Reviewer's report

Title: Relapsing macrophage activating syndrome in a young patient with still's disease.

Version: 2 Date: 28 November 2008

Reviewer: Karin Beutel

Which of the following following best describes what type of case report this is?: Other

If other, please specify:

The authors claim that they report an unexpected event in the course of treatment of a patient but in my view the disease course is not interpreted correctly.

Has the case been reported coherently?: No

Is the case report authentic?: Yes

Is the case report ethical?: Yes

Is there any missing information that you think must be added before publication?: Yes

Is this case worth reporting?: No

Is the case report persuasive?: No

Does the case report have explanatory value?: No

Does the case report have diagnostic value?: No

Will the case report make a difference to clinical practice?: No

Is the anonymity of the patient protected?: Yes

Comments to authors:

The authors describe a case of apparently new-onset Still’s disease (SD) which is associated with macrophage-activation syndrome (MAS) and argue that discontinuing the treatment with cyclosporine A (CSA) resulted in an early relapse of MAS.

The patient’s history and the given laboratory values are consistent with the
reported diagnosis but it is hard to believe that, despite severe liver disease, the patient never developed hepatosplenomegaly and/or ascitis (typical for either SD or MAS or both). The search for a triggering infectious agent has only been performed by serology at the onset of disease. This obviously is not enough, especially to exclude the well-known triggers of MAS, i.e. pathogens of the herpes virus group. Complete work-up includes PCR analysis and should have been repeated, at least at the time of the “relapse”, because a persistent trigger might have promoted MAS. The authors report that the patient improved after initiation of the recommended treatment for this condition (pulses of high dose steroids and CSA). However, they do not provide enough data to assess disease activity. Biomarkers, i.e. ferritin and sCD25, are lacking completely. Furthermore, the drop of leukocytes at that point might also have indicated ongoing MAS rather than improvement. Thus, it remains unclear if the patient had responded sufficiently at the time of therapy reduction or if there was still ongoing MAS activity possibly caused by insufficient treatment of the underlying condition (infection or rheumatologic disorder). In addition, a relapse after two weeks is very unlikely.

In their introduction and the discussion the authors repeatedly mention the diagnostic and therapeutic guidelines issued by the Histiocyte Society. However, it must be stressed that these guidelines primarily refer to genetic forms of hemophagocytic lymphohistiocytosis (HLH). MAS is grouped into the acquired (secondary) forms of HLH but is considered to be a special entity regarding diagnosis and treatment. Suggestions for specific definitions have been published. Intensity and duration of therapy in any case have to be adapted to the clinical course. In non-responding cases biologicals have already been used with success.

In summary, there is not enough evidence for relapsing MAS and prolonged treatment is not a new recommendation.

Minor comments: CrP values are given without units. Generic names of drugs should be used, not “solumedrol” and “celcept”.

**Quality of written English:** Acceptable

**Declaration of competing interests:**

I declare that I have no competing interests.