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ABSTRACT N. PO60

ASYMPTOMATIC PNH CLONE IN ACQUIRED APLASIA (AA).

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Background: Paroxysmal nocturnal hemoglobinuria (PNH) is a rare, acquired clonal hematopoietic cell disease characterized by destruction of hematopoietic cells through the activation of the complement system with manifestations that can be life threatening including hemolysis, thrombosis, and marrow failure. Small clinically silent PHN clone are found in patients with bone marrow disordes. We describe a case of acquired aplasia (AA) with silent PHN clone.
Case report: A 62-year-old male was admitted to the hospital for anemia and dyspnea. His laboratory tests showed leukopenia (leukocytes 0.84 x10^3/uL, with 45% neutrophilis and 49% lymphocytes), anemia (hemoglobin 8.1 g/dL) and thrombocytopenia (Platelets count was 9.0 x10^3/uL). Intravascular hemolysis was not proven and a direct Coombs test and ADAMTS-13 were negative. An ultrasound of the abdomen showed normal spleen size and echotexture. A bone marrow aspirate revealed hypocellularity of the erythroid and myeloid lineage with absent megakaryocytes and no increase in blasts. PNH flow cytometry reveals 3% PNH granulocytes, 8% PNH monocytes. He was then diagnosed as having AA with coexisting silent PNH. Prednisolone and granulokines treatment was initiated and he was candidate for allogeneic bone marrow transplantation.
Conclusion: It is important to note that the presence of PNH clone does not necessarily equate to a PNH diagnosis. Patients with AA with subclinical PNH clones should be monitored prospectively as most classical PNH arises out of AA.

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