Reviewer’s report:

In their manuscript "Idiopathic pleuroparenchymal fibroelastosis: consideration of a clinicopathological entity in a series of Japanese patients" Kusagaya et al describe a series of five patients whom the authors feel represent cases of a rare disorder, idiopathic pleuroparenchymal fibroelastosis. This entity has been described in a handful of case reports and case series, most notably in the English language literature by Frankel et al in Chest in 2004, and more recently Reddy et al in the European Respiratory Journal in 2012. The patients described by Kusagaya et al, like prior patients described in the literature, have dense upper lobe pleural and parenchymal fibrosis of unknown cause. The combination of upper- but not lower-lobe pleural and associated parenchymal fibrosis, with an abrupt transition to normal lung, is not typical of asbestosis, the most common disease causing pleural and parenchymal fibrosis, and asbestos bodies are not identified in the cases described in this manuscript, or in other cases in the literature. The authors describe the patients’ clinical, radiologic, pathologic, and physiologic features in the current report. They also describe a decline in FVC in the affected patients in follow up, suggesting this is a progressive disorder, unlike the presence of apical pleural capping.

I believe the manuscript is clearly written. Although the study is retrospective, the data as presented is fairly complete. While the methods do not describe any consensus or blinded review of the IPPFE cases by a pathologist(s), which would add to the validity of the study, the methods do state that the specimens were reviewed by a co-author who is a well known pulmonary pathologist. The authors state that the retrospective nature of the study is the major limitation, which is true. As a case series, the data presented does not allow insight into prevalence of the disease, and does not provide a comparison/control group, such as coexistent individuals with IPF. I think the authors do a fairly good job of not overstating the significance of the data presented, which is descriptive. There are no novel insights into pathogenesis of idiopathic pleuroparenchymal fibroelastosis.

Despite the above limitations, this case series does add to the small body of information on this seemingly rare proposed disease entity, which does not fit into the ATS/ERS classification system of idiopathic pneumonias, and I think is therefore worthy of consideration for publication after addressing below comments.
A. Major comments:

1. Were the cases collected here diagnosed with IPPFE only in retrospective review, or was this the actual pathological diagnosis at the time of the open lung biopsy?

2. In the Results, the authors state that "In all cases in this study, the histopathological findings fitted those previously described for IPPFE." The Methods state that the "subjects included 5 patients fulfilling criteria PPFE" and reference the paper by Frankel et al. While the authors adopt wording similar to that of Frankel et al in the description of pathology on page 6 of the Results, I think that all the criteria used for the diagnosis of IPPFE should be delineated in the Methods.

3. Case 5 seems to have significant lower lobe involvement based on the CT images presented. In the text of the Results on page 5, case 5 is described as having "somewhat more prominent involvement of the lower lobes rather than other cases". Yet in the Discussion on page 7, the authors write that "The upper lobes were ALWAYS (emphasis mine) more severely involved, with involvement of the lower lobes being absent or less marked." I think the lower lobe cystic and/or bronchiectatic changes shown in Figure 1 are significant and arguably more pronounced than the upper lobe disease, since the lower lobe disease in case 5, in the image presented, seems to involve more than 50% of the lung area.

4. I disagree with the authors' statement in the Discussion that "Our data indicate that serum SP-D may also be useful indicator for diagnosis and prediction of disease progression of PPFE". The data as presented do not indicate how SP-D levels are different in IPPFE from other forms of interstitial lung disease, and do not indicate how they are a prognostic marker in IPPFE.

Minor:

1. The authors sometimes use the abbreviation "IPPFE" for idiopathic pleuroparenchymal fibroelastosis, and at other times use the abbreviation "PPFE". These terms should be reconciled.

2. The authors state in the Abstract that 4/5 patients had an impairment in DLCO, but in the Results, page 5, state that "In addition, three cases had impairment of DLCO (Table 2)." Table 2 seems to indicate that 4/5 had impaired DLCO (if DLCO <80% predicted is considered impaired), while 3/5 have impaired DLCO/VA. This should be clarified.

3. On page 7, there is a minor wording issue. "Among those cases, four cases had treatment with BMT, and thoses cases presented with histological evidence of obliterative bronchiolitis, which was not a feature in THAT with idiopathic presentation of PPFE..." I think the authors mean THOSE. There are some other minor wording issues throughout.

4. There are some minor formatting issues with Tables 1 and 2 that I assume will be corrected in the proof, such as a value for RV(L) and RV % predicted being on the same line.
Please note that my views expressed above do not necessarily reflect those of the Department of Veterans Affairs.

**Level of interest:** An article whose findings are important to those with closely related research interests

**Quality of written English:** Acceptable

**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.