

## Response to reviewers

Dear Reviewers and Editors:

Thank you very much for your comments. They are very positive and constructive on our manuscript entitled "Cardiac Amyloidosis Presenting as Pulmonary Arterial Hypertension: A Case Report and Review of Literature" (ID: 79044). According to those comments, we modified our manuscripts.

### Reviewer#1:

1. It is necessary to supplement the pathological report of Congo red staining of myocardial samples in this case report. There are limitations in the data related to the absence of right cardiac catheterization in the early and late hospitalization of this case.

Response: We can't perform an endomyocardial biopsy and the patient underwent endomyocardial biopsy and Congo red staining of myocardial samples at another hospital (Figure 1). We also regretted that the patient did not have a right cardiac catheterization at the same time. We add these in the main text.

Case presentation section, line 6 – line 8, page 8

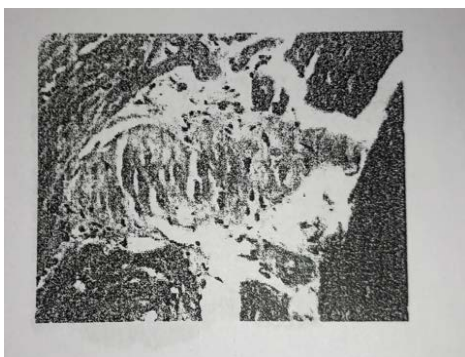


Figure 1. Congo red staining of myocardial samples

2. Only the "PATIENT PHOTOGRAPHIC AUTHORIZATION RELEASE AND DISCHARGE" was seen in the manuscript, but the relevant official ethics documents reviewed and approved by the local ethics committee were not seen in the manuscript.

Response: Thank you for your careful review. We add these to the attachment.

#### **Reviewer#2:**

1. Comment: According to the nomenclature recommendation from the International Society of Amyloidosis (Amyloid 2022; 29: 213-219), the term "primary amyloidosis" is no longer used. I would recommend consistently using "AL amyloidosis" in this manuscript.

Response: We made correction according to the Reviewer's comments.

Abstract section, line 4, line15, line 19, page 2

Introduction section, line 11, page 3

Introduction section, line 19, page 3

2. The authors mentioned that "A myocardial sample stained with Congo red was positive" for the diagnosis of AL amyloidosis. Does this mean amyloid deposits were detected by Congo red staining?

Response: As Reviewer suggested, we have re-written the sentences. The patient underwent endomyocardial biopsy at another hospital. Amyloid deposits were detected by Congo red staining. Myocardial sample was positive, but periumbilical fat aspirates, as well as samples from the tongue, gums and bone marrow were all negative.

Case presentation section, line 6 – line 8, page 8

3. Immuohistochemical examination is important for the confirmation of AL amyloidosis because there are many proteins responsible for amyloidosis. Although serum free light-

chain analysis revealed an increase of lambda light-chain, this issue should be considered.

Response: We made correction according to the Reviewer's comments. Immunohistochemistry of amyloid deposits is used to distinguish TTR from other proteins that may cause amyloidosis. Immunoglobulin light chain associated with light chain (AL) amyloidosis

Case presentation section, line 22, page 7

Case presentation section, line 1 – line 2, page 8

4. How did the authors obtain a myocardial sample?

Response: We can't perform an endomyocardial biopsy and the patient underwent endomyocardial biopsy at another hospital. We also regretted that the patient did not have a right cardiac catheterization at the same time. We add these in the main text.

Case presentation section, line 6 – line 8, page 8

5. In addition to AL amyloidosis, transthyretin (ATTR) amyloidosis is another major cause of cardiac amyloidosis. As this manuscript will attract broad range of readers, the distinction between AL amyloidosis and ATTR amyloidosis should be mentioned in the introduction section, by citing a relevant article describing this issue (Cardiol Ther 2021; 10: 289-311)

Response: We have re-written this part according to the Reviewer's suggestion. In addition to AL amyloidosis, transthyretin-related amyloidosis (ATTR) has been considered a disease in the field of cardiology. Cardiac amyloidosis was confirmed according to results of endomyocardial biopsy, with Congo red staining, nuclear scintigraphy and immunohistochemistry to determine the amyloid type<sup>[5]</sup>.

Introduction section, line 11 – line 15, page 3

References section, line 15 – line 17, page 14



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6. Please reconfirm the use of abbreviations. For example, “MRI” in the main text is not needed because it appears only once.

Response: We are very sorry for our negligence. Thank you for your careful review.

Case presentation section, line 5, page 8

We are truly thankful for your thorough work, and we hope that the corrections will be meted with approval.

Once again, your comments and suggestions were highly appreciated. We look forward to hearing from you.

Best regards.

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