CLINICAL CASE STUDY IN PAROXYSMAL NOCTURNAL HEMOGLOBINURIA

A 37-year-old woman with anemia and undiagnosed PNH who developed hepatic encephalopathy due to hepatic venous thrombosis

PATIENT HISTORY*

Patient is a 37-year-old woman who was noted to have hemoglobinuria on a regular check-up in 2006. Her lab values at that time were significant for a hemoglobin of 9.4 g/dL, and a normal platelet count. Her work-up revealed a mild hemolytic anemia, for which she received a red blood cell transfusion and immunosuppression with an improvement in her hemoglobin to 12.4 g/dL. Over the next few years, she continued to have intermittent episodes of hemoglobinuria, for which she again received RBC transfusions and immunosuppression. No further work-up was performed.

PATIENT ASSESSMENT/DIAGNOSIS

In 2009, she presented with abdominal pain and jaundice. Her hepatitis work-up was negative but an MRI of the abdomen revealed hepatic venous thrombosis. She was referred to a hematologist and high flow cytometry confirmed the suspected PNH diagnosis.

Laboratory Results at Referral

Parameter	Result	Reference Range
High-Sensitivity Flow Cytometry		
RBC (%)	29	>1% PNH clone size for clinical relevance
Granulocytes (%)	88	>1% PNH clone size for clinical relevance
Monocytes (%)	72	0-9
Additional Assessments		
Platelet Count (x 10^9/L)	130	150-400
Hemoglobin (g/dL)	9.8	12.1-15.1
LDH (IU/L)	475	105-333

TREATMENT

The patient received anticoagulation therapy but subsequently developed refractory ascites in spite of placement of a transjugular intrahepatic portosystemic shunt. She developed recurrent hepatic encephalopathy and died while awaiting liver transplant.

*Hypothetical case adapted from actual patient data