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DETECTION OF COEXISTING MYOSITIS-SPECIFIC AUTOANTIBODIES WITH LINE AND DOT IMMUNOASSAYS IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES

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Background: Myositis-specific autoantibodies (MSAs) can be identified in up to 60% of patients with idiopathic inflammatory myopathies (IIM) (1). Based on previous immunoprecipitation studies, MSAs are considered to be mutually exclusive (1), but detection of coexisting MSAs with line or dot immunoassay (LIA/DIA) has been described (2-3).

Objectives: To determine the prevalence of detection of coexisting MSAs with different LIAs/DIAs in patients with IIM, assess the concordance between different assays and describe the clinical phenotype of patients with coexisting MSAs.

Methods: Cross-sectional assessment of prevalence of coexisting MSAs as detected by two LIAs (Euroline Autoimmune Inflammatory Myopathies, Euroimmun, Lübeck, Germany; and ImmcoStripe Myositis Advanced LIA, Trinity Biotech, Buffalo, USA) and 1 DIA (12 IgG Dot, Alphadia, Mons, Belgium), concordance between these assays and clinical phenotype of patients with coexisting MSAs in a single-center cohort of patients with a diagnosis of one of the subtypes of IIM as diagnosed by the treating physician.

Results: Nineteen of 145 patients (12%) had coexisting MSAs on at least one assay: 5 on the DIA (3,5%) and 5 (3,5%) and 11 (7,6%) on the LIA of Euroimmun and Trinity Biotech respectively. The results were concordant between 2 assays for more than one MSA in only 3 patients. The three combinations of these patients were anti-Jo-1 and anti-NXP-2, anti-Jo-1 and anti-TIF1-gamma, and anti-SAE and anti-NXP2 autoantibodies. The first two patients had an antisynthetase syndrome and the last patient an overlap myositis phenotype. All assays combined, anti-TIF1-gamma (9/19), anti-Jo-1 (7/19) and anti-NXP2 (6/19) autoantibodies were the most detected MSAs with concurrent detection of another MSA, though concordance between assays for these MSAs was low to moderate.

Conclusion: Detection of coexisting MSAs with LIA or DIA occurs in a minority of patients with IIM with varying prevalence between assays from different manufacturers. The combination of more than 1 MSA is not concordant between LIAs/DIAs in the vast majority of patients, suggesting that in most patients detection of coexisting MSAs with LIAs/DIAs reflects a problem of specificity of the assays for the involved autoantibodies.

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LOW DOSE CYCLOPHOSPHAMIDE AND PIRFENIDONE MIGHT WORK IN SYNERGY TO RELIEVE INTERSTITIAL LUNG DISEASE WITH CONNECTIVE TISSUE DISEASE: A PRELIMINARY OBSERVATIONAL STUDY

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Background: There are anti-inflammation and anti-fibrosis agents available for connective tissue disease associated interstitial lung disease (CTD-ILD). A clinical study has been initiated to assess the combination of the two (NCT03221257), but whether they can spare dose for each other is unknown.

Objectives: This preliminary study is aimed to observe outcomes of CTD-ILD patients receiving cyclophosphamide plus pirfenidone as a rescue therapy, with each agent at about one third of routine dosage.

Methods: We enrolled CTD-ILD patients who did not improve their symptoms (dyspnea or cough) after at least one-month steroids treatment (prednisone $\geq 1\text{mg/kg}$ daily). Patients who had adjusted immunosuppressive agents other than steroids or had received anti-fibrotic medications within three months before enrollment were ruled out. We switched the treatment into pulse cyclophosphamide (0.4g/m² monthly) combined with pirfenidone (300mg twice per day). Besides, we reduced the steroids to prednisone 0.5mg/kg daily and then tapered routinely. All the patients were followed up for 12 months.

Results: We enrolled seven patients, of whom two had anti-synthetase syndrome, two had Sjögren syndrome, two had scleroderma and one had mixed connective tissue disease. The media DLCO was 51% of prediction (range 47.7% to 63%) and media FVC was 72.3% of prediction (range 39% to 81%). The media 6MWD was 275m (range 202 to 324m). At the end of 12-month follow-up, all the patients regained functional independence with a media 52.7% increase of 6-minute walk distance (range 34.4% - 86.3%). Pulmonary function tests showed improved forced vital capacity (median improvement 13.4%, range 0-35.9%) and DLCO (median improvement 6.3%, range 1.7%-16%). The HRCT score had a median decrease of 20.1% (range 11.7% to 29.6%). For quality of life assessment, the SGRQ total score had a median improvement of 53.3% (range 19.5% to 61.7%). Of note, no adverse events were observed during the 12-month follow-up.

Conclusion: Our Study provided preliminary but promising clinical evidence for a new strategy in treating CTD-ILD, that cyclophosphamide and pirfenidone might work in synergy and spare dose for each other. A well-designed controlled study is needed to further establish its safety and efficacy.

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VALIDATION OF 2017 CLASSIFICATION CRITERIA FOR ADULT AND JUVENILE IDIOPATHIC INFLAMMATORY MYOPATHIES PROPOSED BY EULAR/ACR IN CHINESE PATIENTS

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Background: Idiopathic inflammatory myopathies(IIMs) are heterogeneous disorders characterized by muscle weakness and muscle inflammation. Although the Bohan and Peter criteria proposed in 1975 are most widely used^[1,2], there are some limitations. Firstly, they did not clearly specify how to exclude other forms of myopathy disease. Secondly, each criterion is not well defined either. EULAR and ACR jointly proposed the classification criteria for adult and juvenile IIMs and their major subgroups in 2017. The data-driven criteria exhibited high sensitivity and specificity. But most of the patients (62.6%) in the data were Caucasians, the performance of the criteria in Asian patients is unknown, which was one of the important limitations^[6,7].

Objectives: To evaluate the ability of 2017 EULAR/ACR Classification Criteria for Adult and Juvenile Idiopathic Inflammatory Myopathies (IIM) to