PAROXISTIC NOCTURNAL HEMOGLOBINURIA MANAGEMENT AND EVOLUTION IN AN ELDERLY PATIENT.

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Background: Paroxistic nocturnal hemoglobinuria (PNH) is a clonal acquired stem cell disease, characterized by intravascular complement-mediated lysis of red blood cell, white blood cell and platelet. PNH is a rare disorder that can affect any age, but most frequently occurring in early adulthood, with a median age at the time of diagnosis of 32 years.
Materials and method: We present the case of an 81 years old patient admitted in the Hematology Clinic of Craiova in October 2014 accusing fatigue, dizziness and moderate dyspnea. The medical history, physical examination and laboratory investigations revealed an underweight patient, with dry and pale skin and mucous, dark colored urine (hemoglobinuria), hepatomegaly with the lower edge of the liver at 3 centimeters below the lower edge of the ribs, grade I splenomegaly, anemia (Hb- 9,6 g/dl), thrombocytopenia (42000/mmc), TBIL- 3.96 mg/dl, DBIL- 0.41 mg/dl, LDH- 586 mg/dl, negative indirect Coombs test and positive HAM test. Peripheral blood smear detected poikilocytosis (dacryocyte, codocyte, schistocyte), platelet- 50.000/mmc. Bone marrow examination shows: erythrocytic series 75% with binucleated macroeritroblasts, erythroblasts with intracytoplasmic bridges. Bone marrow biopsy revealed hypercellular bone with erythroblastic hyperplasia, many erythroblastic groups and dyserythropoiesis. Thorax and abdomen computed tomography showed hepatomegaly (left liver lobe= 7.3 cm, right liver lobe= 15 cm) and splenomegaly (12.78 cm).
Results: Based on clinical and laboratory investigations we have established the diagnosis of paroxistic nocturnal hemoglobinuria. Consecutively the treatment consisted of supportive measures and corticotherapy (Dexamethasone 16mg/ day iv divided q 12hr for 4 days) every 21 days. During follow-up the patient showed multiple remissions and acute episodes, and it developed the following complications: candida esophagitis, insulin-dependent type II diabetes mellitus, left basal pleurisy and acute bacterial pneumonia.
Conclusions: The particularity of our case resides in its advanced age and co-morbidities, thus the treatment posing numerous problems. Another particularity lies in the aggressive course of this disease, with the patient presenting multiple relapses in a limited timeframe.