small ubiquitin-like modifications. <i>J Biol Chem</i> (2009); 284(30): 20.340 - 20.348 (PMID: <a href="#">19443651</a> ).