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Failure of Intraoperative Red Cell Salvage: A Patient with Sickle Cell Disease and HELLP (Hemolysis, Elevated Liver enzymes and Low Platelets) Syndrome

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Abstract: Cell salvage is a process whereby the bloodshed from the operative field is collected and returned to the patient. It can be especially useful when allogeneic red blood cell (RBC) units are not readily available such as when the recipient has multiple alloantibodies. We report on the anesthesia and transfusion strategies for managing a pregnant patient with sickle cell disease (SCD) with HELLP (Hemolysis, Elevated Liver enzymes and Low Platelets) syndrome. A pregnant patient with twins at 30 weeks of gestation was admitted in an SCD crisis. She subsequently developed HELLP syndrome and required urgent cesarean delivery; however, she had multiple RBC antibodies complicating the immediate provision of crossmatched RBC units. Cell salvage was used to capture the blood shed during her procedure while the blood bank was searching for compatible RBCs units. Despite multiple interventions designed to optimize the cell salvage procedure for the unique challenges of a patient with SCD, the salvaged RBCs hemolyzed and could not be reinfused. Cell salvage in an obstetric patient with SCD in an acute crisis and superimposed HELLP was unable to recover intact and useable RBCs. Further studies into methods of optimizing the procedure for use in this context are warranted. Close communication between the clinical teams treating the patient and the transfusion service is required so that the RBC transfusion requirements can be anticipated; this is especially important when the patient has multiple antibodies. **Keywords:** sickle cell disease, HELLP syndrome, cell salvage, thromboelastogram, anticoagulation. *JECT. 2014;46:314–316*

A patient with sickle disease (SCD) in an acute sickle crisis at 30 weeks of gestation with dichorionic–dichorionic (di-di) twins developed HELLP (Hemolysis, Elevated Liver enzymes and Low Platelets) syndrome, a critically low hemoglobin (Hb) concentration, and acute chest syndrome requiring urgent cesarean delivery. She also had multiple red blood cell (RBC) antibodies, which complicated the provision of crossmatched RBCs from the blood bank. We describe the blood transfusion strategy for this patient. The patient is a young woman with SCD who presented at 30 weeks of gestation with di-di twins. At presentation she was 163 cm tall and weighed 82 kg. The patient initially presented to the outpatient triage unit at a large maternity hospital with concern for an acute pain crisis. She was admitted for hydration and pain management. Her medical history also included asthma, chronic anemia, and thrombocytopenia with admission values of 9.6 g/dL and 75,000/μL, respectively, recently diagnosed pulmonary hypertension, and schizophrenia. The patient had stopped taking her prescribed hydroxyurea 13 weeks before this presentation as a result of a concern about its teratogenicity; however, she was still taking quetiapine (Seroquel) and albuterol. A type and screen was ordered on the day the patient was admitted. It revealed that the patient had four clinically significant RBC antibodies (Anti-C, K, Fyb, Jka) suggesting that crossmatched RBCs would not be immediately available should they be needed urgently. Despite recognition of the antibodies, an order to actually crossmatch RBCs or to put several units on hold was not placed along with the type and screen order.

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OPERATIVE PLAN AND TREATMENT

Two days after admission the patient developed preeclampsia with proteinuria and consistently elevated systolic blood pressure over 135 mmHg with diastolic blood

pressures consistently above 80 mmHg. At this time her laboratory studies demonstrated a further decline in her Hb concentration and platelet count to 7.2 g/dL and 41,000/μL, respectively, as well as rising liver enzyme concentrations consistent with HELLP syndrome. A peripheral blood film showed sickle cells, schistocytes, and target cells. The obstetric plan was to transfuse RBCs and perform a cesarean delivery.

Abruptly, before the patient could be transfused, she decompensated and her Hb concentration fell to 6.5 g/dL and her platelet concentration dropped to 30,000/μL. She developed acute respiratory distress with her oxygen saturation decreasing to 83% on room air. Clinically it was felt that she was experiencing an acute chest crisis, which is a common event in patients with SCD. In addition to these clinical findings, the diagnosis of acute chest syndrome can also be suggested by the appearance of new infiltrates on radiography. However, as a result of the acuity of her decline, a chest radiograph was not obtained. At this time she also experienced some altered mentation and the decision was made to urgently take her to the operating room for a cesarean delivery. Before moving her to the operating room, a platelet transfusion was started because the RBC antibodies do not complicate the provision of platelets.

When the decision to take the patient to the operating room was made, an order for crossmatched RBCs was sent to the blood bank, and an extensive search of the local RBC inventory revealed two compatible and leukoreduced units; one unit was in liquid form and could be transfused immediately, whereas the other would be available after deglycerolization, a process requiring several hours. The blood bank was able to find a total of six antigen-negative RBCs at two surrounding blood centers; however, there would be an approximately 24-hour delay in receiving these six additional units as a result of the geographic distance between the patient's hospital and these blood centers.

The intraoperative plan focused around management of her anemia, acute chest syndrome with respiratory distress, and thrombocytopenia. When the patient arrived in the operating room, her oxygen saturation was 91% on 6 L/min by facemask. The patient had a rapid sequence induction with 1 mg/kg Propofol followed by 2 mg/kg succinylcholine. The patient's blood pressure throughout her surgery ranged from 125 mmHg to 140 mmHg systolic and the diastolic ranged from 80 mmHg to 90 mmHg. To minimize surgical blood loss, 1 g tranexamic acid was given before the surgery started. The patient was kept euthermic with the use of warming forced-air blankets, an intravenous fluid warmer, and additional preheated blankets on her lower extremities.

Because only two compatible RBC units were available at the time of the surgery, a cell salvage device was prepared and brought into the room. From the start of surgery, blood was collected in the cell salvage device (COBE BRAT, Arvada, CO). In an attempt to minimize sickling, several changes in standard processing were made. To keep the shed blood from getting cold, a forced-air warming blanket was wrapped around the collection reservoir and set to 43°C. In an attempt to minimize acidosis, Ringer's lactate solution was heparinized as the anticoagulant and run at standard rates. Ringer's lactate solution was also prepared as the wash solution. Throughout the case, approximately 1 L of blood was collected; however, on inspection, the suctioned blood appeared to be clotted. Given that adequate heparin had been mixed with the suctioned blood, it was hypothesized that the RBCs had sickled. Thus, the salvaged product was not suitable for retransfusion.

After both fetuses were delivered, the patient had ongoing uterine atony, which was clinically diagnosed by failure of the uterus to constrict in size postplacental delivery. In addition to an oxytocin infusion, intramuscular methylergovine at 200 μg was given and the anesthesia gas concentration was reduced. An arterial blood gas (ABG) was drawn that showed a Hb concentration of 6.2 g/dL. Both of the crossmatched RBCs were administered during the surgery. The patient continued to have uterine atony and 250 μg of carboprost was given intramuscularly.

Because of her preoperative chest crisis, the decision was made to keep her intubated after the surgery was completed. Immediately postoperatively, the patient had a chest radiograph that revealed a pulmonary infiltrate consistent with her clinical findings of an acute chest syndrome. After repeated ABGs, which showed a mixed metabolic and respiratory acidosis, a trend to normalization of her acid-base status was observed; however, she still required a FiO_2 of 0.7. An additional four units of RBCs were administered over the ensuing 24 hours, which led to an increase in her Hb to 10.2 g/dL. On postoperative day (POD) 1, the patient was able to be extubated and placed on facemask oxygen. She was discharged home on POD 7.

DISCUSSION

This case highlights several interesting practice points. One of the primary therapeutic modalities for patients in a sickle cell crisis is RBC transfusion. Patients with SCD are known to have significantly higher rates of RBC alloimmunization compared with non-SCD RBC recipients, approaching 44% in the former (1). When a recipient has multiple antibodies, or even a single antibody against a high-incidence antigen that is present on most donors' RBCs, finding RBC units that lack the antigen(s) and are thus safe for transfusion becomes a challenge; the blood center has to screen its inventory to determine if compatible units can be found and if none are locally available,

then other blood centers have to be queried to determine if suitable RBCs can be obtained. This process is time-consuming. In this case, two units of RBCs were available within approximately 12 hours of the order for RBCs and more arrived later in the day. Earlier notification of the blood bank of the patient's clinical status would have led to an earlier search for RBCs and would potentially have increased the number of units available during the case. Thus, when a patient with SCD with a suspected sickle crisis is admitted and the probability of requiring an RBC transfusion is high, the blood bank should be notified as soon as possible, even if RBCs are not immediately required. The details of the patient's condition and the potential need for RBCs can be discussed with the blood bank's medical director, and a plan for locating units and a timeline for having them available can then be developed.

Because only two compatible RBCs were available for the surgery, a cell salvage device was used to collect any shed blood and return it back to the patient. The use of cell salvage in SCD is understudied and only few commentaries are available (2–4). Several reports have been published describing cell salvage use in patients with sickle cell trait (5–7). There are only two case reports describing the use of salvaged blood during surgery in a patient with SCD (8,9). In one of these cases, no useable RBCs were recovered and a high percentage of cells showed the characteristic sickle shape when viewed under a light microscope. To our knowledge there have been no previous reports describing the use of cell salvage in a patient with sickle cell crisis. Thus, with few alternatives, a strategy was developed to try to minimize sickling by minimizing acidosis of the shed blood using lactated Ringer's (LR) solution instead of normal saline, maintaining the patient's temperature at nearly normal levels, and recognizing that exposure to room air should maintain high oxygenation and prevent sickling. Heparinized LR was used instead of heparinized saline or citrate solution for the anticoagulation solution. Lastly, to maintain a near physiologic temperature of the shed RBCs, a forced-air warming blanket was wrapped around the collection reservoir. Despite these interventions, the salvaged blood rapidly became unusable. Thus, it would appear that cell salvage has no benefit in a patient with SCD in an acute crisis.

The use of a lysine derivative, tranexamic acid, was administered to reduce blood loss, although its use has not been studied in the setting of SCD and HELLP syndrome. However, because it is known to be effective in reducing bleeding in other surgeries, a low dose was administered. There was no noticeably observed adverse effect of its use noted in the intraoperative or postoperative period.

The management of pulmonary hypertension in a patient with SCD with superimposed HELLP syndrome warrants discussion. The patient had a recent diagnosis of pulmonary hypertension, which is not uncommon in the sickle cell population. The mechanism for this is likely the chronic hemolysis that occurs in a patient with SCD with the subsequent liberation of free Hb that scavenges nitric oxide leading to chronic pulmonary vasoconstriction (10). Perhaps synergistically, HELLP and sickle cell crisis could lead to increases in free Hb and worsening of pulmonary hypertension. Intravascular hemolysis seen with HELLP syndrome may be interlinked with a sickle cell crisis leading to worsening pulmonary hypertension.

CONCLUSION

Overall, this unique case posed numerous challenges to the transfusion and surgical services. Obtaining a type and screen and potentially ordering RBCs for transfusion should be done early in the patient's admission and in consultation with the blood bank's medical director in case extra time is required to identify compatible units. Additionally, cell salvage appears to not be useful in a parturient in a sickle cell crisis.

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