Ask a Pathologist: Hyperhemolysis

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ASK A PATHOLOGIST

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Question: How can hyperhemolysis syndrome be distinguished from a delayed hemolytic transfusion reaction in a patient with sickle cell disease?

Answer: Hyperhemolysis syndrome (HS) is an uncommon but potentially life-threatening complication of blood transfusion that can occur in patients with sickle cell disease (SCD). Similar to a classic delayed hemolytic transfusion reaction (DHTR), HS typically manifests approximately 7 days after an index transfusion. Unlike DHTR, HS is generally associated with significant hemolysis of both transfused and patient cells, resulting in a post-transfusion hemoglobin that is significantly lower than the pre-transfusion hemoglobin. The most common presenting symptoms of HS are fever and pain, which may overlap with the symptoms of both vaso-occlusive pain crisis and delayed hemolytic transfusion reaction. An accurate diagnosis is important, however, since additional blood transfusion may exacerbate hemolysis in HS.

Whenever HS is suspected, it is important to order a hemoglobin level, direct antiglobulin test (DAT), antibody screen, reticulocyte count, serum bilirubin, LDH, and hemoglobin electrophoresis. The DAT is typically negative in HS, with no new alloantibodies detected on antibody screen. Hemolysis labs are positive and a relative reticulocytopenia is often seen. In contrast, DHTRs will classically be associated with the identification of a new red blood cell alloantibody on the antibody screen, a positive DAT, positive hemolysis labs, and a drop in hemoglobin consistent with hemolysis of only the recently transfused cells and not of patient cells.

Transfusion is to be avoided in the management of HS, as it may worsen the degree of hemolysis and increase the likelihood of mortality. Instead, corticosteroids are often utilized. If the anemia becomes severe enough to warrant transfusion with RBCs, then co-administration of IVIG, erythropoietin, and folic acid has been effective in some cases. Conversely, transfusion does not need to be avoided after a delayed hemolytic transfusion reaction if appropriate red blood cell units that are compatible with the patient's newly identified alloantibody can be acquired.

References:

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