THE ISRAELI SOCIETY OF RHEUMATOLOGY ANNUAL MEETING 22-23.4.2021 | virtual meeting

Characteristics of interstitial lung disease in pts with systemic sclerosis during long-term follow-up, single center experience.

Shiri Keret[1], Yolanda Braun-Moscovici[2,5,6], Doron Markovits[2,5], Adi Abshalom-Mashiah[2,5], Vika Shataylo[2,5], Samy Giries[2,5], Kochava Toledano[2,5], Yonit Tavor[2], Katya Dolnikov[2], Alexander Rozin[2,5,6], Mordehai Yigla[3,5], Luda Guralnik[2,5,6] and Alexandra Balbir-Gurman[2,5,6]
[1]Internal Medicine C, [2]B Shine Rheumatology Institute, [3]Pulmonary Institute, [4]Imaging Department, [5]Rambam Health Care Campus, [6]Rappaport Faculty of Medicine, Technion, Haifa

Background: ILD is one the leading causes of morbidity and mortality in patients (pts) with SSc. Diagnosis of SSc-ILD is based on signs of fibrosis on HRCT and measurements of lung volumes by FVC and gas exchange by DLCO supports the diagnosis. Clinically significant SSc-ILD is associated with male gender, age, DcSSc, topoisomerase antibodies (ATA), low FVC at baseline, widespread lung involvement on baseline HRCT, and decline rate of PFT during follow-up. A standardized approach to assessing and treating SSc and SSc-ILD is proposed. The data on survival in SSc generally and SSc-ILD are conflicting.

Objectives: To analyze demographic and clinical features, survival rates and mortality of pts with SSc-ILD.

Methods: We performed a retrospective study on prospective cohort of SSc pts for the period between January 2000 and September 2020. Pts were recruited at their early visits to the clinic; they underwent baseline and serial HRCT and PFT. Pts' demographic (age, gender, ethnicity, date of SSc diagnosis and ILD diagnosis, disease duration) and clinical (subset, pulmonary, cardiac, renal, and muscle involvement, treatment used, autoantibodies, FVC, DLCO, and pulmonary artery pressure) data were extracted via hospital electronic records. Comparison between SSc-ILD and SSc-non ILD pts using t-test, Pearson's Chi-squared test, Fisher's test, Kaplan-Mayer curve and Cox regression analysis with p value less than 0.05 as significant

Results: Among 446 SSc pts (female 82.3%, mean age 46.5 and disease duration 11.6 years, DcSSc 39.2%; 27.4% dead during follow-up), 141 pts had ILD. Comparison between SSc-ILD and SSc-non ILD showed significant differences in term of nationality (Arabs 34% vs 18.7%), SSc related death (78.3% vs 50.7%), DcSSc (68.8% vs 25.6%), ATA (61.7% vs 24.9%), myopathy (21.3% vs 10.2%) and pulmonary hypertension (PAH, 34.8% vs 22.3%). More SSc-ILD pts received with DMARDs and biologicals. Kaplan-Meier curve showed reduced survival in SSc-ILD ($p<0.01$). Five years survival during years 2001-2005, 2006-2010, 2011-2015 in SSc was 91.8%, 91.2% and 87.3% and in SSc-ILD was 85.7%, 89.7% and 81.6% (NS). Mortality risk (Cox regression) in the SSc-ILD group was significantly higher in males, Arabs, DcSSc, elder age, heart and muscle involvement. In the SSc-ILD group, the mortality was significantly higher in Arabs (X3.3), elder age (X8.9), and PAH (X3.1). Appearance of PAH at 6 and 12 months, reduction of DLCO at 12 months from disease onset ($p<0.001$, $p< 0.05$) and high calculated monthly median decline rates of FVC (40% vs 16.3%, $p<0.012$) and DLCO (38.1% vs 14.6%, $p<0.015$) correlated with mortality.

Conclusions: In our cohort, SSc-ILD affected about third of pts and had major impact on pts' outcome. Male gender, Arab nationality, elder age, DcSSc, ATA antibodies, heart and muscle involvement indicated risk for ILD; Arab nationality, elder age, heart and PAH defined the worst prognosis. Early onset of PAH and fast monthly reduction of FVC and DLCO strongly correlated with mortality. Survival rates in SSc and SSc-ILD pts were stable during last decades and compatible with world experience. Early and proactive recognition of SSc-ILD using baseline and serial HRCT and PFT is crucial; coming to the clinic new therapies may change the fate and prognosis of SSc pts in general and SSc-ILD in particular.