Emergent Cesarean Section in a Patient with Systemic Lupus Erythematosus and Atypical HELLP Syndrome

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Introduction:
HELPP syndrome is a condition in pregnancy characterized by hemolysis, elevated liver function tests, and thrombocytopenia. Atypical HELLP syndrome is described as HELLP syndrome without evidence of hypertension or proteinuria. We describe the case of a multigravida with known systemic lupus erythematosus (SLE) and anti-phospholipid antibody syndrome (APS), presenting for emergent cesarean delivery due to atypical HELLP syndrome.

Case Description:
28 year old female patient, G5P0220, presented for emergent cesarean section at 31 weeks gestation due to a diagnosis of atypical HELLP syndrome. Her past medical history was significant for systemic lupus erythematosus (SLE), anti-phospholipid antibody syndrome (APS), deep venous thrombosis and pulmonary embolism, alpha-1 antitrypsin deficiency, MTHFR mutation, intra-hepatic cholestasis of pregnancy, two prior miscarriages, two prior classical cesarean sections secondary to HELLP syndrome (resulting in fetal demise post-delivery at 22 and 25 weeks), and morbid obesity.

She was initially admitted to the hospital 5 days prior with nausea, vomiting, right upper quadrant (RUQ) pain, elevated liver function tests, and mild thrombocytopenia. Her history and physical exam suggested the possibility of an acute severe lupus flare. During her hospital course, her RUQ pain worsened despite increasing doses of corticosteroids and analgesics in the setting of elevated but stable liver function tests and stable thrombocytopenia (> 100,000/uL). On the morning of hospital day five, laboratory findings were significant for a 10-fold increase in AST, a 7-fold increase in ALT, a rise in total bilirubin, and worsening thrombocytopenia (< 100,000/uL). She was then taken to the OR for emergent cesarean section with a diagnosis of atypical HELLP syndrome. Blood products were readily available due to patientâ€™s thrombocytopenia and recent therapeutic dose of enoxaparin.

The patient was placed supine on the operating room (OR) table, with left uterine displacement. Standard monitors were applied. While the obstetricians were prepping and draping the surgical field, 100% of oxygen was given to the patient by a face mask. After five minutes of preoxygenation, general anesthesia was induced using a rapid sequence intravenous technique (propofol and succinylcholine), and patient was uneventfully intubated. After endotracheal intubation, an arterial line was placed for continuous blood pressure monitoring and serial blood gas sampling, and a classical incision was made with delivery of a live neonate 8 minutes after anesthesia induction.
Anesthesia was maintained with sevoflurane, fentanyl, hydromorphone and midazolam. Estimated blood loss for the procedure was 1L and the patient remained hemodynamically stable throughout, receiving a total of 1.8L of crystalloids. At the conclusion of the surgery, the patient was uneventfully extubated and transferred to the postoperative care unit (PACU) for further hemodynamic monitoring. Her post-operative course was significant for down trending of her liver function tests and resolution of her thrombocytopenia. However, her RUQ pain continued and two days later an MRI of the abdomen was significant for right hepatic lobe infarction.

Discussion:

This case demonstrates the importance of considering HELLP and atypical HELLP syndrome in patients with complex rheumatologic and hematologic histories, recognizing and preparing for intra-operative complications from HELLP syndrome, and how it affects anesthetic technique.