Autoantibodies: The mystery revealed

Connective Tissue Diseases

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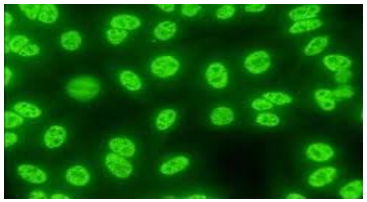
Anti-SAE antibodies

- Firstly reported in **2007**
- **Name**: Anti-small ubiquitin-like modifier activating enzyme (SAE) antibodies
- **Family:** Myositis specific antibodies (MSA)
- **Target antigen:** Two enzymatic sub-units: SAE2 (90 kDa) / SAE1 (40 kDA) involved in the post-translational modification of specific proteins known as **SUMOylation.**

- Reported in **dermatomyositis (DM) cases exclusively with significant titers**, mostly in **adults**
- **Prevalence :** 1% to 8%
- Included in the **239th European Neuromuscular Center (ENMC) criterias**

Screening technique:
Indirect Immunofluorescence pattern on HEp-2 cells: Fine speckled nuclear
Not used as a routine method for detection.

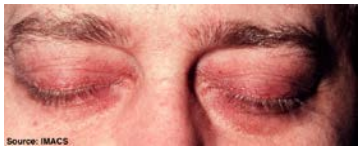


Confirmation techniques:

- **Immunoprecipitation (IP): Gold standard assay** for the detection of anti-SAE, with a **high sensitivity and specificity,** although is not routinely used.
- **ELISA or comparable methods** e.g. fluoroenzyme immunoassay (FEIA) has a **good sensitivity and specificity** when compared to IP
- **Multispecific immunoassays**: Line/dot blot immunoassays, Addressable Laser Bead immunoassays (ALBIA), and solid-phase antigen microarrays. **Most commonly used**

Main clinical features :

- **Cutaneous involvement (94%)**: Heliotrope sign, Gottron's papules, shawl sign and periungual erythema



Heliotrope sign

- **Amyotrophy/mild muscular weakness** + normal or slightly elevated CK levels
- **Interstitial lung disease (20%) mild**
- Associated **dysphagia** have been described 39%

Prognosis:

- **Good response to treatment**
- **Risk of cancer**: considered **intermediate** according to American College of Rheumatology (ACR) guidelines (up to 16% in a french cohort)

References