

March 1, 2014

Dear Editor,

Please find enclosed the edited manuscript in Word format (file name: 9128-review_final.doc).

Title: First Case of IgG4-related Sclerosing Cholangitis Associated with Autoimmune Hemolytic Anemia

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In response to the reviewers' comments, we have revised the manuscript as described below. In the revised manuscript, revisions are in red letters, and parts to be deleted are indicated by a strike-through.

1. The format has been updated
2. Revisions have been made according to the suggestions of the reviewer

Response to the reviewers' comments

Reviewer: No. 00057447

1. IgG4 were already markedly increased at the first observation. Authors should discuss why they ignored this finding.

Response: We measured serum IgG4 concentrations at initial presentation (Table 1). However, there was a time lag of about 1 week until the test results became available (because it was a specialized test). Although the patient had elevated serum IgG4 levels, endoscopic retrograde cholangiography showed no typical findings of IgG4-SC, but revealed findings suggesting cholangiocarcinoma or PSC. Treatment with ursodeoxycholic acid temporarily improved liver dysfunction, and this clinical course may be the reason why IgG4-SC could not be diagnosed at an early stage. These findings are already described in the text. We believe that IgG4-SC in the absence of AIP is usually not easy to differentiation from PSC or bile-duct cancer. We have added the reason why the elevated serum IgG4 concentration did not directly lead to diagnosis to the first paragraph of the Discussion section.

2. The last paragraph of the discussion (from: Many cases...) is redundant and could be omitted.

Response: We have deleted the last paragraph of the Discussion.

3. Table 1 should be omitted.

Response: We have integrated Table 1 and Table 2.

4. In table 2 the range of normal values should be inserted (at least IgG values).

Response: We added the range of normal values of IgG to Table 1.

Reviewer: No. 00055108

1. It would be interesting if the authors could implicate data (if available) according to the conclusions of the just e-published paper in Hepatology – by Boonstra et al, Serum IgG4 and IgG1 for Distinguishing IgG4-Associated Cholangitis from Primary Sclerosing Cholangitis. If possible speculate if the IgG4/IgG1 ratio could have helped the authors to make the diagnose IgG4-SC.

Response: As you suggested, Boonstra et al. reported that the IgG4/IgG1 ratio is useful for differentiating IgG4-SC from PSC. However, the IgG4/IgG1 ratio was unavailable because we did not measure serum IgG1 levels in our patient. These facts were added to the first paragraph of the Discussion section and the report by Boonstra et al. was added to the Reference section.

2. Regarding figures 1-4, it would be appropriate to mark the specific findings the authors are referring to in the figures legends.

Response: We have marked the specific findings in the Figures.

3. Please also control in the text and the legend of fig 2 *contrast effect* is that correct or are the authors meaning * contrast defect*?

Response: We revised “contrast effect” to “contrast enhancement”.

4. Regarding the tables – always nice to be reminded of how to convert mg/dl to g/l, which also is a frequently used unit?

Response: We converted the units to “The International System of Units” in Table 1.

5. In table 1 the authors have written dilect bilirubin, I think they are meaning direct bilirubin (or conjugated).

Response: We changed “dilect bilirubin” to “direct bilirubin” in Table 1.

3 References and typesetting were corrected

Thank you again for publishing our manuscript in the *World Journal of Gastroenterology*.

Sincerely yours,

Hironori Masutani, M.D.

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