Conclusion: Anti-FHL1 autoantibodies were detected in 20.5% of IIM patients. In IBM and IMNM, the presence of anti-FHL1-autoantibodies was associated with a severe myopathy as suggested by presence of dysphagia and muscle atrophy.

REFERENCES:

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Background: Antisyntehase syndrome (ASS) is characterized by inflammatory myopathy, interstitial lung disease, arthritis, mechanical hands and Raynaud phenomenon, among other features. Recent studies have shown that idiopathic inflammatory myopathies (IIM) may develop cardiac involvement, either ischemic (coronary artery disease) or inflammatory (myocarditis). We wonder if characteristic inflammatory interstitial involvement (interstitial lung disease) that appears in patients with the ASS may also affect the myocardial interstitial tissue. New magnetic resonance mapping techniques could detect subclinical myocardial involvement, mainly as edema (increase extracellular volume in interstitium and extracellular matrix), even in the absence of visible late Gadolinium enhancement (LGE).

Methods: Our aim was to describe the presence of interstitial myocarditis in a group of patients with ASS.

Results: MRI cardiac T1 mapping on 14 patients with ASS revealed mild to moderate LGE (Focal LGE in 5 out of 14 patients, 36%). The patients were consecutively selected from our outpatient myositis clinic. Myositis specific and associated antibodies were performed by means of line immunoblot (EUROMIMUN®). Cardiac magnetic resonance (CMR) was performed on all patients. The study protocol includes functional cine magnetic resonance and standard late gadolinium enhancement (LGE), as well as novel parametric T1 and T2 mapping sequences (modified look locker inversion recovery sequences - MOLLI) with extracellular volume (ECV) calculation 20 minutes after the injection of a gadolinium-based contrast material. LGE was analyzed on the end-systolic cardiac cycle using a dedicated CMR software. The percentage of LGE in the total left ventricular mass was calculated and compared to healthy controls. The CMR analysis was performed by two radiologists in consensus.

Conclusions: LGE was present in 36% of the patients with ASS. This study demonstrates that the ASS may affect the myocardium in a subclinical and mild extent. This also represents a potential new and novel manifestation, which could be a feature of the ASS. More studies are necessary in order to assert the prevalence of myocarditis in ASS.