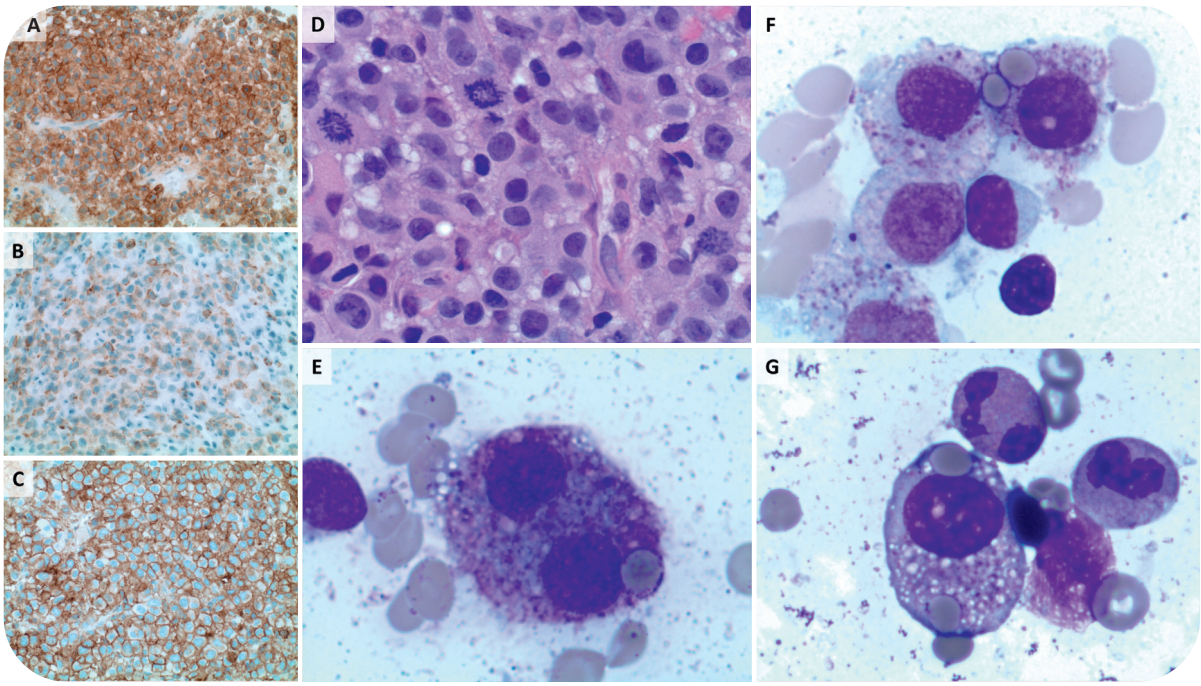
blood work
images in hematology

Aggressive mast cell leukemia/sarcoma with CD4 expression and hemophagocytosis

Anna L. Heimes Dillon and Sarah M. Kelting, University of Kansas Medical Center



A 63-year-old woman presented with a destructive lesion of L2 with soft tissue extension. Biopsy revealed sheets of cells with abundant eosinophilic cytoplasm, moderate nuclear pleomorphism, and frequent mitoses (panel D, 50× objective, ×500 magnification). Malignant cells were positive for CD45, CD117, tryptase, CD4, and CD30 and negative for CD2, CD25, CD34, and myeloperoxidase by immunohistochemistry, consistent with mast cell sarcoma (MCS) (panels A [CD117], B [tryptase], and C [CD4]; 20× objective, ×200 magnification). Serum tryptase was elevated (132 ng/mL). Iliac crest bone marrow aspiration showed mast cell leukemia (MCL) with 41% malignant mast cells; morphologic features included abnormal granulation, binucleation, vacuolization, and frequent hemophagocytosis (panels E-G, 100× objective, ×1000 magnification). Features of an associated hematologic neoplasm were absent. A *KIT* mutation was not detected by next-generation sequencing on the aspirate; conventional cytogenetics was normal. Due to poor functional status, the patient received single-agent midostaurin but died 2 months after diagnosis.

MCL is an aggressive form of systemic mastocytosis defined by ≥20% immature mast cells in the bone marrow, which occasionally represents the disseminated phase of MCS. It frequently lacks the *KIT* D816V mutation but may harbor other *KIT* mutations. The typical immunophenotype includes CD117, tryptase, and frequent aberrant expression of CD2, CD25, and CD30; CD4 expression is uncommon. This case highlights a rare MCL/MCS with unusual morphologic and immunophenotypic findings.

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.

https://doi.org/10.1182/blood.2022018256