A Fatal Case of Severe Hemolytic Disease of Newborn Associated with Anti-Jkb

The Kidd blood group is clinically significant since the Jk antibodies can cause acute and delayed transfusion reactions as well as hemolytic disease of newborn (HDN). In general, HDN due to anti-Jkb incompatibility is rare and it usually displays mild clinical symptoms with a favorable prognosis. Yet, we apparently experienced the second case of HDN due to anti-Jkb with severe clinical symptoms and a fatal outcome. A female patient having the AB, Rh(D)-positive boodtype was admitted for jaundice on the fourth day after birth. At the time of admission, the patient was lethargic and exhibited high pitched crying. The laboratory data indicated a hemoglobin value of 11.4 mg/dL, a reticulocyte count of 14.9% and a total bilirubin of 46.1 mg/dL, a direct bilirubin of 1.1 mg/dL and a strong positive result (++) on the direct Coomb's test. As a result of the identification of irregular antibody from the maternal serum, anti-Jkb was detected, which was also found in the eluate made from infant's blood. Despite the aggressive treatment with exchange transfusion and intensive phototherapy, the patient died of intractable seizure and acute renal failure on the fourth day of admission. Therefore, pediatricians should be aware of the clinical courses of hemolytic jaundice due to anti-Jkb, and they should be ready to treat this disease with active therapeutic interventions.

Key Words : Anemia, Hemolytic, Autoimmune; Erythrobastosis, Fetal; Infant, Newborn; Anti-Jkb

CASE REPORT

A female patient having the AB, Rh(D)-positive boodtype was admitted for jaundice on the fourth day after being born. The baby was born following a normal vaginal delivery by 39-yr-old mother having birth history of gravida 4, para 2. Her body weight was 3,000 g, and her gestational age was 37 weeks and 5 days. The mother's past history was unremarkable. There was no history for transfusions of blood, plasma or any blood derivatives. The first and second siblings were treated with phototherapy due to neonatal hyperbilirubinemia. At the time of admission, the patient was lethargic and exhibited high pitched crying, she had a normal Moro-reflex, but there was decreased respond to stimulation. The laboratory data indicated a hemoglobin value of 11.4 mg/dL, a reticulocyte count of 14.9% and a total bilirubin of 46.1 mg/dL, a direct bilirubin of 1.1 mg/dL and a strong positive result (++) on the direct Coombs test. As a result of the identification of irregular antibody from the maternal serum, anti-Jkb was detected. Despite the aggressive treatment with exchange transfusion and intensive phototherapy, the patient died of intractable seizure and acute renal failure on the fourth day of admission. Therefore, pediatricians should be aware of the clinical courses of hemolytic jaundice due to anti-Jkb, and they should be ready to treat this disease with active therapeutic interventions.

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Cyte Plus Kit (Median Diagnostic GmbH, Dudingen, Switzerland) (Table 1), which was also found in the eluate made from infant’s blood.

She was treated with exchange transfusion and intensive phototherapy for 3 days. A total bilirubin was decreased (16 mg/dL), but she had a convulsion with azotemia and electrolyte imbalance; BUN 41 mg/dL, creatinine 2.2 mg/dL, sodium 134 mEq/L, potassium 8.2 mEq/L, chloride 98 mEq/L. The emergent peritoneal dialysis was initiated under the diagnosis of acute renal failure. Despite the aggressive treat-
ment with peritoneal dialysis and anticonvulsant therapy, the patient died of intractable seizure and acute renal failure on the fourth day of admission. The results of tandem mass screening for metabolic disease revealed normal values. All the specimen cultures showed no growth.

Consequently, the HDN was very likely due to Jk(c−) incompatibility since the fetal blood type was AB, Rh-positive and Jk(c−) (+), while the mother was indicated as AB, Rh-positive and Jk(c−) (−).

**DISCUSSION**

The Kidd blood group is a major antigenic system in human erythrocytes, and this antigen system is defined by two antithetical specificities, Jka and Jkb, and a third rare recessive gene, Jk, that produces neither Jka nor Jkb antigens (7). The Kidd antigens are localized on a 43 kDa red blood cell integral membrane protein that functions as a urea transporter (8). JKA and JKB are alternate, autosomal inherited codominant alleles. The Kidd blood group is clinically significant since Jk antibodies can cause acute and delayed transfusion reactions as well as HDN.

Anti-Jkb was first described by Plaut et al. in 1953 (5). Most of the reports on anti-Jkb have stated that this condition followed after repeated blood transfusions (9, 10). The first case of anti-Jkb related HDN was reported by Kornstad and Halvorsen in 1958 (11). Up to the present time, only eleven cases of anti-Jkb related HDN have been reported in the medical literature. Although approximately 20-29% of the Caucasian and Asian population have the phenotype Jk(a+b−) (12-14), introduction of the Jkb antigen into such individuals is rarely associated with clinically manifesting disease. This is apparently due to the fact that Jkb is a poor antigen. Table 2 summarizes the pertinent clinical and serologic data concerning the cases of HDN due to anti-Jkb that have been reported to date (15-22). In all the reported cases, the disease was usually mild to moderate with a benign outcome. Therefore, HDN due to minor blood group incompatibility must be ruled out for all the cases of jaundice occurring 24 hr after birth. Screening and quantification of irregular antibodies are required for early diagnosis as a medical measure to prevent kernicterus through phototherapy and exchange transfusion, and close attention should be paid for possible delayed hemolytic anemia.

As a therapeutic measure, prenatal genotyping for Jk(b−) of the fetal amniotic cells should be done to identify those high risk pregnancies with HDN that are due to anti-Jkb incompatibility, and the genotyping can be done by allele-specific polymerase chain reaction. An allele-specific polymerase chain reaction (ASPCR) assay for prenatal genotyping of the Kidd antigen system in order to identify pregnancies at risk for HDN was developed. The availability of this assay, which can accurately genotype the Kidd blood group system, even in the presence of extensive maternal contamination, provides an important tool in managing pregnancies at risk for Kidd-related HDN (14). For the high risk fetus, intrauterine exchange transfusion (25), serial follow up of the serum bilirubin level and the use of erythropoietin for anemia (26) should be considered.

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