blood work
images in hematology

Oxidative hemolysis due to Wilson disease

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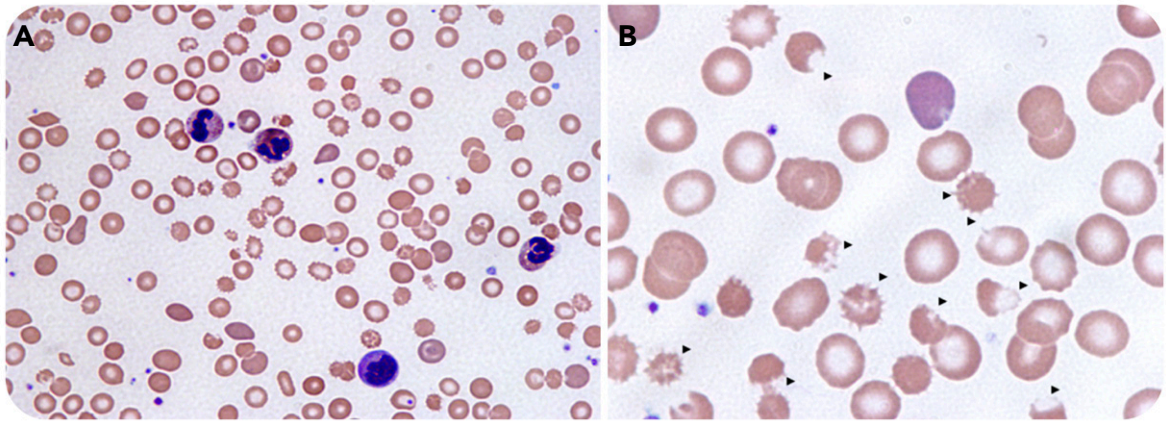

A 9-year-old boy with presumed attention-deficit hyperactivity disorder presented with jaundice/fatigue/headache/elevated creatinine/loose stools. He was investigated for acute hemolytic anemia (HA) including hemolytic uremic syndrome. His complete blood count showed: white blood cells, 16.7 $\times$ 10^9/L; hemoglobin, 69 g/L; reticulocytes, 214 $\times$ 10^9/L; platelets, 168 $\times$ 10^9/L (normal, 180 $\times$ 10^9/L to 440 $\times$ 10^9/L); bilirubin, 471 μM/L; lactate dehydrogenase, 896 U/L; haptoglobin, <0.07 g/L; plasma-free hemoglobin, 1.28 g/L (normal, 0.01-0.05 g/L). The blood smear (BS) demonstrated marked echinospherocytosis/target cells/polychromasia (panel A; Wright stain, original magnification ×40)/bite/blister cells (panel B; Wright stain, original magnification ×100) with no schistocytosis, consistent with oxidative hemolysis. Direct antiglobulin test/hepatitis serology/glucose-6-phosphate dehydrogenase/molecular tests for enteric pathogens including *Escherichia coli* O157 were negative. The patient developed fulminant hepatic failure (FHF) with: aspartate aminotransferase, 256 U/L; alkaline phosphatase, 45 U/L; γ-glutamyl transferase, 156 U/L; albumin, 21 g/L; international normalized ratio, 3.1; partial thromboplastin time, 53 seconds; and fibrinogen, 1.1 g/L. Serum ceruloplasmin was decreased 6.9 mg/dL (normal, 24-50 mg/dL); serum copper was elevated 48.4 μM/L (13.2-21.4 μM/L). A clinical diagnosis of Wilson disease with FHF was made based on the BS/laboratory tests/clinical presentations. The patient was transferred for urgent liver transplantation.

HA was the presenting manifestation in ~5% to 6% of patients with Wilson disease, especially in children/young adults with FHF. It is believed that HA is related to the release of copper into the blood from hepatocytes necrosis, causing oxidative damage to the erythrocytes.

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