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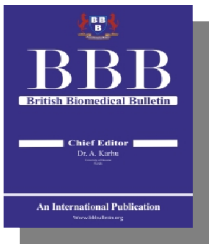

Case Report

Delayed Hemolytic Reaction Due to Anti-jka Antibodies in Thalassemic Child – A Case Report

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A R T I C L E I N F O

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A B S T R A C T

Kidd blood group, though minor, carries a special importance in transfusion medicine. Kidd antibodies are heterogeneous, as they have a weak sociological nature and goes to undetectable levels, so may miss routine serological tests. However, they can mount a secondary amnestic reaction leading to delayed hemolytic reaction, especially in frequently transfused individuals. We report a case of thalassemia child with delayed hemolytic reaction secondary to Kidd antibodies. **Conclusion:** screening for Alloantibodies is of prime importance, especially in frequently transfused individuals like thalassemic children.

Case Report

An Eleven year old male child presented to us with hepatosplenomegaly and hemolyticfacies with no past history of blood transfusion in the past. In view of no blood transfusion we suspected thalassemia intermedia and investigated for the same, and he was diagnosed as Thalassemia Intermedia. The child was transfused with packed cells in view of low haemoglobin (6gm %) and discharged. Transfusion was uneventful. After 15 days the child was taken to a private hospital in view of breathlessness, his haemoglobin was 3gm%, so he underwent one more transfusion. During transfusion the child had developed chills and fever with passing of dark colored urine, so was referred to us.

We suspected acute febrile hemolytic reaction and investigated. He had haemoglobin of 5.5gm%, with peripheral smear feature suggestive of haemolysis. His bilirubin was raised to 8gm%; creatinine was 1mg/dl. LDH was elevated. Direct and indirect Anti-globulin test were negative. The child had features of acute febrile hemolytic reaction. After 24 hours we planned for transfusion and transfused with fresh packed cells compatible with his blood group. Transfusion was uneventful. Next day child started passing dark colored urine. His haemoglobin dropped. We suspected delayed hemolytic reaction due minor blood group incompatibility and sent for investigation.

His serum was positive for JKa (ISBT-JK1) antibodies, but negative for JKb (ISBT-JK2) (Figure). We requested for Kidd antigen negative O Rh negative blood and transfused, the transfusion was uneventful.

This case clearly demonstrates the secondary immune response which had been provoked by the exposure to the Jka antigen by transfused Jka+ RBCs. The secondary, anamnestic response involves the recognition of the antigen by the persisting B lymphocyte memory pool, followed by proliferation, differentiation and maturation into plasma cells, which produce specific alloantibodies in sufficient quantity capable to cause the clinical manifestation of DHTR. In our case the whole process of anamnestic reaction was completed within 48h. Haemolysis of allogeneic Jka+ RBCs persisted in the next 24h, with the clinical symptoms and laboratory findings which confirmed clinical DHTR.

This case points to the need for meticulous screening of Alloantibodies in recurrently transfused individuals, especially thalassemic kids, as they are the most vulnerable candidates.

Discussion

In 1951, a patient called Mrs. Kidd was found to have produced antibodies targeted against a then unknown red cell antigen during her pregnancy. The marker was present on the RBCs of her fetus, and the maternal antibodies targeted against it caused fatal hemolytic disease in her newborn child. The protein was given the name Jka and was the first antigen to be discovered in the Kidd blood group system. Since this time, two other antigens, Jkb and Jk3, have been found.

In 1959, the first example of the null phenotype, i.e., Jk (a-b-), was found in a woman who had become jaundiced after a blood transfusion. Her serum was found to contain an antibody that recognized both Jka and Jkb. This antibody was subsequently named anti-Jk3.[1,2] The Kidd blood group system gene is located at chromosome 18q11-q12. The gene, SLC14A1, also known as JK or HUT11, is distributed over 30 kbp and contains 11 exons. It encodes for the urea transporter hUT-B1. The principal phenotypes of the Kidd blood group system and their frequencies. The antigens Jka and Jkb are found at relatively the same frequencies in the white populations, but

differ in other ethnic groups such as blacks and Asians. The Jk (a-b-) phenotype is rare and is found primarily in the Polynesian population. These null red cells have been demonstrated to be resistant to lysis by 2M urea; however, this phenotype is not associated with shortened red cell survival or clinical symptoms.[3]

The prevalence of JKa (ISBT-JK1) and JKb (ISBT-JK2) are 81.4 % and 67.6% respectively.[4]

The Kidd antibodies are very heterogeneous, clinically significant immune antibodies which are produced as a consequence of immunization by transfusion, pregnancy, rarely by transplantation or as a result of an autoimmune process. [5,6] Despite the fact that Kidd antigens are poor immunogens, Kidd antibodies have been frequently implicated in DHTRs but extremely rare are the cause of AHTRs. Following immunization, Kidd antibodies fall rapidly to undetectable levels in the plasma, therefore they are often difficult to detect. The Kidd antibodies are mainly of the class IgG, subclasses IgG1 and IgG3, capable to bind complement up to C3 stage. These antibodies have weak serological nature and they exhibit dosage phenomena.[7,8]

Although blood transfusion is a lifesaver for thalassemia patients, it may be associated with some complications such as iron overload, platelet and RBC alloimminizatio. [9,10] Repetitions of transfusions for the treatment of thalassemia major provokes the patient's immune system and produces anti-erythrocyte antibodies (alloantibodies and/or autoantibodies). Erythrocyte autoantibodies appear less frequently, but they can result in clinical haemolysis and in difficulty in cross-matching blood. Alloimmunization against red blood cell antigens increases the need for transfusion and can be significantly complicated transfusion therapy. Some alloantibodies are hemolytic and may cause hemolytic transfusion reactions and limit the availability of further safe transfusion. Others are clinically insignificant.[11,12]

The exact prevalence of alloimmunization against each minor blood group antigen in thalassemic children is not known, however, these children are at increased risk of developing alloantibodies.

Conclusion

It is known that the minor blood group is responsible for DHTR, routine screening of minor blood group antigens are not done. This case marks the importance of screening for alloantibodies, at least in recurrently transfused individuals.

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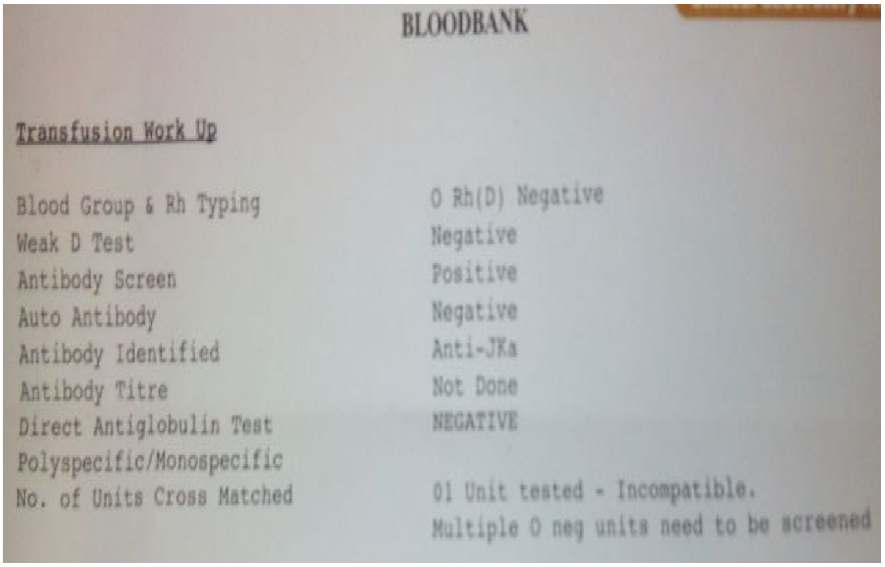
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Transfusion Work Up

Blood Group & Rh Typing	O Rh(D) Negative
Weak D Test	Negative
Antibody Screen	Positive
Auto Antibody	Negative
Antibody Identified	Anti-JKa
Antibody Titre	Not Done
Direct Antiglobulin Test	NEGATIVE
Polyspecific/Monospecific	
No. of Units Cross Matched	01 Unit tested - Incompatible. Multiple O neg units need to be screened

Figure 1. Antibodies positvie for JKa antigen, (JKa+, b-). (original)