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INTERSTITIAL LUNG DISEASE IN ANTI-U1RNP SYSTEMIC SCLEROSIS PATIENTS: A EUROPEAN SCLERODERMA TRIALS AND RESEARCH (EUSTAR) ANALYSIS

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Background: Intestinal lung disease (ILD) has become the leading cause of mortality in systemic sclerosis (SSc) patients (1). The presence of anti-U1RNP antibodies is primarily associated with mixed connective tissue disease (MCTD) however these autoantibodies may be found in up to 10% of SSc patients (2). This study aims to determine if anti-U1RNP antibodies are associated with ILD in patients with SSc.

Methods: Patients positive for anti-U1RNP were compared to controls within the EUSTAR cohort from 1st January 2007 to 31st December 2018. The primary outcome of interest was mortality in SSc patients with ILD identified by HRCT, including lung involvement >20% (4.4 vs 4.2%, p=0.39), or the presence of ILD (based on EUSTAR ILD definitions) during a mean follow-up of 20.8±20.5 months. Mean expected decline in %FVC was calculated as a loss of 5-10% in %pFVC/year. Demographic and clinical parameters were compared between patients positive and negative for anti-U1RNP antibodies in univariate analysis. Associations between anti-U1RNP status and lung involvement were tested in multivariate logistic regression models. The presence of SSc-ILD using machine learning algorithms based on decision tree models was independently associated with more severe disease in SSc patients. However, so far, little is known about the influence of anti-U1RNP antibodies specifically on lung outcomes in SSc-ILD patients.

Results: To describe the clinical features, outcomes and prognosis of anti-U1RNP SSc-ILD patients.

Methods: SSc patients with available data on their autoantibody profile were identified from the EUSTAR database. Those with IBD identified by high-resolution chest tomography (HRCT) were included for analysis. Baseline demographic and disease features were compared between patients positive and negative for anti-U1RNP antibodies in univariate analysis. Associations between anti-U1RNP status and lung involvement were tested in multivariate logistic regression models. Results: A total of 5676 SSc-ILD patients were included for the analysis, among which 320 (5.6%) were positive for anti-U1RNP antibodies. Mean age was 56.4±13.5 years and 4645 (81.6%) were women. Anti-U1RNP+ SSc-ILD patients had more frequently limited cutaneous SSc (52.5% vs 43.3%, p=0.001), higher frequency of joint synovitis (20.0% vs 12.9%, p=0.001) and myositis (22.2% vs 18.2%, p<0.03) and lower incidence of scleroderma renal crisis (0.3% vs 2.0%, p=0.02). Anti-U1RNP+ patients had a baseline lower mean %FVC (81.1% vs 85.3%, p=0.002) and lower mean %predicted diffusing capacity for carbon monoxide (%DLCO) (58.9% vs 60.2%, p=0.004) than anti-U1RNP- SSc-ILD patients. Significant differences were found at baseline HRCT, including involvement >20% (4.4 vs 4.2%, p=0.39), the presence of ground glass opacities (38.1% vs 33.6%, p=0.14). A total of 2697 (50.3%) patients with SSc-ILD had at least one FVC measurement available during a mean follow-up of 20.8±20.5 months. Mean expected decline in %FVC per year was not different between U1RNP positive and negative patients (-0.21% vs -0.68%, p=0.70). Mortality or severe IBD progression was not statistically different between the two groups (35 (21.6%) vs 677 deaths (26.7%), p=0.43, and 23 (14.0%) vs 283 severe progressors (11.0%), p=0.25, respectively). Mild IBD progression was also not statistically different between the two groups (11.0% vs 10.0%, p=0.20). The proportion of patients with FVC<80% at last follow-up was similar between U1RNP positive and negative patients (50.6% vs 44.3%, p=0.13).

Conclusion: Our results from the EUSTAR database show that SSc patients positive for anti-U1RNP antibodies have more impaired baseline lung function but similar rate of progression during follow-up. This suggests that early stages might be important in RNP+ SSc-ILD patients who may require specific management and follow-up.

REFERENCES:
LUNG ULTRASOUND FOR INTERSTITIAL LUNG DISEASE DETECTION IN A COHORT OF SYSTEMIC SCLEROSIS PATIENTS: ROLE OF B-LINE AND PLEURAL LINE MODIFICATIONS

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Background: Lung ultrasound (LUS) is a technique that showed a high diagnostic accuracy for interstitial lung disease (ILD) detection in systemic sclerosis (SSc) patients and currently in progress of standardization. Traditionally, B-lines represented the finding of ILD, with the ≥10 total cut-off reported by Tardella et al., who had a diagnosis of ILD on high resolution chest computed tomography (HRCT). Recently, Fairchild et al. proposed novel diagnostic technique.

Methods: We enrolled 55 consecutive patients affected by SSc according to ACR/EULAR 2013 criteria who underwent respiratory functional tests (RFTs) during 2021, excluding smokers and those with arterial pulmonary hypertension. Twenty-four of them carried out a HRCT during a 6-months’ time. In the same time of RFTs, two certified blinded operators performed LUS for each patient applying the 4-areas technique proposed by Gutiérrez et al., looking for the presence of total ≥10 cumulative B lines and the fulfilling of Fairchild’s criteria for pleural line. A 3-13 MHz operating linear probe was used. Clinical-demographic data and ongoing therapies were collected.

Results: Among 55 total SSc patients, the agreement between the two operators for Fairchild’s criteria was almost perfect (Cohen’s kappa (κ) =0.81) and substantial for ≥10 cumulative B-lines count (κ=0.74). Fairchild’s criteria showed a higher diagnostic accuracy compared with ILD detected on HRCT, with an overall specificity (SP) and a positive predicted value (PPV) of 100% (Table 1). A negative correlation emerged between total lung capacity values (TLC%) and both B-lines cut-off (first operator (IO): p =0.04, r =0.27; second operator (IO): p =0.042, r =0.28) and pleural line criteria (IO: p =0.009, r =0.35; IO: p =0.08, r =0.36), but only the latter negatively correlated also with forced vital capacity values (FVC%) (IO: p =0.04, r =0.27; IO: p =0.03, r =0.28). The ≥10 total B lines amount correlated positively with anti-centromere antibodies (IO: p =0.005, r =0.37) and negatively with anti-centromere antibodies (IO: p =0.002, r =0.3; IO: p =0.009, r =0.34). The presence of digital ulcers showed a positive correlation with pleural line criteria (IO: p =0.03, r =0.29; IO: p =0.005, r =0.37), with a significant association on multivariate analysis (IO: p =0.03, IO: p =0.01).

Conclusion: We confirmed the feasibility and reliability of Fairchild’s recently proposed pleural line LUS criteria, that showed a higher diagnostic accuracy versus ≥10 cumulative B-lines count for ILD detected on HRCT, presenting SP and PPV values of 100% in SSc. Furthermore, these LUS features seem to differently associate with other aspects of the disease such as autoantibody specificity and vascular lesions, thus deserving future deeper evaluations. For the first time, we found that Fairchild’s criteria were associated with a clinical variable such as digital ulcers. Our results highlight the relevance of pleural line evaluation for ILD detection in SSc on LUS and its possible role towards a standardization of this diagnostic technique.

REFERENCES:

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POS0880
CHARACTERISTICS AND DISEASE COURSE OF UNTREATED PATIENTS WITH INTERSTITIAL LUNG DISEASE ASSOCIATED WITH SYSTEMIC SCLEROSIS

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Background: Intestinal lung disease (ILD) is the leading cause of death in systemic sclerosis (SSc). Scar-like changes can lead to guidelines recommending that some patients with mild disease might not need pharmacological treatment (1). Up to now, the disease characteristics and the disease course of non-treated SSc-ILD patients remain unknown.

Objectives: To describe disease characteristics and the disease course of non-treated SSc patients with ILD.

Methods: We included patients from our local EUSTAR center registered since 2008, who had a diagnosis of ILD on high-resolution computed