

Peer Review File

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**Reviewer Comments**

Comment 1: - GPA is not a c-ANCA positive autoimmune systemic disorder, since it can be c- or p-ANCA (it is, PR3 or MPO), as well as an ANCA-negative vasculitis (abstract and introduction). It is an ANCA-associated vasculitis.

Reply 1: Agreed

Changes in the text: We have changed it to ANCA-associated vasculitis as suggested both in abstract and introduction

Comment 2: - Regarding the latter, how many of the reported patients are MPO-positive? (or p-ANCA) This information can be included in table 2 (column of clinical data).

Reply 2: Agreed, can be helpful information

Changes in the text: Where in literature this information was available, we have added this information to all the cases under the “Clinical” column of Table 1 (Previously was Table 2).

Comment 3: - A review of radiological findings with no images is really odd. Permission of previously reported work is recommended.

Reply 3: Agreed

Changes in the text: Figures of CT and MR have been added. (Figure 1 and 2)

Comment 4: - - RCC (in brackets) should be placed after renal cell carcinoma (line 62).

Reply 4: Agreed

Changes in the text: Has been added

Comment 5: - - Until when the literature review has been performed? (It should be included in the methods) I found at least a couple of additional cases:

Reply 5: Thank you for pointing this out. Earlier it included cases from 1978 to 2019. But based on your comment, we extended the search till 2021. We found 4 more cases (including the 2 you suggested).

Changes in the text: These 4 cases have been added to the Table 1

Comment 6: - - Table 1 and 2 should be quoted before, ideally in a paragraph of results (after of the methods,) including the summary of reported cases.

Reply 6: A result section has been added after methods in the body section where

these tables have been quoted, as suggested

Changes in the text: Result section added, and Tables quoted there.