FEVERS AND FATIGUE: A Case of Paroxysmal Nocturnal Hemoglobinuria in the Setting of Aplastic Anemia

Julia Kostka, MD, Sylvester Dorobisz, MD, Faisal Al Bahrani MD, David J Regelmann, MD

Department of Medicine, St Vincent’s Medical Center Bridgeport, CT

Background

- Fever in the setting of pancytopenia has a wide range of etiologies, including PNH, HIV, CMV, malignancy, autoimmunity and HLH.
- PNH occurs with aplastic anemia in up to 25% of patients.

Case

- A 21-year-old man presented with a month’s history of fatigue and a week of fevers, chills, and sore throat.
- He had no known past medical history.
- He moved to the US from Haiti and was sexually active.
- He was found to be febrile with labs significant for leukocytes 1600/mcl, hemoglobin 3.5 gm/dL and platelets 4000/mcl. Neutrophil count was 400/mcl.
- He was started on cefepime, vancomycin, acyclovir and fluconazole and blood was transfused.
- His fevers improved with antibiotics, but no clear source of infection was identified. Multiple studies were sent, including COVID-19 testing, ANA, SPEP/UPEP, HIV, hepatitis serologies, EBV IgG, and parvovirus IgM/IgG.
- Bone marrow biopsy was performed and showed hypocellular marrow, consistent with aplastic anemia (AA). Flow cytometry showed evidence of paroxysmal nocturnal hemoglobinuria (PNH) clone.

Discussion

- AA is closely related to PNH, and approximately 20% of patients with AA also have PNH at diagnosis.
- While PNH can occur in patients with acquired AA, it rarely occurs in patients with inherited AA.
- The mechanism of PNH in patients with AA is thought to be due to autoimmune effects that can target hematopoietic stem cells and possibly GPI anchors.
- The abnormal blood cells are thought to initiate an immune response that damages hematopoietic stem cells and other hematopoietic precursors.
- Patients benefit from PNH testing when AA suspected because the presence of PNH cells is associated with a superior response to immunotherapy.

References