

Hemolytic Uremic Syndrome

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A.P., a 42-year-old woman, presented with rectal bleeding to her primary care physician. Endoscopic examination revealed a mass in the anal canal. Biopsy was positive for squamous cell carcinoma. A.P. underwent local excision and was established as stage II (T2N0M0). After surgery, she received 5-fluorouracil (FU) 1,000 mg/m² by continuous infusion on days 1–4 and on days 29–32, mitomycin-C 10 mg/m² days 1 and 29, and radiation therapy (4c500 cGY). Minimal nausea was the only side effect. A.P. achieved complete remission.

Approximately eight weeks after completing therapy, A.P. presented for an implantable port flush. The nurse noted that A.P. was pale, weak, and short of breath. A complete blood count (CBC) was obtained and revealed a hematocrit of 17.1% (normal range 37%–48%), hemoglobin of 4.9 (normal range 12.3–15.3 g/dl), and platelet count of 50,000 (normal range 150,000–450,000 cells/ml). A physical examination revealed paleness with generalized anasarca. A.P. was afebrile; her blood pressure was 180/100 mm Hg, pulse rate was 72, and respiration was 24. A.P. was admitted to the hospital for further evaluation and possible blood transfusion.

Other laboratory results included a serum creatinine 3.5 mg/dl (normal range 0.7–1.4 mg/dl), lactic dehydrogenase (LDH) 342 units/L (normal range 140–280 units/L), and serum haptoglobin < 38 mg/dl (normal range

60–270 mg/dl). Peripheral blood smear demonstrated prominent schistocytes with overall low blood cells. The coagulation profile was within normal limits. Mitomycin-C–induced hemolytic uremic syndrome (HUS) was diagnosed.

A.P. initially underwent plasmapheresis every other day with daily plasma infusions and IV methylprednisolone. Her blood pressure continued to increase and was treated successfully with a combination of antihypertensive agents, including a calcium channel blocker, an angiotensin-converting enzyme, and a beta blocker. Weekly erythropoietin-stimulating agent (ESA) injections were initiated. Within three weeks, A.P. had significant improvement of hematologic parameters and daily plasma infusion was stopped.

Plasmapheresis continued twice a week. A.P.'s blood pressure had stabilized. Unfortunately, her renal function continued to worsen and she was placed on renal dialysis three times per week with an initial creatinine of 5.6 mg/dl, potassium of 5.6 mEq/l, and LDH of 580 units/L. Symptoms improved with diuretic administration, and A.P.'s creatinine was maintained at 3.2 after one week on dialysis. After eight weeks in the hospital, A.P.'s hematologic parameters stabilized as well as her renal function on dialysis. She was discharged with close monitoring of her counts and continued dialysis three times per week.

What is hemolytic uremic syndrome?

HUS is a rare condition with a clinical triad of acute renal failure, microangiopathic hemolytic anemia, and thrombocytopenia. Thrombotic microangiopathy dominated by renal impairment usually is referred to as HUS. Systemic hypertension and noncardiogenic pulmonary edema commonly occur during the course of the syndrome. Variable signs of organ failure occur because of platelet thrombi in the microcirculation (Pisoni, Ruggenti, & Remuzzi, 2001).

Little is known about the disease. HUS was first described in 1924 (Wu et al., 1997; Zakarija & Bennett, 2005), but little nursing literature exists concerning risks and management. Because of the complexities identified with this syndrome, a cancer-associated HUS national registry was established in 1984 and ran through 1986 (Lesesne et al., 1989). The registry defined HUS as patients with hematocrit less than 25%, a platelet count less than 100,000, and a serum creatinine greater than 1.6 mg/dl. Clinical characteristics were common among 85 patients identified through the registry, including diagnosis of adenocarcinoma (particularly of the gastrointestinal tract), partial to complete tumor response from treatment, use of mitomycin-C, noncardiogenic pulmonary edema associated with blood product transfusions, and poor response to treatment of HUS with significant mortality.

Do You Have an Interesting Clinical Experience to Share?

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