Paroxysmal nocturnal hemoglobinuria

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Serial urine samples were collected at variable intervals during the day-time from a 66-year-old patient who presented with anemia (Hb 6.6 g/dL), leukopenia (WBC 2.5×10^9/L) and hyperbilirubinemia (total 3.9 mg/dL, indirect 3.3 mg/dL). The color changes seen in these images represent the resolving hemoglobinuria after an overnight episode of intravascular hemolysis. A diagnosis of paroxysmal nocturnal hemoglobinuria (PNH) was made upon demonstration of red cell fragility (positive Ham test) and absence of decay accelerating factor (CD55) and membrane inhibitor of reactive lysis (CD59) surface membrane proteins (by flow cytometry). These proteins deactivate complement in normal subjects by inhibiting C3b and C5b-9, respectively. Therefore, in their absence, hematopoietic cells become more susceptible to complement-mediated lysis secondary to changes in pH, infections etc. Although all the three cell lines are affected (thus the pancytopenia); the overnight intravascular hemolysis (due to lowering of body pH during sleep) results in early morning dark urine, constituting the most striking feature of this disorder and hence its name.

Figure 1. Serial urine samples collected during the day-time.

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