POST-OPERATIVE, POST-TRANSFUSION
PAINFUL CRISIS IN A PATIENT WITH SICKLE CELL DISEASE: ANSWERS

Case study by Jim Perkins (©2009)

1. What antibody(ies) is(are) present in the patient's post-transfusion serum? What is present in the eluate? Is any further serologic workup needed to prove your answer?

   The serum appears to have anti-C plus anti-E. The eluate only has anti-C. Anti-S is not ruled out in either the serum or the eluate.

2. What immunohematologic problem is occurring based on this sequence of events and the patient's laboratory data? What additional laboratory tests would help make this diagnosis?

   This patient appears to be having a delayed hemolytic transfusion reaction (DHTR) due to anti-C and anti-E. The anti-C typing on the post-transfusion sample reveals that some C-positive RBCs of donor origin are still circulating, but E-positive cells are not detected. The fact that anti-C is present in the eluate, but anti-E is not, suggests the same conclusion. In order to make the diagnosis of a DHTR, one would like to demonstrate that the patient indeed received RBCs bearing the C and E antigens. One might also want to repeat the pre-transfusion antibody detection test to rule out a serologic error.

   The diagnosis of any immune hemolytic process requires demonstration of both immunity (the blood group antibodies discussed above) and hemolysis. Proving the latter criterion is difficult in patients with SCD, since SCD itself is a hemolytic anemia. For example, when a SCD patient has a warm autoantibody it may be difficult to determine if the antibody is contributing to the patient's anemia. In the current case the fall in hematocrit within 10 days after the first transfusion demonstrates hemolysis. Note that the hematocrit fell below the patient's pre-exchange baseline since a significant portion of his own cells had been removed. DHTRs after exchange transfusion can result in severe anemia.

3. Is there any connection between this patient's reaction to transfusion and his painful sickle cell crisis? Do you think that this exchange transfusion was indicated? Assuming that the transfusion was appropriate, how might this transfusion reaction have been avoided?

   It is a general observation that occurrence of a DHTR in a patient with SCD may be an antecedent to painful sickle cell crisis. Moreover, blood group alloimmunization is common in multiply transfused SCD patients, and DHTRs are correspondingly common. At the time of this case it was standard therapy in the United States to perform exchange transfusion prior to operations such as cholecystectomy in an effort to prevent sickle cell crises that caused by operative complications such as transient hypoxemia. However, a randomized study demonstrated that prophylactic pre-operative exchange transfusion was not superior to simple transfusion to achieve a hgb level of 10gm/dL (Vichinsky et al, New Engl J Med, 1995) in preventing acute chest syndrome (10% of each group) or painful crisis (5-7%). Moreover, new blood group antibodies were twice as common in the exchange group, and DHTRs were 5 times as common after exchange. Since publication of this article many physicians have reserved prophylactic exchange transfusion for specific operations such as retinal surgery.

   Since formation of unexpected blood group alloantibodies is caused by exposure to RBCs expressing "not-self" antigens, transfusion of RBCs selected to LACK blood group antigens that are NOT present on the recipient's RBCs has the potential to reduce both primary and secondary alloimmunization. This strategy will be most effective for the most immunogenic blood groups. For these reasons many hospitals have elected to provide RBCs that lack Rh and Kell system antigens that the recipient also lacks (Vichinsky et al Transfusion, 2001). In this case that policy would have meant transfusing the patient with C-neg, E-neg, and K-neg RBCs. Such units would have been readily identified by among the Rh negative RBCs in stock.