

Response to the comments

Dear Editors

Thank you for giving me the opportunity to submit a revised draft of my manuscript titled **“Unilateral pleuroparenchymal fibroelastosis as a rare form of idiopathic interstitial pneumonia: A case report”** to *World Journal of Clinical Cases*. We appreciate the time and effort that you and reviewers have dedicated to providing your valuable feedback on our manuscript. We are grateful to the reviewers for your insightful comments on my case report.

Followings are the response to the reviewers’ comments and concerns.

Reviewer 1

Comment 1: Is there a difference between bilateral and unilateral prognosis?

Response 1: Thank you for your comment. There are not enough evidence and data about the difference of prognosis between bilateral and unilateral PPFE. However, I presume that involvement of lower lung or progression of PPFE will be more important factors for prognosis in patients with PPFE rather than unilaterality or bi-laterality.

Comment 2: How Uncommon is the Unilaterality of this Disease?

Response 2: Thank you for your comment. The prevalence of the unilateral PPFE is not uncertain and case of unilateral have hardly been reported. In a literature search, I found just one report. But with increasing recognition of PPFE, I guess the reports will be increasing.

Comment 3: Reference 17 also reports localized PPFE in the upper right lobe. Is this illness more common on the right side?

Response 3: Thank you for your comment. In reference of 17, the authors reported that 6 cases of unilateral PPFE and all the lesions were to the right. All the patients had a relevant conditions related to the PPFE, lung injury after thoracotomy or operations. However, in our case, there was no confirmed direct lung injury with only suspected history of exposure. It is still difficult to conclude that the right side is common in patients with unilateral PPFE than left side. Therefore, in cases of secondary PPFE, I assume that the lesions of PPFE with unilaterality will be related to the site of lung injury.

Comment 4: Did you check markers for interstitial pneumonia such as KL-6 and SPD

Response 4: Thank you for pointing this out. Serum biomarkers of KL-6 and SP-D have been widely used in ILD patients. I'm also very curious about the role of KL-6 and SP-D for predicting prognosis in patients with PPFE. However, unfortunately, test of KL-6 and SP-D are not available yet in South Korea, so we could not check.