Characterisation of patients with interstitial pneumonia with autoimmune features

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ABSTRACT Patients with interstitial lung disease (ILD) may have features of connective tissue disease (CTD), but lack findings diagnostic of a specific CTD. A recent European Respiratory Society/American Thoracic Society research statement proposed criteria for patients with interstitial pneumonia with autoimmune features (IPAF).

We applied IPAF criteria to patients with idiopathic interstitial pneumonia and undifferentiated CTDILD (UCTD). We then characterised the clinical, serological and morphological features of the IPAF cohort, compared outcomes to other ILD cohorts and validated individual IPAF domains using survival as an endpoint.

Of 422 patients, 144 met IPAF criteria. Mean age was 63.2 years with a slight female predominance. IPAF cohort survival was marginally better than patients with idiopathic pulmonary fibrosis, but worse than CTD-ILD. A non-usual interstitial pneumonia pattern was associated with improved survival, as was presence of the clinical domain. A modified IPAF cohort of those meeting the clinical domain and a radiographic or histological feature within the morphological domain displayed survival similar to those with CTD-ILD.

IPAF is common among patients with idiopathic interstitial pneumonia and UCTD. Specific IPAF features can identify subgroups with differential survival. Further research is needed to replicate these findings and determine whether patients meeting IPAF criteria benefit from immunosuppressive therapy.