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IMAGES IN HEMATOLOGY

Hemophagocytosis as the initial presentation in a patient with myelodysplastic syndrome and coexisting NK/T cell lymphoma involving bone marrow

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A 60-year-old man with uremia undergoing regular hemodialysis was admitted due to general weakness and dizziness persisting for 1 month. He denied any history of smoking, alcoholism and exposure to chemical or radiation. Initial physical examination noted marked pallor of the conjunctiva and skin, distended but nontender abdomen with splenomegaly (10 cm below the left costal margin) and grade 2 pitting edema in lower extremities. No peripheral lymphadenopathy was found. He presented with the following on workup: complete blood count (CBC) including hemoglobin, 6.3 g/dL; white blood cell count (WBC), 21.77 $\times$ 10^9/L (1 % blast, 4 % band form, 38 % segmented neutrophils, 19 % lymphocytes, 32 % monocytes) and platelets, 29 $\times$ 10^9/L. Biochemical data were all within normal limits, apart from ferritin, 865 ng/mL (normal range 38–280 ng/mL); sodium, 128 mEq/L; blood urea nitrogen, 148 mg/dL; creatinine, 11.26 mg/dL; albumin, 3.3 g/dL; prothrombin time, 12.3 s; activated partial thromboplastin time, 38.4 s, and prothrombin time of international normalized ratio (INR) 1.19. Lactate dehydrogenase level was 176 U/L (normal range 131–250 U/L). Computed tomography detected multiple enlarged lymph nodes in gastrohepatic ligament, celiac trunk root, mesenteric root, aortocaval and left para-aortic retroperitoneal regions, as well as splenomegaly. Bone marrow (BM) aspirate showed a hypercellular marrow with increased tissue histiocytes and evident hemophagocytic activity (Fig. 1). Hematological malignancy such as leukemia or lymphoma was first considered. Marked dysplastic features involving megakaryocytes, myeloid (Fig. 2, arrow) and erythroid (Fig. 3, arrow) series and left shift maturation with 11 % myeloblast, 9 % promyelocytes were noted in BM aspirate. Excess blasts in BM aspirate were also noted (Figs. 2, 3, arrowhead). In addition, aggregates of immature lymphoid cells, about 19 %, coexisted with the dysplastic marrow. Immunocytochemical stain confirmed concomitant presence of myeloperoxidase-positive myeloblasts and acid-phosphatase-positive T-lymphoblasts. Iron staining showed no increased iron storage or ringed sideroblasts. Cytogenetic analysis of the bone marrow revealed monosomy 7, a common myelodysplastic syndrome (MDS)-associated cytogenetic abnormally indicating poor prognosis. Flow cytometry immunophenotype analysis of BM revealed two abnormal clones of cells: myeloid precursor cells resembling myeloblasts were positive staining for human leukocyte antigen (HLA)-DR (97 %), CD34 (35 %), CD13 (94 %) and CD33 (96 %); abnormal lymphoid populations showed positive for CD2 (80 %) and CD7(83 %). BM biopsy showed vaguely nodular infiltration of atypical medium to large lymphoid cells in the marrow by hematoxylin and eosin (HE) stain (Fig. 4), excess of myeloblasts (10 %) with positive staining in CD34 (Fig. 5) and myeloperoxidase. In addition, BM biopsy revealed focally nodular infiltration of atypical medium to large lymphoid cells with

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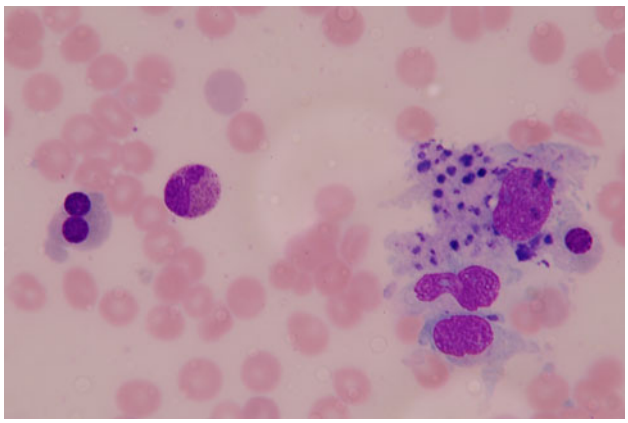

Fig. 1 Hemophagocytosis and dyserythropoiesis in bone marrow

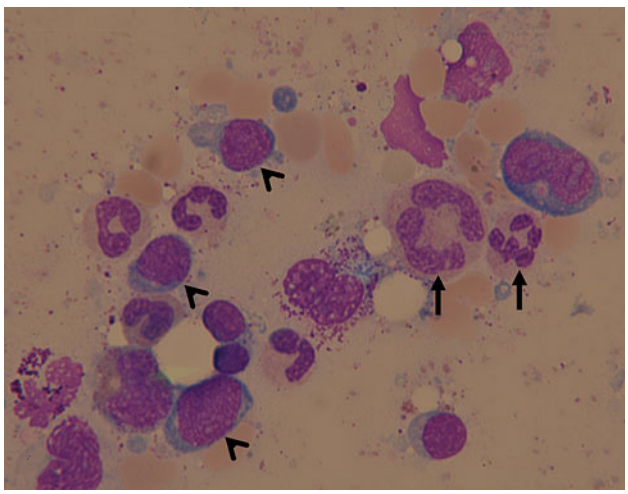

Fig. 2 Myloid dysplasia of giant band and hypersegmentation with excess blasts

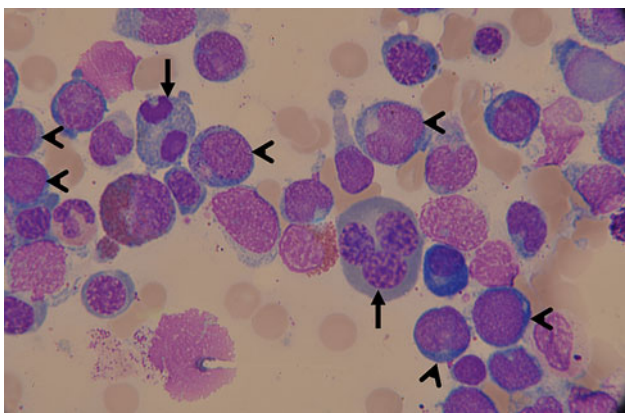

Fig. 3 Dyserythropoiesis with excess myeloid and lymphoid blasts

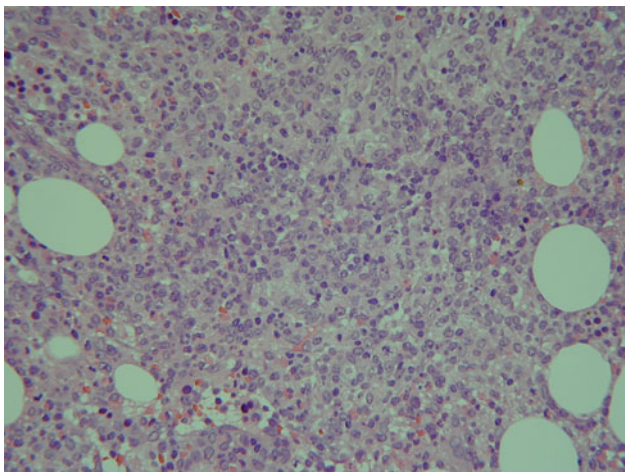

Fig. 4 Nodular infiltration of atypical large lymphoid cell

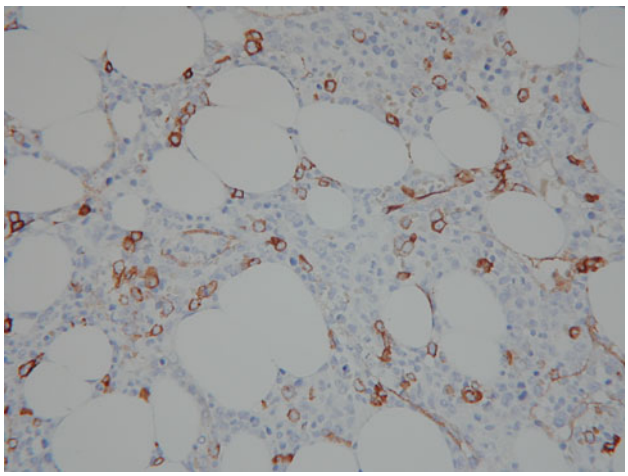

Fig. 5 CD34 stain in bone marrow

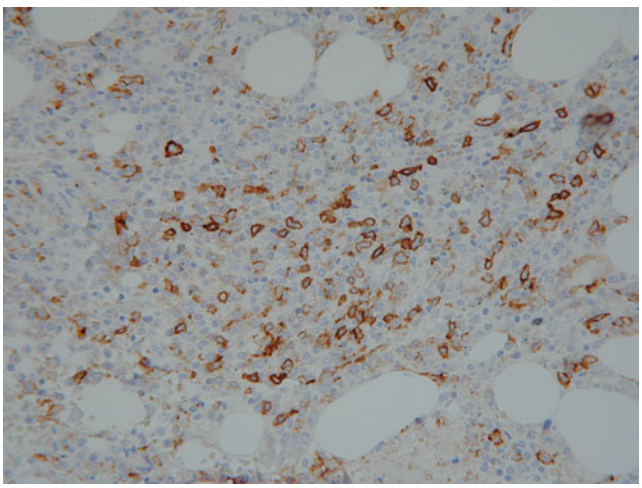

Fig. 6 CD56 stain in bone marrow

immunoreactivity for CD7 and CD56 (Fig. 6) and focally positive stain for CD2 and CD3. Concurrent existence of de novo MDS-RAEB II and NK/T cell lymphoma in BM was finally diagnosed. This is a rare presentation in bone marrow. Though the frequency of HPS in adults with hematological malignancy is typically associated with lymphomas or leukemias of the T or NK cell lineages [1, 2], it is worth to analyze the activated macrophage derived by NK/T cell lymphoma or MDS by FISH analysis.

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Conflict of interest The authors declare no conflict of interest.

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