Author's Response Letter to Reviewer's Comments

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Column: Case Report

Title: Systemic Mastocytosis: a rare cause of non-cirrhotic portal hypertension

Authors: Cláudio Martins, Cristina Teixeira, Suzane Ribeiro, Daniel Trabulo, Cláudia

Cardoso, João Mangualde, Ricardo Freire, Élia Gamito, Ana Luísa Alves, Isabelle Cremers,

Cecília Alves, Anabela Neves, Ana Paula Oliveira

Correspondence to: Cláudio Martins, Department of Gastroenterology, Hospital de São

Bernardo, Centro Hospitalar de Setúbal E.P.E., 2910-446 Setúbal, Portugal

Reviewer code: 00069458 and 00070310

First decision: 2016-05-27 11:09

Scientific editor: Yuan Qi

June 6th, 2016

Dear Editor,

Enclosed, please find our revised manuscript entitled "Systemic mastocytosis: a rare

cause of non-cirrhotic portal hypertension". This manuscript is being submitted for

consideration as a review article in World Journal of Gastroenterology. In this letter you will

find a point-by-point response to each of the reviewer's concerns. We have completely

followed the comments made by the reviewers and have carefully revised the manuscript.

We appreciate the thoughtful and positive reviews from the reviewers. We hope that the

revised manuscript will now be suitable for publication, and thank you to the reviewers

for their effort and time.

Sincerely yours,

Cláudio Martins,

Department of Gastroenterology,

Setúbal Hospital Center - São Bernardo Hospital,

Camilo Castelo Branco Street, Setúbal 2910, Portugal.

E-mail: cmartins1@campus.ul.pt

Telephone: +351265549000

Fax: +351265238066

Authors' Rebuttal to Reviewers' Comments:

In the following we discuss the points raised by the reviewers and the changes in the manuscript to address these points. We appreciate these thoughtful comments, and hope that our responses and in particular our revisions have allowed this paper to achieve priority sufficient for publication in *World Journal of Gastroenterology*.

Reviewer Code: 00069458

Reviewer's Comments: This is a well-written manuscript and a very interesting case report. Minor points. **1**. Discussion is too long and should be shortened. **2**. Another relevant case report [Pathol Int. 2009 Nov;59(11):817-22. doi: 10.1111/j.1440-1827.2009.02451.x.] should be commented in the discussion section. **3.** There are a few grammatical errors eg. 'Minor Criterion' instead of 'Minor criteria' and 'agressive' instead of 'aggressive'.

Authors' Response: Thank you for all your comments.

- 1. Regarding the "Discussion" section we tried to shorten it by removing some information that was not essential to the overall comprehension of the case. In this process, for example, we removed the 2th paragraph and shortened the 4th, about hematologic abnormalities. We believe that all the remaining information is crutial to a proper understanding of the pathology and the clinical case.
- 2. We introduced the suggested case report in the discussion section. It was commented in th 4th paragraph concerning liver involvement: "Yoshida M et al^[8] reported a case of mast cell leukemia, a rare form of SM, complicated with rapidly progressing non-cirrhotic portal hypertension due to blocking of sinusoidal and venous flow".
- 3. Grammatical errors had been corrected.

Reviewer Code: 00070310

Reviewer's Comments: This paper shows a rare case of systemic mastocytosis with non-

cirrhotic portal hypertension. This is very interesting, and will be acceptable for

publication except one. 1, Please show exactly the data in 'case report' according to

WHO diagnostic criteria for SM.

Authors' Response: Thank you for all your comments. We introduced in "Case Report"

section the exactly data according to WHO diagnostic criteria for SM: "These findings were

fully consistent with SM fulfilling the WHO diagnostic criteria for this entity, namely the presence

of the major and three minor criteria: >25% of atypical MC in bone marrow and liver, coexpression

of CD25 and CD117 by atypical MC, and high serum tryptase levels". Regarding

subclassification in aggressive systemic mastocytosis, this was commented in "Discussion"

section.

Once again thank you very much for the comments and opportunity to revise the

manuscript

Yours sincerely,

Cláudio Martins