Reviewer's report

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

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Reviewer: Kiyosawa Kendo

General

This article described the pathological study of 12 retroperitoneal fibrosis cases in special interest to IgG4-positive plasma cell infiltration. From this study, authors proposed the concept of hyper-IgG4 disease, which is an important condition to recognize as the diagnosis can be readily verified and the outcome with treatment is very good.

Though authors implied important systemic disease characterized by IgG4-positive plasma cell infiltration and fibrosis, results concerning this were too poor to support the new concept of hyper-IgG4 disease. In addition, authors should discuss the concept of hyper-IgG4 disease, not the differential diagnosis of retroperitoneal fibrosis or FUO.

Specific comments

1) In Table 4, Fig.4 and Fig. 5, authors should describe and compare the pathological findings of retroperitoneal fibrosis and other organs. Authors only described the findings of retroperitoneal fibrosis. Details of other organs were obscure. If authors implied the systemic disease of hyper-IgG4 disease, they should do systemic examination.

2) How many samples for each organ were authors examined in Fig.4 and Fig.5. Authors examined the pathological findings for all tissues of liver, pancreas, salivary gland, retroperitoneal tissue and kidney for all cases?

3) In Fig.5, the proportion of IgG4-positive plasma cell in each organ was over 80%, which is unreliable. Please examine the immuno-staining of other IgG subclass, such as IgG1, IgG2, and IgG3, and compare IgG4-staining with other IgG subclass stainings, especially that of IgG1. The number of IgG4-positive plasma cell is possibly equivalent to that of IgG1 positive plasma cell even in the condition of hyper IgG4 disease.

4) Similar concepts to hyper IgG4-disease have been proposed previously in connection with autoimmune pancreatitis. Please discuss the association with these equivalent concepts.

5) In discussion, descriptions for the differential diagnosis of retroperitoneal fibrosis or FUO are meaningless.

Which journal?: Appropriate or potentially appropriate for BMC Medicine: an article of importance in its field

What next?: Accept for publication in BMC Medicine after discretionary revisions

Quality of written English: Acceptable

Statistical review: No