POSTSPLENECTOMY EVOLUTION IN ONE CASE OF HYPOPLASTIC MYELODYSPLASTIC SYNDROME

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We are showing the clinical observation of the patient having a severe aplastic anemia wich evolution after immunosuppresion went towards PNH.The patient G.O.37 years old had diagnosis in 2006 with Aplastic anemia based on pancytopenia and bone marrow trephine biopsy showing reduction in all haemopoietic tissue replaced by fat spaces. After immunosuppression with Cyclosporin and prednisone the patient had obtained complete remission sustained, until April 2009, when became pancytopenic with bone marrow aspirate showing hipercellularity. The further treatment is based on transfusions of erithocytary mass, cyclosporine and prednisone with increased dose. In October 2009 the patient develops a superficial femoral thrombosis with urinary haemosiderin positive. Examinations done at Hematology Department of the Emergency Universitary Hospital- Dr. Horia Bumbea-, Bucharest: Cellular immunophenotype - have confirmed the PNH diagnostic. The further treatment is based on enoxaparinum s.c., followed by oral Acenocumarolum, transfusions of erithocytary mass, danazol and prednisone. It is interesting to note that 2 of the late clonal complications of IS treated AA may actually coexist with AA (the AA/PNH syndrome and hypoplastic MDS). The treatment of late clonal complications of AA is often difficult. Allo - SCT and the recent introduction of anticomplement monoclonal antibodies are the only effective current treatments. The bone marrow aspiration showed persistence hypoplastic myelodysplastic syndrome (hMDS) - refractory cytopenia with multilineage dysplasia. The patient received transfusion of eritrocites and platelets, methylprednisolon and dexamethason but no persistent improvement and even worsening of the thrombocytopenia were registered. Splenectomy has been performed when the platelets were approximately 5000mm3, without intraoperative incidents. After splenectomy, no other therapy or transfusions have been applied and a slow but continuous improvement of the peripheral blood counts up to normal values has been noted. In the bone marrow, a notable increase of cellular density was registered almost one years from splenectomy, with the persistence of the other morphological dysplastic features.