Autoantibodies: The mystery revealed

Connective Tissue Diseases

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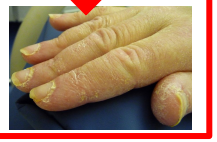
Anti-EJ antibodies

- Firstly described in 1990 in patients with myositis and interstitial lung disease (ILD) and was named after a patient called "EJ".
- **Target antigen:** anti-glycyl tRNA synthetase (75 kDa)

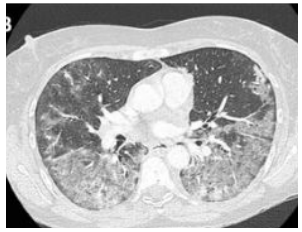
Idiopathic inflammatory myopathies: **Prevalence <5%**
Associated with **Antisynthetase Syndrome (ASS)**

Mechanic's hands is most frequently noted in anti-EJ ASS.
- Other frequent clinical features : rash, arthritis, fever.



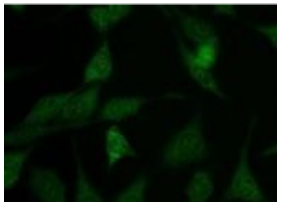
ASS associated ILD

- The predominant clinical manifestation, reported as an early manifestation in the majority of patients.
- **ILD patterns**: Non specific interstitial pneumonia
  Usual interstitial pneumonia
  Organizing pneumonia
  Diffuse alveolar damage.



- Although a good response to initial treatment, **recurrence of ILD is frequent**.
- Prognosis is poorer than ASSD with anti-Jo-1 antibody, mainly depending on the severity of ILD which is the first cause of death.

Screening technique:
- **Indirect Immunofluorescence (IIF)** on HEp-2 cells: Fine granular cytoplasmic fluorescence in all cells.



Cytoplasmic or negative staining on IIF does not exclude MSAs

- **ELISA screen (Jo1, PL7, PL12, EJ and KS)**

Confirmation techniques:

- **Immunoprecipitation (IP)**: Gold standard technique, impractical method for widespread diagnostic use
- Immunoenzymatic techniques (Superior specificity compared to IIF) :
  - **Line/dot blot immunoassay**
  - **ELISA** : highly sensitive method, but needs pure recombinant protein. Poor purification leads to false positives.
  The use of ELISA is comparable to IP in the detection of anti-EJ in a European population (sensitivity 90and specificity equal to 100%).

References