Author's response to reviews

Title: Complicated Crohn's-like colitis associated with Hermansky-Pudlak syndrome, treated with Infliximab: a case report and brief review of the literature

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Manuscript entitled:

Complicated Crohn’s-like colitis associated with Hermansky-Pudlak syndrome, treated with Infliximab: case report and brief review of the literature.

COVER LETTER
Giving a point-by-point response to the concerns.

Referee 1

Revision 1: It seems that the case is a real Crohn’s colitis case thus the treatment success can not be generalized to rare HPS associated colitis cases in which ceroid deposition can be found in the granulomatous colitis. This limitation should be clearly included in the manuscript.

The necessary remark has been done and it is included in “Conclusions” in red.

Besides all these facts, the treatment success with infliximab is still based on limited experience and only in patients with HPS-associated Crohn’s-like enterocolitis and should not be generalized as the optimal therapeutic approach to all patients with granulomatous colitis complicating HPS.

Revision 2: A further important point is that authors should include some details on the differential diagnosis of colitis, especially details should be included on TB Testing.

Differential diagnosis including TB colitis is mentioned in “Case presentation- The patient” in red.

Differential diagnosis of colitis included granulomatous enterocolitis found in HPS-patients, ulcerative colitis, Crohn’s disease, tuberculous (TB) colitis and other forms of inflammatory bowel disease. For the possibility of tuberculosis of the intestine, at first, a tuberculin skin test was performed, which was negative.

All the mucosal biopsies from ileum and different colonic segments (included those from ulcer bases) were negative for Mycobacterium tuberculosis.

Revision 3: Please use x-ray instead of roentgenograms and mg/kg instead of mg/Kgr

Done
Referee 3

Revision 1: Pictures illustrating the case should be added (colonoscopy photos before and after treatment, image of the iris of the patient)

We added photos of colonoscopy before and after treatment but we don’t have image of the iris of the patient, since our ophthalmologist found only an horizontal nystagmus with reduced vision, with no pigmentation of the iris in both eyes.

Revision 2: How was HPS confirmed concerning: a. Tyrosinase-positive OCA?, b. Platelet dysfunction?

The diagnosis of HPS was done in another hospital three years before our involvement with the patient, with Ty-pos OCA and platelet dysfunction.

According to her history, HPS was diagnosed elsewhere in 2002 [tyrosinase-positive oculocutaneous albinism (Ty-pos OCA) and normal platelets count with small quantity of dense bodies (DB), without genetic linkage analysis].

In our hospital hematologists found that:
Baseline laboratory values were within normal range, including a normal platelet count \((401 \times 10^9/L)\) and no sings of hemorrhagic diathesis were observed. However, platelets were small in size (MPV 8fL). Platelet aggregation tests (PFA with collagen/ADP and collagen/epinephrine) were within normal range. Bone marrow aspiration revealed the presence of pseudo-Gaucher like appearance histiocytes.

Revision 3: (SI) is used in the text (forgive us but we didn’t found any other term)

Thank you very much for your useful and accurate remarks.
With best regards
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