Author’s response to reviews

Title: An unusual case of acute lupus haemophagocytic syndrome: a test of diagnostic criteria

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Author’s response to reviews:

Reviewer #1:

Concerns

1. As the patient showed negative ANA and ds-DNA antibody at the first admission, it is possible that infectious disease induced the onset of SLE. There might be several cases or reviews that describe infection-induced autoimmune diseases, and it is recommended that the authors mention the possibility of infection-induced SLE, then discuss this point with referring previous articles.

Response:

It is possible that an infection may induce the onset of SLE. But we have discussed that an infection in this patient was unlikely in the 1st admission with the given clinical presentation and investigation results (5th paragraph of the discussion). But we agree that this differential diagnosis should be discussed. I have included a paragraph in the discussion giving references.

2. Also, there still exists a possibility that the symptoms at the second admission is lupus-like syndrome associated with other diseases.

i. Usually, SLE patient with organ involvement show at least low grade fever. How was the body temperature of this patient?

Response:
The patient had intermittent fever for 3 weeks. I added this to the manuscript as a revision.

ii. What was the dose of prednisolone before the second admission?
Response:
She was on prednisolone 5 mg daily.

iii. Did the authors tested HIV again? HIV antibodies sometimes become positive after several months.
Response:
Screening for HIV infection (antibodies for HIV) was done in the second admission. It was negative.

iv. The authors show ANA was '>1/80', but x80 of ANA is not 'strongly positive'. What exactly was the titre of ANA? Also, it is recommended to show the pattern of ANA.
The lab report has given the result as ‘positive (>1/80)’ which is the norm for this lab. They haven’t given the exact titre. The pattern of ANA was homogenous. Since the exact titre is not available, I replaced the term ‘strongly positive’ with ‘positive’.

v. ds-DNA is usually shown by IU/ml. How did the authors measure the ds-DNA, and what was the normal limit of this measurement?
Response:
The anti Ds-DNA is reported as ‘positive (>1/10)’ which is again the norm for this lab. The exact titre has not been given. This was measured by the indirect immunofluorescence method. Although Ds-DNA can be given as IU/ml, it can be also given as a titre where >1/10 is considered positive in literature including many research studies.

3. In the last paragraph of case presentation, the authors said the patient remained asymptomatic, but did not show how long. Please add the period of her remission.
Response:
I have added this as a revision.

Minor points

1. It seems better to delete '0' in each numbers shown in the article- for example, 03 days → 3 days, 06 months → 6 months- because there is no reason to add 0 in this part.

   Response:

   Revision done.

2. There is also a possibility that she had asymptomatic SLE before her first admission. How was the result of urine test at her first admission?

   Response:

   I have added this as a revision. It was normal without any active sediment.