

2nd January, 2019

Dear Editor,

Thank you for the review, feedback, and consideration of our manuscript entitled "Crohn's-like acute severe colitis associated with Hermansky-Pudlak syndrome: a case report" to be considered for publication in World Journal of Gastroenterology.

We have addressed all comments of reviewers and listed them below item-wise. All edits are manually highlighted in yellow and cited below. All authors concur with the revision.

Please find below the edits that have been done to the manuscript:

- Added manuscript number in the title page.
- Change in the format of the paragraph "Author contributions".
- Change in the "Informed consent statement".
- We added the names of the authors and the manuscript title at the end of the core tip.
- We changed the format of references, and we checked and confirmed that there are no repeated references.
- We distinguished the three levels of subtitles.
- We submitted the figures as image files on the system, and removed the abbreviations from the figure legends.

The manuscript was reviewed by Filipodia Publishing for language polishing.

Sincerely yours,

Catherine Le Berre, MD

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Point-by-point response to reviewers

Reviewer 1 (01467363):

Title and running title: accurately reflects the topic and contents of the paper.

- We thank reviewer 1 for her/his comment.

Abstract: is appropriate, structured, 240 words. Key words: 7 key words, precisely define the content of the paper.

- We thank reviewer 1 for her/his comment.

Core tip: is appropriate, 139 words.

- We thank reviewer 1 for her/his comment, however our core tip consists of 100 words only.

Introduction: is informative, short, 139 words, the reader is acquainted with the known facts about basic characteristics of this syndrome and the associations with diseases of other organs are explained. HPS types 1 and 4 can be associated with inflammatory bowel disease (IBD)-like disorder of the gastrointestinal tract.

- We thank reviewer 1 for her/his comment.

Case report: informative, 481 words, key data about the female patient are presented: chief complaints, history of present/past illness, clinical examination at admission, laboratory data, results of imaging procedures (Fig 1, 2), treatment and follow-up and unfortunately, a fatal outcome 3 months later due to advanced pulmonary fibrosis.

- We thank reviewer 1 for her/his comment.

Discussion: 739 words, the characteristics and the incidence of this syndrome are presented (high in Puerto Rico: 1/1,800 persons, approximately 1/22 persons are carriers of the gene; Western world: 1/500,000 and 1/1,000,000). The authors emphasize the fact that since 1980 they have already been several descriptions of granulomatous colitis associated with this syndrome. In the international literature are 9 cases of HPS where patients received anti-TNF α therapy. The authors also point to an important problem in dealing with these patients: the delay in diagnosing this rare condition. The discussion is appropriate, relevant.

- We thank reviewer 1 for her/his comment.

Conclusion: short, 51 words, the authors conclude on the effectiveness of corticosteroids, azathioprine and infliximab in the described female patient with severe colitis and HPS.

- We thank reviewer 1 for her/his comment.

References: 15, from the period 1959 (Blood) - 2018 (Nat Rev Gastroenterol Hepatol), references are appropriate.

- We thank reviewer 1 for her/his comment.

Conflict of interest: the authors declared no conflict of interest.

- We thank reviewer 1 for her/his comment.

Informed consent statement: the patient died before writing the report which is in accordance of the Declaration of Helsinki and its latter amendments.

- We thank reviewer 1 for her/his comment.

Opinion of the reviewer The manuscript is very interesting, I suggest to accept the contribution with minor language corrections.

- We thank reviewer 1 for her/his comment.

Reviewer 2 (01557050):

Dr. Girot, et al. reported 'Crohn's-like acute severe colitis associated with Hermansky-Pudlak syndrome: a case report'. The manuscript is informative and well-presented. The reviewer has no comments.

- We thank reviewer 2 for her/his comment.