PBC or other autoimmune liver disease or manifested elevated liver function tests.

Conclusion: The expression of the PDC antigenic components is variable both in SSc patients with AMA positive PBC and in SSc patients AMA positive without PBC or altered liver function tests, but we could not identify a clinical significance of this variability. It may be necessary to maintain a strict follow-up of these patients and to perform longitudinal studies to determine the prognostic value of this variable expression of PDC components in the onset of PBC.

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FRI0317

NOVEL CLASSIFICATION OF IDIOPATHIC INFLAMMATORY MYOPATHIES BASED ON DISTINCTIVE FEATURES AND AUTOANTIBODIES: ANALYSIS OF 67 KOREAN PATIENTS

Sang Wan Chung¹, In Seol Yoo², Su-Jin Yoo², Jinhyun Kim², Seong Wook Kang², Seungcheol Shim², Miye Kwon³, Chung-Il Joung³, Seung-Jae Hong¹, Yeon-Ah Lee¹. ¹Kyung Hee University Medical Center, Seoul, Korea, Rep. of (South Korea); ²Chungnam National University College of Medicine, Daejeon, Korea, Rep. of (South Korea); ³College of Medicine, Konyang University, Daejeon, Korea, Rep. of (South Korea)

Background: Since Bohan and Peter first described their diagnostic criteria for idiopathic inflammatory myopathies (IIM) in 1975, new discoveries such as myositis-specific and myositis-associated autoantibodies (Abs) have been made.

Objectives: To investigate correlations between specific myositis Abs and their frequencies and clinical associations across different IIM groups, collectively demonstrating the utility of the new clinicoserologic classification in Korean adult patients with IIM.

Methods: We conducted a multicenter cohort study including 67 adult patients (age ≥ 18 years) who have been diagnosed as IIM by ENMC criteria. Immunoblot assay with Euroline strip (EUROIMMUN, Germany) was performed using the sera of definite dermatomyositis (DM, n=36), definite polymyositis (PM, n=25), amyopathic DM (n=4), DM sine dermatitis (n=1), and immune mediated necrotizing myopathy (IMNM, n=1). Patients were classified based on three classifications: 1) novel clinicoserologic classification suggested by Troyanov et al. in 2017. 2) 2017 EULAR/ACR classification criteria. 3) 2004 European neuromuscular center (ENMC) criteria. Associations of myositis Abs and clinical subsets of IIM were investigated.

Results: The distribution of the various IIM differed strikingly from those using the 3 classifications (Fig1). According to the 2004 ENMC classification and 2017 EULAR/ACR classification criteria, DM and PM was the most and the second frequent entities (DM: 55.2%, 56.7%; PM: 35.8%, 37.3%). But, using the new clinicoserologic classification, overlap myositis (OM) is the major type of IIM and the frequency of PM is significantly decreased. Anti-ARS Abs specificity included anti-Jo-1 (16.4%), -OJ (4.6%), -EJ (6.2%), -PL-7 (3.1%), and -PL-12 (4.6%). Interstitial lung disease was closely associated with anti-MDA5, and anti-ARS Abs, while DM-specific skin lesion was frequently observed in patients with anti-TIF1γ and anti-ARS Abs. Seven patients with cancer-associated DM were identified. They were positive for anti-TIF1γ (5/7) and anti-SRP (3/7) (table 1).

Table 3. Associations between myositis autoantibodies and distinctive clinical features

	Anti-ARS N=25	Anti-SRP N=24	Anti-MDA5 N=14	Anti-TIF1γ N=9	Anti-MG2 N=8	Anti-SAE N=3	Total N=87
ILD	20*	5	11*	0	3	1	36
Malignancy	1	3	0	5*	1	0	7
DM specific skin lesion	8*	6	10	9*	5	2	39
Dysphagia	2	2	2	3	2	1	16

*P<0.05, *P<0.001

Fig 1. Distribution of 67 patients according to 3 classifications for idiopathic inflammatory myopathies

Conclusion: Novel classification based on distinctive features and new myositis Abs reflects the clinical phenotype of IIM better. Establishment of a system routinely available to screen myositis Abs is needed. This will be beneficial to provide more precise diagnosis and proper management for patients with IIM.

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FRI0318

THROMBOCYTOPENIA AND MULTI-ORGAN INVOLVEMENT MIGHT BE POOR PROGNOSTIC FACTORS FOR SYSTEMIC SCLEROSIS CARDIOMYOPATHY

Xiuning Wei, Lijuan Yang, Donghui Zheng, Lie Dai. Sun Yat-Sen Memorial Hospital, Sun Yat-Sen University, Department of Rheumatology, Guangzhou, China

Background: Systemic Sclerosis (SSc)-cardiomyopathy is associated with a high mortality in SSc. It is important to identify potential prognostic factors for SSc-cardiomyopathy.

Objectives: To describe the clinical characteristics and identify potential prognostic factors for patients with SSc-cardiomyopathy.

Methods: We retrospectively reviewed the clinical data of SSc-cardiomyopathy patients hospitalized at Sun Yat-Sen Memorial Hospital from January 1992 to November 2018. SSc-cardiomyopathy was defined as structurally and functionally abnormal of heart muscle in SSc patients with cardiac symptoms. Coronary artery disease, hypertension, valvular disease and congenital heart disease were excluded. The Chi-square test was used to compare the differences between the death group and the survival group.

Results:

1. There were 903 SSc patients recruited and 20 patients (2.2%) of them were SSc-cardiomyopathy. Among these 20 patients, 65% were females, age was 52.8±13.2 years, disease duration was 3.1±2.3 years, duration from SSc onset to cardiomyopathy occurred was 2.8±2.3 years.
2. SSc-cardiomyopathy developed in 18 patients (90%) with diffuse cutaneous SSc (dcSSc), and in 2 patients (10%) with limited cutaneous SSc (lcSSc). Eleven patients (55%) had pulmonary fibrosis, 7 patients (35%) had pulmonary arterial hypertension (PAH), 5 patients (25%) had scleroderma renal crisis (SRC) and esophageal involved respectively, 6