Survival in fibrosing alveolitis associated with rheumatoid arthritis is better than cryptogenic fibrosing alveolitis

SIR, Hubbard and Venn [1] have used a novel approach by using the general practice research database to show that the survival in fibrosing alveolitis associated with connective tissue disease (FA-CTD) is as bad as in lone cryptogenic fibrosing alveolitis (LCFA). The result is contrary to current view [2] that FA-CTD is more benign than LCFA. Rheumatoid arthritis (RA) accounts for the major proportion (80%) of the CTD cohort in this study. The median survival of 2.6 yr in the FA-CTD cohort compared with 2.4 yr in the LCFA cohort is quite alarming. This is an important finding in view of new evidence that suggests that respiratory disease contributes to considerable mortality even in early RA [3].

However, we recommend caution in interpreting the conclusions of this study [1] as it is based on the following assumptions.

First, the causes of death in both the groups have not been studied; hence it cannot be assumed that the patients in the CTD group died of fibrosing alveolitis. Ischaemic heart disease [3, 4] and cancer are major causes of death in RA. A second control group comprising patients with RA without fibrosing alveolitis would help us identify the mortality attributable to fibrosing alveolitis in RA. We suspect that the median survival rate in RA without FA would be less than the healthy controls.

Second, we do not know the exact duration and severity of fibrosing alveolitis in both cohorts. The duration was assumed to be the date of ‘first recorded diagnosis’. Many patients with RA have subclinical interstitial lung disease (RA-ILD) [5, 6]. Rheumatologists do not necessarily screen their rheumatoid patients for early ILD and hence this diagnosis is likely to be unreported to primary care practitioners. The prevalence of ILD in RA patients is around 5% [7]. Hubbard and Venn report 86 cases of RA-ILD without information on the total number of patients with RA in the GP research database. Given the fact that this is the largest primary care database in the UK it is likely that 86 patients with RA-ILD is a marked underestimate of the true number. We feel patients with RA in this study represent a group with more severe interstitial lung disease.

Third, the pathological and radiological features of FA associated with RA and CTD are known to be different to those of LCFA. Patients with RA are thought to have more ground-glass shadows on HRCT lung [8] and ‘non-specific interstitial pneumonia’ pattern (NSIP) on histology is more common in systemic sclerosis [9]. This is in contrast to established fibrosis on HRCT and ‘usual interstitial pneumonia’ pattern (UIP) on histology in LCFA. It is, however, not known if the former picture is more benign and amenable to immunosuppression [10].

We have recently completed a 5-year follow-up of patients with RA-ILD (n = 18) and LCFA (n = 18) described in our earlier study [8], identified at baseline with matching degrees of pulmonary dysfunction. This reveals major differences in both the survival and causes of death. At 5 yr, 44% of RA-ILD but only 11% of LCFA patients are alive. Death was due to fibrosing alveolitis in 20% of RA-ILD and 62% of LCFA. There was a wide range in the rate of progression of ILD in both the groups.

Hubbard and Venn’s study must be interpreted with caution and further prospective work is needed to identify the true course of FA associated with RA.

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