Pediatric Rheumatology

Poster presentation

Macrophage activation sydrome with systemic onset juvenile idiopathic arthritis (SOJIA) in Chinese childern

C Li*, X He, W Kuang, T Han, Y Zhou, J Zhang and X Tan

Address: Beijing Children's Hospital, Beijing, PR China
* 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):P33 doi:10.1186/1546-0096-6-S1-P33

This abstract is available from: http://www.ped-rheum.com/content/6/S1/P33

Background

Macrophage activation syndrome (MAS) is a complication of systemic onset JIA (SOJIA), which is characterized by fever, hepatosplenomegaly, lymphoadenopathy, pancytopenia, liver dysfunction, pulmonary damage and CNS dysfunction. We did the study to analyze the clinical features, treatment, and outcome of MAS with SOJIA.

Materials and methods

Review of cases of MAS from a prospectively collected database of children with SOJIA in Beijing Children's Hospital from the year of 2003 to 2007.

Results

42 patients were diagnosed MAS with SOJIA from 159 SOJIA cases. The duration prior to MAS is 11 months. High fever, hepatosplenomegaly, pancytopenia, liver dysfunction were in all cases. Bleeding was in 12. 45% has had CNS dysfunction. 24% were with ARDS. 2 suffered from renal damage. The lab.tests revealed elevated live enzymes and ferritin, decreased value of ESR, album, CBC and fibrinogen in all. Bone marrow examination supported the diagnosis with definite haemophagocytosis in 42 cases. Lymph node biopsy was done for one case and found out it was filled of activated macrophage. In the treatment, thirteen only received high dose steroids (four of thirteen died), twenty-one got high dose steroids plus cyclosporine (four died), five were steroids plus cyclosporine and etoposide (none died). The causes of death were ARDS and CNS involvement.

Conclusion

MAS is a rare and potentially fatal complication of SOJIA. Most of our patients were male. Bone marrow studies support the diagnosis. CNS involvement and ARDS are poor prognostic signs. Early diagnosis and aggressive therapy is essential.