Reviewer's report

Title: Idiopathic Pleuroparenchymal Fibroelastosis: Consideration of a Clinicopathological Entity in a Series of Japanese Patients

Version: 3 Date: 9 September 2012

Reviewer: Keith C. Meyer

Reviewer's report:

The entity described by the authors, idiopathic pleuroparenchymal fibroelastosis (IPPFE) appears to be an emerging entity that falls under the category of ILD. This report is straightforward and relatively well-written.

Major Compulsory Revisions

1. Abstract:: Conclusions: "uniformity and largely overlapping with ..." This sentence needs revision (suggest .... ".... owing to its relatively distinct features."). I recommend removing "largely overlapping with previously described idiopathic upper lobe fibrosis (IPUF) or change the sentence to state that IPPFE has some features in common with IPUF.

2. Abstract: Conclusions: "... IPPFE should be clearly distinguished from other types of interstitial lung disease, because it is highly progressive." I recommend softening this to "our limited experience with a cohort of 5 subjects suggests that IPPFE can be rapidly progressive." Other forms of ILD can progress rapidly (e.g. IPF), and a larger number of patients would be needed to make firm conclusions concerning the natural history of the disease.

3. Discussion: First paragraph, page 6: It is stated that "our study was an attempt to identify common features between these two entities...." This is not mentioned in the abstract as an objective of the study, although IPUF is mentioned in the Conclusion.

4. Discussion & Conclusions: It is mentioned that IPPFE is a distinct clinicopathological syndrome that largely overlaps with previously described IPUF. However, the similarities and differences of "IPUF" as compared to IPPFE are not well described. What are the pathologic manifestations of IPUF? Is fibroelastosis present in IPUF? Is it only the radiologic features/regional distribution of the disease that are similar? The histopathology of IPUF should be described and compared to the features of the IPPFE cases. A goal of the study given in the "Background" section on Page 4 is to "clarify its relationship with IPUF," but the reader is left wondering what the histopathologic changes are in subjects with IPUF (is fibroelastosis present in the cited reports of cases of IPUF?).

Minor Essential Revisions

N/A
Discretionary Revisions
N/A

**Level of interest:** An article of importance in its field

**Quality of written English:** Needs some language corrections before being published

**Statistical review:** No, the manuscript does not need to be seen by a statistician.

**Declaration of competing interests:**
I declare that I have no competing interests