São Paulo Medical Journal
REVISTA PAULISTA DE MEDICINA

Case Report

Antonio Fabron Junior
Gilberto Moreira Junior
José Orlando Bordin

Delayed hemolytic transfusion reaction presenting as a painful crisis in a patient with sickle cell anemia

Universidade Federal de São Paulo/Escola Paulista de Medicina, São Paulo, Brazil

ABSTRACT

Context: Patients with sickle cell anemia (SCA) are frequently transfused with red blood cells (RBC). Recently, we reported that the calculated risk of RBC alloimmunization per transfused unit in Brazilian patients with SCA is 1.15%. We describe a delayed hemolytic transfusion reaction (DHTR) presenting as a painful crisis in a patient with SCA.

Case report: A 35-year-old Brazilian female with homozygous SCA was admitted for a program of partial exchange transfusion prior to cholecystectomy. Her blood group was O RhD positive and no atypical RBC alloantibody was detected using the indirect antiglobulin technique. Pre-transfusional hemoglobin (Hb) was 8.7 g/dL and isovolumic partial exchange transfusion was performed using 4 units of ABO compatible packed RBC. Five days after the last transfusion she developed generalized joint pain and fever of 39°C. Her Hb level dropped from 12.0 g/dL to 9.3 g/dL and the unconjugated bilirrubin level rose to 27 mmol/L. She was jaundiced and had hemoglobinuria. Hemoglobin electrophoresis showed 48.7% HbS, 46.6% HbA_1, 2.7% HbA_2, and 2.0% HbF. The patient's extended RBC phenotype was CDe, K-k+, Kp(a-b+), Fy(a-b-), M+N+s+, Le(a+b-), Di(a-). An RBC alloantibody with specificity to the Rh system (anti-c, titer 1:16.384) was identified by the indirect antiglobulin test. The Rh phenotype of the RBC used in the last packed RBC transfusion was CcDEe. The patient was discharged, asymptomatic, 7 days after admission.

Key Words: Sickle cell anemia. Transfusion reaction. Painful crisis.

INTRODUCTION

Red blood cell (RBC) transfusions are widely used in the management of patients with sickle cell anemia (SCA). We reported recently that the calculated risk of RBC alloimmunization per transfused unit in Brazilian patients with SCA is lower than that reported by studies from North America (1.15 vs. 3.10%), reflecting a more concordant RBC phenotype between blood donors and recipients in Brazil.[1,2] Therefore, we have not been routinely performing extended RBC phenotyping on all SCA patients prior to initiating a transfusion program. Nevertheless, in the present report we describe the occurrence of a delayed hemolytic transfusion reaction (DHTR) presenting as a painful crisis in a patient with SCA in Brazil.

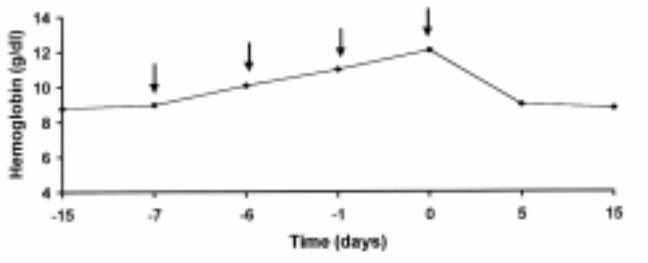

Figure 1 - Delayed hemolytic transfusion reaction presenting as a sickle cell crisis in a Brazilian patient with sickle cell anemia. The arrows indicate the packed RBC units transfused.

CASE REPORT

A 35-year-old Brazilian female with homozygous SCA was admitted for a program of partial exchange transfusion prior to cholecystectomy. Her blood group was O RhD positive and no atypical RBC alloantibody was detected using the indirect antiglobulin technique. Pre-transfusional hemoglobin (Hb) was 8.7 g/dL and isovolumic partial exchange transfusion was performed using 4 units of ABO compatible packed RBC. Figure 1 shows the serial Hb levels in relation to the blood transfusion program. Five days after the last transfusion she developed generalized joint pain and fever of 39°C. Her Hb level dropped from 12.0 g/dL to 9.3 g/dL and the unconjugated bilirrubin level rose to 27 mmol/L. She was jaundiced and had hemoglobinuria. Hemoglobin electrophoresis showed 48.7% HbS, 46.6% HbA_1, 2.7% HbA_2, and 2.0% HbF.

The patient's extended RBC phenotype was CDe, K-k+, Kp(a-b+), Fy(a-b-), M+N+s+, Le(a+b), Di(a-). An RBC alloantibody with specificity to the Rh system (anti-c, titer 1:16.384) was identified by the indirect antiglobulin test. The Rh phenotype of the RBC used in the last packed RBC transfusion was CcDEe. The patient was discharged, asymptomatic, 7 days after admission.

DISCUSSION

It has been reported that the prevalence of DHTR in patients with SCA ranges from 0.7 to 4%.[2,3] Moreover, there are few published reports of cases in which the development of DTHR presents as a painful crisis.[2,4] Although the RBC alloimmunization rate among multitransfused Brazilian patients with SCA is lower than previously reported rates in the United States,[1,2] the present case illustrates that DHTR occurs and can easily be mistaken for a painful sickle cell crisis, even in countries with low antigen mismatching between blood donors and SCA recipients. Interestingly, in contrast to the fact that about 40% of the potentially hemolytic RBC alloantibodies are usually not detected even after long-term follow-up, the presence of the above-mentioned RBC alloantibody (anti-c, titer 1:16) is still demonstrable in our patient's serum 5 years after her last blood transfusion. In conclusion, this case reveals some of the problems associated with the prevention and diagnosis of DHTR in patients with SCA transfused in countries where the rates of phenotypic incompatibility between donors and recipients are low.

REFERENCES

1. Moreira Jr G, Bordin JO, Kuroda A, Kerbauy J. Red blood cell alloimmunization in sickle cell disease: The influence of racial and antigenic pattern differences between donors and recipients in Brazil. Am J Hematol 1996;52:197-200.
2. Cox JV, Steane E, Cunningham G, Frenkel EP. Risk of alloimmunization and delayed hemolytic transfusion reactions in patients with sickle cell disease. Arch Intern Med 1988;148:2485-9.
3. Vichinsky EP, Earles A, Johnson RA, Hoag MS, Willians A, Lubin B. Alloimmunization in sickle cell anemia and transfusion of racially unmatched blood. N Engl J Med 1990;322:1617-21.
4. Cummins D, Webb G, Shah N, Davies SC. Delayed haemolytic transfusion reactions in patients with sickle cell disease. Postgrad Med 1991;67:689-91.

From Disciplina de Hematologia e Hemoterapia da Universidade Federal de São Paulo/EPM, São Paulo, Brazil

Authors

Antonio Fabron Junior - Docente da disciplina de Hematologia e Hemoterapia da faculdade de Medicina de Marília e pós-graduando da UNIFESP/EPM

Gilberto Moreira Junior - Médico da disciplina de Hematologia e Hemoterapia da Universidade Federal de São Paulo

José Orlando Bordin - Profesor livre-docente da disciplina de Hematologia e Hemoterapia da Universidade Federal de São Paulo

Sources of Funding: Not declared
Conflict of interest: Not declared
Last received: 28 September 1997
Accepted: 11 September 1998

Address for correspondence:
José Orlando Bordin
Disciplina de Hematologia e Hemoterapia
Universidade Federal de São Paulo/EPM
Rua Botucatu, 740
São Paulo/SP - Brasil - CEP 04023-092

RESUMO

Contexto: Pacientes com anemia falciforme recebem com freqüência concentrados de glóbulos vermelhos (CGV). Recentemente, nós relatamos que o risco de alo-imunização eritrocitária por unidade transfundida em pacientes brasileiros com anemia falciforme é 1,15%. **Relato de caso:** Nós descrevemos uma reação transfusional hemolítica tardia (RTHT) apresentando-se como uma crise dolorosa em um paciente com anemia falciforme.