However, whether the nailfold capillaroscopy pattern is distinct in SSc patients with malignancy remained unknown.

Objectives: The aim of this study was to investigate the incidence rate of malignancy in SSc patients. Nailfold capillaroscopy morphology patterns in SSc patients with cancer were also compared to those without cancer.

Methods: During 2004 to 2014, consecutive 310 SSc patients who visited outpatient clinics at Taichung Veterans General Hospital, Taiwan were enrolled. Nailfold capillaroscopy was performed at a magnification of 200x. Abnormal morphology description were analysed and categorised by the scoring system proposed by Dr. Cutolo². SSc with malignancy was defined if the subject had a cancer diagnosis during the follow-up period.

Results: Among 310 SSc patients, 28 (10.9%, 13 males, 25 females) patients had cancer. The mean age of SSc with malignancy is 62±10.9 years. Sixty-four percent SSc patients with cancer is diffuse type, but only 42% of them were tested positive for anti-Scl-70 antibodies. The most common cancer were genitourinary tract and gastrointestinal tract, accounting for almost two-thirds patients. Cancer and SSc were frequently diagnosed at the same year. In SSc patients with cancer, 19 patients received nailfold capillary microscope exams when SSc was diagnosed. The assessment of nailfold capillaroscopy morphology patterns in SSc patients with cancer demonstrated neither enlarged loop, giant loop, microhemorrhage nor angiogenesis. However, prominent avascular areas could be observed universally in SSc patients with malignancy.

Conclusions: Rheumatologists should be aware of malignancy in SSc patients, especially those with diffuse type and within the 1st year of symptoms onset. SSc patients with malignancy tend to present atypical capillaroscopic pattern of prominent avascular area without loop dilatation, microhemorrhage and angiogenesis.

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AB0818 THE COMPARATIVE STUDY OF SYSTEMIC SCLEROSIS AND SYSTEMIC LUPUS ERYTHEMATOSUS-ASSOCIATED PULMONARY ARTERIAL HYPERTENSION

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Background: Systemic Sclerosis(SSc) is the main cause of Connective Tissue Disease-associated pulmonary arterial hypertension (CTD-PAH) in western countries, while Systemic Lupus Erythematosus(SLE) is the first cause in Asian.

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Objectives: This study aimed to identify the clinical difference between SSc-PAH and SLE-PAH.

This study aimed to identify the clinical difference between SSc-PAH and SLE-PAH.

Methods: SSc and SLE patients with pulmonary arterial hypertension who visited Guangdong General Hospital in China from 2009 to 2017 were recruited. PAH was diagnosed by transthoracic echocardiography or right heart catheterization and clinical data of patients was collected. Comparative study between SSc-PAH and SLE-PAH was conducted to identify the difference.

SSc and SLE patients with pulmonary arterial hypertension who visited Guangdong General Hospital in China from 2009 to 2017 were recruited. PAH was diagnosed by transthoracic echocardiography or right heart catheterization and clinical data of patients was collected. Comparative study between SSc-PAH and SLE-PAH was conducted to identify the difference.

Results: Twenty-nine SSc-PAH and 55 SLE-PAH patients were enrolled. The baseline characteristics of two groups was shown in table 1. In comparative study: the gender($p=0.018$), the age of PAH diagnosis($p=0.001$), Raynaud's phenomenon($p<0.001$), WHO function state($p=0.019$), PASP ($p=0.013$), the diameter of RA($p=0.045$) and RV($p=0.029$) showed significant difference between SSc and SLE-PAH.

Abstract AB0818 – Table 1. Baseline Characteristics of SSc-PAH and SLE-PAH Patients

	SSc-PAH(29)	SLE-PAH(55)	<i>p</i>
Female	23(79.3%)	53(96.4%)	0.018
Age	46.0±14.3	34.2±10.3	0.001
Course ^a (year)	3.0[0.5, 6.4]	0.8[0.0, 9.5]	0.833
Raynaud's phenomenon	28(96.6%)	22(40.0%)	<0.001
Anti-U1RNP antibody	11(37.9%)	27(49.1%)	0.329
WHO Fc(III/IV)	18(62.1%)	45(84.9%)	0.019
NT-proBNP(pg/ml)	2051[489, 5227]	1392[113, 3632]	0.176
TTE			
PASP(mmHg)	67.1±25.9	81.1±24.0	0.013
Diameter of RA(mm)	48.9±7.7	53.0±8.5	0.045
Diameter of RV(mm)	52.9±6.6	55.9±6.7	0.029
LVEF(%)	63.0±10.5	67.4±7.6	0.081
Pericardial effusion	11(37.9%)	22(41.5)	0.752
RHC^b			
SPAP(mmHg)	88.3±17.5	75.9±22.2	0.109
DPAP(mmHg)	28.0±9.2	29.5±11.6	0.764
mPAP(mmHg)	48.3±11.7	47.5±14.8	0.773
PVR(WU)	13.1±7.2	11.4±6.1	0.363
CI(L·min ⁻¹ ·m ⁻²)	2.9±0.8	3.2±1.1	0.687

TTE: transthoracic echocardiography; PASP: pulmonary arterial systolic pressure; RA: right atrium; RV: right ventricle; LVEF: left ventricle ejection fraction; RHC: right heart catheterization; PAP: pulmonary arterial pressure; PVR: pulmonary vascular resistance; CI: cardiac index.

^a the time between the onset of CTD and PAH;

^b 6 SSc patients and all SLE patients received RHC;

n=number at risk, SpA=Spondyloarthritis; PsA=Psoriatic arthritis

Conclusions: The SSc-PAH patients tend to be older, have more angiopathic characteristics and more silent clinical course than SLE patients, which might bring some insight into the different pathophysiological process between SSc and SLE-PAH.

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AB0819 TFH CELLS CONTRIBUTE TO ABNORMAL B CELL PROFILES IN DERMATOMYOSITIS

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Background: CD4 +CXCR5+PD-1+T follicular helper (Tfh) cells assist B cells in their proliferation, differentiation and antibody class switch.⁽¹⁾ Several studies indicate that Tfh cells play important roles in autoimmune diseases such as SLE, RA and pSS, which are characterised by the production of multiple antibodies.⁽²⁻⁴⁾ The frequency of Tfh cells in the peripheral blood from patients with dermatomyositis(DM) and whether they participate in the development of DM remain to be elucidated.

Objectives: To investigate the frequencies of Tfh cells and B cell profiles in DM patients. To further determine the association of Tfh cells and B cells in DM patients and clarify the possible mechanism.

Methods: Peripheral blood mononuclear cells (PBMCs) were isolated from DM patients and age, gender-matched healthy controls (HCs), respectively. The frequency of Tfh (CD4 +CXCR5+PD-1+) cells, total B (CD19+) cells naïve B (CD19 +CD27-) cells, memory B(CD19 +CD27+) cells and plasmablasts (CD19 +CD38+) were examined by flow cytometry. The serum levels of IL-21, IgG, IgM, IgE and IgA were tested by enzyme linked immunosorbent assay (ELISA).

Results: The percentages of circulating Tfh cells are significantly higher in DM patients than HCs ($p<0.0001$). Compared to HCs, the absolute numbers of circulating Tfh cells also upregulate markedly in DM patients ($p<0.05$). The mRNA expression levels of Bcl-6, a typical transcription factor of Tfh cells, increase apparently in PBMC from DM patients ($p<0.05$). Serum levels of IL-21, a Tfh-specific cytokine, are obviously higher in DM patients ($p<0.01$). The percentages of total B cells ($p<0.01$) and Naïve B cells ($p<0.01$) upregulate significantly in DM patients while Memory B cells ($p<0.05$) downregulate markedly when compared with HCs. The absolute numbers of plasmablasts ($p<0.05$) and Naïve B cells ($p<0.05$) increase notably while memory B cells decreased obviously ($p<0.01$). Serum levels of IgG ($p<0.01$), IgM ($p<0.0001$), IgE ($p<0.01$) and IgA ($p<0.05$) are obviously higher in DM patients ($p<0.05$). The frequencies of Tfh cells are positively correlated with total B cells ($r=0.633$, $p<0.001$) and Naïve B cells ($r=0.643$, $p<0.01$).

Conclusions: Tfh cells may contribute to abnormal B cell profiles and antibodies production in DM and participate in the pathogenesis of DM. Tfh cell-targeted therapy might be a potential strategy for DM.

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AB0820 ANTI-KS AUTOANTIBODY IS ASSOCIATED WITH SICCA SYNDROME AND INTERSTITIAL LUNG DISEASE

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Background: Anti-aminoacyl-tRNA synthetase (anti-ARS) autoantibodies have been found in patients with polymyositis/dermatomyositis (PM/DM). Anti-KS, an anti-ARS antibody, often presents with interstitial lung disease (ILD) without other clinical symptoms of connective tissue disease (CTD). However, the clinical manifestations of anti-KS-positive patients are not well-studied.

Objectives: To clarify the clinical and laboratory characteristics of Japanese patients with antibodies against anti-KS antibody.

Methods: Sera from 326 patients with CTD (including PM/DM) or ILD at Tokai University were screened for anti-KS antibody using RNA and protein immunoprecipitation assays. Demographic data, clinical symptoms, laboratory findings and chest computed tomography (CT) scan results were retrospectively reviewed from medical charts.

Results: Five patients with anti-KS autoantibody were identified. All five patients were female with a mean age ($\pm$ SD) of 59.4 ($\pm$ 13.9) years at onset, presenting with respiratory symptoms without any sign of myositis. In all patients, ILD was chronic and chest CT scan revealed a non-specific interstitial pneumonia pattern in three patients and the remaining two showed a usual interstitial pneumonia pattern. Three patients (60%) had arthritis, mechanic's hand, while one (20%) had Gottron's sign and was diagnosed as amyopathic DM with ILD. Interestingly, three patients (60%) showed symptoms of the Sicca syndrome with presence of anti-SSA antibody and two of those were diagnosed as Sjögren's syndrome. The frequency of Sicca syndrome in anti-KS-positive patients was significantly higher compared with other anti-ARS antibody-positive patients (60% vs. 14%, respectively, $p=0.031$).

Conclusions: These results highlight that the presence of anti-KS antibody is associated with the Sicca syndrome as well as ILD without muscle symptoms.

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AB0821 EARLY DETECTION OF THE CHANGES IN PULMONARY ARTERIAL PRESSURE AND VASCULAR FUNCTIONS IN SYSTEMIC SCLEROSIS: EXPLORING NON-INVASIVE CLINICAL TEST METHODS AND UNDERLYING GENE EXPRESSIONS

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Background: Pulmonary arterial hypertension (PAH) is prominent as a vascular involvement in systemic sclerosis (SSc), which remains a leading cause of death in spite of current best treatments. Although recent studies focused on early diagnosis of established PAH, it is known that more than a half of the pulmonary circulation is impaired before early PAH is detected. However, there is little study about the changes of vascular functions or the underlying gene expressions during its subclinical stage.

Objectives: I. To detect the pathological changes in pulmonary arterial pressure (PAP) and vascular functions before PAH is manifested. II. To explore the changes in its underlying gene expressions of peripheral blood.

Methods: Total of 103 cases without PAH symptoms (NYHA I) with either Raynaud phenomenon (RP: $n=87$), skin sclerosis ($n=65$) or SSc-related autoantibody ($n=68$) were enrolled. To detect the pathological change of PAP, exercise Doppler echocardiography was carried out, and exercise induced pulmonary hypertension (exPH) group was segregated from normal response group (exN) with using the definition described in R. Naeije *et al.*¹⁾ Vascular function was evaluated with thermography after 0°C-stress and determination of ankle-brachial index (ABI) and cardio-ankle vascular index (CAVI). Furthermore, reactive hyperemic index (RHI), augmentation index (AI) and second derivative of photoplethysmogram ageing index (SDPTGAI) were assessed with using EndoPAT. Micro-vascular changes were also recorded with nailfold videocapillaroscopy. Meanwhile, genome-wide gene expression analysis was performed with using whole peripheral blood. The genes correlated with each vascular function tests were analysed by weighted gene co-expression network analysis (WGCNA) and pathway enrichment analysis (PathVisio).

Results: There were significant differences between exPH and exN group in the result of thermography after 0°C-stress test, CAVI and AI normalised to heart rate of 75bpm (AI@75bpm). As the CAVI and AI are known to correlate positively with age, careful interpretation was necessary because the mean age of exPH group was higher as compare with exN group (69.05 $\pm$ 11.04 vs. 60.23 $\pm$ 14.73). However, the fact that recovery of blood-flow from RP was significantly delayed in exPH group suggested the additional pathological changes of vascular and endothelial functions. Gene expression analysis revealed that several mutual pathways such as "type2 interferon signalling", "oxidative damage" and "fatty acid omega oxidation" seemed to underlie some vascular changes.

Conclusions: The results of vascular function tests including thermography after 0°C-stress, CAVI and AI@75bpm were significantly different between exPH and exN group. On the other hand, gene expression analysis showed that many factors such as ageing, arteriosclerotic and immunological mechanisms were involved in the changes of these vascular functions. Although further prospective study is required to select appropriate set of the tests, it is possible that evaluation of these vascular functions may be useful as a non-invasive test to assess the pulmonary vascular disease before PAH is manifested.

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AB0822 A MEASUREMENT OF ANTI-ARS ANTIBODIES, ANTI-MI-2 ANTIBODY, ANTI-TIF1 GAMMA ANTIBODY AND ANTI-MDA5 ANTIBODY BY ENZYME-LINKED IMMUNOSORBENT ASSAY AS A DIAGNOSTIC TOOL OF IDIOPATHIC INFLAMMATORY MYOPATHY AT RHEUMATOLOGY DAILY PRACTICE

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Background: Enzyme-linked immunosorbent assay (ELISA) tests of anti-ARS antibodies, anti-Mi-2 antibody, anti-TIF1 γ antibody and anti-MDA5 antibody

became available at daily clinical practice on ^{Oct. 2016} in Japan. Diagnostic accuracy study of myositis-specific autoantibodies (MSAs) except for anti-Jo-1 antibody in daily clinical practice is scarce^[1].

Objectives: This study investigated the diagnostic utility of newly available myositis-specific autoantibodies (ELISA) in idiopathic inflammatory myopathy (IIM) suspected patients at a rheumatology clinic with muscle and skin biopsy as a reference standard.

Methods: This study is a retrospective, cross-sectional study. The electrical medical records of patients who visited the department of rheumatic diseases at ^{Tokyo} Metropolitan Tama Medical Centre between Nov. 2016 and Aug. 2017 were retrospectively searched for clinical variables and laboratory data. Patients with serum creatine phosphokinase (CK) greater than two-fold of normal upper limit (over 300 IU/ml) or suspected dermatomyositis skin rash were included. Anti-ARS antibodies, anti-Mi-2 antibody, anti-TIF1 γ antibody and anti-MDA5 antibody were measured by ELISA^[2-4]. Two muscle pathologists interpreted muscle biopsy. The skin biopsies with interface-dermatitis were considered positive. The biopsy results of patients who did not have biopsy procedure but whose symptoms and signs improved without immunosuppressive therapy were assumed negative. IIM diagnosis was based on 2017 EULAR/ACR classification criteria^[5]. We calculated likelihood ratio of MSAs for biopsy results and IIM diagnosis.

Results: Eight hundred and eighty-six new patients visited an outpatient clinic of rheumatic disease department and 38 patients met inclusion criteria. 14 patients were excluded because of apparent causes (1 each for sepsis, rhabdomyolysis, macro CK, serotonin syndrome, motor neuron disease, statin myopathy, SSc, idiopathic IP, seborrheic dermatitis, 3 for symptoms not compatible with IIMs.) and no biopsy results (2 patients). 17 (71%) of 24 patients analyses were female. The median age was 56.5 (IQR 41–65) years, median CK 658 (IQR 105–1866) IU/ml. 11 (46%) patients had the positive biopsy result (6 muscle biopsy and 5 skin biopsy) and IIM diagnosis. 10 (42%) patients were positive for MSAs (anti-ARS antibodies in 3, anti-Mi-2 antibody in 1, anti-TIF1 γ antibody in 2, anti-MDA5 antibody in 3 and double positive for anti-ARS and anti-MDA5 antibody in 1). The positive likelihood ratio of MSAs for the biopsy result and IIM diagnosis was ∞ (95% CI cannot be calculated). The negative likelihood ratio was 0.091 (95% CI 0.086–0.097).

Conclusions: Our analysis demonstrated that MSAs measured by ELISA efficiently differentiated biopsy-proven IIMs from other medical conditions at rheumatology daily practice.

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AB0823

PATIENTS WITH CONNECTIVE TISSUE DISEASE HAVE MORE ANXIETY IN COMPARISON WITH PATIENTS WITH OTHER RHEUMATIC DISEASES

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Background: Rheumatic diseases often coexists with psychological disorders. Psychological symptoms not only have a substantial negative impact on the quality of life, but also on the course and outcome of the chronic disorders.¹

Objectives: Our study was to investigate psychological disorders in Chinese patients with rheumatic diseases and to compare the differences of psychological disorders among different diseases.

Methods: Patients with rheumatic diseases were enrolled from July to December in 2017 in rheumatology department of the Third Affiliated Hospital of Sun Yat-sen University. Participants were required to complete a set of questionnaires and examinations, including demographic and clinical information, Zung self-rating anxiety scale (SAS), and Zung self-rating depression scale (SDS). The Statistical Package for Social Sciences (SPSS) software version 21 was used for all data management and analysis. The Mann-Whitney U test and Student's t test were used to compare patients with different rheumatic diseases and psychological disorders. Stepwise linear multiple regression was used to determine the contributors to SAS and SDS.

Results: Of all the 153 patients, 49 (32%) were male patients. 14.4% had only primary education, while 32% received education in university. Numbers of the patients were stated as follows. Lupus, 44; rheumatoid arthritis, 11; Sjogren's syndrome, 11; systemic sclerosis, 9; myositis, 7; vasculitis, 6; spondyloarthritis, 9; gout, 10; other diseases 46. Mean age was 37.64 $\pm$ 15.23 years. Mean disease

duration was 3.78 $\pm$ 5.07 years. Mean SAS scores were 43.44 $\pm$ 8.51, and mean SDS scores were 46.66 $\pm$ 12.43. 35 (22%) of the patients had anxiety, while 6 (3.9%) had moderate anxiety. 50 (32.7%) patients suffered from depression, of which 5 (3.3%) had severe depression, and 13 (8.5%) had moderate depression. Patients with connective tissue diseases had more anxiety (30.5%) than average score ($p < 0.05$), especially in patients with lupus. SDS ($B = 1.073$, $p < 0.001$), educational scale ($B = -2.147$, $p < 0.05$) contributed to SAS scores.

Conclusions: Psychological disorders could concur with rheumatic diseases, especially in connective tissue diseases. Physicians should be aware of psychological status in these patients.

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AB0824

EVALUATION OF THE DETECTION OF MYOSITIS-SPECIFIC ANTIBODIES BY LINEBLOT AS A TOOL FOR THE CHARACTERISATION OF A PROSPECTIVE MULTICENTER COHORT

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Background: Traditionally, the diagnosis of idiopathic inflammatory myopathies (IIM) is based on clinical findings, muscle enzyme levels, electromyography and biopsy. Newer approaches include the detection of myositis-specific autoantibodies (MSA), with MSA identified in about 50% of patients.

Today, several commercial platforms for MSA detection have become commercially available. In the context of the start-up of a prospective multicentre cohort, we evaluated the usefulness of a multiparameter line blot for the serological characterisation of patients with clinical suspicion of IIM.

Objectives: To evaluate the usefulness of a multiparameter line blot for the serological characterisation of patients with clinical suspicion of IIM.

Methods: A total of 225 samples (118 consecutive routine samples for which MSA were requested and 107 controls [50 systemic sclerosis (SSc), 29 systemic lupus erythematosus (SLE) and 28 rheumatoid arthritis patients (RA)]) was analysed on indirect immunofluorescence (HEp-2000, Immunoconcepts) and lineblot (MYO12 blot, D-Tek). All consecutive samples were retrospectively categorised by the treating medical specialist as definite IIM ($n = 16$), probable IIM ($n = 10$), immune mediated inflammatory disease (IMID) – myositis overlap not excluded ($n = 28$), myopathic features without IIM ($n = 1$), IIM excluded ($n = 44$), and lost from follow-up/no data available ($n = 19$). Data to calculate the EULAR/ACR probability score were also collected.

Results: MSA were detected in 12% of the 225 samples, showing no multiple reactivities. A sensitivity of 50% in the IIM patients (definite and probable $n = 13/26$ – based on judgement of the clinician) was observed, with the highest frequencies observed for anti-HMGCR (15%, $n = 4$), anti-Mi2 (12%, $n = 3$) and anti-Jo1 (12%, $n = 3$). Less frequent antibodies were anti-EJ ($n = 1$), anti-MDA5 ($n = 1$) and anti-NXP2 ($n = 1$). All these patients had a MSA-compatible clinical IIM subtype. The median minimal EULAR/ACR probability score was 97% (range 6%–100%, no biopsy data entry), with the lowest score observed in the anti-EJ patient. In contrast, MSA were also observed in 8 patients with no convincing clinical diagnosis of IIM or myositis overlap syndrome (anti-TIF1 γ in 4 [3 SSc and 1 SLE]; anti-SRP in 1 [1 RA]; anti-SAE in 3 [1 SSc, 1 RA and 1 IIM excluded], mostly showing low antibody titres ($n = 6/8$) and no compatible immunofluorescence pattern on HEp-2000 ($n = 7/8$). For 2 other positive samples the results were not conclusive (anti-PL7 in patient with myopathic features and anti-SRP in an unspecified IMID patient).

Conclusions: The detection of MSA by lineblot will be useful for the prospective serological characterisation of patients with clinical suspicion of IIM. Nevertheless, careful interpretation in correlation with the clinical findings and other technical examinations is necessary in case of low titres and absence of a compatible immunofluorescence pattern.

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