Author’s response to reviews

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

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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 revision of the paper, the BMC editors gave us the opportunity to revise our manuscript again. So, we are so thankful for that.

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

Valeria Tonini, MD PhD (Reviewer 1): The new revised paper now covers all the points I personally suggested to revise. Any other minor essential revision is present on this manuscript. For this reason I think it could be published on a scientific journal.

Dear Reviewer (Valeria Tonini, MD, Ph.D.), we are so grateful for your comments and we hope the revised article will be valuable for the audience.

Reviewer 2 (Reviewer 2): REVISION ASSESSMENT FROM THE ACADEMIC PEER REVIEWER: Has the author addressed your concerns sufficiently for you to now recommend the work as a technically sound contribution? No

Reviewer comments: I have appreciated the effort of the authors to improve the manuscript but I still see major points that need to be addressed:
1) The treatment of desmoid tumors is reported in the discussion but, in my opinion, what the authors report is not completely in line with current recommendations and practice.

2) There are a number of sentences that are not clear and even if the authors had been supported by a professional translator the manuscript still needs language revision.

We appreciate your comments and recommendations. About the first comment, our goal was not to present a detailed review of all treatment strategies because we believed it should be the purpose of the original and review articles. We emphasized the difficulties in diagnosis and treatment of such an unusual case, presenting with the clinical picture of acute surgical abdomen.

Regarding the language, even the manuscript had been already revised, we sent it to AJE for language editing. Several corrections were made by the editors, so now we attached a certificate for the quality of the language of the manuscript as supplemental material. The certificate may be verified on the AJE website using the verification code 321D-4941-4423-D57C-65DA.

REQUESTED REVISIONS:

I report below some examples:

Page 3 Line 12-13: … and have dual behaviour. Please explain what the authors mean by dual behavior (indolent or aggressive course?)

Dear reviewer, thank you for your comments. The phrase “dual behavior” was explained in the next sentence in the manuscript “Although these tumors are benign neoplasms without metastatic potential, they can affect every part of the body, can be locally aggressive and have a high recurrence rate”. So, aggressive fibromatosis is a benign disease with a possible local “malignant”-type course.

Page 3 Line 20-21: Consider cancelling "Despite the progress in medicine" (this is a very generic expression)

We appreciate your recommendation, so we canceled this phrase.

Page 3 Lines 23 to 25: "because of their low incidence……." this is a confusing sentence. At least I would cancel the word extensive and I do not think the word respectively is correct here.

Thank you for your recommendation – we revised the whole sentence.

Page 3 Line 25-28 "even more…"authors added this sentence but, in my opinion, it does not fit well here

We do not agree with this comment. Desmoid tumors are rare and the disease can have different behavior (asymptomatic to locally aggressive with a high recurrence rate). Because of this, even in published guidelines, there are so many possible treatment options. Even more challenging in diagnosis and treatment are situations in which the aggressive
fibromatosis leads to peritonitis, requiring emergency operations. This sentence is the bond to our unusual case.

Page 3 Line 51 "before two days" I think should be "two days before"

We agree with the recommendation and the sentence was revised.

Page 3 Line 58: "were in referent limit". Is this correct? probably better within normal ranges

We agree with the recommendation and the sentence was revised.

Page 4 Line 37 ...."in negative margins..." i.e. resection margins with no evidence of tumor: this is established by the pathologist so should go in the next paragraph

We agree with the recommendation and we made corrections in the order of these sentences.

Page 4 line 55: I am not aware that laboratory tests are useful for a desmoid tumor follow up

The blood test cannot show specific changes that are characteristic of desmoid tumors or their recurrence. But these tests are valuable because they are part of the assessment of the quality of the patients' health. Therefore, the laboratory tests are included in the protocol for follow-up.

Page 5 line 23. The diagnosis usually is delayed.... I think the authors here refer to intraabdominal desmoid, please specify

We agreed with this comment, so we specified that the diagnosis of intraabdominal desmoid tumors is usually delayed due to the lack of specific symptoms and often asymptomatic behavior.

Page 5 line 23: I think that "a case" should be "any case", please check

Thank you for your recommendation. We replaced “a case” with “any case”.

Page 6 line 13 to 27: Very confusing sentence. When the authors write about differential diagnosis they should clarify better. In the clinical differential diagnosis of bowel masses other more frequent tumors should be taken into account (i.e. G.I. carcinoma, lymphoma,…). If this means histological differential diagnosis, they should better specify this.

According to your comment, we revised the sentence.

Page 7 line 5: I think asymptomatic here should be symptomatic, otherwise it is in contradiction with the previous paragraph

We agreed with this comment. We had missed the space between “a” and “symptomatic” and this mistake changed the meaning. We corrected it.

Page 7 line 12-14: the authors suggest the use of radiotherapy in case of R1 or R2 resection. I think the use of radiotherapy should be considered taking into account the presence of
symptoms and the risk of acute and late toxic effect and only after medical treatment has been considered.

The paragraph was revised. We specified that radiotherapy is rarely used for the intraabdominal localization of the disease because of bowel toxicity.

Line 24: The authors use the term high risk without explaining what this means (symptomatic patients? when the tumor mass invades or is at risk to invade organs?)

The term “high-risk patients” means cases where surgery would be unacceptably morbid. The term is not related to the tumor features.

Discussion: Authors should clarify if they are discussing treatment options for desmoid in general or for abdominal desmoid

We discussed briefly treatment options for desmoids with an emphasis on the management of abdominal desmoid tumors. As we mentioned above, our goal was not to present a detailed review of all treatment strategies because we believed it should be the purpose of the original and review articles. We emphasized the difficulties in diagnosis and treatment of such an unusual case, presenting with the clinical picture of acute surgical abdomen.

In my opinion tamoxifen is no longer the first line medical therapy. MTX and vinorelbine/vinblastine are increasingly used

According to your comment, we revised the whole paragraph.

Please note that a very recent paper has been published in Lancet Oncology that may be of interest for the discussion: Toulmonde M et al, Lancet Oncol. 2019 Sep;20(9):1263-1272.

Thank you for that suggestion. We included this reference in the manuscript.

Thankfully,

Yavor Asenov