Table 1.

NCM	SSc PH (n=40)	SSc noPH (n=38, 2 missing)	P value
Mean capillary density/mm (±SD)	5.0 (±1.4)	5.4 (±1.6)	0.39
Mean number of fingers with density <3 (±SD)	2.3 (±1.9)	1.7 (±2.1)	0.20
Mean abnormally shaped capillar- ies/ mm (±SD)	1.2 (±0.46)	0.99 (±0.53)	0.07
NCM pattern (n)			0.103
Normal/aspecific	1	3	
early	0	3	
active	4	7	
late	34	25	
Not interpretable	1	2	
Biomarkers*	SSc PH (n=33, 7 missing) (median, IQR)	SSc noPH (n=40) (median, IQR)	P value
IL6 pg/ml	4.8 (1.7-8.2)	2.5 (1.3-5.6)	0.047
PDGF-AB-BB pg/ml	29535 (24777-33458)	32221 (29409-37549)	0.0034
sTRAIL pg/ml	36.6 (20.4 - 50.6)	60.8 (40.7 - 82.4)	<0.0001
sVCAM1 ng/ml	311 (254-374)	265 (228 - 323)	0.0236
Endostatin ng/ml	53.8 (41.9 - 72.6)	39 (28.6 - 46)	<0.0001
sVEGFR2 pg/ml	10388 (7276 - 13323)	13736 (10282 - 16554)	0.0006
CXCL4 ng/ml	3347 (2546-4708)	5309 (4089-6322)	<0.0001
VEGF-A pg/ml	672 (248 - 955)	346 (175-703)	0.0424
VEGF-D pg/ml	671 (421-1012)	515 (262-734)	0.0296

* only significant values with a p<0.05 are shown

Disclosure of Interests: Jacqueline Lemmers: None declared, C.H.M. van den Ende: None declared, R. Smeets: None declared, Brigit Kersten: None declared, Arjan van Caam: None declared, Sander van Leuven: None declared, Jolanda van Haren-Willems: None declared, Arie van Dijk Speakers bureau: Actelion(-Janssen), Consultant of: Actelion(Janssen), Grant/research support from: Unrestricted educational Grant for PhD student from Actelion(Janssen), Madelon Vonk Speakers bureau: Actelion(Janssen), Boehringer Ingelheim, Roche, Consultant of: Advisory Board from Actelion(Janssen) and Boehringer Ingelheim, Grant/research support from: Unrestricted Educational Grant and research support from Actelion(Janssen), research support from Ferrer
DOI: 10.1136/annrheumdis-2021-eular.2722

AB0442 CLINICAL FEATURES OF POLYMYOSITIS AND DERMATOMYOSITIS PATIENTS WITH SEVERE DYSPHAGIA

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Background: Polymyositis (PM) and dermatomyositis (DM) are autoimmune inflammatory diseases characterized by proximal myositis. Dysphagia has been reported to develop in 35 to 62% of PM/DM patients and known as poor prognosis factor.

Objectives: The purpose of this study is to determine the clinical characteristics of PM/DM patients who present with deglutition disorder.

Methods: Consecutive patients with PM/DM who visited National Hospital Organization Tokyo Medical Center between April 2010 and January 2021 are included in this study. We compared clinical features between the patients with and without dysphagia. The diagnosis of dysphagia was based on videofluorography swallow study, and dysphagia requiring gastrostomy was defined as severe dysphagia. The clinical characteristics compared in this study were following: age of onset, levels of serum creatine kinase (CK) and lactate dehydrogenase(LDH), sense of dysphagia, manual muscle test (MMT) score, and complication of malignancy or interstitial pneumonia.

Results: A total of 73 patients with PM/DM were identified. Among them, 12 patients were diagnosed with dysphagia, and 5 patients developed severe dysphagia. Patients with dysphagia had the following characteristics compared to patients without dysphagia: higher levels of serum LDH (833.7 ± 500.1 U/L vs 471.9 ± 321.0 U/L, p = 0.0088), higher levels of serum CK at initial examination (6070.3 ± 7184.8 IU/L vs 1534.7 ± 2978.8 IU/L, p = 0.0086) and more frequent sense of dysphagia (90.9% vs 10.6%, p < 0.0001), lower MMT score (3.18 ± 1.07 vs 4.31 ± 0.75, p = 0.0017). In addition to those, patients with severe dysphagia presented older age of onset (mean age 69.4 ± 12.0 vs 51.7 ± 14.8, p = 0.014), more frequent complication of malignancy (80.0% vs 14.8%, p = 0.0048) and less frequent complication of interstitial pneumonia (0.0% vs 55.5%, p = 0.023).

Conclusion: These results indicate that dysphagia develops frequently in PM/DM patients with higher levels of serum LDH or CK, sense of dysphagia and low MMT score. Among them, patients with elderly onset or malignancy are at risk for severe dysphagia, and should be treated carefully.

Disclosure of Interests: None declared.

DOI: 10.1136/annrheumdis-2021-eular.2757

AB0443

BIOMARKER SUCH AS IL-17, IL-21 AND TIMP-1, IS USEFUL FOR PREDICTING THE PATHOPHYSIOLOGY OF CONNECTIVE TISSUE DISEASE-ASSOCIATED PULMONARY HYPERTENSION

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Background: Connective tissue disease-associated pulmonary hypertension (CTD-PH) is constructed by a variety of pathologies, including cardiac, pulmonary, and vascular involvement, as well as immune abnormalities. Because of its various constructs, it is difficult for many respiratory physicians, cardiologists, and rheumatologists to determine a treatment strategy for CTD-PH. In addition, CTD-PH has different pathologies from iPAH, and there are cases in which immunosuppressive therapy is effective. These suggests that the two PAHs may have different pathogenesis, including inflammation in the pulmonary artery. However, there are not enough biomarkers to distinguish pathologies. On the other hand, it has been reported that various cytokines such as TIMP-1, Interleukin (IL)-6, IL-17, and IL-21 are involved in the pathogenesis of CTD-PH or vasculitis. (1,2) However, the relationship between these cytokine expression and the pathogenesis or treatment of CTD-PH has not been fully clarified.

Objectives: To clarify the relationship between cytokine profile and clinical features, change in cytokines and hemodynamics by treatment, association with the effectiveness of immunosuppressive therapy.

Methods: Patients suspected PH was included. At the time of cardiac catheterization(RHC), sera in pulmonary pre and post-capillary were collected and TIMP-1, MCP-1, IL-17 and IL-21, IL-12p70 and IL-6 were analyzed by ELISA(ABCAM UK, Ella simple plex USA). The following clinical data were collected: age, gender, underlying disease, complication of interstitial lung disease, treatment (immunosuppressant and pulmonary vasodilator), hemodynamics. Furthermore, we investigated the relationship between cytokines and clinical data.

Results: 15 cases of CTD-PH, 13 cases of non-CTD-PH, and 6 cases of non-PH were analyzed. (SSc 12 cases, MCTD 7cases, SLE 2 cases, and others 13 cases) 28 cases were diagnosed with PH by RHC. There was a positive correlation between IL-6 and mean pulmonary arterial pressure in all PH case. In addition, MCP-1, IL-6, and TIMP-1 tend to be high in SSc-PH cases. On the other hand, in Non-SSc-PH, IL-12p70 and IL-17 were high. In cases who pulmonary vascular hemodynamics improved by treatment, IL-17, IL-21, and TIMP-1 decreased.

Conclusion: Biomarker profiles in pulmonary capillaries may differ depending on the disease. Furthermore, it suggested that IL-17, IL-21 and TIMP-1 may be biomarkers of therapeutic effect.

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Disclosure of Interests: None declared.

DOI: 10.1136/annrheumdis-2021-eular.2765

AB0444

CLINICAL-IMMUNOLOGICAL CHARACTERISTICS OF PATIENTS WITH INFLAMMATORY MYOPATHIES

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Background: Inflammatory disorders of the skeletal muscle include polymyositis (PM), dermatomyositis (DM), amyopathic dermatomyositis ADM, (immune mediated) necrotizing myopathy (NM), overlap syndrome with myositis (overlap myositis, OM) and including anti-synthetase syndrome (ASS).

The detection of myositis-specific antibodies, the clinical effect of which remains to be determined, may be of great importance for diagnosis.

Objectives: To study clinical-immunological characteristics of patients with inflammatory myopathies.

Methods: 57 pts were enrolled in this study: 28 (47%) pts were diagnosed with DM, 14 (23%) pts – OM, 5(8%) pts – PM, 5 (8%) pts-NM, 4 (7%) pts-ADM, 1 (2%)- ACC. Duration of disease in average 11,29 (2-48) month. Standard clinical examination and laboratory immunological evaluation including myositis-specific autoantibodies were performed.

Results: There were 3 age groups: 18-39 years old – 16 (28%) pts, 40-49 yo – 28 (49%) pts and > 60 yo – 13 (33%) pts. Symptoms of myositis were muscle weakness 57 (100%) pts, dysphagia 29 (51%) pts, arthritis -12 (21%) pts, Raynaud's phenomenon 21 (37%) pts, skin lesions- 37 (65%) pts (Gottron signs- 7 (12%) pts, digital ulcers -11 (19) pts, panniculitis – 6(11%) pts, hand of mechanic 16(28%) pts). MMT 8 was 55.57 (SD 18,49) (20-80) score. Clinical features of myositis were myocarditis 8 (14%) pts and arrhythmia 9 (16%) pts, interstitial lung