LEUKEMIAS

• ALL – “Acute Lymphoblastic Leukemia”
  a) occur at any age
    i) peak incidence < 5 yrs; incidence again rises in elderly
  b) ~ 20% of all leukemias
  c) most common of leukemias in children
  d) characterized:
    i) bone marrow infiltration of immature lymphoid cells (i.e., “BLASTS”)
      - precursor B and T lymphocytes
      - spill over into circulation
      - therefore, blood contains ↑ malignant lymphoid cells
e) rapid course
   i) recurrent infections
   ii) weakness
   iii) bleeding into skin and organs
   iv) enlarged lymph nodes
   v) mild splenomegaly

• AML – “Acute Myelogenous Leukemia”
  a) ↑ myeloblast in marrow and spill over into blood
  b) diagnosis of AML
     i) must have at least 20 % myeloblast in bone marrow
c) characteristics:
   i) most common type of acute leukemia in adults (~ 70 %)
   ii) “Auer rods” – specific for myeloid

d) ALL and AML cannot be differentiated without further biochemical techniques !!

• **CLL – “Chronic Lymphocytic Leukemia”**

  a) older patients (> 50 yrs)
  b) CLL cells not distinguishable from mature normal lymphocytes
     i) “acute CLL” – immature lymphocytes
  c) CLL is suspected when lymphocytes > 5,000/μL
d) progress slowly – not responsiveness to Tx

e) B-cell = 95% of all cases (T-cell 5 %)
  i) B-cell markers = CD19, CD20
  ii) T-cell marker = CD5

f) “smudge” cells or “parachute” cells

• CML – “Chronic Myelogenous Leukemia”
  a) leukocytosis
  b) disease of adulthood (~ 30 yrs; 85 %)
  c) three phases:
    i) chronic - ↑ WBC, Basophils, Eosinophils
    - < 10 % blasts
ii) accelerated - > 10 % blasts, > 20 % Basophils, usually develops into “blasts” crisis
   - > 20% blasts, develops into acute leukemia profile

d) Philadelphia chromosome in ~ 90-95% of CML patients
   i) $BCR/ABL$ gene rearrangement
   e) ↓ leukocyte alkaline phosphatase
   f) Most significant splenomegaly
A 38-year-old man complains of increasing weakness and fatigue. He has recently noted abdominal bruising and nosebleeds. He then developed fever and a cough. The spleen is not palpable. The peripheral blood reveals a normocytic, normochromic anemia without polychromasia. Abnormal-appearing, immature cells are seen in the peripheral blood smear. The most likely diagnosis is

(A) acute myelogenous leukemia
(B) acute lymphoblastic leukemia
(C) chronic lymphocytic leukemia
(D) chronic myeloid leukemia
The prognosis in Hodgkin disease, relative to survival, is best predicted by

(A) peripheral leukocyte blood picture
(B) clinical staging
(C) patient age at time of diagnosis
(D) intensity of systemic symptoms
(E) estimation of size of lymph node groups by lymphangiography
A 27-year-old woman has persistent, low-grade fever and occasional night sweats for the past month. She has seemed unusually fatigued and has lost her appetite. Pruritus has become troublesome. She has firm 1 cm enlarged lymph nodes in her lower anterior neck bilaterally and in her right axilla. Which of the following is most likely?

(A) acute lymphoblastic leukemia/lymphoma
(B) cutaneous T cell lymphoma
(C) follicular center cell lymphoma
(D) Hodgkin disease, nodular sclerosis type
(E) small noncleaved cell lymphoma
A patient with Hodgkin disease has involvement of mediastinal and retroperitoneal lymph nodes. The stage is

(A) I
(B) II
(C) III
(D) IV
(E) V
A characteristic age peak occurs in which two diseases?

(A) acute lymphoblastic leukemia and Hodgkin disease
(B) acute granulocytic leukemia and immunoblastic lymphoma
(C) acute lymphoblastic leukemia and acute granulocytic leukemia
(D) acute granulocytic leukemia and Hodgkin disease
(E) immunoblastic lymphoma and Hodgkin disease
The type of Hodgkin disease with the lowest 5-year survival is

(A) lymphocytic predominance
(B) nodular sclerosis
(C) mixed cellularity
(D) lymphocytic depletion
In the age groups 1 to 3, 30 to 40, and 60 to 70 years respectively, which types of leukemia are most common?

(A) chronic lymphocytic, acute lymphocytic, acute granulocytic
(B) acute lymphocytic, chronic lymphocytic, acute granulocytic
(C) acute lymphocytic, chronic myelogenous, chronic lymphocytic
(D) acute lymphocytic, chronic lymphocytic, chronic granulocytic
A patient with Hodgkin disease involving cervical lymph nodes and spleen is clinically in stage

(A) I
(B) II
(C) III
(D) IV
(E) V
A 25-year-old man has involvement of a cervical lymph node and an inguinal lymph node by Hodgkin disease. An exploratory laparotomy, splenectomy, liver biopsy and bone marrow are negative. The extent of disease is classified as stage

(A) 0
(B) I
(C) II
(D) III
(E) IV
Renal failure is characteristically associated with

(A) acute myelogenous leukemia
(B) multiple myeloma
(C) Hodgkin disease
(D) reticulum cell sarcoma
(E) chronic lymphatic leukemia
Bence-Jones protein may best be defined as an

(A) light chain produced in excess in some cases of multiple myeloma
(B) heavy chain produced in excess in some cases of multiple myeloma
(C) intact immunoglobulin produced in excess in some cases of multiple myeloma
(D) protein responsible for hypersensitivity
A 65-year-old man presents with a neck mass. Biopsy shows nodular mixed cellularity Hodgkin disease. Physical exam reveals axillary nodes, no abdominal or inguinal nodes, and nonpalpable spleen. Chest x-ray reveals enlarged mediastinal nodes. A bone marrow biopsy is negative. The clinical stage of the disease is

(A) I
(B) II
(C) III
(D) III E
(E) IV
A 43-year-old man complains of an enlarging painless right palatine tonsil. Biopsy of the affected tonsil reveals complete effacement of normal architecture by increased numbers of follicles composed of small cleaved lymphocytes. The most likely diagnosis is

(A) diffuse lymphoma
(B) follicular lymphoma
(C) Hodgkin disease
(D) reactive lymphoid hyperplasia
(E) tonsillar abscess
Lymphocytosis for a period of months, in people over 50 years of age is usually caused by

(A) allergic reactions
(B) chronic lymphocytic leukemia
(C) idiosyncratic drug reactions
(D) infectious lymphocytosis
(E) recovery from an aplastic crisis
The t(14;18) translocation associated with follicular non-Hodgkin lymphoma results in:

(A) activation of c-myc and unregulated cell proliferation
(B) expression of CD21, the Epstein-Barr virus receptor
(C) hybrid BCR-abl RNA transcription
(D) inappropriate production of ALK protein and T-cell activation
(E) inhibition of apoptotic cell death due to deregulated bcl-2 protein production
The patient with which of the following neoplastic disorders is most likely to develop acute leukemia?

(A) agnogenic myeloid metaplasia  
(B) chronic myelogenous leukemia  
(C) essential thrombocythemia  
(D) polycythemia vera  
(E) refractory anemia
A patient has a lymphocytosis of mature-appearing lymphocytes. Ninety-nine percent of the B lymphocytes express kappa light chain while only one percent of the B lymphocytes express lambda light chain. The most likely diagnosis is

(A) chronic lymphocytic leukemia  
(B) Hodgkin disease  
(C) mononucleosis  
(D) reactive lymphocytosis
Which of the following set of blood findings is most consistent with chronic myelogenous leukemia?

(A) high leukocyte alkaline phosphatase, blasts 4%, WBC 55,000/mm³
(B) low leukocyte alkaline phosphatase, blasts 4%, WBC 9,000/mm³
(C) low leukocyte alkaline phosphatase, blasts 4%, WBC 55,000/mm³
(D) low leukocyte alkaline phosphatase, blasts 35%, WBC 9,000/mm³
(E) low leukocyte alkaline phosphatase, blasts 35%, WBC 55,000/mm³
Which of the following types of leukemias are most difficult to separate morphologically (without special studies or historical information)?

(A) acute lymphocytic leukemia/acute myelogenous leukemia (M1-without maturation)
(B) acute myelogenous leukemia (M3-hypergranular promyelocytic)/acute lymphocytic leukemia
(C) chronic lymphocytic leukemia/acute lymphocytic leukemia
(D) chronic lymphocytic leukemia/chronic myelogenous leukemia
(E) chronic myelogenous leukemia/acute myelogenous leukemia (M2-with maturation)
A 53-year-old man presents to your office with complaints of fevers, weight loss and early satiety. Physical exam is remarkable for a spleen that is palpable about 5 cm below the left costal margin. There are no findings to suggest a source for his fever. His CBC is remarkable for WBC 13,500/mm³, Hct 44% and platelets 730,000/mm³. There are myelocytes and metamyelocytes present in his peripheral blood. Some giant platelets are seen. Which of the following tests would be most helpful to you in establishing a diagnosis
(A) blood cultures x 6
(B) leukocyte alkaline phosphatase score
(C) liver / spleen scan
(D) serum protein electrophoresis
(E) serum uric acid
Leukemic blood cells in chronic myeloid leukemia have which of the following chromosomal abnormalities?

(A) trisomy-21  
(B) D1-trisomy  
(C) Philadelphia chromosome  
(D) chromosome deletion  
(E) 15/21 translocation chromosome
HISTORY: This 35-year-old woman presented with severe fatigue for 3 weeks.

PHYSICAL FINDINGS: She was obese with blood pressure 160/50 mm Hg, pulse 120/min, and temperature 40 C. There were petechial hemorrhages in the retina, mouth and skin. Tonsils were greatly enlarged. Liver and spleen could not be felt due to obesity.
LABORATORY RESULTS:
  hemoglobin: 8 g/dl
  WBC: 14,300 /cu mm
  WBC diff:
  3% seg, 39% lymph, 3% mono, 55% blast

CLINICAL COURSE: Repeated bouts of septicemia occurred for the next 4 months until her death. She also developed diabetes mellitus which required large amounts of insulin to control.
At autopsy the bone marrow most likely will show

(A) hyperplasia
(B) multiple myeloma
(C) myelogenous leukemia
(D) myelofibrosis
(E) lymphocytic lymphoma
A patient with a history of mycosis fungoides presents develops Sezary cell leukemia. Which of the following would most likely be found by molecular analysis of the leukemic cells in the patient's peripheral blood?

(A) bcl-6 gene rearrangement
(B) BCR/ABL gene rearrangement
(C) monoclonal immunoglobulin gene rearrangement
(D) monoclonal T-cell receptor gene rearrangement
(E) PML/RARA gene rearrangement
In what age range is acute lymphoblastic leukemia most likely to be found?

(A) 0 - 9 years
(B) 10 - 29 years
(C) 40 - 59 years
(D) 60 - 75 years
Which of the following items is of most help in distinguishing chronic myelogenous leukemia from myeloid metaplasia?

(A) anemia
(B) a peripheral blood smear with 10% myelocytes
(C) marked splenomegaly
(D) decreased leukocyte alkaline phosphatase
Basophilic leukocytes in peripheral blood are most commonly seen in

(A) congenital spherocytosis
(B) acute lymphocytic leukemia
(C) chronic myelogenous leukemia
(D) non-specific infections
A 64-year-old white man presents in your office with vague, non-specific symptoms. A CBC with differential shows a white blood cell count of 40,000/mm³ of which 85% are lymphocytes. Further work-up discloses no other significant physical findings. This patient probably has

(A) chronic lymphocytic leukemia  
(B) cytomegalovirus infection  
(C) lymphocyte predominate Hodgkin disease  
(D) myelodysplastic syndrome  
(E) Waldenstrom macroglobulinemia
This 26-year-old housewife was in excellent health until ten days following delivery, when she developed rectal bleeding associated with weakness and faintness. Sigmoidoscopy revealed an area of oozing inside the rectum. Hemoglobin-8 g/dl; WBC-150,000/mm³. Bone marrow examination was similar to the findings on the slide which was obtained thirteen months later at autopsy.

The disease process which best characterizes this slide is
(A) hyperplasia of bone marrow
(B) multiple myeloma
(C) myelogenous leukemia
(D) myelofibrosis
The American type of Burkitt lymphoma is characterized by which of the following?

(A) Associated with EBV
(B) involves the maxilla and mandible
(C) commonly involves the abdomen
(D) characteristic t(11;18) translocation
(E) is a large cleaved lymphoma
A white blood cell count of 200,000/mm³ with 5% blast cells is most characteristic of

(A) chronic lymphocytic leukemia
(B) acute myelogenous leukemia
(C) leukemoid reaction
(D) chronic myelogenous leukemia
(E) acute lymphoblastic leukemia
The mechanism of death in leukemia most commonly relates to

(A) congestive heart failure
(B) bone marrow failure
(C) hypersplenism
(D) thrombosis
(E) hepatic failure
Acute leukemia is most reliably distinguished from chronic leukemia on the basis of

(A) number of blast cells in peripheral blood and/or bone marrow  
(B) level of elevation of white blood cell count  
(C) degree of hypercellularity of bone marrow  
(D) presence of enlarged lymph nodes  
(E) presence of anemia and thrombocytopenia
With present day therapy, the leukemia with the best prognosis is

(A) chronic granulocytic leukemia
(B) acute granulocytic
(C) acute lymphocytic, T-cell type
(D) acute lymphocytic, B-cell type
(E) acute lymphocytic, pre B-cell type (null)
The differential count on a peripheral blood smear shows myeloblasts. Bone marrow examination shows an increase in the percent of myeloblasts. Auer rods are present. The patient has

(A) chronic lymphocytic leukemia
(B) myelomonocytic leukemia
(C) pure monocytic leukemia
(D) acute myelogenous leukemia
A diagnosis of multiple myeloma has been made on an adult male. The medical resident requests the immunology laboratory to perform a serum protein immunoelectrophoresis. The most likely result is the demonstration of a paraproteinemia exhibiting

(A) IgA
(B) IgD
(C) IgE
(D) IgG
(E) IgM
The leukocyte alkaline phosphatase is usually decreased in

(A) chronic myelogenous leukemia
(B) leukemoid reactions
(C) myeloid metaplasia with myelofibrosis
(D) polycythemia vera
Which one of the following immuno-phenotypic subtypes of acute lymphoblastic leukemia is associated with the longest survival?

(A) pre-B cell
(B) B cell
(C) pre-T cell
(D) T cell
(E) mixed myeloid/lymphoid
Reed-Sternberg cells are characteristic of which disease?

(A) Burkitt lymphoma  
(B) cat scratch disease  
(C) Hodgkin disease  
(D) large cell lymphoma
Which type of Hodgkin disease is most likely to be diagnosed at an early stage?

(A) lymphocyte predominance
(B) mixed cellularity
(C) nodular sclerosis
(D) lymphocyte depletion
An example of a lymphoma derived from T-lymphocytes is

(A) follicular center cell
(B) hairy cell leukemia
(C) mycosis fungoides
(D) Waldenstrom macroglobulinemia
A 43-year-old man received radiation to his chest shortly after birth because of an enlarged thymus. He comes to see you because of sore swollen gums. His hemoglobin is 8 gm/dL, white blood cell count is 2,400/mm³ and his platelet count is 60,000/mm³. You correctly suspect which of the following:

(A) acute leukemia
(B) aplastic anemia secondary to radiation
(C) iron deficiency anemia
(D) megaloblastosis due to B12 deficiency
In the U.S., the majority of non-Hodgkin lymphomas in adults are

(A) B lymphocytes
(B) histiocytes
(C) null cells
(D) macrophages
(E) T lymphocytes
Multiple myeloma is frequently accompanied by

(A) carcinoma of the colon
(B) osteoblastic skeletal lesions
(C) renal cell carcinoma
(D) renal excretory impairment
In multiple myeloma the immunoglobulin most commonly elevated

(A) IgA
(B) IgG
(C) IgM
(D) IgD
(E) IgE
A 61-year-old man has had dull, constant back pain for 3 months. He recently developed a cough productive of yellowish sputum. On physical examination there are crackles at the right lung base. A plain film radiograph of the spine reveals several 1 to 2 cm lytic lesions of the vertebral bodies. A sputum culture grows Streptococcus pneumoniae. Which of the following pathologic findings is most likely to be seen in a bone marrow biopsy from this man?
A) Scattered small granulomas
B) Nodules of small mature lymphocytes
C) Occasional Reed-Sternberg cells
D) Numerous plasma cells
E) Hypercellularity with many blasts
Hairy cell leukemia would best be diagnosed with which of the following?

A) Myeloperoxidase
B) tartrate-resistant acid phosphatase
C) CBC and physical exam (massive splenomegaly)
A) t(8,14) genetic testing
B) plasma calcium and physical exam
A) t(9,22)  F) CD 20
B) t(8,14)  G) CD 5
C) t(11,14)  H) CD 23 +
D) t(14,18)  I) CD 10 -
E) CD 19

1) Burkitt lymphoma (B)
2) CML (A)
3) Follicular lymphoma (D)
4) Mantle cell lymphoma (C)
5) CLL (E,F,G,H,I)
A 16-year-old man presented to the emergency room following the development of swelling of his face and neck accompanied by respiratory difficulty. Mediastinal widening was appreciable by chest x-ray, accompanied by evidence of a right pleural and pericardial effusion. If this is a lymphoproliferative disorder, which one of the following is likely?

(A) acute lymphocytic leukemia, pre B type
(B) hairy cell leukemia
(C) lymphoblastic lymphoma/leukemia
(D) malignant lymphoma, follicular center cell
(E) malignant lymphoma, small lymphocytic
The viscosity of the blood is increased in patients with

(A) hemolytic anemia
(B) nephrotic syndrome
(C) monocytic leukemia
(D) Waldenstrom macroglobulinemia
(E) pernicious anemia
A 63-year-old man presents for evaluation of decreased stamina, a sense of abdominal fullness, and early satiety. His skin is pale, and he has an enlarged spleen. A CBC reveals pancytopenia. The most likely diagnosis would be

(A) acute lymphocytic leukemia, pre B type
(B) hairy cell leukemia
(C) lymphoblastic lymphoma/leukemia
(D) malignant lymphoma, small lymphocytic type
(E) malignant lymphoma, follicular center cell type