Author's response to reviews

Title: Hyper-IgG4 disease: Report and characterisation of a new disease.

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Author's response to reviews:

Dear Editors,

Reviewer Kendo has made a further series of comments - some of which are inappropriate. However, we have considered them all and made some changes to the text.

1. We have removed the words 'In this review..' from the start of our final paragraph of the Background (see Kendo's Revision 2).

2. We have changed the patients' details in Table 4 from date of birth to age - we agree this was an error (Kendo revision 3).

3. Also in Table 4 we have added a line to the Legend, which had been omitted, regarding the scoring of 'Fibrosis'.

4. When we showed the manuscript to the 'Index Case' and obtained his signed permission for publication, he pointed out that he had been in the UK 31 years not 10 - we have now changed this.

5. We have added to the Case report, the values of the IgG4 level at the time of diagnosis. (see also Kendo comment on "hyper").

Other comments:
The Reviewer made some further comments about the meaning of 'Hyper' (in Hyper-IgG4 disease) and the meaning of 'systemic disease'. He also commented that not all patients do well with treatment.

We believe it is clear from our presentation that we are pointing out that this disease is not limited to one organ, but can be found in different organs of the body; and in some patients the disease may simultaneously involve several organs; and thirdly, some patients may even have a systemic illness with fever and constitutional symptoms. The term 'systemic' is appropriate in all these cases.

We also believe that it is clear what we mean by 'Hyper IgG 4 disease', but we have added in the data for our Index Case (point 5 above) and added a sentence in the Conclusion - see italicized comment below.

Finally, we have already responded to the question of treatment in our reply to Kamisawa's comments (first review). However, we have now added in ..' during the acute phase '.. in the first sentence of the Conclusion - see below.

"Conclusions
We believe that hyper-IgG4 disease is an important condition to recognise as the diagnosis can be verified by histology and simple blood tests, and the outcome with treatment during the acute phase is very good. A raised IgG concentration in the absence of hypergammaglobulinaemia is typical, but ideally IgG4 should also be measured. Even when pathology appears localised, as with RPF, constitutional symptoms can be present with evidence of an acute phase response."

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