World Journal of Clinical Cases

RE: A Case Review of Undifferentiated Intimal Sarcoma of the Pulmonary Artery

Dear editor:

We would like to thank **World Journal of Clinical Cases** for giving us the opportunity to revise manuscript. We have carefully taken the comments into consideration in preparing our revision, which has resulted in a paper that is more clear and compelling. The point-by-point responses are attached after this letter. The revisions were highlighted to the text with tracked, have been prepared.

The manuscript has not been published previously, in any language, in whole or in part, and is not currently under consideration elsewhere. None of the authors have any competing financial interest to report.

Thank you for considering our manuscript for publication in your esteemed journal.

Response to Reviewer #1

General comments: The manuscript by Li et al. describes an interesting case of pulmonary arterial sarcoma (PAS). While the clinical case is worth reporting because PAS is such a rare disease, the manuscript is not well written. I have a number of questions in which I can't tell if the authors are describing the current patient or reviewing the literature. These questions will need to be resolved to consider acceptance of the manuscript.

Question 1: I think my biggest concern is that I am not certain why the authors seem to indicate throughout the manuscript that the diagnosis of PAS was in question at the time the patient presented to their institution. Was not the biopsy proving PAS done in 2017? If so, the authors' radiographic findings are of value to confirm the diagnosis, but the diagnosis was already clear. Since the diagnosis was already known, the authors could use this case as an example of the difficulties of diagnosing PAS – their 4 features in the Core Tip are quite useful – but should extensively revise the manuscript accordingly, and not make the reader feel as if the biopsy-proven diagnosis is in question.

Response: We appreciate the reviewer given this kindly comment. Patients was diagnosed with pulmonary artery sarcoma by pathological biopsy in other hospitals before receiving echocardiography examination in our hospital. Therefore, this study focused on the retrospective analysis for the treatment of pulmonary artery sarcoma. We have already made corresponding changes in the title and the contents of manuscript with tracked.

Question 2: I have difficulty following which imaging/procedures were done for this patient during his initial presentation to the outside hospital in 2017, and which were done at the authors' institution in 2019. The authors need to make this clear.

Response: Thanks for this suggestion. We have already made these changes into: 'In July 2017, PET-CT scanned in another hospital discovered space-occupying lesions in the right lung and multiple metastases in both lungs. The lesions of the right lung were biopsied, and pathology revealed undifferentiated sarcoma. Chemotherapy had been performed since July 2017. Doxorubicin and ifosfamide was administered for 20 courses of treatment, followed by radiotherapy for the right lung lesion in November 2019. In November 2019, the PET-CT images showed that the tumor activity was inhibited after chemoradiotherapy for hilum of right lung. But multiple nodules and patchy images of both lungs and nodules at superior vena cava were found, indicating occurrence of metastasis. The patient was admitted to our hospital in December 2019，whose CT images scanned in our hospital showed multiple nodules and patchy images in the right lung.'

Question 3: Since this submission is for the World Journal of Clinical Cases, I think readers would like to know what happened to the patient after the authors confirmed the diagnosis. Any history of treatment after his initial presentation in 2017 would be welcome too, since the patient did not present to the authors until over 2 years from his initial diagnosis, which is much greater than the median survival time for PAS without therapy.

Response: Thanks for this suggestion. Chemotherapy had been performed since July 2017. Doxorubicin and ifosfamide was administered for 20 courses of treatment, followed by radiotherapy for the right lung lesion in November 2019. In November

2019, the PET-CT images showed that the tumor activity was inhibited after chemoradiotherapy for hilum of right lung. But multiple nodules and patchy images of both lungs and nodules at superior vena cava were found, indicating occurrence of metastasis. Moreover, In December 2019, he received CyberKnife treatment in our hospital, he was discharged in January 2020 and died in the following month.

Question 4: I cannot tell in the last two paragraphs of the Discussion regarding references 8, 9 and 10 whether the authors are discussing aspects of this particular case, or are describing general characteristics of PAS patients from the literature. The authors need to clarify whether they are discussing their patient or findings from the literature.

Response: Thanks for this suggestion. The last two paragraphs of the Discussion section have already changed in the revised manuscript.

Question 5: Abstract Background, page 2, last line and also Introduction, page 3, last line: the word “by” makes it sound as if the patient is choosing which imaging study to get, not the physicians.

Response: Thanks for this suggestion. We have already made these changes in the revised manuscript.

Question 6: Abstract Case Summary, line 5 and also Case Presentation, page 5, line 5: By “punctured” do you mean biopsied, or more specifically a fine needle aspiration? Please clarify.

Response: Thanks for this suggestion. We have already changed “punctured” into “biopsied”.

Question 7: Abstract Case Summary, last line and also Case Presentation, page 6, last line of top paragraph: As noted above, the radiographic studies only make one suspicious of the diagnosis; the diagnosis is not confirmed until the biopsy is done. The authors should revise the wording in these sections to something like “suspicious of PAS” not that PAS was confirmed radiographically.

Response: Thanks for this suggestion. We have already made these changes in the revised manuscript.

Question 8: Core Tip, page 3 and Conclusion, page 8: To use consistent language,

point 4 should say something like “ultrasound of the lower extremity and inferior vena cava can be used to exclude fails to demonstrate thrombus”

Response: Thanks for this suggestion. We have already made these changes in the revised manuscript.

Question 9: Introduction, page 4: I would add myxoma as another possible diagnosis.

Response: Thanks for this suggestion. The myxoma have already added in the revised manuscript.

Question 10: Case Presentation, first paragraph, page 5: Are any of the original imaging studies from 2017 available? If the biopsy in Fig. 3 was done in 2017, the authors should present that here, not in the Discussion. Line 7, nodules, not nodes. Line 8, should say “At that time” rather than “currently”. Did the patient receive any treatment in 2017? Two year survival is a long time for PAS patients without treatment.

Response: Thanks for this suggestion. We have already made these changes in the revised manuscript. Moreover, the treatment strategies have already presented in Results section.

Question 11: Case Presentation, second paragraph, page 5: The video is good for showing the mass in the RVOT and its movement, but it is difficult to see blood flow in such a short clip. Second to last line: recommend deleting “with RVOT-MPA of mild stenosis as well as development of the mass”.

Response: Thanks for this suggestion. We have already made these changes in the revised manuscript. Moreover, the “with RVOT-MPA of mild stenosis as well as development of the mass” have already removed in the revised manuscript.

Question 12: Page 6: The last two paragraphs of the Case Presentation seem better suited for the Discussion.

Response: Thanks for this suggestion. These 2 paragraphs have already moved into Discussion section.

Question 13: Case Presentation, page 6, bottom paragraph, sentence beginning with “Second”: I again am having difficulty determining if the authors are referring to their patient, or PTE patients in general. Did this patient ever have an elevated D-dimer,

and if so, when?

Response: Thanks for this suggestion. The purpose of this sentence indicated mostly patients presented with pulmonary embolism was complicated with lower extremity vein or inferior vena cava thrombosis. This patient was pathologically diagnosed as PAS in 2017, then he admission in our hospital. Therefore, the data regarding D-dimer were not available.

Question 14: Discussion, page 7, first paragraph: I am not familiar with the term "sarciniform"; please use another descriptive word.

Response: Thanks for this suggestion. We have already made these changes in the revised manuscript.

Question 15: Discussion, page 7, second paragraph: Once again, are the authors referring to CT images of their patient, or of PAS patients in general since they cite reference 8? If the authors have such images from their patient, please include them. Please insert (Fig. 2) on the bottom line of page 6 after MPA.

Response: Thanks for this suggestion. Mostly PAS patients showed signs of pulmonary artery filling defect in different degrees on CT. This patient just admits in our hospital for 2 weeks, and the imaging modality and data were not available.

Question 16: Discussion, page 8, top paragraph: Again, it is unclear if the authors are referring to PA wall erosion and tumor/thrombus volume in their patient, or reviewing PAS vs. PTE characteristics in general from the literature; please clarify.

Response: Thanks for this suggestion. PAS could increase the secondary right ventricular resistance load, pulmonary artery blood flow speed increased, hemodynamics like pulmonary artery stenosis, and central pulmonary embolism although also cause right heart enlargement, but more complicated with pulmonary hypertension, pulmonary artery hemodynamic characteristics can be used as the differential diagnosis clues of pulmonary artery thromboembolism and pulmonary artery primary tumor. We have already addressed this question in Discussion section.

LANGUAGE QUALITY

Please resolve all language issues in the manuscript based on the peer review report. Please be sure to have a native-English speaker edit the manuscript for grammar,

sentence structure, word usage, spelling, capitalization, punctuation, format, and general readability, so that the manuscript's language will meet our direct publishing needs.

Response: Thanks for this suggestion. The language of the manuscript have already performed by native English speaker.

Response to editor

Science editor

Scientific quality: The manuscript describes a case report of the undifferentiated intimal sarcoma of the pulmonary artery diagnosed by echocardiography and analysis of its differential diagnosis.

The topic is within the scope of the WJCC. (1) Classification: Grade C; (2) Summary of the Peer-Review Report: The manuscript describes an interesting case of pulmonary arterial sarcoma. While the clinical case is worth reporting because pulmonary arterial sarcoma is such a rare disease. The questions raised by the reviewers should be answered; and (3) Format: There are 3 figures. A total of 10 references are cited, including 7 references published in the last 3 years. There are no self-citations.

Language evaluation: Classification: Grade C. A language editing certificate issued by FSeditor was provided.

Academic norms and rules: The authors provided the written informed consent. No academic misconduct was found in the Bing search.

Supplementary comments: This is an unsolicited manuscript. No financial support was obtained for the study. The topic has not previously been published in the WJCC.

Issues raised: (1) The language classification is Grade C. Please visit the following website for the professional English language editing companies we recommend: https://www.wjgnet.com/bpg/gerinfo/240; (2) The authors did not provide original pictures. Please provide the original figure documents. Please prepare and arrange the figures using PowerPoint to ensure that all graphs or arrows or text portions can be reprocessed by the editor; (3) PMID numbers are missing in the reference list. Please

provide the PubMed numbers to the reference list and list all authors of the references. Please revise throughout; (4) The “Case Presentation” section was not written according to the Guidelines for Manuscript Preparation. Please re-write the “Case Presentation” section, and add the “FINAL DIAGNOSIS”, “TREATMENT”, and “OUTCOME AND FOLLOW-UP” sections to the main text, according to the Guidelines and Requirements for Manuscript Revision; and (5) Authors should always cite references that are relevant to their study. Please check and remove any references that not relevant to this study. 6 Recommendation: Conditional acceptance.

Response: We appreciate the editor given these kindly comments. All of changes in the revised manuscript have already performed with tracked.

Company editor-in-chief:

I have reviewed the Peer-Review Report, the full text of the manuscript, and the relevant ethics documents, all of which have met the basic publishing requirements of the World Journal of Clinical Cases, and the manuscript is conditionally accepted. I have sent the manuscript to the author(s) for its revision according to the Peer-Review Report, Editorial Office’s comments and the Criteria for Manuscript Revision by Authors. The title of the manuscript is too long and must be shortened to meet the requirement of the journal (Title: The title should be no more than 18 words). Before final acceptance, uniform presentation should be used for figures showing the same or similar contents; for example, “Figure 1Pathological changes of atrophic gastritis after treatment. A: ...; B: ...; C: ...; D: ...; E: ...; F: ...; G: ...”. The quality of the English language of the manuscript does not meet the requirements of the journal. Before final acceptance, the author(s) must provide the English Language Certificate issued by a professional English language editing company. Please visit the following website for the professional English language editing companies we recommend: https://www.wjgnet.com/bpg/gerinfo/240.

Response: We appreciate the editor given these kindly comments. All of changes in the revised manuscript have already performed with tracked.

Dear editor:

We would like to thank **World Journal of Clinical Cases** for giving us the opportunity to revise manuscript. We have carefully taken the comments into consideration in preparing our revision, which has resulted in a paper that is more clear and compelling. The point-by-point responses are attached after this letter. The revisions were highlighted to the text with tracked, have been prepared.

The manuscript has not been published previously, in any language, in whole or in part, and is not currently under consideration elsewhere. None of the authors have any competing financial interest to report.

Reviewer #1: The authors have addressed my previous comments. I have a few additional clarifications I would like the authors to make, but these are minor and I do not need to review the manuscript again. 1. Core Tip, page 3 last line, and also Conclusion, page 8: Please delete the phrase "failure to demonstrate" (I think this was in my previous review; I apologize this was an error and the sentence reads better without "failure to demonstrate").

Response: Thanks for your suggestion. We have already made these changes in the revised manuscript.

2. Chief Complaint, page 4: delete "has been".

Response: Thanks for your suggestion. We have already removed it.

3. History of Present Illness, page 4, line 3: Please deleted "scanned in" and simply say "scan from" another hospital; line 4: Regarding the outside hospital scans, it would be good to add (images not available) at the end of the sentence.

Response: Thanks for your suggestion. We have already made these changes in the revised manuscript.

4. Discussion, page 8, last paragraph: Please delete "is enough inside" and change to "has a blood supply", since PAS will have a blood supply and PTE will not.

Response: Thanks for your suggestion. We have already corrected it in the revised manuscript.

5. Conclusion, page 8: There appear to be extra computer commands at the beginning and end of the Conclusion paragraph; please delete.

Response: Thanks for your suggestion. These have been removed in the revised manuscript.