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Letter to the editor

To the Editor:

It was with great interest we read the article by Dr. Collins and colleagues published in the August 2018 edition of Respiratory Medicine, titled "Sarcoidosis and IPF in the same patient-a coincidence, an association or a phenotype?" The authors describe the characteristics of a disease entity they coined "combined sarcoidosis and idiopathic pulmonary fibrosis (CSIPF)" [1]. We appreciate the authors' efforts to shed light on this clinical entity, which to date has sparked debate in terms of proper classification of patients who share radiographic and histopathologic features of sarcoidosis and idiopathic pulmonary fibrosis (IPF). The article highlights similarities in such clinical characteristics as survival time and decline in respiratory function between patients with CSIPF and selected patients with lone IPF. The authors go on to conclude that since patients can manifest sarcoidosis and IPF concurrently, this suggests a mere coincidence as opposed to a clear association between the two; and given the presence of UIP features in patients with CSIPF, the clinical course behaves much like lone IPF and should be regarded similarly when considering candidacy for antifibrotic therapy and/or lung transplantation.

We are uncertain about the term CSIPF since the diagnostic criteria of IPF requires the exclusion of any other interstitial lung disease, based on the most recent diagnostic guidelines [2]. It is especially difficult to apply the term CSIPF in the seven patients that were diagnosed concurrently. To the authors' credit they mention the possibility of CSIPF being a phenotype of sarcoidosis. Based on our experience, we favor this explanation or that the UIP pattern represents an advanced stage of sarcoidosis. Within our own cohort, patients presented with severe lower lobe honeycombing and minimal upper lobe peribronchovascular fibrosis on imaging with pathology demonstrating granulomatous inflammation in the areas of honeycombing. For these patients diagnosed concurrently, we suggest the term "Sarcoidosis with UIP pattern".

It was intriguing to see that the clinical course of the CSIPF patients closely resembled that of lone IPF patients when compared to patients with pulmonary sarcoidosis, suggesting that the presence of honeycombing is an important prognostic factor in sarcoidosis patients, as has been shown in other ILDs [3].

Our patients had similar outcomes as those in the study. Whether or not these patients should be considered for antifibrotic agents, disease modifying antisarcoid drugs, or both, remains to be determined. Based on the findings in this paper, it would be premature to suggest a similar treatment strategy for sarcoidosis with honeycombing as IPF patients, as studies in this population are lacking. We wait with anticipation for the results of the ongoing phase 4 trial comparing pirfenidone versus placebo for patients with advanced fibrotic sarcoidosis which may better inform how to approach these patients.

References

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