A 32-Year-Old Pregnant Woman With Malaise, Headache, Nausea, Right Upper Quadrant Abdominal Pain, and Facial Swelling

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A 32-year-old woman in her 36th week of pregnancy is admitted to your ICU after presenting with complaints of malaise, headache, nausea, right upper quadrant abdominal pain, and facial swelling. She denies alcohol abuse, fevers, chills, and hepatitis exposure. She is not on any medication at home. She is alert and oriented. Blood pressure is 160/98 mm Hg, pulse is 100 beats/min, respirations are 20 breaths/min and unlabored, and temperature is 37.2°C. Sclerae reveal mild icterus, and there is mild facial edema. Lung examination is normal. The abdomen is consistent with a gravid uterus; it is soft and nontender and bowel sounds are decreased. Fetal heart tones are normal and regular. There is diffuse, 2+ pitting edema. Neurologic examination is nonfocal. Laboratory tests are notable for a hematocrit of 32% and WBC count of 8,400/mm³ (8.4 × 10⁹/L) with a normal differential. Platelet count is 85,000/mm³ (85 × 10⁹/L). Prothrombin time is 13 s, and partial thromboplastin time is 41 s. Glucose and electrolytes are within normal limits. Creatinine is 1.4 mg/dL (124 μmol/L), and BUN is 25 mg/dL (8.9 mmol/L). Liver function tests reveal an alanine aminotransferase of 100 IU/L, an aspartate aminotransferase of 95 IU/L, a bilirubin of 3.2 mg/dL (55 μmol/L), and lactate dehydrogenase of 820 IU/L. Urinalysis shows 2+ protein. The peripheral blood smear is shown (Fig 1). Which of the following diagnoses most likely accounts for the patient’s findings?

A. Thrombotic thrombocytopenic purpura
B. Disseminated intravascular coagulation
C. Hemolytic uremic syndrome
D. HELLP syndrome
E. Acute fatty liver of pregnancy

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(A Critical Approach to an Emergency Department Patient)
Answer: D. HELLP syndrome

The patient described above has the HELLP syndrome (H = hemolysis; EL = elevated liver enzymes; LP = low platelets). This condition typically develops in the third trimester of pregnancy and occurs in 3 to 12% of patients with preeclampsia and eclampsia. However, 30% of cases of the HELLP syndrome can develop in the first 24 to 48 h postpartum. Patients often complain of progressive malaise, nondependent edema, and, occasionally, right upper quadrant pain. Early on, significant hypertension may be absent.

Although the pathogenesis remains incompletely defined, it is thought that the HELLP syndrome results from vasospasm leading to microangiopathic hemolysis and endothelial cell damage, with subsequent platelet consumption and fibrin deposition. The primary organ of involvement is the liver and, in severe cases, hematomas can develop in the subcapsular area, contributing to hepatic necrosis.

Diagnostic criteria include the following: (1) microangiopathic hemolytic anemia with schistocytes on peripheral blood smear (Fig 1); (2) bilirubin ≥ 1.2 mg/dL (≥ 20 μmol/L); (3) lactate dehydrogenase ≥ 600 IU/L; (4) aspartate aminotransferase ≥ 70 IU/L; and (5) thrombocytopenia, with a platelet count < 100,000/mm³ (100 × 10⁹/L). Treatment of the HELLP syndrome in those pregnancies with viable fetuses is immediate delivery. The immature fetus can be monitored conservatively, although there is increased risk for catastrophic placental abruption, renal failure, or hepatic rupture. In those patients who develop an acute abdomen, shock, or severe right upper quadrant pain, rupture of a subcapsular hematoma should be suspected and surgical intervention recommended. HELLP syndrome has been reported to recur in subsequent pregnancies.

The HELLP syndrome can be difficult to distinguish from other causes of microangiopathic hemolytic anemias, including thrombotic thrombocytopenic purpura or the related disorder, hemolytic uremic syndrome, and disseminated intravascular coagulation. Other causes of liver disease in pregnancy include acute fatty liver of pregnancy, which often presents with fulminant hepatic failure including hypoglycemia, markedly elevated bilirubin levels, and coagulopathy. Hemolysis is not a common feature in acute fatty liver of pregnancy. Other characteristic features of these disorders are outlined in Table 1.

**Suggested Readings**


Table 1—Characteristic Features of HELLP, HUS, TTP, DIC, and AFLP

<table>
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<th>Features</th>
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<th>HUS</th>
<th>TTP</th>
<th>DIC</th>
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</table>

*HELLP = hemolysis, elevated liver enzymes, and low platelets; HUS = hemolytic uremic syndrome; TTP = thrombotic thrombocytopenic purpura; DIC = disseminated intravascular coagulation; AFLP = acute fatty liver of pregnancy.