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UNCLASSIFIABLE INTERSTITIAL LUNG DISEASE OR UNDIFFERENTIATED CONNECTIVE TISSUE DISEASE? A CHALLENGING DIFFERENTIAL DIAGNOSIS

A. manfredi, M. Sebastiani, S. Cerri, G. Della Casa, D. Giuggioli, C. Vacchi, M. Colaci, A. Spinella, F. Luppi, C. Ferri
University of Modena and Reggio Emilia, Modena, Italy

Background: Interstitial lung disease (ILD) includes a group of disorders of the pulmonary parenchyma including ILD secondary to environmental exposure, to sarcoidosis and connective tissue diseases, idiopathic pulmonary fibrosis, nonspecific interstitial pneumonitis. Unclassifiable-ILD (U-ILD) is defined when ILD cannot be included in one of these subtypes, because of inadequate findings or impossibility to complete diagnostic iter. Undifferentiated connective tissue disease (UCTD) is a systemic autoimmune diseases characterized by clinical and serological features typical of other CTD, but not fulfilling any of the existing classification criteria. It has been recently suggested that UCTD should be responsible for ILD, although the available classification criteria do not consider lung manifestations. Differential diagnosis between U-ILD or ILD secondary to UCTD (UCTD-ILD) can be difficult, but fundamental for therapeutic implications.

Objectives: To evaluate the clinical and serological features of patients with ILD secondary to UCTD compared to unclassifiable ILD, to improve differential diagnosis and identify patients candidate to immuno-suppressive therapy. Secondary outcome was to construct a clinical algorithm, using a priori variables, helpful to predict ILD-UCTD in clinical practice.

Methods: From September 2011 to November 2014, 50 patients referred to our Center were diagnosed for UCTD (26/50) or U-ILD (24/50), after a multidisciplinary discussion according to standard available criteria.

Results: Main features and comparison between UCTD-ILD and U-ILD are reported in the table. An inconsistent with usual interstitial pneumonia (UIP) pattern at high resolution computerized tomography (HRCT) was more frequently detected in UCTD-ILD compared to U-ILD. A predictive model based on Raynaud's phenomenon, ocular dryness, and antinuclear antibodies showed a predictive value of 85.7% (UCTD-ILD were correctly classified in 90.5% and U-ILD in 78.6%).

Clinical and demographic features of patients with interstitial lung disease

	UCTD	**U-ILD**	**p**
Number of patients	26	24	
Sex	19F/7M	14F/10M	ns
Age at diagnosis	61.7±12.7	67.1±9.1	ns
Raynaud's phenomenon	68.0%	29.4%	0.027
Oral dryness	60.0%	35.3%	ns
Ocular dryness	60.0%	13.3%	0.007
Schirmer test	41.6%	4.2%	0.006
Arthritis	19.2%	0	0.05
Skin manifestations	30.4%	0	0.029
Thrombocytopenia	16.7%	18.2%	ns
Anemia	33.3%	18.2%	ns
Antinuclear antibodies	87%	52.4%	0.02

	UCTD	U-ILD	p
ENA	38.1%	10%	ns
Rheumatoid factor	13.6%	9.1%	ns
UIP pattern to HRCT	41.2%	58.8%	ns
Inconsitent with UIP pattern to HRCT	66.7%	33.3%	ns

Conclusions: Lung involvement is a possible presenting symptom of UCTD; therefore, differential diagnosis with U-ILD is crucial due to the relevant therapeutic implications; a multidisciplinary approach, including rheumatologist, pulmonologist, radiologist, and pathologist, is mandatory. Some clinical-serological features potentially helpful in differential diagnosis should be carefully evaluated

Disclosure of Interest: None declared

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