Reviewer's report

Title: Pancytopenia as an early indicator for Stevens-Johnson syndrome complicated with Hemophagocytic lymphohistiocytosis: a case report

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Reviewer: ALBERT CATALA

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Dear Editors:

Pancytopenia as an early indicator for Stevens-Johnson syndrome complicated with Hemophagocytic lymphohistiocytosis: a case report
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The case report describes the exceptional presentation of HLH in Steven Johnson disease. So far, rarely reported. A clinical observation that can modify the clinical approach in a disease. It has educational value per se.

However, several issues must be addressed before publication:

Minor revisions
1) Spelling:
   - Abstract: hyperferritinaemia: hyperferritinemia, pluse: pulse
   - Case presentation: second paragraph: chlamydia pneumonia: chlamydia pneumoniae
   - Fourth paragraph: leucopenia: leukopenia

2) Case presentation third paragraph
   - They talk firstly of interleukin-2 receptor and after about CD25. If they want to mention both names it must be specified in brackets that both are equivalent terms.
   - CD25 level, since many of the clinicians are not familiar with normal range they should provide the normal value range

Fourth paragraph: they mention that clinical and lab parameters have improved before discharged. If NK cells activity or CD25 levels were assessed then, it could be provided. If they have measured the activation parameters (ferritin, NK activity or CD25 levels) along the outbreak they could include a diagram graph showing biochemical evolution.

3) Discussion: fifth paragraph: mention HLH-2004 protocol. This protocol must be cited.

Compulsory revisions:

Introduction
1) Second paragraph: it mentions that “secondary HLH typically develops when infections activate immune system”. It is widely acknowledged that some viral infections (EBV, CMV..) can trigger HLH in primary settings. Review this sentence.

Case presentation

2) Third paragraph: “the diagnosis of HLH was established since the patient fulfilled 6/8 HLH 2004 diagnostic criteria”. Could be more illustrative if a table is included. They can adapt it from Henter et al.: Diagnostic and Therapeutic Guidelines for Hemophagocytic Lymphohistiocytosis. Ped blood and cancer 2007.

Discussion

3) Third paragraph: “the diagnostic reference standard remains the hemophagocytosis in bone marrow”, hemophagocytosis is neither sensitive nor specific for HLH. Some authors have considered the less important diagnostic criteria (How I treat HLH, Jordan M, et al. Blood 2011)

Conclusion:

4) First sentence: according to the background provided it is hard to say that HLH in SJS is a fatal disease. Only one case published.

5) Same comment for "with excellent prognosis when treated promptly".

In my opinion they must remark that HLH in SJS is very uncommon. However pancytopenia can be a precocious indicator and enables to start a prompt diagnosis and treatment.

Due to the rarity of such presentation in SJS this patient must be closely followed in order to detect an underlying primary genetic defect that predisposes to HLH.

Level of interest: An article of limited interest

Quality of written English: Acceptable

Statistical review: No, the manuscript does not need to be seen by a statistician.

Declaration of competing interests:

I declare that I have no competing interest