

## Comment and reply

### Reviewer 1

**1. Comment:** In the introduction section, more information about the disease's morbidity, diagnosis, prognosis etc. Should be also briefly introduced and reviewed.

**Reply:** Thanks for your advices. We have added the disease's morbidity, diagnosis, prognosis of P-EHE in the introduction section.

**2. Comment:** In the discussion section, more references should be cited and reviewed to elucidate the physiological and pathological features of the disease and then its response to the recommended therapy.

**Reply:** Thank you very much. We have cited some references for physiological and pathological features of disease in the discussion section.

**3. Comment:** The manuscript displays some language errors that need to be carefully rectified.

**Reply:** Thank you. We have made changes in some language errors.

### Reviewer 2

**1. Comment:** Please provide more detail regarding treatment outcome and follow-up.

**Reply:** Thanks for your advices. We have added relative information in the “TREATMENT OUTCOME AND FOLLOW-UP” section.

**2. Comment:** In the current version of the manuscript it was stated that “The clinical status of the patient showed dramatic improvement with resolution of chest pain, chest tightness, and cough”. It would be very important to state how long it took after the start of apatinib + doxorubicin + cyclophosphamide for the patient to start experiencing clinical improvement in terms of improvement / resolution of chest pain and cough.

**Reply:** Thanks for your advices We have added relative information in the “TREATMENT OUTCOME AND FOLLOW-UP” section.

**3. Comment:** It would also be very important to provide information regarding how long the patient was under the combined therapy and when the patient stopped taking doxorubicin + cyclophosphamide.

**Reply:** Thanks for your advices. We have added relative information in the “TREATMENT OUTCOME AND FOLLOW-UP” section.

**4. Comment:** It is not clear whether and when (?) the patient became completely asymptomatic during follow-up.

**Reply:** Thanks for your advices. We have added relative information in the “TREATMENT OUTCOME AND FOLLOW-UP” section.

**5. Comment:** Page 6, second paragraph: “The P-EHE patient who received apatinib for a month showed dramatic improvements in clinical presentation and on CT imaging.” Please substitute “A patient” for “The patient” in that sentence because there was no prior information about the case of that patient.

**Reply:** Thanks for your advices, We have made changes according to your advices.

**6. Comment:** Was it because of that case report that the authors decided to try apatinib in their own patient?

**Reply:** Thanks for your advices. P-EHE with metastatic potential has an epithelioid appearance; apatinib selectively binds to VEGFR-2 as a tyrosine kinase inhibitor, and is therefore considered as a treatment option.

**7. Comment:** Page 2, Abstract, case summary: Please substitute “gefitinibe” for “Iressa”. The same observation applies to the last paragraph of page 3. Page 5, first paragraph: please substitute “confirmed the diagnosis of P-EHE” for “conformed the diagnosis of P-EHE”.

**Reply:** Thanks for your advices. We have made changes according to your advices in manuscript.

**8. Comment:** Page 5, first paragraph of the discussion section: “EHE demonstrates a low-to-intermediate malignancy and has metastatic potential...”. That sentence is not clear. What does low-to-intermediate malignancy mean?

**Reply:** Thanks for your advices. We have made explanation about “low-to-intermediate malignancy” meaning in the discussion section. EHE not only has composition of solid nests and short cords of epithelioid endothelial cells in myxohyaline stroma, but also shows the presence of increased mitotic activity and necrosis and greater nuclear atypia. So, EHE is considered as low-to-intermediate malignancy.

**9. Comment:** Page 6, first paragraph: please substitute “Thereafter, the patient did not undergo…” for “Thereafter, the patient had not undergo…” Additionally, please describe in that sentence for how long that patient was followed up.

**Reply:** Thanks for your advices. We have made changes according to your advices and added followed up time in the “TREATMENT OUTCOME AND FOLLOW-UP” section.