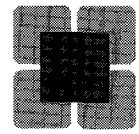

DAVID L. LONGWORTH, MD, AND JAMES K. STOLLER, MD, EDITORS

The elevated peripheral white blood count

Q1 A 50-YEAR-OLD MAN PRESENTS with an abnormal complete blood count. His hemoglobin is normal; the white blood cell count is markedly elevated to 200 000/μL, with 5% neutrophils and 90% lymphocytes; and the platelet count is normal. The patient has diffuse lymphadenopathy and splenomegaly. He feels fine. The patient likely has:

a. Acute myelogenous leukemia
b. Chronic myelogenous leukemia
c. Chronic lymphocytic leukemia
d. No hematologic pathology; the parameters described are appropriate for the clinical situation
e. Myelofibrosis with myeloid metaplasia

Q2 A 50-YEAR-OLD WOMAN PRESENTS WITH LEFT upper quadrant abdominal pain. Her white blood cell count is 150 000/μL with 60% neutrophils, 15% band cells, 8% metamyelocytes, 5% basophils, 5% myelocytes, and 2% promyelocytes; the hemoglobin is normal, but the platelet count is elevated to 700 000/μL. She has mild splenomegaly but no lymphadenopathy. She has no other symptoms. Her clinical situation is consistent with:

a. Acute myelogenous leukemia
b. Chronic myelogenous leukemia
c. Chronic lymphocytic leukemia
d. No hematologic pathology; the parameters described are appropriate for the clinical situation
e. Myelofibrosis with myeloid metaplasia

Q3 A 50-YEAR-OLD MAN PRESENTS WITH A 3-DAY history of cough and progressive fever. He has minimal cervical lymphadenopathy. The white blood cell count is elevated to 25 000/μL with 70% neutrophils and 20% band cells. The hemoglobin and platelet count are normal. Physical examination reveals rales at the base of the left lung. Chest roentgenographic findings are consistent with pneumonia. This clinical situation is compatible with:

a. Acute myelogenous leukemia
b. Chronic myelogenous leukemia
c. Chronic lymphocytic leukemia
d. No hematologic pathology; the parameters described are appropriate for the clinical situation
e. Myelofibrosis with myeloid metaplasia

Q4 A 50-YEAR-OLD WOMAN PRESENTS WITH A history of fatigue over several weeks, and fever and cough for 3 days. Physical examination reveals no lymphadenopathy; rales are auscultated at the base of the left lung. Her hemoglobin is 9.2 g/dL; the white blood cell count is 50 000/μL, with 1% neutrophils and many immature cells. The platelet count is 20 000/μL. Chest roentgenographic findings are consistent with pneumonia. This clinical situation is most consistent with:

a. Acute myelogenous leukemia
b. Chronic myelogenous leukemia
c. Chronic lymphocytic leukemia
d. No hematologic pathology; the parameters described are appropriate for the clinical situation
e. Myelofibrosis with myeloid metaplasia

Q5 A 54-YEAR-OLD MAN PRESENTS WITH increasing abdominal girth and left upper quadrant abdominal pain. His white blood cell count is 25 000/μL, with 60% neutrophils, 20% band cells, 8% myelocytes, and 2% promyelocytes. The hemoglobin is 9.8 g/dL, and the platelet count is normal. Physical examination reveals massive splenomegaly, with the spleen palpable 10 cm below the left costal margin. Peripheral blood smear analysis reveals abnormal red blood cell morphology, with many nucleated red blood cells and many teardrop forms. This clinical situation is compatible with:

a. Acute myelogenous leukemia

b. Chronic myelogenous leukemia

c. Chronic lymphocytic leukemia

d. No hematologic pathology; the parameters described are appropriate for the clinical situation

e. Myelofibrosis with myeloid metaplasia

Discussion

A1 There are many causes of an elevated white blood cell count. Chronic lymphocytic leukemia (CLL) is characterized by an absolute lymphocytosis of the peripheral blood and lymphocytosis in the bone marrow. The most frequent physical finding is lymphadenopathy. Approximately 70% of patients with CLL present with a normal hemoglobin value and a normal platelet count. Hypogammaglobulinemia is common, either initially or during the course of the disease. Immune-mediated cytopenias occur occasionally in patients with CLL. Autoimmune hemolytic anemia is indicated by a positive Coombs' test. Autoimmune thrombocytopenia is occasionally seen as well.

A2 Chronic myelogenous leukemia is characterized by an elevated white blood cell count that includes increased numbers of metamyelocytes, myelocytes, promyelocytes, and, frequently, an increased number of basophils. Elevations of the platelet count at presentation are common. Most patients with a white blood cell count > 150×10^9/L have a normochromic or normocytic anemia. The Philadelphia chromosome is pathognomonic for chronic myelogenous leukemia. This chromosomal abnormality involves a reciprocal translocation between the long arms of chromosome 9 and 22. This translocation juxtaposes the C-ABL oncogene from chromosome 9 within a specific breakpoint cluster region (BCR) and chromosome 22. Allogeneic bone marrow transplantation remains the only curative form of therapy.

A3 The third patient has an appropriate hematologic response to a pneumonia. He has a moderately elevated white blood cell count that is "left-shifted," but he has no other described hematologic pathology. Mild thrombocytosis is occasionally seen in patients with infections.

A4 Patients with acute myelogenous leukemia generally present with neutropenia, anemia, and thrombocytopenia. The peripheral white blood cell count is variable. This particular patient has an elevated white blood cell count, with the majority of cells being blast forms. Prompt medical attention is required for this patient with neutropenia, fever, and a pulmonary infiltrate. Appropriate cultures and empiric broad-spectrum intravenous antibiotic therapy should be initiated immediately. Combination chemotherapy would then be required in an attempt to obtain a complete remission.

A5 Myelofibrosis with myeloid metaplasia is a clonal stem cell disorder that is one of the myeloproliferative syndromes. It is characterized by excessive fibroblastic proliferation of marrow and myeloid metaplasia in the liver, spleen, and (occasionally) lymph nodes, which become active sites of extramedullary hematopoiesis. One hundred percent of patients present with splenomegaly. Fifty percent also present with hepatomegaly. Peripheral blood smear analysis reveals teardrop-shaped red blood cells characteristic of myelofibrosis, and nucleated red blood cells. A bone marrow biopsy will provide the diagnosis.

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