Background: Antisynthetase 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- istic lung 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).

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

Methods: Cross-sectional, observational study performed in a tertiary care center. We included 13 patients diagnosed with ASS (7 male, 53%, mean (SD) age at diagnosis 56,8 years (±11,8)). The patients were consecutively selected from our outpatient myositis clinic. Myositis specific and associated antibodies were performed by means of line immunoblot (EUROIMMUN©). 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.

Results: CMR could not be performed in one patient due to anxiety. All patients studied (12) had a normal biventricular function, without alteration of segmental contraction. A third (4 out of 12, 33%) of the studied patients showed elevated T2 myocardial values without focal LGE, half of them (2/4) with an elevated ECV, consistent with myocardial edema. Two patients with normal T2 values showed unspecific LGE focal patterns, one in the right ventricle union points and another with mild interventricular septum enhancement (Figure 1). None of the patients studied refer any cardiac symptomatology. All the four patients with T2 mapping alterations (100%) had interstitial lung involvement, but only 4 out of 8 (50%) of the rest ASS patients without T2 mapping positivity. The autoimmune profile was as follows: 10 anti-Jo1/Ro52, 1 anti-EJ/Ro52, 2 anti-PL12.

Conclusion: Myocarditis, although subclinical, appears to be a feature in ASS patients. T1 and T2 mapping sequences might be valuable to detect and monitor subclinical cardiac involvement in these patients. The possibility that the same etiopathogenic mechanism may be involved in the interstitial tissue in lung and myocardium is raised. More studies must be done in order to assert the prevalence of myocarditis in ASS.

REFERENCES:
Background: Idiopathic inflammatory myopathies are a heterogeneous group of systemic autoimmune diseases. Several phenotypes have been linked to specific autoantibodies. Clinically amyopathic dermatomyositis with rapidly progressive interstitial lung disease, the most severe form of ILD, is associated with the anti-MDA5 antibodies. However not all the patients with dermatomyositis and anti-MDA5 positive antibodies develop this severe condition.

Methods: We retrospectively analyzed the clinical and immunological data of 90 anti-MDA5 patients [50 female, 55.6%, mean (SD) age at diagnosis 47 (15.4) yrs.] with dermatomyositis recruited from a multicenter register in Spain (MEDRA5) including 30 hospitals. All the patients fulfill the International Myositis Classification Criteria (EULAR/ACR) for dermatomyositis (score >90%). Anti-MDA5 were detected by means of commercial immunoblot (EUROMIMUN®). The chi-square test was used to assess the relationships between qualitative variables. The Kruskal-Wallis test was used to compared medians between groups.

Results: Sixty-six patients (73.3%) were diagnosed with clinically amyopathic dermatomyositis. Three different phenotypes linked with the anti-MDA5 antibody were identified: group 1: patients with rapidly-ILD phenotype (28 patients, 31.1%), group 2: antisynthetase-like phenotype (23 patients, 25.5%), and group 3: non-ILD phenotype (39 patients, 43.3%). Clinical and immunological comparison between the groups disclosed that age at disease onset was higher (median, IQR) in patients from group 1 [53 (43-60)] vs. group 2 [46 (40-56)] vs group 3 [42 (41-51)] (p<0.01); disease onset was more frequent in spring in patients from group 1 (46.5%) than in the rest of the groups (21.7% and 28.9%) (p<0.01). Cancer was detected in 7 patients, only associated with myositis in 3 cases (3 years interval between cancer and dermatomyositis) without significant differences between phenotypes. Vascularity (one case ANCA positive) was detected in 9 cases (6 limited to skin, 1 renal and 1 intestinal), 6 of them in the group 3 (statistical significance, in comparison with group 1 and 2, p<0.01). Mortality rate was higher in group 1 (51.9%, 16 out of 17 due to refractory respiratory failure) vs group 2 (12.5%) or 3 (0%) (p<0.01). Anti Ro52 positivity was more frequent in group 1 (65.4%) vs. group 2 (25%) or 3 (35.5%) (p<0.017), although it did not reach statistical significance in terms of mortality (p=0.173) or patients admitted in the intensive care unit (p=0.173). Mechanic hands were more frequent in group 2 (40.6%) than in groups 1 (25%) and 3 (34.4%) (p=0.05). Fever was significantly most frequent in group 1 (52.6%) than in group 2 (21.1%) and 3 (28.3%) (p=0.001). Other clinical or immunological features such as arthritis, myositis, or the number of characteristic skin lesions among others were not more frequent in one group or another.

Conclusion: Three different phenotypes of patients positive to anti-MDA5 were identified. The presence or not of ILD, or the different type (rapidly progressive or not) of ILD were the main feature that allow to differentiate these phenotypes, which are relevant in clinical practice.

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