1-4-2018

Cabot rings and marked anisopoikilocytosis in Imerslund-Gräsbeck syndrome.

Derrick L. Goubeaux
Children's Mercy Hospital

Weijie Li
Children's Mercy Hospital

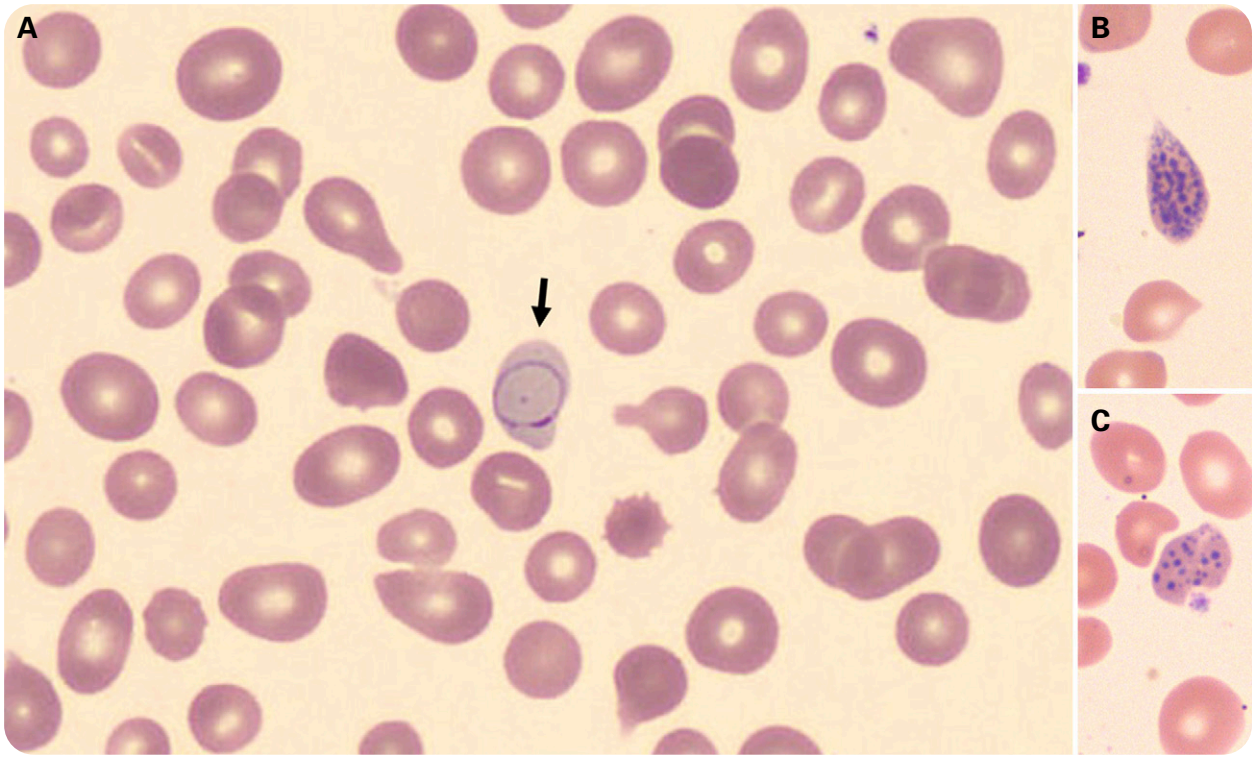
Recommended Citation

Goubeaux DL, Li W. Cabot rings and marked anisopoikilocytosis in Imerslund-Gräsbeck syndrome. Blood. 2018;131(1):153. doi:10.1182/blood-2017-10-809178

blood® work
images in hematology

Cabot rings and marked anisopoikilocytosis in Imerslund-Gräsbeck syndrome

Derrick L. Goubeaux and Weijie Li, Children's Mercy Hospital



A 12-year-old female presented with fatigue and weight loss. She appeared pale and had gray streaks in her hair. She had a history of chronic anemia, and some family members also had anemia. Laboratory tests showed macrocytic anemia (hemoglobin 5.2 g/dL, mean corpuscular volume 107.3 fL) and thrombocytopenia (platelet count 129 × 10^9/L), increased blood homocysteine (111.6 μmol/L, reference range 4.7-10.3), and decreased vitamin B_{12} (111 pg/mL, reference range 260-935). Wright-stained peripheral blood smear showed macrocytosis, anisopoikilocytosis, a few polychromatophilic red blood cells with rare purple Cabot rings (panel A, arrow; original magnification ×1000), and coarse basophilic stippling (panels B and C; original magnification ×1000). Workup for vitamin B_{12} deficiency identified a homozygous pathogenic variant in the *AMN* gene, which is consistent with Imerslund-Gräsbeck syndrome (IGS), a rare autosomal recessive disorder caused by a defect in the receptor of vitamin B_{12}–intrinsic factor complex in terminal ileum. This receptor is composed of 2 proteins, amnionless (*AMN*) and cubilin (*CUBN*). Mutations of either the *AMN* or *CUBN* gene can cause IGS.

Cabot rings are thin, thread-like ring- or "figure eight"-shaped red blood cell inclusions, likely remnants from mitotic spindles. They are rarely seen in peripheral blood, and their presence indicates a defect in erythrocyte production, especially in pernicious anemia and lead poisoning.

ASH® Image Bank

For additional images, visit the ASH Image Bank, a reference and teaching tool that is continually updated with new atlas and case study images. For more information, visit http://imagebank.hematology.org.

DOI 10.1182/blood-2017-10-809178

2018 131: 153
doi:10.1182/blood-2017-10-809178

Cabot rings and marked anisopoikilocytosis in Imerslund-Gräsbeck syndrome

Derrick L. Goubeaux and Weijie Li

Blood (print ISSN 0006-4971, online ISSN 1528-0020), is published weekly by the American Society of Hematology, 2021 L St, NW, Suite 900, Washington DC 20036.