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Pathology and Therapy of Interstitial Lung Illness Caused by Connective Tissue Disease

Ben Duffy*

Department of Pathology, Unversity of West London, London, United Kingdom

***Corresponding author:** Ben Duffy, Unversity of West London, London, United Kingdom, E-mail: dr.jacobduffyben@gmail.com

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Description

Interstitial Lung Disease (ILD), also known as diffuse parenchymal lung disease, is a diverse collection of lung illnesses characterized by diffuse involvement of the pulmonary parenchyma by various degrees of inflammation and/or fibrosis. ILDs can have a known cause, such as drug-related, environmental and/or occupational, and systemic illnesses, or they can have an unclear cause, such as Idiopathic Interstitial Pneumonia (IIP), granulomatous diseases, and other disorders like lymphangioleiomyomatosis. Connective Tissue Disease (CTD) is one of the most prevalent causes of ILD among systemic illnesses.

To determine the diagnosis of a particular CTD, the classification criteria for that CTD must be fulfilled. ILD affects 40%-50% of CTD patients and is the leading source of morbidity and death. The stated incidence of ILDs in patients with CTDs (CTD-ILDs) differs by categorization standards and research databases for particular conditions, with SSc and idiopathic inflammatory myopathies (Dermatomyositis (DM) and Polymyositis (PM)) having a greater frequency and SLE having a lower frequency.

Recent report are the relative frequency of various CTD-ILDs and proportion estimates of patients with progressive fibrosing phenotype (percentages of people having each trait of CTD-ILD and percentages of individuals among whom a progressive fibrosing phenotype is developing): RA-ILD accounted for 39% of the whole CTD-ILDs and 40% of RA-ILDs showed progressive fibrosing phenotype; demonstrated progressive fibrosis.

ILDs linked with CTDs have the same histopathologic and radiologic characteristics as their idiopathic peers. Although not specific, some histopathologic findings, such as lymphoid hyperplasia (follicular hyperplasia), are indicative of a link with CTDs.

Previous research has shown that the existence of CTD in ILD patients affects prognosis, and that therapy choices are contingent on the underlying CTD. Guidelines emphasize aetiology-based ILD categorization and have consistently suggested searching for evidence of CTD in recently identified ILDs. However, due to the intricacies of CTD identification and therapy, as well as a dearth of proof, existing recommendations do not explicitly provide methods for CTD-ILD assessment and management, despite their importance.

Recently, the clinical significance of Interstitial Lung Abnormalities (ILA) identified inadvertently on CT has been recognized, which appear in 4%-9% of smokers and 2%-7% of nonsmokers. ILA is defined as the occurrence of non-dependent abnormalities such as ground-glass, reticular abnormalities, architectural distortion, traction bronchiectasis, and non-emphysematous cysts involving at least 5% of a lung zone in people who do not have ILD. The treatment of ILA in CTD patients (CTD-ILA) is unknown. TSCT gives detailed images of the lungs by using thin (1-1.5 mm) slice thickness and high-frequency reconstruction methods. TSCT is typically done on a multi-detector CT scanning with volumetric rather than axial incremental acquisition, covering the complete thorax in near isotropic resolution. Volumetric TSCT raw data can be used to generate not only radially rebuilt thin-section images, but also other planar images such as sagittal and coronal images. TSCT is an essential technique for detecting and characterizing lung pathology of ILDs in CTD patients.

A study of 203 individuals with CTD-ILD, undifferentiated CTD-ILD, and idiopathic pulmonary fibrosis, found that CT results did not vary substantially between the three groups. IPF was associated with more lung symptoms, whereas CTD-ILD and Undifferentiated Connective Tissue Disease (UCTD-ILD were associated with more extrapulmonary symptoms. CTD-ILD patients had more aberrant antibody assays than UCTD-ILD and IPF patients.

When encountering individuals with ILD or ILA features on CT, the presence of certain demographic (younger ages, never smokers, and women), histopathologic (fibrosing NSIP), or laboratory (autoantibodies) characteristics, namely those favoring CTD-ILD rather than idiopathic ILD or ILA, raises the possibility of CTD.

ILDs linked to CTD are most common in individuals with SSc, followed by DM/PM, SS, RA, and SLE. Despite the fact that the proportions of ILDs differ histologically, the NSIP pattern accounts for a significant percentage, particularly in SSc, DM/PM, and Mixed Connective Tissue Disease (MCTD). Interstitial Pneumonia with Autoimmune Features (IPAF) is described as ILD in individuals who have clinical, serologic, and/or morphologic autoimmunity but no characteristic CTD. ILDs with comparable characteristics to CTD-ILD are found in IPAF patients during TSCT or surgical lung biopsy. Straight edge sign, exuberant HC sign, and anterior upper lobe sign are considerably more prevalent on CT in patients with CTD-ILD and Usual Interstitial Pneumonia (UIP) pattern than in those with IPF and UIP pattern. The presence of CTD-ILA in CTD patients can be identified using CT findings of ILA in individuals without CTD.

However, it is reasonable to presume that the possible treatment plan of CTD-ILA is modified in light of the extent and stage of the disease itself. ILAs in RA patients have been shown to be radiologically advancing in 35%-45% of them. Sub-pleural spread and increased initial ILA length are risk factors for progression. Asymptomatic CTD-ILD individuals with normal lung function and no signs of disease progression are typically followed up on without therapy. In individuals who require therapy, immunosuppressive or anti-fibrotic drugs for symptomatic and/or fibrosing CTD-ILD can be chosen.