Autoantibodies: The mystery revealed

Connective Tissue Diseases

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- •Name: Anti-transcriptional intermediary factor 1-γ antibodies
- •Family: Myositis-specific autoantibody
- •Target antigen: E3-ligase, transcriptional factor-1-γ hetero complex 155/140 kDa; the larger tripartite motif (TRIM) family: TIF1-γ (TRIM33), TIF1-α (TRIM24), and, less commonly, TIF1-β (TRIM28)
- •Role in DNA repair, transcriptional elongation, differentiation of cells, mitosis and embryonic development.
- •It can act as tumor suppressor (Breast cancer), or tumor promotor (Cervical and pancreatic cancer)

Prevalence in: Adult dermatomyositis (DM): 7-31%

Juvenile DM: 22-32%

Connective tissue disease (CTD)-associated myositis: 15%

Juvenile CTD-associated myositis:33%

Anti-TIF1-y antibodies

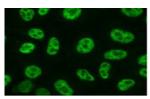
- **Skin involvement**: Severe hyperkeratotic Gottron papules (82,2%), psoriasiform lesions, 'Red on white' patches, shawl sign rash
- Muscle involvement: Moderate, Low CPK levels Dysphagia (18-40%)
- Muscle histology: Severe lesions Vasculopathy
- Diffuse interstitial lung disease, Arthritis, Raynaud phenomena: Infrequent

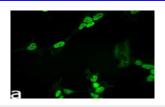
Screening technique: Indirect Immunofluorescence on HEp-2 cells. **IIF pattern:** Nuclear , fine speckled

Confirmation techniques: ELISA, WB (Immunoblotting)

-Strip test ,Immunoprecipitation..

IgG2 autoantibodies in particular were associated with mortality and malignancy in adult.





High risk factor of IIM-associated cancer:



- -The presence of anti-TIF1 gamma antibodies is associated with an increased risk of cancer in adults (27-fold higher)
- Cancer-associated DM in adults: VPN=97%, VPP=42%
- -There is no association with cancer in juvenile DM.
- -Its presence may be related to the initial diagnosis of cancer or the recurrence/metastasis of a known cancer.
- -Most frequently associated cancers are: **Pulmonary** cancer, **Ovarian** and **Breast** cancer, **Digestif tract** cancer, **Lymphoma** and **leukemia**.

All patients with anti-tif1- γ antibodies should undergo systematic cancer screening: Basic screening + CT scan of the neck, thorax, abdomen and pelvis

+ Cervical screening + Mammography + PSA + CA 125 + Pelvic ultrasonography + Faecal occult Blood + Nasoendoscopy.

