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HRCT patterns in a cohort of connective tissue disease patients with interstitial lung disease.

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BACKGROUND

Connective tissue disease (CTD) is a group of chronic inflammatory immune mediated disorders which comprise systemic lupus erythematosus, rheumatoid arthritis (RA), Sjögren's syndrome, polymyositis/dermatomyositis (PM/DM), systemic sclerosis (SSc), and mixed connective tissue disease (MCTD). The respiratory system is frequently affected in CTD. Airways, vessels, parenchyma, pleura, and respiratory muscles may be affected. Among the diverse pulmonary manifestations of CTD, interstitial lung disease (ILD) play a particular role leading to unfavorable prognosis and high mortality. Clinical manifestations of CTD-ILD are heterogeneous and include non-specific interstitial pneumonia (NSIP), typical idiopathic pulmonary fibrosis (IPF)/usual interstitial pneumonia (UIP), organizing pneumonia, obliterative bronchiolitis and desquamative interstitial pneumonia. High resolution computed tomography (HRCT) scans are currently used to diagnose ILD in CTD patients and to assess disease improvement or progression. HRCT scans may show a focal or a diffuse lung involvement, especially at the periphery, appearing as fine and coarse inter- and intra- lobular septal thickening and honeycomb, cyst formation with distortion of the pulmonary parenchyma and associated traction bronchiectasis, reticulation, ground-glass opacities, and nodules.

OBJECTIVE

To distinguish the different HRCT patterns in a cohort of CTD-ILD patients.

METHODS

We examined the anonymized HRCT scans of 94 CTD-ILD female patients (mean age 58 ± 13 years) from the OSIC-ILD Data Repository including at present 941 patient cases and 1411 HRCT scans. ILD was diagnosed based on the radiological imaging pattern distinguishing the following five subsets: IPF, NSIP, IPF associated with NSIP, IPF with emphysema and NSIP with emphysema. According to ATS guidelines criteria typical IPF was characterized by honeycombing, reticular pattern, and bronchiectasis; NSIP was identified by ground glass and bronchiectasis; emphysema was detected by bullous lung pattern and bronchiectasis.

RESULTS

Based on the radiological imaging, we detected 51 NSIP (54.2%), 23 IPF (24.5%), 10 IPF/NSIP (10.6%), 6 IPF/emphysema (6.4%) and 4 NSIP/emphysema with cyst (4.2%). The HRCT patterns and CTD diagnosis are shown in Table 1.

TABLE 1

	HRCT pattern		
CTD diagnosis	*IPF* *IPF + emphysema*	*NSIP* *NSIP + emphysema*	*IPF/NSIP*
SSc	14	29	7
RA	4	4	
PM/DM	3	9	1
MCTD	2	6	1
Other	6	7	1
Total	29	55	10

We examined forced vital capacity (FVC) percent predicted and volume, forced expiratory volume in 1 second (FEV1), and predicted diffusing lung capacity for carbon monoxide (DLCO) value. The comparison between HRCT pattern and pulmonary function tests (PFT) is shown in Table2.

TABLE 2

	Pulmonary Function Tests*			
HRCT pattern	*FVC predicted (%)*	*FVC volume (Lt)*	FEV1 (Lt)	*DLCO*
IPF + IPF/emphysema	81.14 ± 24.72	2.05 ± 0.72	1.64 ± 0.53	10.62 ± 7.87
NSIP + NSIP/emphysema	80.05 ± 20.98	2.17 ± 0.63	1.76 ± 0.50	10.26 ± 4.58
IPF/NSIP	75.60 ± 19.12	2.05 ± 0.89	1.64 ± 0.67	± 4.90

** Values are Mean ± SD*

The results of PFT analysis demonstrate a decrease of all evaluated parameters, especially FVC % and DLCO, with respect theoretical predicted values.

DISCUSSION AND CONCLUSIONS

The main finding of the present research on HRCT scans in a cohort of CTD-ILD patients is the prevalence of NSIP pattern with respect to IPF pattern. Additional finding is the association of IPF and NSIP pattern in a significative percentage of patients. Emphysema was detected in about 10 percent of both IPF and NSIP patients. These data are in accordance with the literature (*Shao T et al., Interstitial Lung Disease in Connective Tissue Disease: A Common Lesion with Heterogeneous Mechanisms and Treatment Considerations. Front. Immunol. 2021; 12: 684699)* and confirm that NSIP pattern is most common in CTD-ILD patients. PFT data do not identify clear differences in functional airways parameters among HRCT patterns. A possible confounding factor of this study is the impossibility to perform a more detailed analysis, as the data are anonymized. The availability of a diagnostic algorithm to evaluate the extension and pattern of lung injury in HRCT images would greatly improve ILD diagnostic accuracy, disease monitoring and treatment as well as reduce the need of invasive procedures.