Gene expression profiles may distinguish patients that can benefit from imatinib³. Also, Notch signalling could be exploited to increase imatinib uptake into fibroblasts, thereby increasing efficacy⁴.

References:

- [1] Pattanaik D et al. Pathogenesis of Systemic Sclerosis. *Frontiers in Immunology*. 2015;6: 272
- [2] Distler JHW et al. Tyrosine kinase inhibitors for the treatment of fibrotic diseases such as systemic sclerosis: towards molecular targeted therapies. *Annals of the Rheumatic Diseases*. 2010;69: 48-51
- [3] Chung L et al. Molecular framework for response to imatinib mesylate in systemic sclerosis. *Arthritis and Rheumatology*. 2009 Feb;60(2):584-91
- [4] Harrach S et al. Notch Signaling Activity Determines Uptake and Biological Effect of Imatinib in Systemic Sclerosis Dermal Fibroblasts. *Journal of Investigative Dermatology*. 2019; 139(2):439-4475

Acknowledgments: Skin Research Institute of Singapore

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.666

AB0627

PROGNOSIS AND MORTALITY OF DERMATOMYOSITIS AND POLYMYOSITIS PATIENTS WITH MALIGNANCY

X. Yang¹, Y. Hao², Z. Zhang¹ on behalf of Peking University First Hospital; Department of Rheumatology and Clinical Immunology. ¹Peking University First Hospital, Beijing, China; ²Peking University First Hospital, Beijing, China

Background: Previous studies indicate that cancers in DM/PM patients are associated with increased mortality. Hence, identifying predictors of malignancy in PM and DM is crucial. However, few large series studies have reported prognostic and predictive factors of malignancy in patients with PM and DM. Moreover, in recent years, several published studies also allow us to better understand the clinical characteristics of malignancy in PM and in DM.

Objectives: To analyze the mortality and identify the major independent risk factors for death in patients with dermatomyositis/polymyositis (DM/PM) complicated with malignant tumor.

Methods: The clinical data of all patients with DM/PM in Peking University First Hospital from January 2007 to Jan 2019 were retrospectively reviewed. All patients were followed up to confirm whether they had malignant tumors. According to the statistics of the National Bureau of Statistics of China, the standard mortality (SMR) and life lost years (YLL) of patients with DM/PM were combined with malignant tumors. The Kaplan-Meier method was used to analyze the 10-year survival of DM/PM patients with malignant tumors. Cox multivariate regression was used to predict independent risk factors for DM/PM patients with malignant tumors.

Results: A total of 334 patients with dermatomyositis and 69 patients with polymyositis were enrolled in the study. The mean age of onset was 50.5 ± 14.8 years and 48.9 ± 16.1 years, with a median follow-up of 40.6 (11.6-77.6) months. Among them, 320 patients were successfully followed up, including 69 patients with death, 46 DM/PM with malignant tumors (38 with dermatomyositis and 8 with polymyositis). The average age of onset of DM/PM patients with malignant tumors was 55.4 ± 15.1 years and 59.5 ± 4.7 years, respectively, of which 17 died. The age-sex adjusted SMR of DM/PM patients without malignant tumors was 9.0 (95% CI 6.8-11.2). The age-sex adjusted SMR of DM/PM with malignant tumors was 12.3 (95% CI 9.0-14.7). The life loss of male patients with dermatomyositis complicated with malignant tumors was 30.1 years, and that of females was 38.6 years; the life loss of male patients with polymyositis was 27.6 years, and that of females was 22.1 years. A 10-year survival analysis showed that DM/PM patients with malignant tumor had significantly worse prognosis than patients without malignancy (p=0.001 Log-rank). The 1-, 5-, and 10-year survival rates of DM/PM patients who did not have malignant tumors were 87.9%, 81.9%, and 78.4%, respectively. DM/PM Patients with malignant tumors 1, 5, and 10 years The survival rates were 73.3%, 56.0%, and 45.7%, respectively. The independent risk factors for death in DM/PM patients with malignant tumors were advanced age (HR=1.11 95% CI 1.02-1.20, p=0.014) and infection (HR=17.07 95% CI 1.66-175.75, p= 0.017).

Conclusion: Malignant tumor is a common in patients with DM/PM, and the mortality of DM/PM patients with malignant tumors is high. The independent predictors of mortality for PM/DM patients with malignant tumors were age at disease onset and infection.

survival rates	1 year	5 years	10 years
Without malignancy	87.9%	81.9%	78.4%
Malignancy	73.3%	56.0%	45.7%

Acknowledgments: We thank our patients with PM/DM participating in this study.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.4048

AB0628

EMERGING AUTOANTIBODIES PANEL (MYOSITIS ASSOCIATED AND MYOSITIS SPECIFIC ANTIBODIES) IN INFLAMMATORY MYOPATHIES: THE FREQUENCIES OF AND RELATIONSHIP WITH CLINICAL FEATURES

G. K. Yardımcı¹, E. Erul², E. Bilgin¹, B. Farisoğulları¹, L. Kılıç¹, Z. Saribas³, U. Kalyoncu¹, A. Akdoğan¹, B. Sener³, Ş. A. Bilgen¹, O. Karadag¹. ¹Faculty of Medicine; Hacettepe University, Rheumatology, Ankara, Turkey; ²Faculty of Medicine; Hacettepe University, Internal Medicine, Ankara, Turkey; ³Faculty of Medicine; Hacettepe University, Department of Medical Microbiology, Ankara, Turkey

Background: The idiopathic inflammatory myopathies (IIM) are characterized by muscle weakness, skin disease and various internal organ involvement and they can overlap with other autoimmune diseases. Recent autoantibody panels improve understanding and management of inflammatory myopathies. Myositis specific autoantibodies including Anti-TIF-1, Anti-NXP2, Anti-MDA5, Anti-SAE1, anti-HMGCR and anti-cN1A are regarded as key biomarkers aiding the diagnosis of patients. On the other hand, myositis-associated autoantibodies (MAAs) are also found in other autoimmune rheumatic diseases.

Objectives: To investigate the clinical meaning and impact of new myositis autoantibodies panel in real life data.

Methods: A total of 110 subjects (77 female, 33 male) admitted to Hacettepe University Hospitals with the signs and symptoms of IIM were screened by a line immunoblot assay (EUROLINE: Autoimmune Inflammatory Myopathies16 Ag) between 2017 and 2020. Only moderate or strong reactivity results were reported as positive. Demographic, clinical, laboratory, therapeutic data and imaging features were obtained by the retrospective review of medical records. IIM patients were diagnosed by Bohan and Peter's criteria and classified according to the EULAR/ACR classification criteria for adult and juvenile IIM and their major subgroups.

Results: IIM was diagnosed in 61 patients (42.6% DM/JDM and 57.4% PM) and patients with overlap were in decreasing order, Scleroderma (n=8), RA (n=5), Sjogren (n=4), SLE (n=3), autoimmune hepatitis (n=2).

Myositis-specific autoantibodies (MSAs) were found in 60%, myositis-associated autoantibodies (MAAs) in 41% of inflammatory myositis patients. The most common MSAs were anti-Jo-1 (16.4%) and anti-MDA-5 (13.1%) and the most common MAA was Ro-52 (32.7%) (Table 1). MAAs were more common in patients with polymyositis (54% vs 23% p=0.013). Twenty-one (34.4%) patients (61.0% females) had interstitial lung disease (ILD). Anti-Jo-1 (38.1%) and anti-Ro-52 (52.4%) was the most common MSA and MAA in patients with IIM and ILD.

MSAs were also determined in 10 of the 49 patients who were not diagnosed with IIM. Five patients with Anti-SRP, Anti-Mi-2 alpha, Anti-Jo-1 Anti-PL7, Anti-MDA-5 autoantibodies diagnosed as connective tissue disorder, one of them also had bladder cancer. Three patients with Anti-NXP2, Anti-PM-Scl 75, Anti-Ro-52, Anti-Mi-2 beta, Anti-PL7, Anti-PL12 autoantibodies had ILD. One patient (Anti-Mi-2 alpha, Anti-NXP2 and Anti-Ku autoantibodies positive) had viral myositis and one patient (Anti-Mi-2 alpha, Anti-Ro-52 autoantibodies positive) had inflammatory polyneuropathy.

Conclusion: Our study revealed relationship of anti-Jo-1 and anti-Ro-52 but not anti-MDA-5 in ILD-inflammatory myopathies. Even though new autoantibodies panel give opportunity to a closer look for inflammatory myopathies, larger series of patients should be evaluated to determine the association of specific antibodies in the differential diagnosis and prediction of outcome of IIM. MSA positivity in non-IIM diagnosed patients should be monitored to determine whether this positivity is related to a future disease development.

Table 1: Frequency of MSAs and MAAs in idiopathic inflammatory myositis

	JDM/DM (n=26 patients)	PM/ASS (n=35 patients)	All (n=61 patients)
Myositis-specific autoantibodies, n(%)			
Anti-Jo-1	1 (3.8%)	9 (25.7%)	10 (16.4%)
Anti-MDA-5	5 (19.2%)	3 (8.6%)	8 (13.1%)
Anti-SRP	2 (7.7%)	4 (11.4%)	6 (9.8%)
Anti-TIF1g	4 (15.4%)	2 (5.7%)	6 (9.8%)
Anti-NXP2	4 (15.4%)	2 (5.7%)	6 (9.8%)
Anti-SAE1	2 (7.7%)	1 (2.9%)	3 (4.9%)
Anti-Mi-2 alpha	2 (7.7%)	1 (2.9%)	3 (4.9%)
Anti-Mi-2 beta	2 (7.7%)	0	2 (3.3%)
Anti-PL7	0	3 (8.6%)	3 (4.9%)
Anti-PL12	0	2 (5.7%)	2 (3.3%)
Anti-EJ	1 (3.8%)	3 (8.6%)	4 (6.6%)
Anti-OJ	1 (3.8%)	0	1 (1.6%)
Myositis-associated autoantibodies, n(%)			
Anti-Ro-52	4 (15.4%)	16 (45.7%)	20 (32.8%)
Anti-PM-Scl 100	1 (3.8%)	4 (11.4%)	5 (8.2%)
Anti-PM-Scl 75	1 (3.8%)	4 (11.4%)	5 (8.2%)
Anti-Ku	0	4 (11.4%)	4 (6.6%)

Disclosure of Interests: Gözde Kübra Yardımcı: None declared, Enes Erul: None declared, Emre Bilgin: None declared, Bayram Farisoğulları: None declared, Levent Kılıç: None declared, Zeynep Saribas: None declared, Umut Kalyoncu Consultant of: Abbvie, Amgen, Janssen, Lilly, Novartis, UCB, Ali Akdoğan: None declared, Burcin Sener: None declared, Şule Apraş Bilgin: None declared, Omer Karadag: None declared

DOI: 10.1136/annrheumdis-2020-eular.6437

AB0629

VASCULAR MANIFESTATIONS OF SYSTEMIC SCLEROSIS: SIMILAR IN PATHOGENESIS, DIFFERENT IN FREQUENCY

N. Yudkina¹, A. Volkov², E. Nikolaeva², E. Nasonov^{2, 1}. ¹V.A.Nasonova Research Institute of Rheumatology, Moscow, Russian Federation; ²V.A.Nasonova Research Institute of Rheumatology, Moscow, Russian Federation

Background: Pulmonary arterial hypertension (PAH) is one of the main manifestations of vascular involvement in systemic sclerosis (SSc). The association of PAH with Raynaud's phenomenon (RP) and digital ischemic disorders is assumed.

Objectives: The aim of the study to detect of the possible relationship of pathogenetically similar processes and the predictor role in the early diagnosis of PAH and digital ischemic disorders by the nailfold videocapillaroscopy (NVC).

Methods: 111 patients with SSc (51 patients with PAH (SSc-PAH) and 60 patients without PAH) include in this study. In all patients, the diagnosis of SSc was validated according to the 2013 ACR-EULAR criteria. PAH was diagnosed by right heart catheterization. NVC was performed in all recruited subjects. Capillary quantitative parameters (loops length and width, capillary density, neovascularization) were evaluated and a semi-quantitative scoring was used (specific patterns - early, active and late) to define microvascular alterations. The test evaluated the presence of capillaroscopic changes in the nailfold bed on 2-5 fingers of both hands. The normal capillaroscopic pattern was characterized by the presence of 7-11 capillaries in the form of hairpins per 1 mm. Pathological patterns were characterized by morphological and structural changes, such as expanded and giant capillaries, hemorrhages, avascular fields, neovascularization. The capillaroscopy pattern (normal, non-specific, early/active/late) was determined qualitatively. Decreased capillary density, dilated, giant or branched capillaries, microhemorrhages were evaluated semi-quantitatively.

Results: RP was detected in 100% of cases in both groups. In the analysis of capillaroscopic patterns in both groups, the early and late scleroderma types of changes prevailed, but no significant differences were noted. Typical scleroderma patterns were found in 51 patients (100%) with SSc-PAH. In 3 patient with SSc without PAH, the abnormalities were regarded as non-specific. The NVC pattern was detected to be early in 8 patients with SSc-PAH and in 11 with SSc without PAH. The NVC pattern was found to be active in 16 patients with SSc-PAH and in 18 with SSc without PAH. The NVC pattern was detected to be

late in 27 patients with SSc-PAH and in 28 with SSc without PAH. In addition to RP, the development of digital ulcers was noted with equal frequency in history (25 patients with SSc-PAH and 32 with SSc without PAH). Also, the time to their appearance from the first symptom of SSc was the same (56 (16; 84) months and 44 (23; 72) months, respectively). Severe forms of digital ischemic disorders were observed rarely and with the same frequency in the studied groups. Ischemia in 2 patients with SSc-PAH and in 5 patients with SSc without PAH, gangrene in 2 patients only in the SSc group without PAH, amputation in 1 of each group.

Conclusion: In the course of the study, it was not possible to identify differences between the NVC patterns, the frequency and severity of digital ischemic disorders in the compared groups. That fact does not allow using the NVC as an early diagnosis of PAH in SSc. However, the NVC can help predict the development of digital ischemic disorders.

References: No

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.6656

AB0630

IMBALANCE BETWEEN TH17 AND REGULATORY T CELLS IN PATIENTS WITH PM/DM COMBINED WITH EBV/CMV VIRAEMIA

X. Zheng¹, R. Su¹, Y. Liu¹, X. Li¹, C. Wang². ¹The Second Hospital of Shanxi Medical University, Department of Rheumatology, Tai Yuan, China; ²The Second Hospital of Shanxi Medical University, Department of Rheumatology, Tai Yuan, China

Background: Dermatomyositis and polymyositis (DM/PM) are associated with muscle weakness and inflammatory infiltration within the skeletal muscle. The numerical and functional defects of immune cells, due to long-term uses of glucocorticoids and disease-modifying anti-rheumatic drugs (DMARDs) together with immune disturbances associated with disease itself, lead to high risks in opportunistic infections, such as Epstein-Barr virus (EBV) and cytomegalovirus (CMV).¹⁻² We want to observe changes of peripheral lymphocytesubsets in PM/DM patients with EBV and/or CMV infection, especially whether there is imbalance between Th17 and Treg cells.

Objectives: To investigate the characteristics of peripheral lymphocyte subsets in PM/DM with EBV and/or CMV infection, especially the Th17 and Treg cells.

Methods: From February 2016 to November 2019, PM/DM patients with EBV and/or CMV viremia (infection group, n=34) and without infection (non-infection group, n=31) as well as healthy adult controls (n=20) were enrolled in our study. Absolute numbers of total T, total B, NK, CD4 + T, CD8 + T cells, and CD4 + T subsets (Th1, Th2, Th17 and Treg cells) in peripheral blood by flow cytometry combined with standard absolute counting beads.

Results: (1) Compared with PM/DM patients without infection, 34 PM/DM patients with EBV and/or CMV infection, including 12 patients with EBV, 20 patients with CMV, 2 patients combined EBV and CMV, the absolute number of total T lymph cells (P=0.019), total B lymph cells (P=0.037), NK cells (P=0.033), CD4 + T cells (P=0.000), Th1 cells (P=0.014), Th2 cells (P=0.003), Th17 cells (P=0.003), Treg cells (P=0.004) lower than its of (P=0.003) patients without infection, the absolute number of CD8 + T cells (P=0.427) has no obvious difference between them.

(2) And its the absolute number of total T lymph cells (P=0.000), total B lymph cells (P=0.003), NK cells (P=0.000), CD4 + T cells (P=0.000), CD8 + T cells (P=0.006), Th1 cells (P=0.000), Th2 cells (P=0.001), Th17 cells (P=0.000) and Treg cells (P=0.000) significantly lower than healthy control.

(3) Compared with the healthy control, the absolute number of total T lymph cells (P=0.000), NK cells (P=0.000), CD4 + T cells (P=0.031), CD8 + T cells (P=0.000), Th1 cells (P=0.002), Th2 cells (P=0.031), and Treg cells (P=0.000) in PM/DM without infection evidently lower, but there is no significant difference in absolute number of total B lymph cells (P=0.19) and Th17 cells (P=0.171).

Conclusion: We show that the absolute number of peripheral blood lymphocytes and CD4+T subsets in patients with PM/DM with EBV and/or CMV viremia is further reduced. In addition to Treg cells, a decrease in Th17 cells may also be an important feature of EBV and/or CMV infection in DM/PM. These cell reductions may be the cause and risk indicator of viral infections.

References:

- [1] Yang X, Hao Y, Zhang X, et al. Mortality of Chinese patients with polymyositis and dermatomyositis. Clin Rheumatol. 2020 Jan 4. doi: 10.1007/s10067-019-04910-w. [Epub ahead of print]. PMID: 31902027
- [2] Matsushita T, Kobayashi T, Kano M, et al. Elevated serum B-cell activating factor levels in patients with dermatomyositis: Association with interstitial lung disease. J Dermatol. 2019;46:1190-6. doi: 10.1111/1346-8138.15117