Response to Reviewer's Comments

1) This case report was well organized and well investigated. This paper will give us a new information especially in the field of IBD, I have no claim in the present study.

Dear Reviewer, we thank you for your comments.

2) This is a very interesting case and well documented. I thought that a photomicrograph of the colitis and the ileal pathology might be very useful. In addition, is it possible to provide some figure or photograph on the NBT test?

Dear Reviewer, histopathologic view of colectomy specimen and NBT test results are provided and added to the case report as you recommended.

3) Dear Associate Editor, Thank you for sending the article (ID: 31525) for review. The manuscript reports a case of CGD that was treated as ulcerative colitis for a long period of time. Finally it was diagnosed of CGD was confirmed biologically by a nitroblue tetrazolium test (NBT) and flow cytometry-based dihydrorhodamine (DHR) or 2′7′-dichlorofluorescein diacetate (DCFDA) assays. There are some comments to improve the quality of the article as the followings: a-Please define clearly the criteria used for the diagnosis of ulcerative colitis. The diagnosis is based on the clinicopathological basis. Please explain this statement: "Surgical material found to be compatible with ulcerative colitis." b-Please demonstrate the pathological slides of colectomy sample in the manuscript. c-Please provide the Pelvic MRI and fistulography. d-Crohn disease is not ruled out in this case. In my opinion investigation about the mal absorption profile, ASCA assay, and MR enterography is of great importance in this case. e-The figure one and four could be substituted with colonoscopic view, MR fistulography and pathology slides.

Dear Reviewer, thank you for your recommendations. Here are answers for your comments.

a-b) Ulcerative colitis diagnose depends on clinical state, laboratory tests, colonoscopy, and histopathologic examinations. Bloody and mucoid diarrhea was present, acute phase reactants were elevated, pancolitis was noted during colonoscopy. Light microscopic examination of colectomy specimen revealed crypt distortion, cryptitis, crypt loss and crypt abscesses throughout whole colonic mucosa. Epithelial ulcerations, regenerative changes and pseudopolyps were also noted. Neutrophils were present in lamina propria; dense and mixed type inflammatory cell infiltration was seen. These changes were limited thorough mucosa and submucosa. Granulomas and dysplasia were not encountered. Necessary explanation and picture were added to the manuscript (Figure 2).

c) Pelvic MRI and fistulography were performed in other gastroenterology center and unfortunately we failed to provide these tests.

d-e) The diagnosis is made with the presence of a homozygous mutation in the NCF2 gene. The most likely diagnosis in a patient with ulcerative colitis with ongoing complaints following colectomy is Crohn's disease and must be strictly excluded. However, the absence of lesions in the terminal ileum and upper gastrointestinal system, the absence of granulomas and the involvement of the mucosa-submucosa in histopathologic examinations, the absence of intact tissues and presence of pancolitis in colonoscopy, any positive sign for Crohn's disease were not found, thus; Crohn's disease was ruled out. The lesions in the perianal region were not seen at the onset of the disease (case report section 2) and developed after colectomy. Immunodeficiency state which is detected later causes delay in wound healing.

4) Very interesting case report, but some points are not clear to me. - Were there any data about previous suppurative infections in the case history of the patient? If not, it is quite unusual in case of CGD. - Was the presence of any Mycobacterial infection excluded? - How was the diagnosis of UC establihed? - How were the patient's basic immunological parameters like ? (exact blood cell counts and percentages, immunoglobulin levels, complement levels) - Include the histological examination of the bowel. - Which type of CGD was diagnosed? - Was a cytochrom C reduction assay performed? What was its result? English language needs minor polishing. After major revision I suggest to accept the manuscript for publication.

Dear Reviewer, we thank you for your comments and questions. Here are the answers for your questions.

In patient's medical history there was no evidence for recurrent infections, fungi and tuberculosis. There was no consanguinity in family history. We found it appropriate to report this patient. Ulcerative colitis is diagnosed by clinical, laboratory, colonoscopy and histopathologic examinations. Bloody and mucoid diarrhea was present, acute phase reactants were elevated, pancolitis was noted during colonoscopy. Light microscopic examination of colectomy specimen revealed crypt distortion, cryptitis, crypt loss and crypt abscesses throughout whole colonic mucosa. Epithelial ulcerations, regenerative changes and pseudopolyps were also noted. Neutrophils were present in lamina propria; dense and mixed type inflammatory cell infiltration was seen. These changes were limited thorough mucosa and submucosa. Granulomas and dysplasia were not encountered. These findings are consistent with clinical, laboratory, colonoscopy and histopathologic findings of ulcerative colitis. After the patient's genetic screening result, the quantiferon level for Tbc was studied

in adult gastroenterology and found to be positive. Tuberculosis disease was excluded, the patient is considered to be infected with Tbc and under isoniazid treatment.

Percentile values of the patient's development and CBC values are indicated in the case report section paragraph 2. Immunoglobulin and complement values were not mentioned in the article before and added upon your request. The histopathologic findings of the colectomy material are given in the case report paragraph 3. Genetic studies showed NCF2 mutation for CGD which shows an autosomal recessive trend. Consanguineous marriage was not noted in family history, but coming from same village and the CGD disease with an autosomal recessive trait made us think of a distant relativity between parents. Cytochrom C reduction assay was not performed. Genetic analyses and NBT were performed.

English is edited by Enago, Crimson Interactive.

5. This is a paper of a clinical case in which a misdiagnosis of chronic granulomatous disease in an adolescent undergoing total colectomy for suspected ulcerative colitis is reported. The english language needs revision. This paper does add not information to current literature.

Dear Reviewer, thank you for your comments.

English is edited by Enago, Crimson Interactive.