Author's response to reviews

Title: Ruptured Aneurysm at the Fenestration of the Middle Cerebral Artery Detected by Magnetic Resonance Angiography in a Patient with Systemic Lupus Erythematosus and Renal Failure: a case report

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The Biomed Central Editorial Team

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Ruptured Aneurysm at the Fenestration of the Middle Cerebral Artery Detected by Magnetic Resonance angiography in a Patient with Systemic Lupus Erythematosus and Renal Failure. Dr Sadaharu Tabuchi et al.

Thank you for consideration of our manuscript for publication in your journal.
We have reviewed the above manuscript according to your reviewer’s comments.
The manuscript was corrected along with the suggestions and important comments of the reviewers. We would like to express our sincere thanks for these important comments. Points of the revision and responses to the reviewers’ suggestions are summarized as follows;

Reviewer #1 (Dr. Bernhard Schaller)

Comments to authors:
Excellent manuscript

Declaration of competing interests:
I declare that I have no competing interests

Response to reviewer;
I greatly appreciate your comment.

Reviewer #2 (Dr. Yeoungjee Cho)

Comments to authors:
Thank you for an interesting case. Authors describe an uncommon complication of SLE in a 47-year old female who presented with SAH from ruptured aneurym from fenestration of the MCA. The manuscript is clearly written, but I have a few
concerns that require clarification:

1. It is difficult to ascertain the level of SLE activity or any measures were taken to exclude other causes of aneurysm.

Response to reviewer;
An assessment of disease activity is important for the treatment decision as you suggested. Unfortunately I could not mention the precise index or score in this manuscript because there were no descriptions of the activity index such as BILAG, SLAM, ECLAM, SLEDAI in the medical records by physician. Instead of these, I added the information of laboratory data which related to the disease activity. These information was described in the part of case presentation (page 3) as follows;

A 47-year-old female with 23 years history of SLE had sudden onset of severe headache and was referred to our hospital. She had been treated as an outpatient with a daily prednisolone dose of 7.5 mg and cyclophosphamide of 50 mg/day before the onset. GFR was 5.6 ml/min/1.73m² and the stage was classified to G5 (endstage) of chronic kidney disease (CKD) on admission. Laboratory examinations revealed the hypocomplementemia (complement C3: 66 mg/dl, C4:14 mg/dl). Anti-double-stranded DNA antibody (IgG) was 6.5 IU/ml (normal range < 20).

Surprisingly, SLE seems to be involved in the onset of SAH regardless of disease activity. Owada reported , at the time of SAH onset, 26 of 57 patients (45.6%) had high SLE disease activity. However, 13 cases had low or no disease activity [7].
This reference was added in the reference list and manuscript (page 3, discussion).

Although SAH due to ruptured aneurysm has rarely been reported in patients with SLE, SAH in Japanese SLE patients is more frequent than in patients from Western countries and can occur regardless of SLE disease activity [7].

This patient had no family history of subarachnoid hemorrhage. The risk factors for the cerebral aneurysm formation were the renal hypertention and SLE itself. I supposed that local weakness of cerebral vascular walls and cerebral vasculitis were responsible for the aneusymal formation. Other cause of aneurysm could not been obtained in this study.
2. I do not agree with the last sentence, where "as the prognosis of SLE patients is usually poor, both diagnosis and therapy were difficult and important in such conditions" - SLE is a treatable condition. Although prognosis can be poor in some, if compliant and on correct treatment, it is a manageable condition albeit may not curable. If the sentence refers to SAH pts with SLE, then it should be more explicitly clarified.

Response to reviewer;
I think this sentence was inappropriate as you suggested. Although SLE itself is a treatable condition, accompany of SAH to SLE is a difficult condition to treat in neurosurgical practice. According to the several literatures (including case report), the prognosis of such condition was generally poor. The mortality rate was relatively high in the 57 reported Japanese SLE patients with SAH [7]. This information was added in discussion (page 3) as follows;

The mortality rate associated with SAH is relatively high (38.6%) in Japanese SLE patients [7].

I changed the last sentence (page 4) according to your suggestion as follows;

As the prognosis of SLE patients associated with SAH can be poor, both diagnosis and therapy requires special attention in such conditions.

3. It would also be helpful to define what the level of chronic renal failure was - given CT with contrast was contraindicated.

Response to reviewer;
Thank you for your thoughtful comments.
I added the information in the case presentation (page 3) as follows;

Gulomerular filtration rate (GFR) was 5.6 ml/min/1.73m² and the stage was classified to G5 (endostage) of chronic kidney disease (CKD) on admission.
Also in the discussion (page 3) as follows;

CKD (GFR < 60 ml/min/1.73m$^2$) is a risk factor for the contrast induced nephropathy. CKD (GFR < 30 ml/min/1.73m$^2$) without dialysis is generally considered as a contraindication for usage of contrast medium according to the KDIGO 2012 clinical practice guideline for the evaluation and management of chronic kidney disease.

Quality of written English: Acceptable

We hope that the revised manuscript is now suitable for publication of “Journal of Medical Case Reports”.
I do look forward to hearing from you a constructive response at your earliest convenience.

Sincerely yours,

Sadaharu Tabuchi M.D., Ph.D.