Conclusions: This proof-of-principal study demonstrates that DNN processing of stained dermal biopsy sections are sensitive to clinically relevant features of SSc skin. These results suggest that DNNs dramatically expand the quantifiable SSc phenome and that histological samples can now be incorporated into models of SSc. Moreover, our results indicate that the gene expression underlying SSc may be driving histological differences in SSc skin.

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THU0388

DIFFERENCES IN CLINICAL COURSES AND SERUM MARKERS OF INTERSTITIAL LUNG DISEASE ASSOCIATED WITH ANTI-AMINOACYL-TRANSFER RNA SYNTHETASE ANTIBODY AND ANTI-MELANOMA DIFFERENTIATION-ASSOCIATED GENE 5 ANTIBODY-POSITIVE POLYMYOSITIS/DERMATOMYOSITIS

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Background: Polymyositis/dermatomyositis (PM/DM) is a chronic autoimmune disease that is often complicated by interstitial lung disease (ILD). Anti-aminoacyl-transfer RNA synthetase antibody (ARS-Ab) and anti-melanoma differentiation-associated gene 5 antibody (MDA5-Ab) are highly detected in PM/DM with ILD. It was reported that ARS-Ab-positive-ILD (ARS-ILD) is often recurrent¹, and patients with MDA5-Ab-positive-ILD (MDA5-ILD) develop fatal rapidly progressive ILD².

Objectives: To evaluate the differences in clinical courses between ARS-ILD and MDA5-ILD, including the changes in serum ILD markers.

Methods: We retrospectively investigated 25 patients with ARS-ILD and 26 patients with MDA5-ILD who received induction therapy between April 2002 and September 2017 at Kobe University Hospital. The survival rate and relapse-free survival rate were analysed with Kaplan-Meier estimation and the log-rank test. The differences in serum ILD markers between patients with ARS-ILD and MDA5-ILD were evaluated with the Student's t-test.

Results: Disease subtypes at diagnosis with PM/DM-associated ILD were as follows: Eleven ARS-ILD and no MDA5-ILD patients had PM, 10 ARS-ILD and 5 MDA5-ILD patients had DM, and 4 ARS and 21 MDA5 patients had amyopathic DM. The survival rate for MDA5-ILD was significantly lower than that for ARS-ILD ($p < 0.01$, figure 1A). On the other hand, there was no significant difference in the relapse-free survival rate between ARS-ILD and MDA5-ILD ($p = 0.25$, figure 1B). The serum level of Krebs von den Lungen-6 was not significantly different between ARS-ILD and MDA5-ILD (1044.3 ± 768.1 U/ml in ARS-ILD vs $1044.863.1 \pm 566.5$ U/ml in MDA5-ILD, $p = 0.33$), but the serum level of ferritin was significantly higher in MDA5-ILD than in ARS-ILD (286.4 ± 422.3 ng/ml in ARS-ILD vs 696.2 ± 839.5 ng/ml in MDA5-ILD, $p = 0.04$). Although the serum level of surfactant protein D (SP-D) in ARS-ILD was high, the SP-D level in MDA5-ILD was normal (158.9 ± 82.1 ng/ml in ARS-ILD vs 46.3 ± 22.1 ng/ml, $p < 0.01$).

Conclusions: MDA5-ILD patients should be monitored for both rapidly progressive disease and relapsing disease. A normal SP-D level is a feature of MDA5-ILD.

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THU0389

BORDERLINE PULMONARY HYPERTENSION WAS ASSOCIATED WITH REDUCED CARDIAC OUTPUT DURING EXERCISE IN PATIENTS WITH CONNECTIVE TISSUE DISEASES

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Background: In patients with systemic sclerosis (SSc) borderline mean pulmonary arterial pressures (mPAP; 21–24 mmHg at rest) are a frequent finding and could represent an intermediate stage between normal pulmonary pressures and manifest pulmonary hypertension (PH).¹

Objectives: The objective of this prospective study was to compare right ventricular function and pulmonary arterial compliance (PAC) at rest and during exercise between systemic sclerosis (SSc)-patients with normal and borderline mean pulmonary artery pressures, respectively.

Methods: SSc-patients (n=112) underwent clinical assessment, including right heart catheterization at rest and during exercise and were divided in three groups according to their resting mPAP values: normal mPAP (≤ 20 mmHg), borderline mPAP (21–24 mmHg) and manifest pulmonary hypertension (PH, mPAP ≥ 25 mmHg). Results were compared between groups by ANOVA followed by post-hoc student's t-test.

Results: SSc Patients with borderline PAP showed significantly lower cardiac index (CI) increase during exercise and higher PVR values than SSc patients with normal PAP at rest. Six-Minute-walking distance (6MWD) and PAC (stroke volume/(systolicPAP-diastolicPAP)) were significantly lower in the borderline PAP group compared to patients with normal PAP.