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CONTINUING MEDICAL EDUCATION
ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Hematology Quiz – Case 70

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2025, 42(3):428–430

S. Papadakis,
I. Liapis,
C. Misidou,
L. Inglezou,
B. Malkoc,
G. Vrachiolias,
I. Stamatiou,
E. Panagiotopoulos,
S. Karavasilis,
P. Chatziioannou,
F. Anastasopoulos,
E. Charitaki,
T. Bounta,
P. Kolovos,
C. Roubakis,
A. Pentidou,
M. Papoutselis,
Z. Bezirgiannidou,
E. Spanoudakis,
I. Kotsianidis,
K. Liapis

Department of Hematology,
University Hospital of Alexandroupolis,
Democritus University of Thrace,
Alexandroupolis, Greece

A 68-year-old man with no significant past medical history presented to the emergency department with abdominal distension and discomfort of 4 weeks' duration. Abdominal distention was associated with early satiety, fatigue, and exertional dyspnea. There was no history of fever, drenching night sweats or pruritus. He smoked 20 cigarettes daily, drank alcohol rarely, and took no medications. He was thin and looked ill. Physical examination showed distension of the abdomen with shifting dullness, and enlargement of liver and spleen. Laboratory test results showed a hemoglobin level of 11.9 g/dL, hematocrit 36.6%, white blood cell count 4.0×10^9/L (neutrophils: 1.2×10^9/L; lymphocytes: 0.8×10^9/L; monocytes: 1.7×10^9/L), platelets 156×10^9/L, lactate dehydrogenase (LDH) 3,015 U/L (normal: 125–220), creatinine 1.4 mg/dL, urea 59 mg/dL, and normal liver-function tests. A computed tomography (CT) scan of the abdomen showed massive ascites, an enlarged spleen measuring 16.0 cm, and mesenteric, celiac and hepatic hilar lymphadenopathy. A representative image from the peripheral-blood smear is shown in figure 1, whereas representative images from the bone marrow are shown in figures 2–6. Paracentesis revealed increased leukocytes in the ascitic fluid (1,250/mm^3), with a serum-ascites albumin gradient (SAAG) of 0.9.

Comment

The presence of ascites without other symptoms or a history of liver disease raises should obtain concern for possible underlying malignancy. The absence of constitutional symptoms does not rule out this possibility. The blood film shows a circulating nucleated red cell and an immature monocyte. Although there is no evidence of severe anemia or thrombocytopenia, the presence of nucleated red

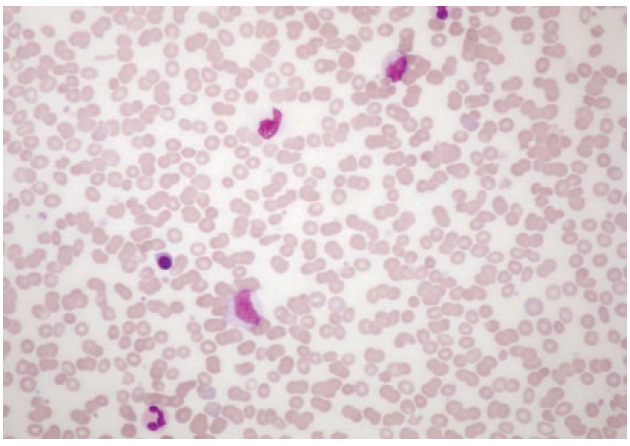

Figure 1.

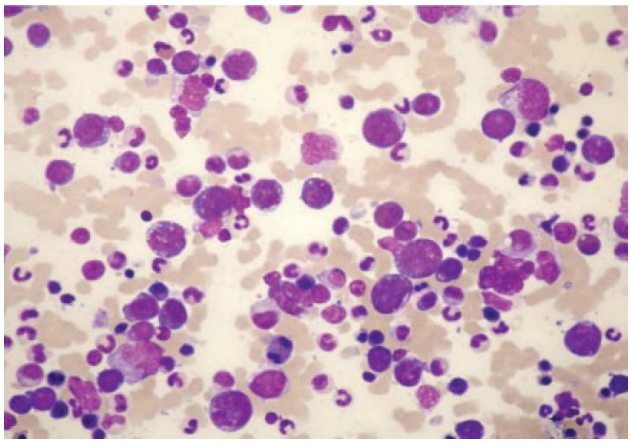

Figure 2.

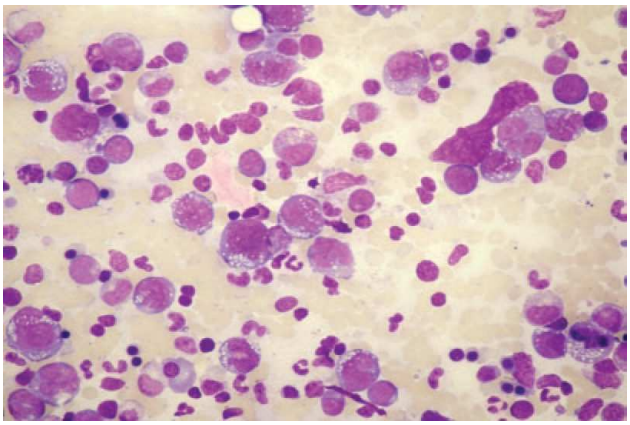

Figure 3

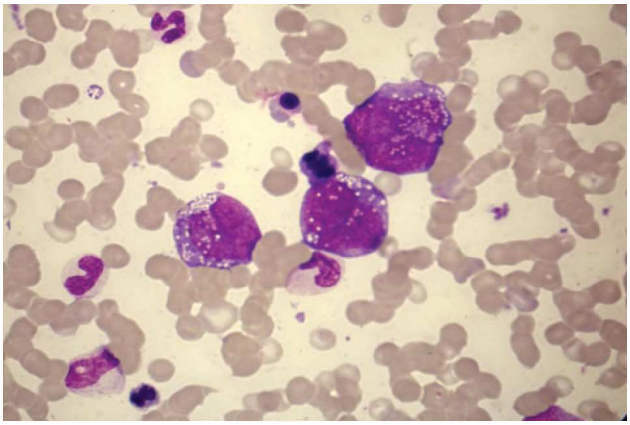

Figure 4

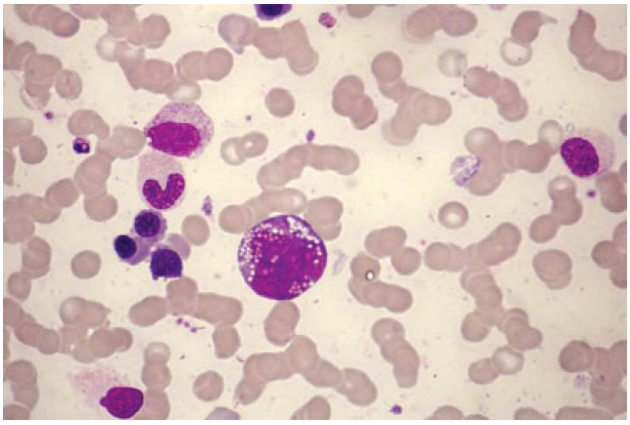

Figure 5

cells in the peripheral blood and the borderline neutropenia are signs of probable bone marrow infiltration. Because of the circulating nucleated red cells and monocytosis, bone marrow aspiration was performed to rule out bone marrow infiltration with foreign cells or a primary myeloid neoplasm.

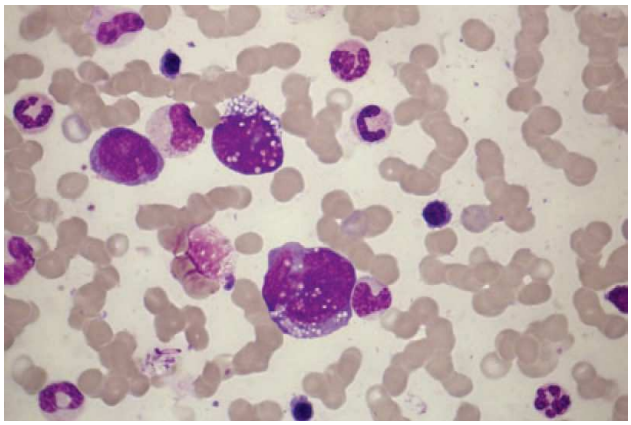

Figure 6

The bone marrow aspirate showed a uniform population of large cells with immature chromatin, nuclear folding, one or more nucleoli, and a moderate amount of basophilic or blue-grey vacuolated cytoplasm without cytoplasmic granules (nuclear:cytoplasmic ratio: 1.5:1) constituting 18% of total marrow cells. An increase in the myeloblast count (about 6%) was also noted (fig. 3). Because of the prominent cytoplasmic vacuoles and markedly increased serum lactate dehydrogenase (LDH), a diagnosis of Burkitt or Burkitt-like lymphoma was initially suspected. The possibility of a solid tumor was also considered, but it was felt to be unlikely in view of the microscopic characteristics of the cells in the aspirate. Burkitt lymphoma is morphologically characterized by large, monomorphic cells (12–20 µm) with prominent cytoplasmic vacuoles (tab. 1).

Burkitt lymphoma is characteristically homogeneous in respect of every one of the seven morphological features listed in table 1, both from case to case, and in an individual case (for each of the features listed, occasional cells [<10%] may depart from the classical morphology). All listed seven features are required for diagnosis of Burkitt lymphoma and differentiation from vacuolated lymphoblasts of B-cell precursor ALL, other high-grade non-Hodgkin lymphoma with vacuolated cells (especially high-grade B-cell lymphoma,

Table 1. Features of Burkitt cells.

Cytological features	Burkitt cells
Cell size	Large and homogeneous
Nuclear chromatin	Dense but finally stippled and homogeneous
Nuclear shape	Regular-oval to round, no clefting, identation or folding
Nucleoli	Prominent; one or usually more vesicular
Amount of cytoplasm	Moderately abundant, surrounds the nucleus, no granules
Basophilia of cytoplasm	Very deep (due to high RNA content)
Cytoplasmic vacuolation	Often prominent in a majority of the cells

double-hit lymphoma, and diffuse large B-cell lymphoma), blastic mantle cell lymphoma (sometimes vacuoles are prominent and c-Myc rearrangement may also occur in blastic mantle cell lymphoma), acute monocytic leukemia (in which vacuoles may be striking), and non-hematological tumors involving the bone marrow e.g., rhabdomyosarcoma (in which vacuoles are typically prominent). Therefore, the presence of vacuoles in immature cells is not enough to justify Burkitt cells.

The cells in figures 2–6 have nuclear folding and indentations and, therefore, are not morphologically consistent with Burkitt cells. In fact, this was a case of acute monocytic leukemia. Cytologic analysis of the ascitic fluid revealed the same population with the cells infiltrating the bone marrow, consistent with immature monocytic-lineage cells (figures 7, 8). The cells were CD13+, CD33+, CD34-, CD14+, CD64+, and HLA-DR+. Ascites is quite rare in acute myeloid leukemia (AML). This case serves as a reminder that we should morphologically examine not only the peripheral blood and bone marrow of a patient, but also all fluid collections where we may find malignant infiltrates. This case also highlights that AML may present with prominent extramedullary manifestations (approximately 17% of cases) such as ascites and intra-abdominal lymphadenopathy, resembling high-grade non-Hodgkin's lymphoma.

The morphologic classification of AML with monocytic element established by the French-American-British (FAB) group includes AML subtypes M4, M5a, and M5b. The defining factor is the number of myeloid versus monocytic progenitors. In acute myelomonocytic leukemia (M4) both neutrophils and their precursors as well as monocytes and their precursors constitute ≥20% of the bone marrow cells, whereas in acute monoblastic/monocytic leukemia (M5) the neutrophils and their precursors are ≤20%. AML M5 subtype can further be subdivided in acute monoblastic leukemia (M5a – monoblasts >80% of monocytic lineage) and acute monocytic leukemia (M5b – monoblasts <80% of monocytic lineage).

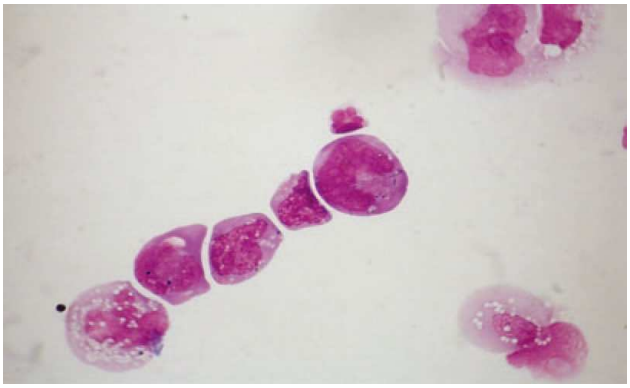

Figure 7

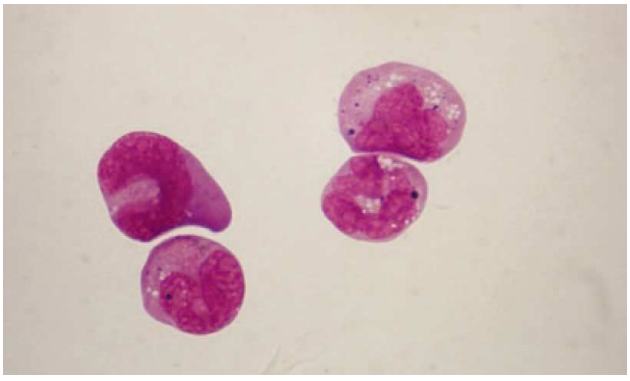

Figure 8

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Corresponding author:

C. Misidou, Department of Hematology, University Hospital of Alexandroupolis, Dragana, 681 00 Alexandroupolis, Greece
e-mail: xmisidou@yahoo.gr