INDEX OF SUSPICION

Case of Hemolytic Disease of the Newborn due to a Rare Red Cell Antibody

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PRESENTATION

An early term, 2,860-g female infant is delivered via caesarean section by a 28-year-old gravida 3, para 1-1-1 woman. Her second pregnancy had terminated in an abortion 4 years ago with the cause unknown.

Results of serologic studies at the time of her previous pregnancies are not available, but the antibody screen (3-cell panel, Immucor Inc, Peachtree Corners, GA) during the present pregnancy is positive. The mother's red blood cells (RBCs) are typed as B Rh (D) positive. The sample reacts with all cells in the 16-cell panel (Capture-R Ready-ID, Immucor Inc) and 11 cell panels (ID DiaPanel, Biorad Labs, Hercules, C). The auto control and direct antiglobulin test results are negative. The reaction pattern seen on screening and identification panels indicates the presence of an RBC alloantibody to a high-frequency antigen. Crossmatch with multiple B-positive and O-positive RBC units are incompatible on Coombs testing. Extended phenotyping of the mother for minor blood group antigens of high frequency reveals her minor blood group phenotype to be K−, k+, Kp (a−b+), Js (a+b−), Lu(a–b+) and In(a–b+).

Allogeneic adsorption studies done using 0.01%-ficin treated RBCs of known phenotype to deliberately adsorb the anti-Js^b from the mother's serum confirms the absence of other common alloantibodies. The antibody is confirmed to be anti-Js^b alloantibody. The anti-Js^b titer using the antiglobulin technique Js^b(+) red cells is 128. The father's phenotype is positive for Js^b. She has no prior history of blood transfusions. Her first child, born 6 years ago, has no history of jaundice requiring treatment. Antenatal ultrasonography with middle cerebral artery Doppler has normal findings.

The neonate is born at 37 weeks and 6 days of gestation, appears pink, and requires only routine care. Investigation shows the neonate's blood type to be B Rh (D) positive, that is, similar to the mother. Direct antiglobulin test (IgG+C3d) is positive (4+) and antibody screen is pan-reactive. Antibody identification using the neonate's plasma and RBC eluate shows pan-reactivity similar to that of the mother, confirming the presence of maternal anti-Js^b alloantibodies in the neonate's plasma. The neonate's RBCs are found to be Js^b positive

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and anti-Js^b titers in her plasma is 16.At about 16 hours after birth, the neonate is started on intensive phototherapy as her total serum bilirubin is 8.17 mg/dL (140 μmol/L) and hemoglobin is 17 g/dL (170 g/L). Because the bilirubin value is found to be near exchange transfusion value (8.8 mg/dL [150 μmol/L]) with positive direct antiglobulin test, the neonate is given a transfusion of intravenous immunoglobulin to prevent exchange transfusion. (1) Exchange transfusion in such a scenario has to be performed only with Js^b-negative blood, which is extremely difficult to find in our population. Repeat bilirubin value 6 hours after commencing phototherapy decreases to 6.15 mg/dL (105 μmol/L) but her hemoglobin concentration drops to 13.5 g/dL (135 g/L). It is decided to give her a transfusion with her mother's RBCs in case the hemoglobin drops to critical levels because they share the same ABO Rh type. Phototherapy is continued, her bilirubin value 40 hours after birth drops to 4.92 mg/dL (84 μmol/L), and hemoglobin remains stable. Phototherapy is stopped on day 3 after birth and rebound bilirubin is 8.69 mg/dL (149 μmol/L) with no further drop in hemoglobin. The infant remains well and is discharged on day 5 after birth. She is followed up on day 7 and bilirubin and hemoglobin values are found to be 12.77 mg/dL (218 μmol/L) and 14 g/dL (140 g/L), respectively.

DISCUSSION

Js^b is a high-incidence antigen in all populations. More than 99.9% of whites and 99.9% of blacks have the presence of Js^b antigen of the Kell blood group system. (2) As with other Kell antibodies, fetal anemia due to anti-Js^b is a result of the suppression of erythropoiesis and hemolysis caused by maternal anti-Js^b antibodies that cross the placenta. Hemolytic disease of the fetus and newborn resulting from anti-Js^b is reported to be mild to moderate and, like other clinically significant antibodies to high-incidence antigens, presents a challenge, especially with the lack of antigen-negative blood for intrauterine transfusion or exchange transfusion. (3) Antibody against this antigen causing hemolytic disease of newborn was first reported by Wake et al and described as a mild disease. (4) In 1978, Purohit et al reported the second case, which was of severe nature requiring exchange transfusions and further top-up transfusions with Js^b-negative RBCs. (5) In 1995, Gordon et al reported even more severe disease requiring intrauterine transfusion followed by top-up transfusions. (6) In 2000, another case of severe hemolytic disease of newborn requiring exchange transfusions and top-up transfusions was reported by Stanworth et al. (7) All these cases were seen in the black population, because the absence of Js^b antigen is slightly more common among blacks compared with other races. All the transfusions performed were either from RBCs donated by the mother or Js^b-negative deglycerolysed frozen RBCs.

In our case, the disease was not severe in spite of the high antibody titers found. The case could have been challenging if the ongoing hemolysis had not been controlled, leading to an exchange transfusion. Initial bilirubin was just below the exchange transfusion level along with a normal hemoglobin. With an aim to prevent exchange transfusion, we treated the neonate with intravenous immunoglobulin along with intensive phototherapy. Initially, her hemoglobin level dropped but stabilized later. She was out of phototherapy by day 3 after birth and did not require any RBC transfusion.

To conclude, such a case represents a difficult setting in neonatal practice. Anti-Js^b is a rare antibody against a high-incidence antigen, which has the potential of causing hemolytic disease of the fetus and the newborn. If this happens, the degree of hemolysis and suppression of erythropoiesis can be significant. In this setting, if ABO Rh compatible, the mother's blood is the only available source that could be used in the absence of a rare donor registry and frozen antigen-negative blood inventory.

Lessons for the Clinician

Cases like that described here necessitate a mandatory maternal antibody screen even in non-Rh(D)–negative mothers followed by antenatal build-up of maternal hemoglobin with frequent antenatal surveillance.

- Transfusion with maternal blood if ABO Rh compatible may need to be considered in such exceptional conditions as allogeneic banked Js^b-negative RBCs is extremely difficult to find.
- Timely intervention during the antenatal and immediate postnatal period and awareness of problems associated with management of such rare cases becomes a cornerstone for success.

American Board of Pediatrics Neonatal-Perinatal Content Specification

- Know the causes of and diagnostic approach to an infant who is anemic at birth

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