Methods: Twenty-six patients with SLE, who had been referred to Niigata University Hospital between 2004 and 2017 and diagnosed as having LN by percutaneous kidney biopsy, were recruited for this study. Flow-cytometric analysis of urinary inflammatory cells was performed for each subject at the time of admission for kidney biopsy. Numbers of urinary T cells or macrophages were determined by multiplying the number of viable cells in the gated mononuclear cell region in each sample by the percentage of urinary CD3-positive or CD14-positive cells in the population, respectively. The histopathological findings of kidney biopsy specimens for each subject were classified according to the 2003 International Society of Nephrology/Renal Pathology Society (ISN/RPS) classification of LN. In addition, they were also evaluated for glomerular and interstitial lesions such as mesangial cell/matrix proliferation, endocapillary hypercellularity, crescent formation, and interstitial cell infiltration and fibrosis. The severity of each lesion was scored from 0 to 4, and the numbers of urinary inflammatory cells were analysed by Spearman's rank correlation coefficient to determine the relationships among these scores. Next, the patients were divided into two groups according to whether the total number of urinary inflammatory cells was significantly increased (more than 120 cells/ml; positive group, n=12) or not (negative group, n=14), and the severities of the histopathological lesions were compared between the two groups.

Results: Disease severity in terms of the ISN/RPS classifications was positively correlated with the severity of mesangial cell and matrix proliferation, cellular crescent formation, and duplication of the glomerular basement membrane. In addition to these four factors, the numbers of CD3-positive and CD14-positive cells were also positively correlated with adhesion, endocapillary hypercellularity, interstitial inflammatory cell infiltration, and interstitial fibrosis. The severity scores for adhesion, endocapillary hypercellularity, cellular and fibrous crescent formation, duplication, and interstitial cell infiltration were significantly higher in the positive group than in the negative group.

Conclusions: Analysis of urinary immune cells reflects the histopathological features of kidney biopsy specimens from patients with LN.

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THU0386

DIFFERENCE OF IMAGE FEATURES ON COMPUTED TOMOGRAPHY BETWEEN LUPUS ENTERITIS AND MESENTERIC VASCULITIS OF OTHER CONNECTIVE TISSUE DISEASES

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Background: Lupus enteritis (LE), lupus mesenteric vasculitis, occurs occasionally in systemic lupus erythematosus (SLE). Not only LE but also other mesenteric vasculitis can lead to bowel haemorrhage or perforation and will be fatal, but it is difficult to demonstrate histologically-proven vasculitis on endoscopic biopsy.¹ Although several computed tomography (CT) features of LE are reported, we have little knowledge about whether they differ from those of other mesenteric vasculitis.

Objectives: To clarify the imaging pattern on CT which can distinguish LE from other mesenteric vasculitis.

Methods: Patients diagnosed with LE and non-LE at our hospital were consecutively registered from January 2009 to August 2017. The diagnosis of LE was made by the criteria of ^{BILAG 2004} which is defined as either vasculitis or inflammation of small or large bowel with supportive imaging and/or biopsy findings.² Non-LE was defined as mesenteric vasculitis of other connective tissue diseases (CTDs). We compared the contrast-enhanced CT patterns of LE with non-LE. Statistical analyses were performed using XLSTAT.

Results: A total of 8 patients were diagnosed with LE and enrolled in this study. The mean age was 41.1 years old, range 23–53, and 7 were females. CT exams of all 8 patients demonstrated small bowel wall thickening, dilatation of intestine and comb sign (indicating engorgement of mesenteric vessels). Severe bowel wall thickening (>8 mm) was observed in 87.5% (n=7), ascites in 75.0% (n=6) and target sign (indicating abnormal bowel wall enhancement) in 62.5% (n=5). 5 patients were enrolled in non-LE (1 eosinophilic granulomatosis with polyangiitis, 1 IgA vasculitis and 3 Behcet's disease). Comparison of CT findings between LE patients and non-LE patients were summarised on table 1. Bowel wall thickness and comb sign were common in both groups, however the prevalence of dilatation of intestine in LE patients was significantly higher than in non-LE patients. Although not significant, complication of large bowel involvement was shown only in LE patients.

Abstract THU0386 – Table 1

	LE(n=8)	non-LE*(n=5)	p-value
Thickening bowel wall	100%	100%	—
Large bowel involvement	50.0%	0%	0.10
Asymmetrical patterns of involvement	12.5%	60.0%	0.21
Dilatation of intestine	100%	20.0%	0.0070
Ascites	75.0%	40.0%	0.29
Target sign	62.5%	40.0%	0.59
Comb sign	100%	100%	—

*non-LE : 1 Eosinophilic granulomatosis with polyangiitis, 1 IgA-vasculitis, 3 Behcet's disease

*non-LE : 1 Eosinophilic granulomatosis with polyangiitis, 1 IgA-vasculitis, 3 Behcet's disease

Conclusions: Dilatation of intestine on CT is significant to distinguish LE from mesenteric vasculitis of other CTDs.

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THURSDAY, 14 JUNE 2018:

Scleroderma, myositis and related syndromes

THU0387

HIGH-THROUGHPUT QUANTITATIVE HISTOLOGY IN SYSTEMIC SCLEROSIS SKIN DISEASE USING COMPUTER VISION

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Background: There are no validated systems to quantify dermal architecture of skin biopsy sections in systemic sclerosis (SSc). Significant advances in computer vision, called deep neural networks (DNNs), have demonstrated human-level pattern recognition abilities using mathematical transformations of images into millions of quantitative features.¹ Publicly available DNN algorithms have the potential to radically augment current histological analyses via robust, reproducible, and high-throughput image quantification.

Objectives: Apply publicly available DNN algorithms to trichrome-stained sections of dermal biopsies from patients with SSc to identify quantitative features of images that correlate with clinical skin fibrosis assessment via the validated modified Rodnan skin score (mRSS) and skin gene expression.²

Methods: One rheumatologist performed local mRSS (lmRSS) assessments (0–3) and two dermal punch biopsies for 7 patients with SSc and one healthy control at baseline and 6, 12, 24, and 36 months. One biopsy was paraffin embedded, sectioned, stained with trichrome, and photomicrographed. Images were transformed into high-level quantitative features using AlexNet and the Matlab Neural Network Toolbox.³ Correlations between quantitative features and lmRSS were determined with Bonferroni-Holm correction. One biopsy underwent gene expression profiling by DNA microarray. The degree of correlation between each gene and lmRSS was assessed and a functional gene network was determined using the GIANT database.⁴

Results: We identified 90 quantitative features that correlated significantly with local skin score (p<0.05, Bonferroni-Holm correction). Using these features, biopsies sorted into three clusters corresponding to low (mean=0.13), intermediate (mean=0.91), and high (mean=1.7) lmRSS. Gene expression for 488 genes in the biopsies correlated significantly with quantitative features (p<0.05, Bonferroni-Holm correction). Among these 488 genes, 185 genes formed a large functional network in the GIANT database including genes associated with the cell cycle, apoptosis, IL12 signalling, and wound healing such as CD44, THBS1, CAV1, and VEGFA.

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.