Acute Fatty Liver of Pregnancy: DX

Highlights: Acute fatty liver of pregnancy, characterized by microvesicular fatty infiltration of hepatocytes, is a disorder which is unique to human pregnancy. It is rare, with an approximate incidence of 1 in 7000 to 1 in 20,000 deliveries. It is more common with multiple gestations and possibly in women who are underweight. Acute fatty liver occurs typically in the third trimester. The disease is always present before delivery, although it is not always diagnosed prior to delivery. The most frequent initial symptoms are nausea or vomiting (approximately 75 percent of patients), abdominal pain (particularly epigastric, 50 percent), anorexia, and jaundice. About one-half of patients have signs of preeclampsia at presentation or at some time during the course of illness. The association of cases of acute fatty liver of pregnancy with one of the inherited defects in mitochondrial beta-oxidation of fatty acids, long-chain 3-hydroxyacyl CoA dehydrogenase deficiency (LCHAD), suggested that some affected women and their fetuses might have an inherited enzyme deficiency in beta-oxidation that predisposes the mother to this disorder.

Dx: The diagnosis of acute fatty liver of pregnancy is usually made clinically based upon the setting, presentation, and compatible laboratory and imaging results. Laboratory tests that are helpful include serum aminotransferases, serum bilirubin, coagulation studies, electrolytes, serum glucose, uric acid level and creatinine, and a white blood cell count. Imaging tests of the liver are primarily used to exclude other diagnoses (e.g. infarct, hematoma)

The major other condition that must be excluded is the HELLP syndrome, which is characterized by hemolysis, elevated liver enzymes, and a low platelet count. There is a large clinical overlap between acute fatty liver of pregnancy and HELLP syndrome, and it may be difficult, even impossible, to differentiate them. However, evidence of hepatic insufficiency such as hypoglycemia or encephalopathy and abnormalities in coagulation studies is more consistent with acute fatty liver of pregnancy.

TREATMENT AND COURSE — Treatment of acute fatty liver of pregnancy is a combination of maternal stabilization and prompt delivery of the fetus, regardless of gestational age. Patients with acute fatty liver of pregnancy are extremely susceptible to developing coagulopathies due to decreased hepatic production of coagulation factors and/or disseminated intravascular coagulation (DIC). As a result, these patients are at high risk for bleeding complications (eg, postpartum hemorrhage). Serial monitoring of the patient's platelet count, international normalized ratio (INR), partial thromboplastin time, and fibrinogen levels should be undertaken to assess for overt or evolving coagulopathy.

Pearl: Acute fatty liver can recur in subsequent pregnancies, even if the search of LCHAD mutation is negative.

Question:

1. A 25 year-old female on her last trimester presents with abdominal pain, nausea, malaise, anorexia and jaundice. Her platelet count is low. Which of the following is more compatible with Acute Fatty Liver of pregnancy than HELLP syndrome?
   a. Hemolysis
   b. Bleeding
   c. Low antithrombin III
   d. Elevated glucose

Answer: D