Pediatric Rheumatology

Poster presentation

Macrophage activation syndrome: a potentially fatal complication of rheumatic disorders

V Ziaee* and MH Moradinejad

Address: Children's Medical Center, Tehran University of Medical sciences, Tehran, Iran
* Corresponding author

from 15th Paediatric Rheumatology European Society (PreS) Congress
London, UK. 14–17 September 2008

Published: 15 September 2008

Pediatric Rheumatology 2008, **6**(Suppl 1):P62 doi:10.1186/1546-0096-6-S1-P62

This abstract is available from: http://www.ped-rheum.com/content/6/S1/P62

Background

Macrophage activation syndrome (MAS) is a rare complication of childhood with rheumatic disease. This syndrome has been reported in association with many rheumatic diseases, especially systemic juvenile rheumatoid arthritis (SoJIA). The aim of this study was evaluation the rate, symptoms and outcome of MAS during 10 years.

Methods

Retrospective review of cases of MAS from the charts of 120 patients with juvenile rheumatoid arthritis and systemic lupus erythematosus (SLE), were reviewed collected data base of 5 children with MAS from 1998 to 2007, in Children's Medical Center, In Tehran University.

Results

Totally 120 patients evaluated in this study including 108 JIA and 12 SLE. Five patients (4 girls and 1 boy) were considered to have evidence of MAS (incidence rate 4.2%). This rate for all JIA patients was 3.7% and for SoJIA, SLE and juvenile idiopaticarthritis (JIA) and polyarticular RF negative JIA was 8.2%, 16.7% and 2.8%, respectively. Mean age of MAS onset was 4.9 years, and duration of rheumatologic disease prior to MAS, 22 months. Four cases (80%) had abnormal liver function during the disease course, and coagulopathy. Bone marrow examination supported the diagnosis with definite haemophagocytosis in four cases (80%). The mortality rate was 40%.

Conclusion

Incidence of MAS in our JIA patients was about other studies, but the mortality rate was higher than other reports. Although MAS is a rare complication, because it is potentially fatal it must be considered in each childhood rheumatic disorders with suddenly changes in general condition and decrease peripheral cells.

References

1. Sawhney S, Woo P, Murray KJ: **Macrophage activation syndrome: a potentially fatal complication of rheumatic disorders.** *Arch Dis Child* 2001, **85:**421-6.
2. Behrens EM, Beukelman T, Paessler M, Cron RQ: **Occult macrophage activation syndrome in patients with systemic juvenile idiopathic arthritis.** *J Rheumatol* 2007, **34(5):**1133-8.