targeting the U1 small nuclear ribonucleoprotein particle (U1RNP), and various clinical features of other connective tissue diseases (1-2). Interstitial lung disease (ILD) is an established complication of the disease that is suspected to affect morbidity and mortality (3). However, little is known about MCTD-associated ILD (MCTD-ILD) phenotype including first presentation, outcomes and predictive factors for progression.

Objectives: To compare two distinct populations of MCTD patients with and without associated ILD and to identify predictive factors for lung progression and severity.

Methods: International multicenter retrospective study (12 tertiary hospitals). To be included, patients were required to fulfill at least one MCTD international classification criteria (4). ILD was defined by the presence of typical chest high-resolution computed tomography (HRCT) abnormalities. Patients were divided into two groups: with or without ILD, at a ratio of 1:1 and matching on disease duration (+/- 2 years).

Results: 300 patients were included. Mean age at MCTD diagnosis was 39.7±15.4 years and 191 (63.7%) were women. At baseline, we identified several variables associated with the presence of ILD: older age (42.2 vs 37.5 years, p=0.01), scleroderma-like phenotype (38.7 vs 27.3%, p=0.03), upper gastro-intestinal (GI) symptoms (54.7 vs 30.7%, p<0.001), forced vital capacity (FVC) <80% (62.4 vs 74.8%, p<0.001), anti-topoisomerase antibodies (6 vs 0 patients, p=0.01), SSA/RO antibodies positivity (29.3 vs 19.3%, p=0.02), cryoglobulinemia (5.3 vs 13.4%, p=0.04) and elevated C-reactive protein (CRP) >5mg/L (54.7 vs 28.7%, p<0.001). Among the previous variables older age (OR 1.03, 95% CI 1.01 to 1.05), upper GI symptoms (OR 1.92, 95% CI 1.03 to 3.58) and CRP >5mg/L (OR 6.77, 95% CI 2.94 to 26.22) remained significantly associated with the presence of ILD by multivariate analysis. Patients with MCTD-ILD were more likely to be treated with immunosuppressive agents (68.7 vs 49.3%, p<0.001) including mycophenolate mofetil (MMF) (7.3 vs 13.1%, p=0.03).

Conclusion: In this large international cohort of patients with MCTD, we identified several factors associated with ILD development. Our findings highlight a high risk of mortality in MCTD-ILD patients and that digital ulceration seems to be at risk of more progressive ILD.

REFERENCES:

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Results: The median (IQR) age of the patients was 52.0 (43.5-58.5) years and 60 (75.0%) were women. Baseline median score of whole lung-QILD and most severe zone-QILD were 28.1% (19.1-43.9) and 68.0% (45.5-81.8), respectively, and QILD score showed significant correlations with pulmonary function tests (r=0.349, p=0.002 for % predicted forced vital capacity; and r=-0.381, p=0.001 for % predicted diffusing capacity for carbon monoxide). The individual time-estimated yearly ∆QILD score between first 2 visits presented that approximately half of the patients showed improvement or stability in QILD scores; however, when patients were sorted by visual assessment in IILD subtype on HRCT, approximately two-thirds of the patients with usual interstitial pneumonia (UIP) pattern were aggrandized in QILD scores and less than half of subjects with nonspecific interstitial pneumonia and organizing pneumonia were aggrandized (Figure 1, 80% for UIP vs. 44.4% for non-UIP, p=0.013). There was no immunosuppressive drugs related to meaningful improvement in QILD scores during first 2 visits. Notably, we observed significant aggrandization of QILD scores in tacrolimus users (n=7, median time-estimated whole lung-yragrand yearly ∆QILD -1.2 (8.3-6.5)). Among 80 patients, 6 (7.5%) were died due to various lung complications. Higher baseline QILD scores were noted in deaths (median whole lung-QILD 45.4 (32.9-56.5)) than in survivors (median whole lung-QILD 26.9 (19.0-42.4)), albeit not significant (p=0.084). Poor survival rate was observed in patients with high grade of ground glass opacity by visual assessment in right upper lobe (log-rank test, p=0.042). Among subgroup of patients with 3 serial HRCT scans (n=41), dynamic changes of four distinct patterns (improving, worsening, convex, and concave) were observed.

Conclusion: The changes in QILD score in IIM-IILD are dynamic and present different by visual assessment. QILD score has the potential for evaluation of the severity changes, prognosis and medication response in patients with IIM-IILD.

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Disclosure of Interests: None declared


POS0069

LONGITUDINAL CHANGES ON QUANTITATIVE CHEST HIGH-RESOLUTION COMPUTED TOMOGRAPHY IN EARLY DIFFUSE SYSTEMIC SCLEROSIS-RELATED INTERSTITIAL LUNG DISEASE AFTER AUTOLOGOUS HEMATOPOIETIC STEM CELL TRANSPLANTATION.

G. Puget1, A. Petermann2, S. Collof2, P. Otaï2, P. Lansiaux2, A. Maria4, N. Alt Abdallah2, G. Lorillot3, M. Resch-Rigon3, Z. Marjanovic2, D. Farge3. 1CHU Toulouse, Service de Médecine Inténe Immunologie Clinique, Toulouse, France; 2CHU Toulouse, Service de Radiologie, Toulouse, France; 3APHR, Hôpital Saint Louis, CRM MATHEC, Paris, France; 4CHU Montpellier, Service de Médecine Inténe, Montpellier, France; 5APHR, Hôpital Saint Louis, Service de Pneumologie, Paris, France; 6APHP, Hôpital Saint Louis, SBIM, Paris, France; 7APHR Hôpital Saint Antoine, Service d’Hématologie, Paris, France

Background: Since Burt et al. first demonstrated in the ASSIST trial (1) that lung disease high-resolution computed tomography (HRCT) volumetric measurement decreased in autologous hematopoietic stem cell transplantation (aHSCT) recipients and increased in controls receiving cyclophosphamide iv one year after starting treatment, limited data have been reported concerning the evolution of Systemic Sclerosis-Interstitial Lung disease (SSc-ILD) since and its relationship with the evolution of esophageal volume is unknown.

Objectives: To evaluate High-Resolution Computed Computed Tomography (HRCT) interstitial lung disease (ILD) and esophageal involvement in early diffuse systemic sclerosis (dSSc)-patients before and after autologous Hematopoietic Stem Cell Transplantation (aHSCT).

Methods: Overall chest HRCT, lung function and skin score changes were evaluated in 33 consecutive dSSc-patients before and after aHSCT during yearly routine follow-up between January 2000 and September 2016.Two independent radiologists blindly assessed the ILD extent using semi-quantitative Goh and Wells method (2), the widest esophageal diameter (WED) and the esophageal volume (EV) on HRCT. Patients were retrospectively classified as radiological responders or non-responders at 24 months after aHSCT according to the stability or a 5% or more decrease of HRCT IILD extent. Two by two time points comparisons after versus before aHSCT were performed using linear regressions.

Results: Twenty-four months after aHSCT, HRCT median [IQR] ILD (-2 [-10.3; 0] points, p=0.0002) and ground-grass opacities (-2.1 [-8.3; 0] points, p=0.02) extent scores had improved, with 18 patients radiological responders (probability of response 0.76, 95% CI (0.580;0.90), whereas median WED (from 24.5 [18; 29] to 28 [19; 33] mm; p= 0.005) and EV (from 19 [13; 33] to 30 [15; 58] mm2, p=0.01) increased significantly. Kaplan-Meier analyses showed a trend towards better 5 years survival rates (100% versus 60%; HR 0.23 95%CI 0.03 to 1.62, P = 0.11 among radiological responders versus non-responders at 24 months.

Conclusion: Real-world data analysis confirmed significant HRCT SSc-ILD improvement 24 months after aHSCT, although esophageal dilatation worsened requiring specific attention.

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Disclosure of Interests: None declared


POS0070

CONTINUED TREATMENT WITH NINTEDANIB IN PATIENTS WITH LIMITED CUTANEOUS SYSTEMIC SCLEROSIS (LCSSC) AND INTERSTITIAL LUNG DISEASE (ILD)

Y. Allarage1, D. Khanna2, V. Smith3, M. Aringer4, A. M. Hoffmann-Völl5, M. Kuwana6, P. A. Merkel7, A. James8, S. Sambevski9, M. Alves10, C. P. Denton11, 1Descartes University, APHC Cochin Hospital, Department of Rheumatology, A Paris, France; 2University of Michigan, Department of Medicine, Ann Arbor, MI, United States of America; 3Ghent University Hospital, Department of Rheumatology and Internal Medicine, Ghent, Belgium; 4University Medical Center and Faculty of Medicine Carl Gustav Carus, TU Dresden, Division of Rheumatology, Department of Medicine III, Dresden, Germany; 5Oslo University Hospital, Department of Rheumatology, Oslo, Norway; 6Nippon Medical School Graduate School of Medicine, Department of Allergy and Rheumatology, Tokyo, Japan; 7University of Pennsylvania, Division of Rheumatology, Department of Medicine, Philadelphia, United States of America; 8Elderbrook Solutions GmbH, Statistics, Bletchingham, Germany; 9Boehringer Ingelheim International GmbH, TA Inflammation Med, Ingelheim am Rhein, Germany; 10Boehringer Ingelheim International GmbH, TA Inflammation Med, Ingelheim am Rhein, Germany; 11Centre for Rheumatology and Connective Tissue Diseases, University College London Division of Medicine, London, United Kingdom

Background: Few data are available on the progression and management of ILD, or the management of adverse events associated with drug treatment, in patients with lcSSc. SENSICS-ON is an open-label extension trial that is collecting data on decline in forced vital capacity (FVC) and adverse events in patients treated with nintedanib over the long term. SENSCIS-ON is an open-label extension trial that is collecting data on decline in forced vital capacity (FVC) and adverse events in patients treated with nintedanib over the long term.

Objectives: To assess decline in FVC and adverse events in patients with lcSSc and IILD treated with nintedanib in SENSSIS-ON.

Methods: Patients with SSc-ILD were eligible to enter SENSSIS-ON if they completed the randomized placebo-controlled SENSSIS trial (in which patients received trial drug until the last patient reached week 52 but for ≤100 weeks) or a drug–drug interaction (DDI) study of nintedanib and oral contraceptive (in which female patients received nintedanib for ≤28 days). Among patients with lcSSc, we analysed changes from baseline in FVC and adverse events over 52 weeks of SENSSIS-ON in patients who received nintedanib in SENSSIS and continued it in SENSSIS-ON (“continued nintedanib” group) and in patients who received placebo in SENSSIS and initiated nintedanib in SENSSIS-ON or who received nintedanib for a short time in the DDI study (“initiated nintedanib” group). Analyses were descriptive.

Results: There were 98 patients with lcSSc in the continued nintedanib group and 127 patients with lcSSc (114 from SENSSIS, 13 from the DDI study) in the initiated nintedanib group. In these groups, respectively, mean (SD) FVC values