LETTER TO THE REVIEWERS

Name of journal: World Journal of Gastrointestinal Surgery

Manuscript NO: 50688

Title: Isolated colonic neurofibroma in the setting of Lynch syndrome: a case report and review of literature

Reviewer's code: 00043396

A case report of an interesting albeit rare problem. The paper is well written and researched.

Response to Reviewer:

Thank you for your comments and feedback.

Reviewer's code: 00182114

This is a very interesting paper about colon neurofibroma with Hereditary nonpolyposis colon cancer (HNPCC). HNPCC is the most common hereditary colon cancer syndrome. It is characterized by multiple colon as well as extracolonic cancers such as endometrial, ovarian and urinary tract cancers. In addition, it is well known that some cases of HNPCC can present with unique tumor spectrums such as sebaceous tumors, which is often referred to as the 'Muir–Torre' syndrome. In recent years there have been a few reports of families presenting with early onset of colon tumors along with café-au-lait spots and/or hematologic malignancies often associated with homozygous mutations involving one of the mismatch repair genes. The available data

clearly highlight such presentations as a distinct clinical entity characterized by early onset of gastrointestinal tumors, hematologic malignancies as well as features of neurofibromatosis (easily remembered by the acronym `CoLoN'; Colon tumors or/and Leukemia/Lymphoma or/and Neurofibromatosis features). Furthermore, there has also been some evidence that the neurofibromatosis type-1 gene is a mutational target of the mismatch repair deficiency that is seen in families with HNPCC, and that mlh1 deficiency can accelerate the development of leukemia in neurofibromatosis (Nf1) heterozygous mice. This case is colon neurofibloma without café-au-lait and neurofibloma in skin. (Syndrome of early onset colon cancers, hematologic malignancies & features of neurofibromatosis in HNPCC families with homozygous mismatch repair gene mutations. Bandipalliam P) I ask some questions to author.
1 .Please comment relationship between HNPCC and Muir-Torre syndrome. 2.How about peripheral blood count?

Response to Reviewer:

Thank you for your comments and suggestions. We have addressed your questions below: (1) We revised our introduction and identified an association between Lynch Syndrome and neurofibromatosis, citing Bandipalliam (highlighted lines 100-101). However, given that our patient did not exhibit signs or symptoms of Nf1, we elected to not elaborate on this further. (2) We did mention the patient had a normal blood count under the investigations section. This would rule out concerns of hematologic malignancies.