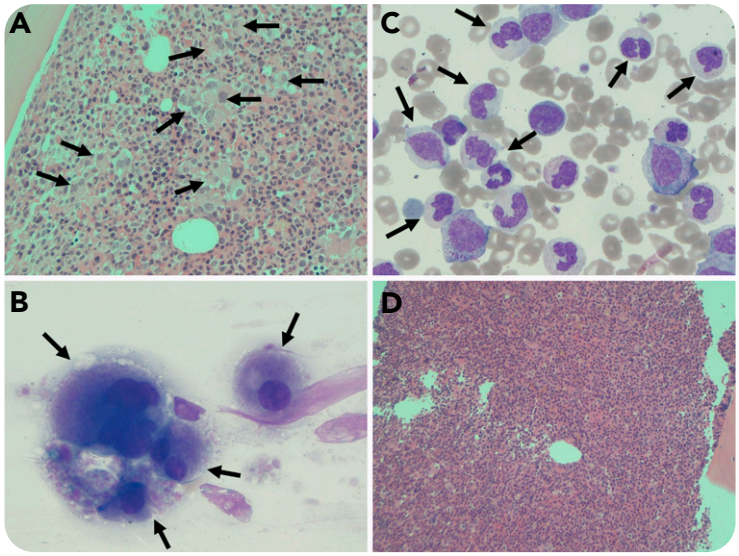
blood work
images in hematology

Leukostasis as a result of rapidly progressive granulocytosis in a patient diagnosed with MDS/MPN-U

Vitaliy Mykytiv and Khalil Alnajjar, Cork University Hospital



A 75-year-old man diagnosed with JAK2$^+$ myelodysplastic/myeloproliferative neoplasm, unclassifiable (MDS/MPN-U), presented with extreme thrombocytosis of 2118 × 10^9/L. Myeloid hyperplasia and increased and frequently clustered hypolobulated megakaryocytes were observed on bone marrow trephine (panel A arrows; hematoxylin and eosin [H&E] stain; original magnification ×20) and aspirate (panel B arrows; Wright-Giemsa stain; original magnification ×40). Thrombocytosis was controlled with hydroxyurea and, for the last 3 months, with ruxolitinib. Two years after diagnosis, he developed rapidly progressive neutrophilia. His leukocyte count increased from 36.8 × 10^9/L to 260.1 × 10^9/L in 19 days; hemoglobin was 8.2 g/dL, and platelet count was 251 × 10^9/L. The peripheral blood film showed increased granulocytes, frequently dysplastic (panel C arrows; Wright-Giemsa stain; original magnification ×60), without increased monocytes or blasts. Bone marrow trephine showed marked myeloid hyperplasia (panel D; H&E stain; original magnification ×10), without fibrosis or increased blasts. His karyotype was 46XY. Next-generation sequencing detected the following mutations: JAK2 p.Val617Phe, ASXL1 p.Glu635ArgfsTer15, TET2 p.Pro656ThrfsTer25, and EZH2 p.Cys548Arg, with respective allele burdens of 36%, 50%, 47%, and 98%. He subsequently developed multiorgan failure as a result of leukostasis. Hydroxyurea was restarted, and leukapheresis was attempted, but we were unable to control progressive leukocytosis. The patient died within 5 days.

Dysplasia is a distinguishing feature of MDS/MPN-U, usually absent in myeloproliferative disorders; however, rapidly progressive granulocytosis and leukostasis are extremely rare.

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.2019002559