blood work
images in hematology

Pancreatitis-associated thrombotic microangiopathic hematolytic anemia with thrombocytopenia in a child

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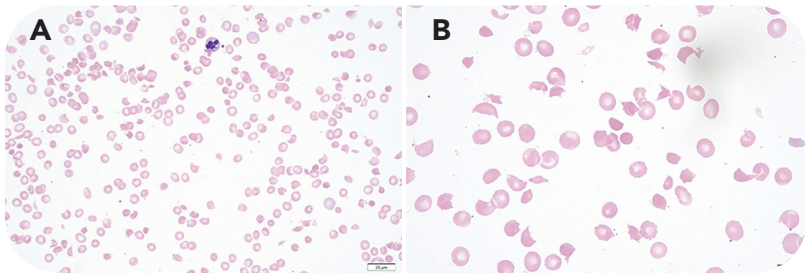

A 4-year-old boy was admitted with recurrent pancreatitis (peak lipase 7315 units/L). On day 3 of hospitalization, he developed altered mental status and laboratory evidence of acute renal injury. Blood count showed an acute decrease in hemoglobin (from 12.7 to 4.0 g/dL) and platelet count (from 213 to 16 × 10^9/L). Lactate dehydrogenase was elevated at 2830 units/L, with direct hyperbilirubinemia (6.4/3.5 mg/dL), and undetectable haptoglobin. A direct Coombs test was negative. Review of peripheral smear showed anisopoikilocytosis (panel A: Wright-Giemsa stain, 50× lens objective) with a predominant morphology of schistocytes, helmet cells, and red blood cell fragments (panel B: Wright-Giemsa stain, 100× lens objective). Given concern for thrombotic thrombocytopenic purpura, daily plasmapheresis, and methylprednisolone were initiated. Stool culture was negative for *Escherichia coli* and *Shigella*. ADAMTS13 activity collected before plasmapheresis was reported to be 91%, following which plasmapheresis (after 5 cycles) and methylprednisolone were discontinued. The patient improved clinically, with complete normalization of hemolysis markers and renal function. Both functional and genetic testing for complement-mediated hemolytic uremic syndrome were negative. Testing for genetic predisposition to pancreatitis identified multiple heterozygous variants in the *CFTR*, *CASR*, and *SLC26A9* genes. A diagnosis of pancreatitis-associated microangiopathic hematolytic anemia with thrombocytopenia (MAHA-T) was made.

MAHA-T, an increasingly recognized complication of pancreatitis, is thought to result from cytokine-mediated endothelial damage. Most reported cases are diagnosed within 3 to 4 days of hospitalization with pancreatitis, have normal ADAMTS13 levels, and undergo plasmapheresis, though the efficacy of plasmapheresis in this cohort remains unclear.

https://doi.org/10.1182/blood.2024026629