Macrophage activation syndrome and acquired hemophagocytic lymphohistiocytosis in adults: A Philadelphia cohort

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Macrophage activation syndrome (MAS) is a form of acquired hemophagocytic lymphohistiocytosis (aHLH) associated with rheumatologic disorders. This cytokine storm syndrome is associated with multiorgan failure and high mortality rate. There is a paucity of literature in the adult form of MAS and aHLH, especially in the U.S. population. We investigated the disease characteristics of adult MAS and aHLH at a single university hospital in Philadelphia. This retrospective cohort study was performed on patients 18 years and older admitted to Temple University Hospital from January 2010 to August 2015 with the diagnosis of MAS or aHLH. Eleven patients were included in our cohort. Our cohort differed from the global literature in that the majority of our patients were male; more commonly associated with autoimmune disorders; and a higher survival rate of 91% to discharge.

Conclusion and significance: The demographic and disease characteristics along with survival rate of MAS and aHLH in our patients differed somewhat from global literature though our conclusions are limited by small sample size.

Biography
Irene J. Tan, MD, FACR, is Professor of Clinical Medicine and the Rheumatology Fellowship Program Director at Lewis Katz Temple University School of Medicine in Philadelphia, USA. She garnered the “Top Doctor” recognition featured in Connecticut Magazine from 2010-2012, “Top Doctor” in Philadelphia Magazine in 2017, 2018, and “Exceptional Woman in Medicine” by Castle Connolly in 2017. She received a number of teaching awards, the most recent one is the Golden Apple Award by American Medical Student Association. Her research interests are rheumatologic emergencies, CTD-ILD, impact of gender and hormones on autoimmunity, sympathetic joint effusion and macrophage activation syndrome.

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