

Peer Review File

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#Reviewer A

Comment 1: I suggest that you should have comments about small sample size and the short follow-up in the end of discussion, as a limitation. Also, I have some comments and suggestions to improve this article as follows.

Reply 1: We have modified our text as advised (see Page 14, line 16-18)

Changes in the text: Page 14, line 16-18

Comment 2: Over 200-350 words (the word is limited within 200-350 words in original article). Please reduce the words under the max according to the author guideline.

Reply 2: Thank you for your reminding, we have declined our abstract to 347 words as advised (see Abstract)

Changes in the text: Abstract

Comment 3: Page 1, line 27: Let me know if you followed the patients for at least 15 months after diagnosis.

Reply 3: Yes, we followed up the patients for at least 15 months after diagnosis.

Comment 4: Page 1, line 29: Let me know if 1-year OS mean after diagnosis (or after treatment).

Reply 4: 1-year OS mean after diagnosis, and we added a sentence "the time since diagnosed" in the text (see Page 5, line 19)

Changes in the text: Page 5, line 19

Comment 5: Page 2, line 19: Malignant: please do not use capital letter here.

Reply 5: Thank you for your reminding, we have modified our text as advised (see Page 3, line 10)

Changes in the text: Page 3, line 10

Comment 6: Page 3, line 18: As shown in Table1, Case 2 is 10 years old and Case 5 is 6 days old. However, you mentioned the patient ages are ranging from 9 days to 8 years. Which is correct?

Reply 6: Please accept my deepest apologies as typing mistakes about the case2(10y27d) and case3(1y11d), we searched the primary data and find that only the "*y*d" mode is wrong, which means "*m*d", we have modified our text as case2(10m27d) and case3(1m11d) (see Table 1, case2/3). Fortunately, they don't influence the results. We have modified our text as "6 days to 8 years" (see Page 5, line 14)

Changes in the text: Table 1, case2/3; Page 5, line 14

Comment 7: Page 2, line 37: Does "survival time" mean survival time, after onset or after

diagnosis, or after treatment?

Reply 7: “survival time” mean survival time after diagnosis. We have modified our text from “Age” to “Age of onset” in Table 1. (see Table 1)

Changes in the text: Table 1

Comment 8: Page 4, line 4: I think that 5 patients with MRTK had lung metastasis, meaning in stage IV. But you said only 3 patients were in stage IV. Do you mean that 5 patients had lung metastasis at diagnosis and the lung metastases in 2 of them were disappeared after chemotherapy/radiotherapy?

Reply 8: Thank you for this valuable suggestion, the staging standards are according to the International Society of Pediatric Oncology (SIOP) of renal tumor preoperatively. Only 3 patients were in stage IV from the beginning, but another 2 patients with MRTK had lung metastasis with the time goes. Our conclusion in the text is” MRTK and EERT are both insensitive to radiotherapy and chemotherapy” (see Page 13, line 22).

Changes in the text: N/A

Comment 9: Many patients are under chemotherapy and observed in too short duration. You should follow-up more time to say the prognosis if you want to evaluate the prognosis.

Reply 9: Thank you for this valuable suggestion. Since rhabdoid tumor is rare and most children died within 1 year after diagnosis. The time for progression is usually short and the patients who relapse generally do not survive. This article aims to sharing about some of the clinical features, treatment and early prognosis of malignant rhabdoid tumor, so we took the cases who are under chemotherapies into the cohort, and the follow up of those patients will be lasted. We have modified our text from “prognosis” to “early prognosis”. (see Page 1, line 16)

Changes in the text: Page 1, line 16

#Reviewer B

Comment 1: There is not adequate assessment of how stage plays into these analyses as most of the EERT patients seem to be low stage while MRTK patients were of higher stage. SIOP/COG data suggest that lower stage MRTK carries a better prognosis so the differences identified may not be there.

Reply 1: Thank you for this valuable suggestion. There is no definite official guideline of the EERT, and also no specific staging standard about the EERT while MRTK followed the SIOP preoperative renal tumor staging standards. We declared that EERT and MRTK are both “highly aggressive” (see Page 3, line 1), and the “average survival time” of MRTK is longer (6.60±3.28m) than EERT (1.40±0.65m) (see Page 28, Table 2), the children who go with tumor total excision in EERT group would have better prognosis than MRTK.

Changes in the text: Page 3, line 1; Page 28, Table 2

Comment 2: Lines 14-15, page 1 - The wording is suggestive that today, we would mistake Wilms for MRTK which is incorrect. I would change the wording to clarify this.

Reply 2: Rhabdoid tumor is very rare and the clinical features aren't clear, this article aims to sharing about some of the clinical features, treatment and early prognosis of malignant rhabdoid tumor, and promote the understanding of MRT.

Comment 3: Line 24, page 1 - The "prognosis is the worst" is not entirely correct. There are other brain tumor entities with poor prognosis such as DIPG. Would clarify this.

Reply 3: Thank you for this valuable suggestion, "prognosis is the worst" refers that the prognosis of AT/RT is the worst among MRTK、EERT and AT/RT. We had changed it in the article. (see Page 3, line 17)

Changes in the text: Page 3, line 17

Comment 4: Studies on MRTK or ATRT etc... usually have follow up time frames longer than 15 months. Can the authors comment on why such a short follow up?

Reply 4: Thank you for this valuable suggestion. Since rhabdoid tumor is rare and most children died within 1 year after diagnosis. The time for progression is usually short and the patients who relapse generally do not survive. This article aims to sharing about some of the clinical features, treatment and early prognosis of malignant rhabdoid tumor, so we took the cases who are under chemotherapies into the cohort, and the follow up of those patients will be lasted.

Comment 5: Was the germline tested for SMARCB1 mutations?

Reply 5: Yes, some of the cases tested for SMARCB1. Most of them were SMARCB1 mutation, a few cases were SMARCB1 deletion.

Comment 6: Line 43, page 3 - I don't understand what the authors mean by "early stage of the disease" There needs to be more of a discussion of staging of your patients and if there was bias of stage of disease based on EERT vs MRTK - right now it is in page 4 lines 13-18. Is the reason the EERT has improved outcomes that EERT patients presented with lower stage?

Reply 6: Please accept my deepest apologies as making you confused, we have modified our text from "stage" to "period", the "stage" didn't mean the tumor stage. (see Page 6, line 4/20)
Changes in the text: Page 6, line 4/20

Comment 7: Line 7, page 5 - medulloblastoma and rhabdomyosarcomas are not known to have loss of SMARCB1/INI1.

Reply 7: Thank you for your reminding, we have modified our text as advised. (see Page 9, line 9)

Changes in the text: Page 9, line 9

Comment 8: Otherwise, in Table 1, would be good to understand which the authors defined as EERT.

Reply 8: Yes, the first ten cases are EERT in Table 1.

Comment 9: Finally, the manuscript should include some discussion on the chemotherapy used to treat these patients (this is in addition to the discussion where you talk about COG/SIOP treatment strategies).

Reply 9: Thank you for this valuable suggestion. We have modified our text as advised. (see Page 13, line 15-16,19-21)

Changes in the text: Page 13, line 15-16,19-21