POSTPARTUM HEMORRHAGE IN AN OBSTETRIC PATIENT WITH PROTEIN S DEFICIENCY

BACKGROUND AND OBJECTIVES: Postpartum hemorrhage is defined as the loss of more than 500 mL of blood after delivery and can result in hemodynamic instability. Even with appropriate obstetric management, approximately 3 percent of vaginal deliveries will result in severe postpartum hemorrhage (> 1000 mL of blood loss) (1,2). Severe postpartum hemorrhage is the most common cause of maternal morbidity and occurs in up to 18 percent of total births (1).

Uterine atony, or failure of the uterus to contract following delivery, is the most common cause of postpartum hemorrhage (3). Uterine atony is usually managed by administration of oxytocin prostaglandins or ergot alkaloids or both. In cases of severe, intractable hemorrhage despite the above medications, uterus-conserving treatments include tamponade procedures and uterine artery embolization.

Coagulation disorders, are a rare cause of postpartum hemorrhage (3) and should be considered in patients who do not respond to the usual measures used to treat postpartum hemorrhage. Deficiencies of natural anticoagulants, such as protein C, protein S and antithrombin, are very rare and, overall, occur in fewer than 2% of the general population. Individuals with protein S deficiency are at risk of deep vein thrombosis and factors such as pregnancy can raise the risk of abnormal blood clots (4). The risk of thrombosis in pregnancy varies from 0–6% and 7–22% among postpartum population with protein S deficiency (5,6).

We present a patient with protein S deficiency and severe postpartum hemorrhage from uterine atony.

Case report: A 32-year-old G5P2A2L2 parturient at 38 plus weeks gestation, with a history of mild intermittent asthma, hereditary protein S deficiency maintained on subcutaneous lovenox which was stopped in anticipation of delivery, presented to the labor and delivery suite for induction of labor. H & P was unremarkable except for a borderline high BP (Sys ~ 140s) and BUN/Cr 04/0.6. Normal values included Hgb 11.1 g/dl, PT 12.1 sec, APTT 43.6 sec, Fibrinogen level 351 mg/dl. An uneventful vaginal delivery was facilitated by placement and subsequent removal of a labor epidural catheter.

About 50 min after the epidural catheter was removed, the patient began to bleed profusely despite the addition of pitocin, methergine and Hemabate in labor suite. Uterine atony was diagnosed. Laboratory values showed Hgb 7.4 g/dl, PT 16.8 sec, APTT 41.1 sec, Fibrinogen level 200 mg/dl.
The patient was moved to the operating room. General Anesthesia was induced with etomidate and succinylcholine. She was easily intubated and a right radial arterial line and a right internal jugular were placed and the massive transfusion protocol was initiated for hemodynamic instability following severe hemorrhage as her Hgb dropped to 6.1g/dl.

The patient subsequently became difficult to ventilate. Differential included TRALI vs flash pulmonary edema vs asthma exacerbation vs bronchospasm. The ventilatory problem resolved gradually with diuretics and bronchodilators.

The patient received a total of 10 units of PRBCs, 12 units of FFP, 2 units of cryoprecipitate, one unit of pooled platelet along with vasopressors to maintain hemodynamic stability. As her bleeding was not completely controlled, she was moved to the interventional radiology suite for bilateral hypogastric embolization. Patient was brought to the IR suite in critical condition and an ICU ventilator was used to control ventilation. Anesthesia was supplemented with ketamine and propofol as tolerated. She continued to require vasopressor support and was transferred to the ICU for post operative care.

Her ICU course was fairly uneventful and she was weaned from vasopressors and was extubated ~ 36 hours after admission. She was discharged home on Lovenox and mother and baby were doing well on initial follow up.

Conclusion and Discussion:

The parturient with coagulation defects presents a unique challenge to the anesthetist. We present a case of a patient with early postpartum hemorrhage due to uterine atony complicated by protein S deficiency. The deficiency could have attributed to widespread blood clots using up all available blood clotting proteins resulting in consumptive coagulopathy leading to further uncontrollable hemorrhage.

Due to the rarity of many of these clotting factor disorders, consensus guidelines are lacking. More studies are crucial on optimum factor-replacement approaches, including the duration of treatment, vigilance in these patient population with new point-of-care tests, and appropriate protocols to ensure prevention of postpartum hemorrhage.