Hemepath Case 11: 5-Year-Old Boy

HISTORY

A 5-year-old boy presents with dark urine after walking home from kindergarten with his mother on a cold, snowy day. The child experienced diarrhea and vomiting 3 weeks ago, and was diagnosed with a GI viral infection. Since then, he has complained of feeling “full all the time”, and experiences occasional abdominal discomfort. His mother also notices that he rarely plays with his siblings, but instead, takes naps on the couch. The child has always been very energetic in the past.

Physical exam reveals a pale-looking boy with an enlarged spleen palpable 3-4 cm below the left costal margin.

CBC

Hgb (g/L)	Low
MCV	N
Reticulocyte Count	High
WBC	N
Plt	N

OTHER LABORATORY FINDINGS

Urine dipstick for blood	(+)
LDH	High
Direct Coombs Test	(+) complement only
Biphasic Donath-Landsteiner Test	(+)

DESCRIPTION OF SLIDE

Peripheral Blood Smear

Anemia with marked spherocytosis (see circles) and polychromasia are seen on the peripheral smear. Leukocytes and platelets are unremarkable.

*** To see the slide annotations in Imagescope, click on VIEW, then ANNOTATIONS, and then on the "eye" icon adjacent to the word "Layers". In the “Layer Attributes” box, a brief description of the annotations is provided. You may also click on individual layer region (e.g. region 1) in the “Layer Regions” box to locate each annotation – this is especially helpful in identifying annotations when the slide is not zoomed in. ***

MORPHOLOGICAL DIAGNOSIS

Autoimmune hemolytic anemia (AIHA) - paroxysmal cold hemoglobinuria

DISCUSSION

Paroxysmal cold hemoglobinuria, also known as Donath-Landsteiner hemolytic anemia, is a rare, self-limited form of cold autoimmune hemolytic anemia (AIHA) usually affecting children younger than 5 years of age. It is typically preceded by a viral infection, at which time autoantibodies (Donath-Landsteiner IgG antibodies) are formed against a viral antigen. These IgG antibodies cross-react and bind to RBCs (frequently to the P antigen) at cold temperatures, e.g. within skin capillaries. As the RBCs return to warmer temperatures, the IgGs fix complements to the RBCs and then dissociate. Direct Coombs assays usually only show complements on the RBCs, as the IgG antibodies have dissociated. The "bind in cold, fix complement in warm" nature of the antibodies gives rise to their "biphasic" or "bithermic" designation.

This condition is usually self-limited. It is much more common in children than adults, and it is probably underdiagnosed.