Case Report

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Alloimmunization in Sickle Cell Disease with Anemia and Pregnancy: A Case Report

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Abstract

Role of anesthesiologist in management of sickle cell disease in pregnancy is related to its complications such vaso-occlusive crisis with severe pain, sepsis, acute chest syndrome, stroke, priapism, avascular necrosis of hip, alloimmunization with severe hemolysis with anemia and fetal complications such as preterm, intrauterine growth restriction (IUGR), hemolysis, hyperbilirubinemia, and intrauterine device. Mainstay of treatment is symptomatic which includes analgesia, hydration, oxygenation, blood transfusion, antibiotics, folate, and hydroxyurea. Herein, we provide brief review of our patient with alloimmunization due to past blood transfusions for sickle cell anemia. We experienced severe hemolysis in our patient due to difficulty to cross match all red cell antigens in donor and antibodies against the K, E, C, Jk^b antigens in our patient. That changed our outlook toward blood transfusion in this patient which requires further red cell antigen phenotype study in donor and antibody study in patient. Furthermore, this patient had difficulty for spinal anesthesia due to decrease mobility of hip due to avascular necrosis and unbearable painful vaso-oclusive crisis requiring good pain relief. Her baby was preterm, IUGR, had hyperbilirubinemia and died on day 3 of life. Such case first time happened in our hospital and was rarely described previously in anesthesia literature to our knowledge. Though blood and hematology literature do mention about this. It is difficult to manage medicolegal aspects of such blood transfusion, where anesthesiologist is directly involved in blood transfusion in perioperative period, pain relief, oxygen, fluid-acid base, and multisystem critical care management.

Key words: Alloimmunization, Blood transfusion, Cross match, Red cell antigen phenotypes, Sickle cell disease

INTRODUCTION

Sickle cell disease (SCD) is a group of inherited single gene autosomal recessive disorders caused by the "sickle" gene, which affects hemoglobin structure.[1] Sickle cell anemia (hemoglobin S [HbS] disease) is due to the production of abnormal hemoglobin due to a single amino-acid substitution in the beta globin chain resulting in glutamic acid being replaced by valine at the 6th position and HbS is produced instead of adult hemoglobin (HbA). Someone with sickle cell trait (a carrier of SCD) will produce both HbA and HbS and is often described as HbAS.[2] Pathology and clinical presentation do not affect until 1st year of life. After that, fetal hemoglobin is replaced by (HbA and HbA2). HbA is made up of abnormal hemoglobin due to which sign-symptoms starts. The pathophysiology of SCD is a consequence of polymerization of the abnormal hemoglobin in low-oxygen conditions, which leads to the formation of rigid and fragile sickle-shaped red cells. These cells are prone to increased breakdown, which causes the hemolytic anemia, vaso-occlusion in the small blood vessels which causes most of the other clinical features including acute painful crises. Other complications of SCD include stroke, pulmonary hypertension, renal dysfunction, retinal disease, leg ulcers, cholelithiasis, and avascular necrosis (which commonly affect the femoral head and may necessitate hip replacement).[3] Sickle trait is asymptomatic, except for a possible increased risk of urinary tract infections and microscopic hematuria. SCD was previously associated with a high early mortality rate, but now the majority of children born with SCD in the UK live to reproductive age and average life expectancy is at least the mid-50s. Women with SCD appear to be susceptible to medical complications including increased infection; an increase in sickle cell related painful episodes,

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increased fetal morbidity, fetal growth restriction, preterm labor, and increased cesarean section rates.[4]

Patients often require supplementary hydration, analgesia, and oxygen. Life-threatening crises may require urgent exchange blood transfusion and management on a critical care unit. Transfusion is not without risk or burden; in particular the risk of alloimmunization (the formation of additional red cell allo-antibodies) can be significant. Patients with SCD are immunogenic and it is not uncommon for them to form antibodies that can lead to delayed hemolytic transfusion reactions, hemolytic disease of the fetus and the new born and also make future cross matching of blood difficult. Blood is routinely fully matched for to reduce this risk.[5]

CASE REPORT

A 37-year-old female, weighing 65 kg presented with severe bilateral upper limb pain and backache. She was 29 weeks pregnant ($G_3P_1L_1A_1$) and known case of SCD with anemia. 15 years back she underwent cesarean section because of sickle cell crisis and delivered a preterm baby, who is living child. She also underwent laparoscopic surgery 10 years back which was uneventful. For above reasons, she received multiple blood transfusions. She was also known case of avascular necrosis of hip and femur head with decreased mobility and painful hip joints. She has multiple episodes of painful crisis of extremity and lower back relived with hospitalization and analgesics which includes paracetamol, fentanyl, tramadol, hydration, and oxygen. In present pregnancy patient gives increased severity and frequency of above symptoms requiring frequent visits to hospital and hospitalization. The patient was admitted intensive care unit for 10 days for same reasons, after treatment she was comfortable and discharged home. However, within 12 h of discharge, she readmitted with unbearable pain in upper limb and backache which required intense care which includes fluid, oxygen, fentanyl, blood transfusion in view of anemia (Hb 7.8 g%) with pregnancy. After blood transfusion patient Hb further dropped to 6 g% and bilirubin levels reached to 10 mg% along with persistent symptoms. At this time, hematologist came into picture and thought process for alloimmunization in SCD started. Meanwhile, worsening condition of patient along with pregnancy make our team to take decision of cesarean section though she was preterm because pregnancy itself is precipitating factor for acute crisis in SCD. The patient and her family were ready for C-section with informed written consent regarding maternal and fetal complications. The patient taken for Cesarean section and spinal anesthesia given with 27 Q spinal needle in left lateral position with all aseptic precautions. We found procedure difficult due to inappropriate position in view of hip immobility and pain. After surgery, patient received two pack cells which are cross matched for different antigens in donor and antibodies in recipient considering alloimmunization due to previous blood transfusions. Post-operative day one patient Hb was 8 g% but symptoms persistent and so patient managed in critical care unit, where she received good care, hydroxyurea, warfarin, heparin along with supportive treatment, and after 3 weeks she shifted in ward and subsequently home.

DISCUSSION

There is published data to support the conclusion that the risk of alloimmunization is greater in patients receiving transfusions for SCD than in patients receiving transfusions for other chronic diseases. In a study conducted by Vichinsky *et al*. in 1990 demonstrated that alloantibodies developed in 30% and delayed transfusion reactions in 11% of patient receiving transfusions in SCD while only 5% in patients with chronic anemia receiving blood transfusion.[6] According to Vichinsky *et al*. this represent low estimate of the rate of alloimmunization, since not all antibodies are detected by standard techniques. 16% of patients have multiple antibodies which resulted in clinical complications (delayed hemolysis) and delayed transfusion therapy due to additional problems in cross matching. Red cell phenotypes most likely to cause antibodies to develop in patient with SC anemia are K, E, C, Jk^b. Activation of immune system may contribute to increased incidence of delayed transfusion reactions. Causes for increased incidence of alloimmunization in SCD are due to lack of phenotypic compatibility between donor and recipients (Giblett demonstrated that when black patient receives blood from white donor the risk of alloimmunization was higher when donor and recipients were matched for race) and racial difference(Kim *et al*., found increased incidence of alloimmunization in black patients compared to white patients).[1] The present case illustrates a problem created due to inadequate cross matching which was not done for red cell antigens which are most likely to cause antibodies in recipients' giving hemolytic reactions. After cesarean section, we read above literature and references.Then, donor's blood was cross matched with recipient for all possible red cell phenotypes in blood bank, and we transfused two packed cell volume to our patient. After 24 h, patient Hb increased with no laboratory evidence of hemolysis confirmed by bilirubin levels, liver enzymes disease etc.[7-10]

CONCLUSION

Most of anesthesiologist are aware of pathophysiology of SCD, its complications and management. Anaesthesiologist managing such cases either in operation room as anesthesiologist or as intensives in intensive care. However, awareness about alloimmunization in SCD is less amongst anaesthesiologist. This may be because this pathology is rarely seen, remain undiagnosed, unnoted or many time this management is done by hematologist as teamwork. Here, we faced difficulty in positioning due to pain and avascular necrosis of hip which was manageable. However, our nightmare was death of baby and severs hemolysis with organ dysfunction in mother who needed intensive care for more than 3 weeks and top on this economical and financial loss to prove that alloimmunization is known with SCD with previous blood transfusions. So, we recommend that all patients with SC anemia who undergo transfusion be matched with donor for the red cell antigens commonly associated with alloimmunization and transfusion reactions, aim is to avoid phenotypic incompatibility between donor and recipient.

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