Peer Review File

Article information: https://dx.doi.org/10.21037/pm-21-28

Reviewer comments

Reviewer A:

The authors describe a very challenging case of desmoid in a young person.

Comment 1: The title is not reflective of what happened to the patient since desmoid grew despite RT - please change the title

Reply 1: Thank you for the comment. The title has been changed to better reflect the outcomes of our patient.

Changes in the text: Line 1-2 now reads “Management of Rapidly Growing Recurrent Extra-Abdominal Pediatric Desmoid Tumor: Case Report”

Comment 2: International guidelines extol the importance of avoiding surgery for desmoids and rather using medical therapy or watchful waiting as first-line approaches. This philosophy needs to be emphasized in the discussion

Reply 2: For patients with asymptomatic and/or stable desmoid tumors, watchful waiting (“wait-and-see”) is recommended and emphasized while being described as the “best option for therapy” in the second paragraph in the discussion (lines 150-155). Line 157-158 we indicate there is ongoing controversy on best mode of therapy for tumors that are symptomatic and continue to progressively grow. The remaining discussion is a review of the literature of outcomes using different modes of therapy that include medical (radiation and chemotherapy) and surgical management.

Changes in the text: No changes were made to the text in this regard.

Reviewer B:

The authors reported a rapidly growing recurrent extra-abdominal pediatric desmoid tumor. The authors described a rare case. I think this is an interesting topic.

Comment 1: 1. Since the tumor showed the best response due to radiotherapy, a detailed description is required. How was the Radiation field? If possible, please show an image of the radiation field. Also, if there is a gross image or MRI image before and

after radiotherapy, show these images.

When was the maximal tumor response after radiotherapy?

“He underwent serial debridement with removal of a significant amount of necrotic tumor with a >70% reduction in tumor size.”: At this time, how long has it passed since the radiotherapy?

Reply 1: Detailed description of the radiotherapy regimen is described in lines 109-115. Additional details were added to the text (lines 115-119), including figures depicting the radiation field and dose distribution. Figure 3 and Figure 5 now depict gross images of the tumor pre- and post-radiation therapy, respectively. Maximal tumor response was within one week of radiation therapy. Line 122-124 we describe debridement of necrotic tumor began 1 week after completion of radiotherapy. Tumor remained stable afterwards, until recurrent growth was noticed 6 months later – these details were added to the manuscript.

Changes in the text: Lines 115-119 have been added to the text as additional details on the radiation field, they read as follows “Figure 4 depicts a summary of the radiation fields and dose distribution. For target delineation, a gross tumor volume (GTV) was contoured including any visible tumor on all available imaging. The clinical target volume (CTV) included GTV with a 2-cm margin constrained by natural anatomic boundaries and a planned target volume (PTV) of 0.5-cm was used to account for potential set up errors.” Figure 3 has been changed, now depicts gross image of the tumor prior to radiotherapy. An additional image as been added to Figure 5 (previously labeled Figure 2), depicting tumor bed post-radiotherapy after the completion of debulking of necrotic tumor and placement of split thickness graft. Lines 126-127 now reads “The tumor remained stable after the dramatic response to radiotherapy, unfortunately it lasted merely six months, when evidence of tumor growth reappeared in the tumor bed.”

Comment 2: Figure 3: How long has it passed since the debridement?

Reply 2: Figure 3 is now labeled Figure 6. Figure depicts tumor 12-months after post-radiotherapy debridement. This detail has been added to the manuscript.

Changes in the text: Lines 129-130 now reads “…requiring re-excision 12-months later and consideration for new experimental trial therapies.”

Comment 3: At first evaluation, the patient had several palpable asymptomatic tumors about the scalp, ribs, back, and sternum, the largest of which was ~4cm.“:
Didn't the aggressive feature of tumors appear anywhere other than the back?
Reply 3: The aggressive features were only present on the back. All other tumors either remained stable or regressed. This detail has been added to the manuscript.
Changes in the text: Lines 87-88 “…whereas other lesions on the scalp, ribs, and sternum remained stable or regressed.”

Comment 4: “At this time, MRI demonstrated the largest mass measuring 20 x 6.5 x104 18.4cm with the involvement of the vertebral lamina and transverse processes with extension into the spinal canal at T10-T11 without spinal cord involvement.”:
Can you show an MRI image?
Reply 4: MRI image has been added to the manuscript.
Changes in the text: Line 96 now references Figure 1.

Reviewer C:
Comment 1: It would be interesting to have more information about the histology characteristics of the recurrent disease.
Reply 1: Histology remained unchanged from previous evaluation (aggressive fibromatosis). This detail was added to the manuscript.
Changes in the text: Lines 101-102 “Pathologic examination of the tumor was consistent with aggressive fibromatosis.”

Comment 2: Furthermore, it would be important specify if there is a difference, in terms of behavior, site of occurrence, biology and response to treatment, between desmoid cancer in FAP and sporadic disease. (line 141-143).
Reply 2: Thank you for this comment. Further details have been added to the manuscript.
Changes in the text: Lines 144-148 now read: “The risk of developing desmoid tumors is much higher in the setting of FAP where they are a significant source of morbidity and mortality as they are commonly found to present earlier in life (compared to sporadic tumors), in males, and located intra-abdominal or within the abdominal wall

(as opposed to extra-abdominal as typically seen in sporadic tumors), with a fondness for previous surgical sites (2).”