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Case Report

P_1^K: A Rare Blood Group Phenotype

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Abstract

P blood group antigen of the GLOB system is a glycolipid structure, also known as globoside on the Red Blood Cells (RBCs) of almost all individuals worldwide. P1PK blood group system antigens include P_1, P and P^K antigens. Among these, P_1^K phenotype is very rare and the RBCS of these individuals express P1, P^K antigens. The high incidence antigen, P, is missing and anti-P antibody is present in the serum. Naturally occurring anti-P is present in the serum of individuals with the rare globoside-deficient phenotypes p, P_1^k, and P_2^k and has been implicated in hemolytic transfusion reactions, Donath-Landsteiner antibody as well as unfavorable outcomes of pregnancy. When an individual with P1K phenotype needs blood transfusion, they can receive only autologous blood or blood from another P1K phenotype or p phenotype (if P1K blood is not available). We report a case of a young boy with P1K phenotype, anti-P and px2 antibodies who developed a severe hemolytic transfusion reaction and were successfully treated conservatively.

INTRODUCTION

The P blood group system was introduced in 1927 by Landsteiner and Levine. The P system antigens are biochemically related to the carbohydrate chains that make up the ABH and I antigens. The P1PK blood group system (formerly known as P blood group system) consists of three antigens P1, PK and NOR whereas P antigen is the member of GLOB blood group system [1]. These antigens are responsible for five different phenotypes; P1, P2, p, P_1^K, P_2^K. The P antigen is present on Red Blood Cells (RBCs) of all individuals except ones with the rare phenotypes p, P_1^k, or P_2^k. Naturally occurring antibodies of IgM or IgG classes are formed against the missing antigen in the same way as they are formed in ABO blood group system [2]. P1k phenotype is very rare with a frequency of less than 1%. The red cells of patients with P1 k phenotype express P1 and Pk antigens and anti-P antibody is present in their serum. Anti-P is clinically significant and may cause acute intravascular hemolytic transfusion reactions [3] and recurrent spontaneous abortions due to damage to placenta [4,5]. Autoanti-P is also associated with the cold reacting IgG autoantibody in patients with paroxysmal cold hemoglobinuria [6]. The Pk and P1 antigens can act as membrane receptors for several pathogens and toxins. Pk and similar glycolipids are receptors for shiga toxins from Shigella dysenteriae and Enterohaemorrhagic E. Coli [7] and both the Streptococcus adhesin and the PA-IL lectin from Pseudomonas Aeruginosa also use Pk (Gb3) and P1 as receptors [8]. Another antigen which has been assigned to the GLOB collection is PX2 [9]. This high prevalence antigen is found in elevated amounts in individuals with p phenotype and has been reported to cause weak or variable cross match reactivity with P1k phenotype [10] (the exact clinical significance of anti-px2 antibodies is not known). In individuals with P1k phenotype and anti-P antibody routine pre-transfusion workup can be a bit challenging and complex. In reverse typing it shows strong hemolyzing reaction with A, B and O group control cells. Whereas during antibody identification, this antibody reacts with all panel cells through a wide thermal range except the auto control and p phenotype [11]. It also shows positive reaction with anti-H antibody. (Reactivity of this antibody can be greatly enhanced by testing with enzyme treated RBCs). Anti-P antibody reacts strongly by all techniques, including direct agglutination techniques at 18°C and 37°C, IAT pa pain. It is recommended that Patients with P1k phenotype should be transfused either with autologous blood or with Pk phenotype RBCs. If these are not available then p phenotype can be tried.

This is one of the rare blood groups and its identification is cumbersome. The case report encourages the experts of Transfusion medicine to take care and should remain vigilant during the identification of blood group. The clinicians should also have adequate knowledge about this blood group and the about the management of Anaphylaxis due to this blood group mismatch transfusion.

CASE REPORT

A 15 years old boy had undergone a road traffic accident leading to left leg fracture and severe blood loss. He was managed

surgically and was transfused with (O blood group) after blood grouping and cross matching at a local hospital. He had developed breathlessness, vomiting, fever and sweating within 20 minutes of commencement of transfusion. Transfusion was immediately stopped with the presumption of transfusion reaction. After 3 days he developed anuria and was referred to nephrology department for management of post transfusion acute renal failure. His CBC showed hemoglobin levels of 6.6 g/dl ,TLC was $10.5x10^9$/l and platelet count was $267x10^9$/l. Coagulation profile was done and showed prolonged PT- 20sec (Normal 10-13sec) and a PTT- 69sec (Normal 24-32sec). His renal function tests were deranged and showed BUN- 144 (Normal 8-22) and serum creatinine -12.4 (Normal 0.7-1.2). Urine complete examination showed the presence blood (+++). The patient required blood transfusion due to markedly low hemoglobin, so a request was sent to our blood bank for grouping and cross matching of the patient's sample. During forward grouping, it did not show any reaction to anti-A and anti-B antibodies just like a normal O blood group. However, during reverse grouping the patient's sample showed strong hemolyzing reaction with A, B and O group control cells. So, a discrepancy was noted between forward and reverse grouping. When cross-matching with the blood bag of group O was done, it showed incompatibility. Patient's close family members almost 15members were also tested but did not matched. Direct ant globulin test was also performed which showed a positive reaction. IAT also showed a positive reaction. Antibody identification showed the presence of a strong antibody which reacted with all panel cells through a wide thermal range with a negative auto control. Furthermore, his sample again showed a positive reaction when tested with anti-H antibody (thus excluding Bombay phenotype). In order to identify the antibody, the patient's sample was sent to a reference laboratory (Bristol Institute for transfusion sciences) for further workup. Their results showed that the patient had P1k phenotype with anti-P antibody reacting strongly by all techniques, including direct agglutination techniques at 18°C and 37°C, IAT papain. The patient's sample also showed weak reactivity with pp phenotype cells, so it was presumed that the patient had antipx2 antibodies in his serum as well. The patient underwent regular dialysis for renal failure after which his renal status improved considerably and became normal. As, the patient's red cells did not showed compatibility with any blood sample; the patient was managed conservatively. He was treated with Haematonics, Iron (300mg daily), Folate (1mg daily) and Vitamin B12 (1mg I/M daily for 1 week and then weekly for 2 months) and Inj. Erythropoietin (100 units/kg three times a week) and he responded well to therapy. The patient's hemoglobin improved on follow up visits and the medications were reduced over a period of two months. During a follow up period of two months and patient's condition was stable. Patient did not develop any other further complaints.

DISCUSSION

P1Pk and GLOB blood group system antigens include P, P1 and Pk^1. Naturally occurring antibodies of class IgG or IgM are present in the sera of individuals lacking the corresponding antigen [2]. P antigen is a very high frequency antigen and is present on the red cells of all individuals except the ones with the rare globoside deficient phenotypes p, P1k and P2k. Among these P1k phenotype expressing P1, Pk antigens and anti-P antibodies is very rare with a frequency of less than 1%. Anti-P is a clinically significant antibody and has been implicated in hemolytic transfusion reactions [3] as well as unfavorable outcomes of pregnancy [4,5].

Another antigen px2, belonging to the GLOB collection [9] has been found to be present in considerably enhanced quantities on the red cells of individuals with p phenotype [10]. The exact clinical significance of this antibody is not known.

Since the diagnostic workup of individuals with P1k phenotype is complex, these individuals can easily be misdiagnosed as O blood group on the basis of forward grouping alone and are at risk of developing fatal hemolytic transfusion reactions [3]. The case of this study developed symptoms and signs of transfusion reaction only a few minutes after starting blood transfusion. Therefore, it is very important to perform the Pretransfusion workup carefully and consider the possibility of a rare phenotype.

Experts recommend if transfusion is necessary and P^k phenotype RBCs are not available , p phenotype can be tried as significance of PX2 is unknown, if not available transfuse with P- positive washed RBCs (to remove complement) that is infused through an approved blood warmer.

Siblings of patients with anti-P should be tested for compatibility, and the patients urged to donate blood for cryogenic storage when his clinical state permits.

Our case report confirms the complexity and difficulty in the diagnosis of P1k phenotype and in the subsequent management of the patient after he had developed transfusion reaction. As compatible blood for this patient was not available, he was managed conservatively.

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