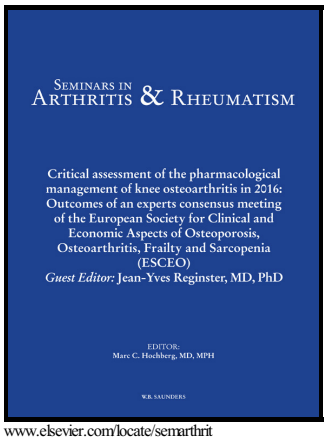
Should refractory Kawasaki disease be considered occult macrophage activation syndrome?

Ui-Yoon Choi, Seung-Beom Han, Soo-Young Lee, Dae-Chul Jeong



www.elsevier.com/locate/semarthrit

PII: S0049-0172(16)30198-6
DOI: http://dx.doi.org/10.1016/j.semarthrit.2016.08.007
Reference: YSARH51092

To appear in: *Seminars in Arthritis and Rheumatism*
Accepted date: 15

Cite this article as: Ui-Yoon Choi, Seung-Beom Han, Soo-Young Lee and Dae-Chul Jeong, Should refractory Kawasaki disease be considered occult macrophage activation syndrome?, *Seminars in Arthritis and Rheumatism,* http://dx.doi.org/10.1016/j.semarthrit.2016.08.007

Should refractory Kawasaki disease be considered occult macrophage activation syndrome?

Ui-Yoon Choi, MD, Seung-Beom Han, MD, Soo-Young Lee, MD, Dae-Chul Jeong, MD

Department of Pediatrics, St. Paul's Hospital, The Catholic University of Korea, 180, Wangsan-ro, Dongdaemun-gu, Seoul, 02559, Korea

Keywords: Refractory, Kawasaki disease, macrophage activation syndrome

Corresponding author: Soo-Young Lee, MD

Department of Pediatrics, St. Paul's Hospital, The Catholic University of Korea, 180, Wangsan-ro, Dongdaemun-gu, Seoul, 02559, Korea

E-mail address: sylee@catholic.ac.kr

We read with great interest the paper entitled "Macrophage activation syndrome in Kawasaki disease: more common than we thought?" by Wang W et al. [1], which brought attention to the underestimated incidence of macrophage activation syndrome (MAS) complicating Kawasaki disease (KD). The incidence of MAS in children with KD was 1.1% (8/719). The authors described the clinical and laboratory findings of KD patients with MAS and compared two diagnostic standards: the hemophagocytic lymphohistiocytosis (HLH) 2009 criteria, and Ravelli's diagnostic criteria for MAS complicating systemic juvenile idiopathic arthritis (sJIA). As shown in their results, the HLH 2009 criteria had low sensitivity and specificity for the diagnosis of MAS complicating KD. We entirely agree with their opinion that Ravelli's criteria have better accuracy for MAS diagnosis in children with KD and those with sJIA, and should be utilized for the diagnosis of MAS complicating KD.

All patients received the standard treatment for KD, but only one patient responded to the first infusion of intravenous immunoglobulin (IVIG). We remain curious, however, about how many of the 719 KD patients remained refractory to the second IVIG infusion. In another study, approximately 4% of children with KD remained refractory to this second infusion [2], and the incidence of MAS complicating KD was 1.1% in their study. Thus, it can be assumed