Abstract

Macrophage activation syndrome (MAS) is a hyper inflammatory, life-threatening complication without a clear pathogenesis. It is considered as a secondary form of hemophagocytic lymphohistiocytosis. MAS is a rare complication of autoimmune inflammatory disorders, such as systemic juvenile idiopathic arthritis and adult-onset Still's disease. MAS is very rarely seen in adults and is a less common complication of systemic lupus erythematosus (SLE).

We report a case of a 23-year-old woman, who had been recently diagnosed with possible sero negative rheumatoid arthritis, presented with multiple joint pain and vomiting, found to have alopecia, generalized lymphadenopathy, photosensitive rash and pallor. On the second day of admission, she developed high grade fever with shortness of breath and her level of consciousness reduced over minutes to hours. She was intubated and transferred to intensive care. Her investigations revealed subarachnoid hemorrhages, ARDS, cardiomyopathy with reduced ejection fraction and other investigations with clinical findings was compatible with SLE complicated with MAS. She was treated with IV methyl prednisolone, cyclosporine, IV Immunoglobulin and plasmaparesis. Despite early treatment she did not improve and succumbed to the illness.