Dear Sir,

The role of hemoglobin in the pathogenesis of acute renal failure (ARF) remains obscure. ARF has been described in paroxysmal cold hemoglobinuria [1] and in other intravascular hemolytic anemias following bouts of severe hemoglobinuria. In paroxysmal nocturnal hemoglobinuria (PNH), progressive declining of renal function has been observed in some patients [2], but very few patients have been recorded as ARF [3, 4]. Even in these we feel there is no clear-cut evidence to show that the spontaneous hemolytic crises were the cause of ARF. We report a case of severe ARF in a patient with previously unknown PNH in which this relationship is clearly defined.

Case Report

On April 26, 1982, a 30-year-old woman was hospitalized because of abdominal pain, dark urine, and oliguria; these symptoms have begun 36 h before. 1 month earlier she had dark urine for 48 h, without other complaints. She denied fever, rash, arthritis, drug intake, or physical exercise. The patient was alert and well hydrated; the temperature was 36.7 °C, the pulse 84/min, and the blood pressure 130/80 mm Hg. Physical examination was normal. Laboratory data on admission showed: blood urea nitrogen (BUN) 20 mg/l00 ml, serum sodium 139 mEq/l, potassium 3.9, chloride 108 mEq/l hematocrit 30%, hemoglobin 10.7 g/100 ml, leukocyte count 3,600/mm3 with normal differential count, no eosinophils were seen, platelet count 75,000/mm3, total bilirubin 3.6 mg/100 ml, reticulocytes 2%, the haptoglobin was undetectable, the urine gave a + + + test for hemoglobin and hemosiderin and + for protein, the sediment contained granular casts, and an urine culture was negative.

She was treated with intravenous fluids and furosemide, but the daily urine output remained below 100 ml, and BUN and serum creatinine (Cr) increased progressively. On April 30 the hematocrit was 21%, BUN 102 mg/100 ml, and Cr 8.5 mg/100 ml. The proteinuria averaged 300–500 mg/24 h. Echography ruled out renal obstruction. Acidified serum test (Ham’s test) and sucrose hemolysis test were repeatedly positive. She received prednisone, 0.5 mg/kg, in tapering doses. Cryoglobulins and antinuclear antibodies
were negative, and the C3 and C4 fractions of complement were normal. Direct antiglobulin tests with anti-IgG and anti complement serum and the Donath-Landsteiner test were negative. She was hemodialyzed six times between April 30 and May 8. One washed red cell transfusion, which was well tolerated, was administered on May 3 because of hematocrit value of 18%. Since May 4 the urine volume increased progressively, its dark color disappearing; BUN and Cr values decreased gradually, and the patient made an uneventful recovery. A urogram done on May 19 was normal. She was discharged on May 21 with hematocrit 30%, hemoglobin 9.2 g/100 ml, BUN 25 mg/100 ml, Cr 1.3, haptoglobin 158, total bilirubin 1.3 mg/100 ml, platelet count 222,000/mm3. On June 15 the patient was normal with hematocrit 36%, BUN 15 mg/100 ml, Cr 1.1 mg/100 ml, and Cr clearance of 103 ml/min/1.73 m2; the urine was normal, without proteinuria. In the previously reported cases of ARF in PNH [3, 4] the patients had a long history of anemia and hemoglobinuria, whole-blood transfusions could be implicated in the ARF, and after recovery of diuresis, renal function did not reach normal values; these patients died some time after ARF. At autopsy, the kidneys showed deposits of hemosiderin in tubules and signs of chronic tubulointerstitial pathology. We think that our case is unique in several aspects: (1) the patient had a recent onset of PNH, with only initially slight crisis of hemoglobinuria 1 month earlier; (2) there were no other clinical factors that could be implicated in ARF, such as hypotension or volume depletion, and (3) the ARF was clear-cut defined, with total recovery of renal function. The clinical picture is very suggestive of acute tubular necrosis, although renal biopsy was not performed because the patient rejected it. The pathogenesis of ARF in PNH, as in other situations of intravascular hemolysis, is not well understood. Deleterious effects of whole-blood transfusions, massive

References