Clinical Image

An Interesting Case of Macrophage Activation Syndrome

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Macrophage Activation Syndrome (MAS) is a form of Hemophagocytic Lymphohistiocytosis (HLH), a condition defined by overactive immune response causing tissue destruction and excessive inflammation. It has been seen in rheumatologic conditions, mainly juvenile-idiopathic-arthritis and systemic lupus erythematosus.

We present a 40 year old obese female with lupus, gastric bypass, and NAFLD initially admitted for alcoholic hepatitis complicated by hepatorenal syndrome due to abdominal pain, nausea, vomiting, and confusion given recent history of alcoholic use. Lab work significant for leukocytosis, anemia, thrombocytopenia, elevated Cr, with mildly elevated AST and liver steatosis on abdominal CT. Differential diagnosis was SLE vs. HRS as IR did not feel a renal or liver biopsy was safe due to body habitus. Subsequently, patient transferred to the medical-ICU for shock requiring intubation. Infectious workup unremarkable, hence cause of decompensation thought to be lupus causing macrophage activation syndrome with anemia, thrombocytopenia, liver function abnormalities, and worsening multi-organ failure. She completed pulse-dose IV Solu-Medrol, after which her clinical status improved, with eventual transition to oral prednisone. She was extubated and returned to floor level of care. Unfortunately, she decompensated as a result of hemorrhage due to peripancreatic drain placement for acute necrotizing pancreatitis, with multiple organ failure that continued to worsen. Patient made comfort care and terminally extubated [1,2].

MAS can present quickly with multiple organ failure. In patients with rheumatological conditions, rapid identification is vital for appropriate treatment. In severe cases, patients may require cyclophosphamide and/or rituximab if there is lack of initial response to treatment or worsening disease. In 2016, the 2016 EULAR/ACR/PRINTO came up with new classification criteria for MAS. The 2016 classification criteria for MAS are useful to identify febrile SLE patients at high risk for in-hospital mortality and have been shown to aid in the early detection of MAS (Figure 1 and 2) [3,4].

References

Figure 1: Genetic factors and the inflammatory milieu created by the underlying rheumatic disease act synergistically to reach the threshold for Macrophage Activation Syndrome (MAS) in the presence of an infectious trigger.

Figure 2: A) Myelocyte within an activated macrophage. In addition, there are multiple adherent red blood cell and myeloid precursors. B) Activated macrophage engulfing a band neutrophil. C) Band neutrophil and metamyelocyte within an activated macrophage. Nuclei of neutrophil band appear condensed. D) Activated macrophage with hemosiderin deposits and a degenerating phagocytosed nucleated cell. H&E stain original magnification × 1000 Macrophage Activation Syndrome (MAS).