ISSN 2307-8960 (online)

# World Journal of *Clinical Cases*

*World J Clin Cases* 2020 June 6; 8(11): 2066-2407

Published by Baishideng Publishing Group Inc

World Journal of Clinical Cases

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Semimonthly Volume 8 Number 11 June 6, 2020

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**NAME OF JOURNAL**
*World Journal of Clinical Cases*

**ISSN**
ISSN 2307-8960 (online)

**LAUNCH DATE**
April 16, 2013

**FREQUENCY**
Semimonthly

**EDITORS-IN-CHIEF**
Dennis A Bloomfield, Bao-Gan Peng, Sandro Vento

**EDITORIAL BOARD MEMBERS**
https://www.wjgnet.com/2307-8960/editorialboard.htm

**PUBLICATION DATE**
June 6, 2020

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World Journal of Clinical Cases

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*World J Clin Cases* 2020 June 6; 8(11): 2406-2407

DOI: 10.12998/wjcc.v8.i11.2406

ISSN 2307-8960 (online)

LETTER TO THE EDITOR

# Macrophage activation syndrome as an initial presentation of systemic lupus erythematosus

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**Author contributions:** Shi LJ and Guo Q contributed equally to this work; Shi LJ and Guo Q performed literature search and wrote the letter; Li SG performed critical comments.

**Conflict-of-interest statement:** The authors declare that they have no conflict of interest.

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**Manuscript source**: Unsolicited manuscript

**Received:** January 15, 2020
**Peer-review started:** January 15, 2020
**First decision:** April 24, 2020
**Revised:** May 5, 2020
**Accepted:** May 13, 2020
**Article in press:** May 13, 2020
**Published online:** June 6, 2020

**P-Reviewer:** Tanaka H

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## Abstract

In a recent article on *World J Clin Cases* 2019; 7: 3859-3865, Sun *et al* reported a case of 36-year-old female with macrophage activity syndrome as an onset of systemic lupus erythematosus. Although this is a very interesting case, some concerns still need to be addressed. First, the patient had an extremely elevated serum ferritin but a normal C-reactive protein level, which was unparallel with the inflammatory condition before she received any treatments. Second, the diagnosis of systemic lupus erythematosus seemed to be insufficient according to the patient's medical information presented, most of which were not specific to lupus but could be explained by macrophage activity syndrome. Hence, more medical information on the patient should be provided, and a profound discussion needs to be addressed.

**Key words:** Systemic lupus erythematosus; Macrophage activity syndrome; Diagnosis; Ferritin; C-reactive protein; Inflammatory

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**Core tip:** The recent report (*World J Clin Cases* 2019; 7: 3859-3865) about a 36-year-old female with macrophage activity syndrome as an onset of systemic lupus erythematosus was rare and interesting. However, the presented patient diagnosed with macrophage activity syndrome should have a high inflammatory status, but reported a normal C-reactive protein level. Furthermore, the medical information on the patient was inadequate for a diagnosis of systemic lupus erythematosus. Therefore, a profound discussion needs to be addressed.

**Citation:** Shi LJ, Guo Q, Li SG. Macrophage activation syndrome as an initial presentation of systemic lupus erythematosus. *World J Clin Cases* 2020; 8(11): 2406-2407
**URL**: https://www.wjgnet.com/2307-8960/full/v8/i11/2406.htm
**DOI**: https://dx.doi.org/10.12998/wjcc.v8.i11.2406

**S-Editor:** Wang YQ
**L-Editor:** MedE-Ma JY
**E-Editor:** Ma YJ

## TO THE EDITOR

In a recent issue of *World Journal of Clinical Cases*, Sun *et al*[1] reported a case of macrophage activity syndrome (MAS) as an onset of systemic lupus erythematosus (SLE). It was a truly interesting case. However, some concerns still need to be addressed by the authors.

First, as we know, MAS is associated with excessive activation and proliferation of T cells as well as macrophages, which leads to a massive release of proinflammatory cytokines[2]. Therefore, in a patient with MAS, it is rational to expect an extremely high inflammatory state that could be accompanied by increased C-reactive protein (CRP). However, the presented patient had a high-grade fever and a very high level of serum ferritin but normal CRP, which was not concomitant with a common condition of MAS. Were the CRP and serum ferritin tests performed at the same time? After the patient's admission to the hospital, did she undergo any CRP monitoring before she received her treatments? Furthermore, did the CRP remain normal during the whole disease duration? Although several retrospective studies have demonstrated that not all MAS patients show increased CRP, we speculate that the results might have been affected by some treatments, especially glucocorticoids[3]. In our centre, a total of 10 MAS patients secondary to different kinds of rheumatic diseases showed increased CRP before treatment was initiated. Once they received treatment, the CRP level could be unparallel with the symptoms and serum ferritin results. Hence, if the CRP level of the presented case was still normal during the whole disease duration, the authors should explain the probable reasons for the unparallel inflammatory index (CRP) and serum ferritin level in the patient with MAS.

Second, could the patient be diagnosed with SLE? We agreed with the diagnosis of MAS in this patient. However, in our opinion, the medical information on the patient was too limited to establish a diagnosis of SLE. For example, whether the patient had any symptoms of mucosal ulcer, hair loss or any abnormality of urine analysis was not mentioned. The clinical manifestations including fever and jaundice were not specific to SLE but could be explained by MAS, which can still present with hematologic involvement and pleural effusion. Given those conditions, we think that it was hard to diagnose SLE in a patient with only a lower level of complement 3, high titre of antinuclear antibody (ANA), positive anti-Ro-52 antibody and anticardiolipin IgM antibody with unknown titre according to the revised American College of Rheumatology classification criteria or Systemic Lupus International Collaborating Clinics classification criteria[4,5]. We still want to know if the authors had repeated immunological tests including C3, ANA, anti-Sm and anti-dsDNA antibodies in this patient in her follow-up visits.

In summary, this interesting case could be more integrated by discussing more profoundly the "normal CRP" and providing more clinical and laboratory data during the periods of the patient's first admission to the hospital and her one-year follow-up.

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