Rare but Deadly TTP Can Masquerade as HELLP

BY SARAH PRESSMAN LOVINGER
Contributing Writer

CHICAGO — You are caring for a gravid woman with very low platelets and hemolytic anemia. You diagnose her with HELLP syndrome and order a platelet transfusion. Is this the best management of a pregnant patient with thrombocytopenia at risk of bleeding?

Not always, according to Dr. James J. Martin Jr. of the University of Mississippi Medical Center in Jackson.

Thrombotic thrombocytopenic purpura (TTP) can masquerade as HELLP (hemolysis, elevated liver enzyme levels, and low platelet count) syndrome. Administering platelets to any patient with this rare but often fatal hematologic disorder can paradoxically worsen the patient's condition, leading to severe disability and death.

If physicians mistake TTP for HELLP syndrome and administer one or more platelet transfusions, they may aggravate the hemolytic state.

Physicians must always consider TTP when caring for pregnant women who present with low platelets.

"Obstetricians-gynecologists are really in a quandary," Dr. Martin said at the annual meeting of the Central Association of Obstetricians and Gynecologists.

"The main problem is differentiating between TTP and HELLP," he said. "You need to do it right away," he added.

While TTP is an extremely rare disease that affects only 4-11 patients per million population, the combination of low platelets, hemolytic anemia, renal failure, and mental status changes can be deadly if undiagnosed.

If physicians mistake TTP for HELLP syndrome and administer one or more platelet transfusions, they may aggravate the hemolytic state caused by absent or severely low levels of a von Willebrand factor–cleaving protease, a potentially fatal error.

Plasma therapy can help replete this protease—known as ADAMTS-13, a disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13—and stop the destructive hemolysis that TTP causes. Maternal mortality caused by TTP can drop to less than 10% in women properly treated with plasma therapy.

Obstetricians lack a straightforward method to differentiate between TTP and HELLP syndrome. "We need a definite test for TTP," said Dr. Martin.

To obtain more information about lab findings in pregnant women that could shed light on possible tests to ease diagnosis of these confounding disorders, Dr. Martin and his colleagues performed an extensive English-language literature search of pregnant patients with TTP, and uncovered 166 reported cases from 1955 to 2006. They excluded patients with hemolytic-uremic syndrome, postpartum renal failure, or severe preeclampsia/eclampsia/HELLP syndrome who lacked reasonable information that could diagnose TTP.

The researchers uncovered only seven papers published between 2002 and 2006 looking at ADAMTS-13 activity levels in suspected TTP patients. Additional analysis revealed that several common lab tests could help differentiate women who have TTP only from women who have TTP together with preeclampsia/HELLP syndrome. The researchers found that the ratio of lactate dehydrogenase (LDH) to serum glutamic oxaloacetic transaminase (SGOT) levels was considerably higher in patients with TTP only, compared to women with combined TTP/preeclampsia/HELLP syndrome.

Dr. Martin recommends testing for ADAMTS-13 levels in pregnant patients who have a LDH/SGOT ratio that exceeds 15. He also recommends this test early in pregnancy in women who have a history of TTP.

A rapid, reliable lab test could help physicians distinguish more easily between the common preeclampsia/HELLP syndromes and TTP, the deadly masquerader. Until then, physicians must keep in mind that pregnant patients who present with low platelets and hemolytic anemia could have TTP and act accordingly, Dr. Martin said. ■