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Hemolytic transfusion reaction due to an anti-H antibody in a sickle cell disease patient

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Background: Clinically significant alloanti-H can be produced only by H-deficient Bombay (Oh) phenotypes and H-partially deficient para-Bombay (Ah, Bh, ABh) phenotypes. Furthermore, autoanti-H/IH can be found in healthy people, most commonly in A1or A1B individuals who express very little H antigen. These antibodies are generally IgM type and are reactive at 4°C and rarely at room temperature (RT). They have been also reported in individuals with cold agglutinin syndrome. However, unusual cases of acute transfusion reactions due to wide thermal range anti-H/IH antibodies have been also described.

Aims: Here we report a case of a sickle cell disease (SCD) patient, blood group B, who presented a hemolytic episode after transfusion of antigen matched packed RBC (pRBC) of blood group O.

Methods: All serologic testing was performed on EDTA patient samples. Panel studies were performed in a gel-card format or by tube testing by a saline method at RT, 37°C, indirect antiglobulin test (IAT) using monospecific anti-IgG reagent and with enzyme-treated reagent cells. The patient's plasma was treated with dithiothreitol to determine the immunoglobulin class of the reacting agglutinins.

Results/Case report: The patients was a 17-year-old young woman with homozygous SCD, blood group B positive, C- E- c+ e+ K- k+ Fya- Fyb+ Jka+ Jkb- M+ N- S- s+ Lan+ Ata+ Jra+ P+, and known with the following alloantibodies: anti-C, anti-E and anti-Jkb from 2009, when she received two O negative pRBC in our institution. She was also transfused in 2013 with one O negative pRBC. She was admitted in May 2017 with painful crisis and severe anemia (hemoglobin (Hb) of 34 g/L) and received 2 antigen-matched (for all clinically significant antigens) and cross-matched O negative pRBC with no adverse events; Hb raised to 68 g/L. Ten days later, she was readmitted with painful crisis and anemia with Hb of 46 g/L. The direct antiglobulin test was negative and the plasma showed a panreactivity at room temperature and at 37°C, in IAT and with enzyme treated test cells with all test cells, except the autologous control. During the following hours the Hb dropped to 35 g/L and one antigen-matched O negative pRBC was transfused in emergency. The transfusion was inefficient as the patient continued to deteriorate and the Hb decreased till 29 g/L. In the meantime, the diagnostic of strong large thermal amplitude anti-H antibodies of IgM type reacting at 37°C in IAT was established. Afterwards, the patient was efficiently transfused with two thawed antigen-matched and cross-matched B negative pRBC and Hb raised to 58 g/L.

Summary/conclusions: This is a rare case of anti-H autoantibodies behaving as alloantibodies and causing severe hemolysis. As the hemolysis appeared 12 days after the last transfusion episode with O negative pRBCs and was accelerated by the latest transfusion of one more O negative pRBC, we could here also stipulate a delayed hemolytic transfusion reaction, a relatively common complication in SCD patients. This case, as the other 5 similar cases with anti-IH from the literature, confirm that ABO-identical matching together with the phenotypical matching should be considered in SCD settings.