Author’s response to reviews

Title: Ruptured Desmoid Tumor Imitating Acute Appendicitis – A Rare Reason for an Emergency Surgery

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Version: 1 Date: 08 Oct 2019

Author’s response to reviews:

Dear Reviewers,

We are grateful for your valuable comments and recommendations. As you rose some important points showing the need for a general revision of the paper, the BMC editors gave us the opportunity to revise our manuscript entitled “Ruptured Desmoid Tumor Imitating Acute Appendicitis – A Rare Reason for an Emergency Surgery.” So, we are so thankful for that.

Addressing the reviewers’ comments and explaining the changes in our revised paper:

Valeria Tonini, MD Ph.D. (Reviewer 1): It is certainly an interesting article but I believe that many things need to be revised even if the study looks well performed. First of all, I found in the whole article both some grammar mistakes that must be corrected and, also, some sentences that need to be rephrased and rewritten because they are difficult to understand. Secondly, my opinion is that before choosing to perform an urgent operation, an abdominal CT scan is always required. It could have shown the tumor and it could have given the possibility to operate the patient with a more specific diagnosis. Having said that, the paper clearly explained what is the best course of action when a soft-tissue tumor is suspected and why surgeons did not include further imaging tests in this specific case. Moreover, despite the article claims that the desmoid tumors can rarely lead to acute surgical abdomen requiring an emergency operation, the literature has a consistent number of case reports about this topic. But even so, to the best of my knowledge, this is the very first reported case in which the disease mimics acute appendicitis or cystitis, adding a relevant clinical manifestation that
could be taken into consideration for further cases. Finally, the authors reported a good number of references about the topic with related imaging.

- The paper was given to a native English speaker for language review. We hope that we rephrased all of the sentences that could be difficult for understanding.

- About the second concern: The problem in our case was the unusual presentation of the disease in the patient, mimicking acute appendicitis with signs of spreading peritonitis. Unfortunately, our ultrasonographic exam did not detect a tumor formation and in our differential diagnosis, we did not think about the possibility of a tumor, even more so rare one. So, for tumors, CT is one of the best imaging tests, but destructive appendicitis with spreading peritonitis was highly probable diagnosis. Because of that, further imaging tests were not considered to be needed and we decided to proceed with an emergency operation. Furthermore, due to the complicated urgent nature of the presented tumor with rupture, signs of peritonitis and the presence of hemoperitoneum an emergency operation was indicated, even in case we had have performed a CT. And because the tumor was resectable, the surgical strategy would be the same.

- Regarding the last point: We agree that there are several reports in the literature about desmoid tumors requiring emergent operation, but to the best of our knowledge, this is the first reported case in the literature with intraabdominal desmoid tumor with hemoperitoneum and peritonitis mimicking acute appendicitis. In the revised version, we included additional review of the similar cases in the literature emphasizing the differences between them and our case.

Reviewer 2: RELEVANCE - Does this case report make a contribution to medical knowledge, have educational value, or highlight the need for a change in clinical practice or diagnostic/prognostic approaches? No, the findings of this report are well known and/or similar reports have already been published. CASE DESCRIPTION - Are the details of the case sufficiently well described to understand the patient's symptoms and course of treatment? Yes, the description of the case is sufficient. DIAGNOSIS/INTERPRETATION - Based on the facts presented, are the diagnosis, interpretation, and course of treatment medically sound? Yes, the work described is medically sound. DISCUSSION OF THE CASE - Does the discussion appropriately analyse the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment? Has an adequate literature review pertinent to the case been included? No – there are major issues. OVERALL MANUSCRIPT POTENTIAL - Could an appropriately REVISED version of this work represent a technically sound contribution? Maybe - with major revision.

PEER REVIEWER COMMENTS: GENERAL COMMENTS: The report is well written, clinical presentation and the diagnostic problem is well discussed. However, the novelty of the case is questionable: previous publications underline the risk of bleeding or bowel perforation due to intestinal fibromatosis (see for instance Li J et al 2019, Georgides et al 2012). There is a general message (that authors correctly underline in the discussion) please do as much as you can before opening and abdomen (i.e. do a CT scan, but this was not performed in the case presented). The weakest part of the manuscript concerns the therapeutic
options for desmoid tumors presented in the discussion: a) surgery is no longer the standard of care and should be considered with some caution due to the high risk of relapse, b) medical treatment is moving more and more from tamoxifen +/- NSAID to low dose chemotherapy as first lien treatment in symptomatic not operable patients, c) there is an increasing evidence that tyrosine kinase inhibitor and especially Sorafenib may be active agents against fibromatosis, d) radiotherapy is much less used than in the past. More importantly in many cases a wait and see strategy may be proposed (not in the case described of course). Please see Kasper et al 2017 (A European Consensus). Some recent references may be considered: i.e. Ebeling et al 2019, Takemoto et al 2019.

REQUESTED REVISIONS: Major issue: Discussion should be carefully revised as I have previously written. Minor issues: guidelines and consensus have been published, so what is written in the background second paragraph is not correct. Why Abdominal CT scan was not performed before surgery. The authors may also comment on the fact that despite tumor rupture no contamination of the abdominal cavity occurred (and they can check if this has been the case also for previously published cases of intrabdominal fibromatosis). This can be a valuable information for clinicians.

- As we have mentioned above, we agree that there are several reports in the literature about desmoid tumors requiring emergent operation, but to the best of our knowledge, this is the first reported case in the literature with intraabdominal desmoid tumor with hemoperitoneum and peritonitis mimicking acute appendicitis. We have reviewed the suggested articles in detail (Li J et al 2019, Georgides et al 2012), but these cases are different from ours. C. Georgiades et al. (2012) presented a tumor strangulating the splenic artery and branches of the artery to the pancreas. We could not find a case presenting with intraabdominal hemorrhage from mesenteric or small bowel fibromatosis, as in our case. Jian Li et al. (2019) reported another unusual presentation of intestinal perforation and purulent peritonitis as an onset of the tumor symptoms. Again, our case is different because there was no perforation of a hollow organ but rupture of the tumor pseudo capsule with subsequent hemoperitoneum. So we include these references in our discussion section to emphasize the rarity of our case.

- Regarding the question about preoperative CT – Of course, the CT is one of the best imaging tests for tumors. But the ultrasound did not detect a tumor formation and in our differential diagnosis, we did not think about the possibility of a tumor, even more so rare one. So destructive appendicitis with spreading peritonitis was highly probable diagnosis. Because of that, further imaging tests were not considered to be needed and we decided to proceed with an emergency operation. Furthermore, due to the complicated urgent nature of the presented tumor with rupture, signs of peritonitis and the presence of hemoperitoneum an emergency operation was indicated, even in case we had have performed a CT. And because the tumor was resectable, the surgical strategy would be the same.

- We appreciate the valuable recommendations about the Discussion section. In the previous version, we have wanted to emphasize the surgical strategy in regards to our case (a resectable intraabdominal tumor, especially with emergent complications). Even the surgery is not a unified treatment of all desmoid tumors, the other options are not applicable in our case. Our goal was not to present a detailed review of all treatment strategies in the previous version because we believed it should be a
purpose for the original and review articles. But now, we understand that the Discussion section might have been misleading, so we revised it and added the other options for treatment of desmoid tumors, the indications for them, according to the latest guidelines and consensuses. We are thankful for the recommendation of Ebeling et al 2019, Takemoto et al 2019 (we used them for the revised article), but it was impossible to be included in our primary version of the manuscript, because it was submitted in 2018.

- About the last comment on the tumor rupture and possible contamination of the abdominal cavity: The aggressive fibromatosis is a benign disease with “malignant” local behavior due to the infiltration of the surrounding tissues, organs, and vessels. So, it cannot metastasize and the tumor rupture does not influence the possibility of tumor relapse. For the surgically treated cases, the status of the resection margins is the most important risk factor for recurrence.

Thankfully,

Yavor Asenov