Background: 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 recurrent 1, and associated gene 5 antibody (MDA5-Ab) are highly detected in PM/DM with ILD. Clinical Immunology, Kobe University Graduate School of Medicine, Kobe, Japan

Results:

Methods: We retrospectively investigated 25 patients with ARS-ILD and 26 MDA5-ILD patients who received induction therapy between 1999 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.

Conclusions:

REFERENCES:

Acknowledgements: none.
Disclosure of Interest: None declared


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

REFERENCES:

Acknowledgements: NIH NIAMS P30 AR057216; NIAMS K23 AR059763; L30 AR054311 and Scleroderma Research Foundation. Chase Correia is supported NIH NIAMS P30 AR057216; NIAMS K23 AR059763l, L30 AR054311.

ACKNOWLEDGEMENTS:

REFERENCES:


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 POLYMYSITIS/DERMATOMYSITIS

K. Akashi, Y. Nose, T. Shirai, Y. Fukushima, T. Nagamoto, T. Okano, S. Takahashi, S. Sendo, A. Onishi, J. Saeugsa, A. Morinobu. Department of Rheumatology and Clinical Immunology, Kobe University Graduate School of Medicine, Kobe, Japan

Background: Polymyositis/dermatomyositis (PM/DM) is a chronic autoimmune disease that is often complicated by interstitial lung disease (ILD). Anti-aminocyl-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 recurrent 1, and patients with MDA5-Ab-negative-ILD (MDA5-ILD) develop fatal rapidly progressive ILD 2.

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 1999 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:

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 phome 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.

REFERENCES:


Acknowledgements: none.
Disclosure of Interest: None declared


THU0389

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

A. Marra 1, C. Nagel 1, B. Egenlauf 1, P. Xanthouli, S. Harutyunova 1, N. Blank 2, H.-M. Lorenz 1, C. Fiehn 1, N. Benjamin 1, C. Fischer 1, E. Bossona 3, A. Cidditani 1, A. Grüning 1, 1Pneumology, Thoraxklinik Heidelberg, 2Rheumatology, University Hospital, Heidelberg, Heidelberg, Germany, 3Kardiology, University Hospital, Salerno

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 pulmonar...
Conclusions: The results of this study suggest that impaired 6MWD in SSc-patients with borderline PAP (and normal RV function at rest) might be caused by reduced RV contractile reserve (reduced RV output) and reduced PAC during exercise rather than by elevated pressures in the borderline range alone. These findings give further evidence for borderline PAP being an early stage of pulmonary vascular disease.

Acknowledgements: Special thanks to the patients that participated in this research.

Disclosure of Interest: None declared


THU0390 PATIENTS WITH SYSTEMIC SCLEROSIS DEVELOP FOCAL FIBROSIS OVER TIME, AND INCREASED ECV, DIFFUSE FIBROSIS SEEN IN POOR PROGNOSTIC GROUPS: A FIRST LONGITUDINAL CARDIAC MRI STUDY

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Background: Subclinical SSc-cardiomyopathy is described in up to 2/3 of the patients when sensitive methods such as cardiac MRI(CMR) are used. The prognostic implications and the natural history of these findings is unknown.

Objectives: To evaluate in SSc patients, free of cardiovascular (CV) risk factors and CV disease (CVD) the prevalence, clinical association and natural history of CMR abnormalities over 3 years.

Methods: 35 SSc patients, fulfilling the ACR/EULAR criteria, with no CVD, diabetes and ≤1 CV risk factor had 2 CMRs, 3 years apart. A 3T CMR with late gadolinium enhancement (LGE), T1 mapping for extracellular volume (ECV) quantification and stress perfusion (data available later) was undertaken. Initial CMR was compared with CMR results of 30 healthy controls (HC).

Results: 35 pts had an initial CMR. 26/35 (74%) female, mean(SD) age 55(10), 15 (43%) dcSSc, 11 (31%) ACA+, 12 (34%) Scl70+, 15 with (43%) interstitial lung disease (ILD) and 13 (37%) with history of digital ulcers(DU), 21 (60%) received any DMARD over the 3 year period and 10 (29%) prior treatment with cyclophosphamide. The first CMR(CMR1) of 35 pts vs HC showed higher ECV% values and comparable left ventricle(LV) volumes(table 1). LGE was present in 9/22 pts vs 1/30 HC. 22/35 pts had a second CMR(CMR2) at year 3(Y3). A further 5 pts had evidence of LGE, total of 14 (40%): 7/14 had dcSSc, 6/14 males, 2 ACA positive, 6 Scl70 positive. LGE distributed in the basal and mid segments, mainly in a linear or patchy pattern. Of those with LGE at CMR1 (5/9 pts with LGE on CMR1 had CMR2) no change in the LGE pattern at CMR2 was observed. None of the initial CMR measures associated with LGE development at Y3 (p=0.05). Whilst ECV had an overall decrease, ECV increased in patients with ILD (mean diff.(CI) 3 (-1,6), p=0.14) and in those with higher mRSS at baseline(r=0.455, p=0.04). A significant decrease over the 3 years was observed in LV end-diastolic volume (LVEDV/BSA), LV end-systolic volume(LVESV/BSA) and left ventricular stroke volume (LVSV/BSA) (table 1). A decrease in LVEDV/BSA was noticed for those with a history of DU(mean diff.(CI) –5(–12,22), p=0.01). ILD(mean diff.(CI) –6(–12,05), p=0.07) and shorter disease duration(=r=0.504, p=0.02).

Abstract THU0390 – Table 1

<table>
<thead>
<tr>
<th>CMR variable</th>
<th>HC mean (SD)</th>
<th>SSc patients mean (SD)</th>
<th>SSc patients change (%CI) in CMR between SSc patients</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>LGE</td>
<td>3% (35/100)</td>
<td>21% (7/35)</td>
<td>-23% (CI -35, 11)</td>
<td>0.003</td>
</tr>
<tr>
<td>ECV%</td>
<td>25(94)</td>
<td>30(94)</td>
<td>0.2 (CI 0, 0.4)</td>
<td>0.053</td>
</tr>
<tr>
<td>T1 native</td>
<td>1202 (1243)</td>
<td>1199 (1423)</td>
<td>47 (CI 1, 94)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>LVEDV/BSA (ml/m²)</td>
<td>80 (78/16)</td>
<td>71 (15)</td>
<td>-7 (CI -10.3, 0)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>LVESV/BSA (ml/m²)</td>
<td>31 (30/9)</td>
<td>26 (9)</td>
<td>-4 (CI -6.2)</td>
<td>0.001</td>
</tr>
<tr>
<td>LVSV/BSA (ml/m²)</td>
<td>49 (48/9)</td>
<td>45 (8)</td>
<td>-3 (CI -6.0)</td>
<td>0.028</td>
</tr>
<tr>
<td>Lymass/BSA (g/m²)</td>
<td>49 (44/13)</td>
<td>50 (21)</td>
<td>6 (CI 2.3)</td>
<td>0.129</td>
</tr>
<tr>
<td>LVEF(%)</td>
<td>62(5)</td>
<td>62(5)</td>
<td>2 (CI 0, 4)</td>
<td>0.08</td>
</tr>
<tr>
<td>Distensibility (10⁻⁶mmHg⁻¹)</td>
<td>5 (2)</td>
<td>5 (3)</td>
<td>0 (CI 1.2)</td>
<td>0.491</td>
</tr>
<tr>
<td>Torsion</td>
<td>15(4)</td>
<td>13(4)</td>
<td>0 (CI 2.3)</td>
<td>0.828</td>
</tr>
</tbody>
</table>

Conclusions: This longitudinal CMR study in SSc patients demonstrates that CMR is sensitive to change over time. More individuals developed LGE (of focal fibrosis) despite immunosuppressive treatment, and ECV% (diffuse fibrosis) appeared to worsen in a poor prognostic group. Functional changes were also observed. These data justify larger studies to inform stratification strategy for CMR in SSc, and also provide new insights for further investigation.

Disclosure of Interest: None declared


THU0391 CLINICAL VALUE OF COMPUTED TOMOGRAPHY FOR THE DIAGNOSIS OF ESOPHAGEAL DYSMOTILITY IN SYSTEMIC SCLEROSIS

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Background: Esophageal dysmotility is common in Systemic Sclerosis (SSc), affecting 50%-80% of patients, usually associated with poor prognosis. SSc leads to atrophy and fibrosis of the smooth muscle of the oesophagus, modifying peristaltic contractions and motility. Manometry is considered the gold standard for the diagnosis of esophageal motility disorders, but dilatation can also be observed with computed tomography (CT), even if its diagnostic validity is still unknown.

Objectives: To compare esophageal dilatation observed with CT to manometry, in patients with SSc and to confirm whether CT can be used in the assessment of esophageal dysmotility.

Methods: Forty six patients meeting the 2013 ACR/EULAR Classification Criteria for SSc, and 33 healthy controls were included and retrospectively studied. Patients with overlapping syndromes, active infections or with longstanding diabetes were excluded. Epidemiological and clinical data were collected from medical records. All patients and controls had undergone at least one manometry and one CT, requested in daily clinical practice for another purpose. The most recent records. All patients and controls had undergone at least one manometry and one CT, requested in daily clinical practice for another purpose. The most recent manometry (aperistalsis, inefficient peristalsis, nonspecific dysmotility and normal motility). Manometry is considered the gold standard for the diagnosis of esophageal motility disorders, but dilatation can also be observed with computerised tomography (CT), even if its diagnostic validity is still unknown.

Objectives: To compare esophageal dilatation observed with CT to manometry, in patients with SSc and to confirm whether CT can be used in the assessment of esophageal dysmotility.

Results: The sample included 76 women (10 dcSSc, 28 lcSSc, 3 MCTD, 35 controls) and 23 men (3 dcSSc, 2 lcSSc, 18 controls). Esophageal dysmotility was seen in 40/46 patients with SSc (87%) by manometry (defined as inefficient peristalsis or aperistalsis). Esophageal dilation (≥10 mm) was present proximally in 23/44 patients (52.3%), distally in 35/46 patients (76.1%), and near the carina in 26/44 patients (59.1%). Esophageal dilatation at any level was statistically associated with esophageal dysmotility (p=0.05). The areas under the ROC curves (figure 1) suggest that the esophageal proximal diameter in the coronal plane is good for detecting esophageal dysmotility (0.798, 95% CI 0.705–0.890), with the distal diameter (0.759, 95% CI 0.661–0.857) and the carinal diameter (0.712, 95% CI