**Author’s response to reviews**

**Title:** A case of atypical anti-GBM disease complicated by CMV pneumonitis and massive hemoptysis

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**Author’s response to reviews:**

Reviewer #1:

Major issue #1: - “Take-home message”.

Response: We have modified the title and text to emphasize the aggressive nature of the renal and pulmonary phenotype in this case, not characteristic of atypical anti-GBM disease. We appreciate the point that the reviewer has made regarding the novelty of the case and we hope that the modifications have made it clearer.

We have added comments and a reference related to the unexpected aggressive clinical course in view of the IgG4 subclass.

Major issue #2: - “The intensity of IF staining in Fig 2 does not match the description in the text. Panels B and C (kappa and lambda) appear similar. Also, the text states strong IgG and weak IgA. However, IgA staining in Fig 2/panel D does not appear weaker than IgG in Fig 2/panel A. This implies that anti-GBM IgA may also contribute to pathology in addition to or instead of
anti-GBM IgG4 (IgG2), which should be considered and discussed (many cases of atypical anti-GBM disease are IgA-mediated).

Response: We realized, upon revisiting the submitted manuscript, that two panels had inadvertently been switched: 2B and 2D. We have now corrected this error. Consequently, the reviewer’s comments regarding the relative strength of the staining of kappa and lambda, and IgA. We feel that with this correction, the images match the description in the text. While IgA is not negative, it is not as positive as IgG. This does not preclude that IgA had a pathogenic effect in this case, and we have adjusted the text to reflect this.

Major issue #3: (wait for MF’s response)

Minor points: The authors should show the staining for all four IgG subclasses in Figure 3 (to document that IgG1 and IgG3 are negative).

Response: unfortunately, photos of these negative stains were not taken at the time.

Also, in Figure 2, consider showing the negative staining for C3 (since C3 staining is usually positive in typical anti-GBM, the absence of C3 serves to exclude one possible effector mechanisms of atypical anti-GBM in this case), and for IgM (as it rules out glomerular deposition of IgM lambda monoclonal).

We have included a photo of IgM and C3. A photo of C1q was not available

Reviewer #2

#1 – we have made changes in the text to reflect these points

#2 - we have made changes in the text to reflect these points

#3 – we have added these points

#4 - we have added these points

#5 - we have added these points

#6 – this has been changed

#7 – We respectfully feel that the pathology description needs to be left unshortened, as it pertains to a central aspect of the case – i.e. the unusual severe clinical presentation in atypical anti-GBM disease.
#8 and 9 We have edited the microphotographs according to the suggestions made by the reviewer.