

JOURNAL CLUB

Interstitial lung disease in patients with mixed connective tissue disease

This cross-sectional study evaluates the prevalence and severity of interstitial lung disease (ILD), with particular emphasis on CT evaluation, in an unselected, nationwide cohort of 126 Caucasian patients with mixed connective tissue disease (MCTD).

Overall, high-resolution CT (HRCT) abnormalities were seen in half the number of patients. Although MCTD is characterised by overlap features between scleroderma (SSc), polymyositis/dermatomyositis (PM/DM) and systemic lupus erythematosus (SLE), the HRCT findings appeared remarkably homogenous. Reticular patterns were the most common, present in a third of patients, with coarse reticulation present only in a minority. As the authors discuss, the findings were consistent with a radiological pattern of non-specific interstitial pneumonia. Interestingly, although myositis occurred in the majority, areas of consolidation suggestive of organising pneumonia were absent, a feature frequently present in PM/DM. The HRCT features of MCTD, when tightly defined, would appear much more similar to a scleroderma-related ILD phenotype than to a PM/DM one. As the unifying feature for MCTD is anti-ribonucleoprotein antibody (anti-RNP) positivity, one could speculate that anti-RNP antibody associated ILD is in fact non-specific interstitial pneumonia, regardless of the associated clinical features, although this will require confirmation. After 4 years of follow-up, lung fibrosis was associated with increased mortality, highest in severe fibrosis (20.8%). Patients with severe fibrosis tended to be older, to have worse lung function/functional status, and to have shorter disease duration compared with those with minor/moderate fibrosis.

This small but well-characterised cohort provides insight into the high prevalence and increased mortality of ILD in MCTD. Similar to ILD associated with other CTDs, patients with severe fibrosis likely represent a more progressive disease subset, and may benefit from early identification and timely immunosuppression.

- Gunnarsson R, Aaløkken TM, Molberg O, et al. Prevalence and severity of interstitial lung disease in mixed connective tissue disease: a nationwide, cross-sectional study. *Ann Rheum Dis*. Published online first: 1 May 2012.

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Provenance and peer review Not commissioned; internally peer reviewed.

To cite Lota HK. *Thorax* 2013;68:186.

Thorax 2013;68:186. doi:10.1136/thoraxjnl-2012-202319