

Whats New in Pathogenesis of IIM

A number of different novel studies on many levels have been performed in the diverse entities of idiopathic inflammatory myopathies recently. The talk will highlight some of those but does not intend to implement a hierarchical validation.

MDA5-myositis is more frequent in Asia than in Europe, and in Europe 3 clusters of MDA5 DM have emerged one of which is associated with catastrophic ILD (30%). Mi2-DM features derepression of NuRD which is a helicase that upon binding upregulates expression of >100 genes not usually expressed in other types of DM.

TIF1gABs in DM are associated with the highest incidence of cancer but additional association with CCAR1 is not.

A recent study targeting C2 complement has not been successful despite clinical and experimental evidence of complement activation being involved in IMNM.

Patients with Antisynthetase syndrome have been successfully treated with CD19 CAR T-cell therapies, for which the morphological basis was laid by studies proving presence of B cells and plasmablasts in skeletal muscles.

IBM features senescent T cells that have been characterized by two groups simultaneously as KLRG1-positive a study targeting this molecule is underway.