Autoantibodies: The mystery revealed

Connective Tissue Diseases

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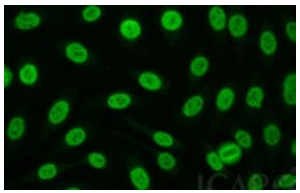
First **Myositis specific antibody** (MSA) to be identified in **1976**

Target antigen: NuRD chromatin remodeling complex (nucleosome remodeling-deacetylase complex).
A 240 kDa autoantigen, with two isoforms: Mi-2α (CHD3) and Mi-2β (CHD4)
Involved in transcription regulation..

Anti-Mi2 antibodies

Screening technique:
Indirect immunofluorescence (IIF) on HEp-2 cells



IIF pattern:
fine speckled staining

Confirmation techniques:

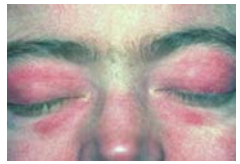
- ELISA
- Line/dot blot immunoassays (Sensitivity , Specificity 95%)

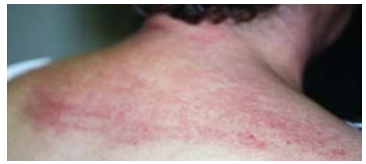
- Limitations: Variable performance, standardization issues
- Advantages: Quick, easy, specific
- Prevalence of anti-Mi-2 varies depending on the technique
- Mi2β are rarely isolated, positive predictive value (PPV) = 7%
- Mi2α PPV = 62% when isolated

Main disease: Dermatomyositis (DM)
Prevalence among adult DM patients: 4% - 35%.
Rare in children (10% of juvenile DM cases)
Included in the new classification of DM according to the **239th ENMC International Workshop**

Main clinical features:

- **Classic cutaneous manifestations:** Facial dermatosis, shawl sign, poikiloderma and flagellate erythema





- Mild **myopathy** with elevated CK levels out of proportion to their degree of muscle involvement
- Low occurrence of **pulmonary involvement**
- Associated with disease activity

Prognosis:

- **Good**
- **No** increased risk of development of interstitial lung disease
- Considered as an **intermediate cancer risk** biomarker according to the American College of Rheumatology (ACR 2022) guidelines
- Mi2β are associated with neoplasms: colon or breast carcinoma

References

Rev Tun Biol Clin 2024 31(2)