Abstract

Macrophage activation syndrome (MAS) is an increasingly recognized life-threatening complication of autoimmune diseases, with a mortality rate as high as 50%, occurring mainly in systemic-onset juvenile idiopathic arthritis and adult-onset Still's disease (AOSD)(1). MAS is a secondary form of hemophagocytic lymphohistiocytosis (HLH), characterized by excessive inflammatory cytokine production and, consequently, leading to multiple organ failure(2). Sulfasalazine, which is used to treat many rheumatological diseases, is one of the triggers of MAS (27).

We report a case of a 47year old female, who was recently diagnosed to have AOSD and treated with sulfasalazine for two weeks, presented with acutely worsen shortness of breath, diagnosed to have MAS. Justification of this case report is that the occurrence of MAS triggered by sulfasalazine is very rare and only a very few cases were reported.

Even though MAS is a rare complication of AOSD, it is potentially fatal. Its presentation is mimic with many other common diseases, a high degree of suspicion is needed to diagnose MAS. Early detection with proper treatment reduces the mortality significantly. As most of the drugs which are used to treat AOSD have the potential to trigger MAS, close monitoring is important.