



PARTICIPANT STATISTICS

CELL IDENTIFICATION

THIRD QUADRIMESTER 2011

Folate Deficiency Q3-2011 Systemic Lupus Erythematosus

The WBC differential for this case revealed that there was an absolute lymphocytopenia.

SLE (often referred in the literature to as “lupus”) is a chronic, autoimmune, multi-system disease of unknown etiology. The number and variety of antibodies that can appear in lupus are greater than those in any other disease. Lupus can be quite mild, or it can be devastating, disabling, or fatal.

Although the majority of patients diagnosed with SLE are young women in their late teens to early 30s, it can occur at any age but appears most often between ages 10 and 50. People of African or Asian ancestry are more commonly affected than those of other racial ancestries.

Because symptoms vary greatly, lupus may resemble many other diseases making SLE somewhat difficult to diagnosis. Because different tissues and organs become inflamed in different people and the severity of the disease ranges from mild to debilitating, depending on the number and variety of antibodies that appear and the organs affected, symptoms vary widely from patient to patient and symptoms often appear and disappear.

Lupus may begin with a fever. A high fever can occur abruptly, or episodes of fever and malaise can come and go, sometimes for years. Joint pain and swelling are the most commonly experienced symptoms. Years of joint symptoms may precede other symptoms. A significant percentage of patients will develop arthritis. About half of patients with SLE demonstrate a characteristic “butterfly” shaped skin rash that covers the cheeks and bridge of the nose. This rash will worsen in the direct sunlight. Additional symptoms may include, fatigue, abdominal pain, nausea and vomiting, chest pain when taking a deep breath, hair loss, mouth sores or Raynaud’s phenomenon.

Anemia, leucopenia and dysproteinemia are common findings. Additionally, patients with SLE can develop glomerulonephritis, pericarditis and arrhythmias, pleuritis, CNS symptoms (headaches, numbness,seizures, vision problems, personality changes), breathing difficulties , splenomegaly and/or lymphadenopathy or hemolytic anemia.

The major pathologic changes appear to be due to circulating autoimmune complexes of antinuclear antibodies and their antigens. The major immune complexes are anti-DNA bound to DNA. All of the clinical and pathologic abnormalities in SLE are presumably related to these autoimmune phenomena.

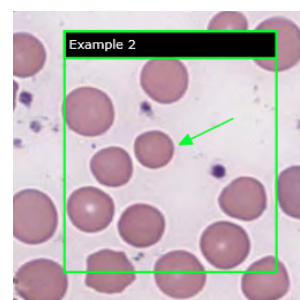
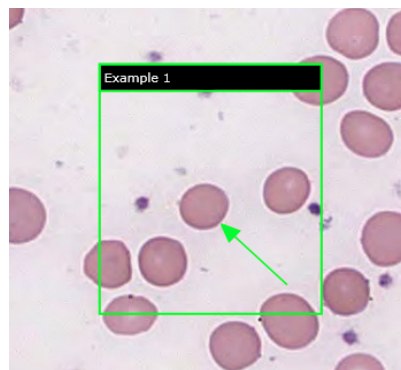
Laboratory tests used to diagnose SLE include an ANA panel, CBC, and urinalysis. Additionally, tests for antithyroglobulin antibody, antithyroid

Specimen 1		Specimen 2		Specimen 3		Specimen 4		Specimen 5	
Code - Result	No. Flag	Code - Result	No. Flag	Code - Result	No. Flag	Code - Result	No. Flag	Code - Result	No. Flag
430-Segmented Neutrophil (PMN, poly)	631	824-Platelet, giant	623	742-Monocyte, any stage	374	112-Polychromatophilic RBC	608	210-Basophil, any stage	655
320-Eosinophil, any stage	19 ***	816-Megakaryocyte	10 ***	200-Band Neutrophil (stab)	227 ***	110-Erythrocyte, normal RBC	22 ***	630-PMN or Band with Toxic Granulation/Vacuolization	3 ***
615-Hypersegmented Neutrophil	8 ***	865-Abnormal Platelet, would refer	6	12-Segmented Neutrophil (PMN, poly)	12 ***	170-Macrocyclic	7 ***	120-Basophilic Stippling	2 ***
630-PMN or Band with Toxic Granulation/Vacuolization	3 ***	100-Abnormal, would refer	3	620-PMN or Band with Dohle Bodies	9 ***	180-Rouleaux	7 ***	100-Abnormal, would refer	1 ***
640-PMN with Pelger-Huet Nucleus	2 ***	818-Metamegakaryocytic Fragment	3 ***	100-Abnormal, would refer	8 ***	100-Abnormal, would refer	4	130-S/C Crystasis	1 ***
Band Neutrophil (stab)	1 ***	820-Platelet, normal	3 ***	334-Metamyelocyte	8 ***	113-Reticulocyte (supravital stain)	4 ***	320-Eosinophil, any stage	1 ***
Total Population:	664	716-Hairy Cell	2 ***	630-PMN or Band with Toxic Granulation/Vacuolization	8 ***	120-Basophilic Stippling	4 ***	824-Platelet, giant	1 ***
Intended result was		826-Platelet Clumping	2 ***	712-Lymphocyte, normal	5 ***	186-Spherocyte	2 ***	Total Population:	664
Segmented Neutrophil (PMN, poly)		110-Erythrocyte, normal RBC	1 ***	714-Lymphocyte, reactive (atypical)	4 ***	870-Abnormal RBC, would refer	2	Intended result was	
		814-Megakaryoblast	1 ***	186-Spherocyte	1 ***	147-Agglutination	1 ***	Basophil, any stage	
		111-Nucleated RBC, any stage	1 ***	640-PMN with Pelger-Huet Nucleus	1 ***	164-Dimorphic RBC	1 ***		
		772-Plasma Cell, any stage	1 ***	855-Immature WBC, would refer	1 ***	168-Hypochromic	1		
		Total Population:	664	875-Abnormal Granulocyte, would refer	1 ***	850-Immature RBC, would refer	1 ***		
		Intended result was		Total Population:	664	Total Population:	664		
		Platelet, giant		Intended result was		Intended result was			
				Monocyte, any stage		Polychromatophilic RBC			
				19 of 22 Referees reported Monocyte					

Correct responses are defined as those reflecting agreement among 80% or more of all participants or referees. Unacceptable responses are indicated by "****" on the Flagging line of each specimen.

EDUCATIONAL CHALLENGES

Specimen 1	No.
Spherocyte	250
Microcytic	59
Erythrocyte, normal RBC	14
Anisocytosis	4
Abnormal, would refer	2
Polychromatophilic RBC	1
Hypochromic	1
Macrocytic	1
Poikilocytosis	1
Total Population:	333
Intended result was	
Spherocyte	



*To see the original full-sized images, please refer to the original CD or sign on to your data entry sheet at <http://www.aab-pts.org/>

Specimen #1: History: A 13-year-old boy is seen by his pediatrician. His mother states that he had a cold approximately 1 week ago and no longer has cold symptoms, but has continued with increasing fatigue and darkening of his urine. He has a history of an unknown blood disorder, for which he has received transfusions in the past. On physical examination, he has sclera icterus and a palpable spleen. A battery of laboratory tests are performed and show the following: WBC 3.8, Hgb 8.1 g/dL, Hct 24.3%, Plts 226,000/ μ L, Total Bilirubin 2.3 mg/dL, Osmotic Fragility tests shows increased red cell fragility. Identify the indicated cells.

The automated CBC results show a mild decrease in the WBC count, accompanied by anemia. Upon review of the peripheral blood smear, the most pronounced abnormalities are seen in the red cells. There is aniso- and poikilocytosis accompanied by polychromasia and the occasional nucleated red cell. In addition, a rare giant platelet can be seen. The red blood cells to be identified are spherocytes. Since spherocytes are round, densely staining, lack an area of central pallor, and are smaller than normal red blood cells, they are often also classified as microcytic. An occasional red cell has central pallor, however the majority of red cells seen are either spherocytic or polychromatic macrocytes (reticulocytes).

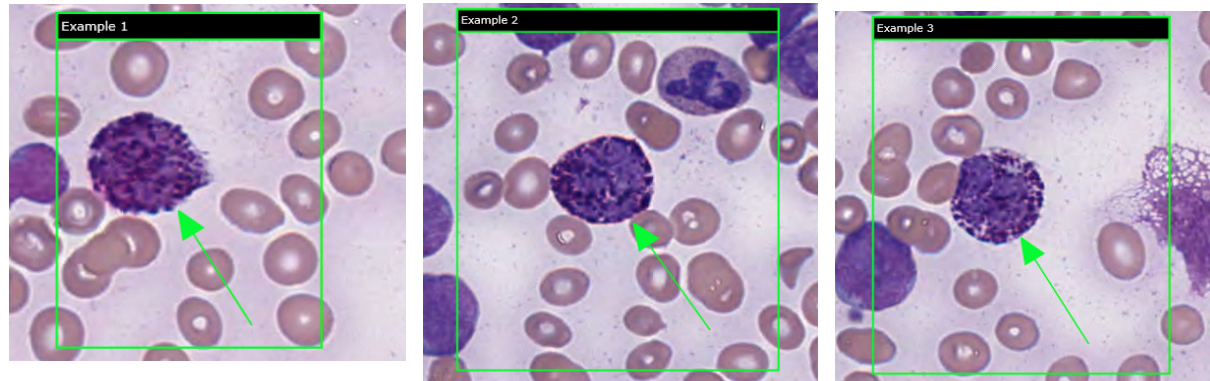
There are a variety of causes of spherocytosis, most of which are acquired rather than inherited. In acquired conditions, damage to the red cell membrane from toxins, infections, snake venom, mechanical trauma, alloantibodies, autoantibodies, and drug-induced antibodies can result in spherocytic red cells. In addition, donor red blood cells become spherocytic as they age during storage, leading to a dimorphic population of cells on the peripheral smear of a transfused patient. This patient was diagnosed with hereditary spherocytosis, an inherited abnormality of the red cell cytoskeleton. A deficiency of certain cytoskeletal proteins – spectrin, ankyrin, and/or band 3 – results in red cell membrane instability, a loss of membrane lipid, and decreased surface area, leading to spherocytic red cells.

Hereditary spherocytosis (HS) is the most common hereditary hemolytic anemia in people of Northern European descent with an autosomal dominant pattern of inheritance. Although anemia, jaundice, and splenomegaly are common clinical features of HS, the clinical manifestations are actually quite varied. Some individuals are completely asymptomatic and diagnosed only when a peripheral blood smear is examined for unrelated reasons. Other individuals have a mild or moderate compensated hemolytic anemia and develop symptomatic anemia only when confronted with a specific stress condition, such as a bacterial or viral (parvovirus B19 and EBV) infection, pregnancy, emotional distress, or exposure to very cold temperatures. Nonetheless, they may develop gallstones as a result of the elevated bilirubin levels and require periodic transfusions during hemolytic episodes. A small percentage of individuals with HS have a severe hemolytic anemia and are transfusion dependent.

The classic laboratory features of HS include anemia, reticulocytosis, spherocytes on the peripheral smear, increased MCHC, elevated bilirubin (indirect) levels, and an abnormal osmotic fragility test. Diagnosis is supported by family history and clinical signs and symptoms. Treatment of HS is supportive through the use of transfusions, splenectomy, and management of complications associated with the hemolysis and splenectomy.

Specimen 2	
Basophil, any stage	293
Promyelocyte	11
Abnormal, would refer	10
PMN or Band with Toxic Granulation/Vacuolization	6
Immature WBC, would refer	3
Abnormal Granulocyte, would refer	3
Eosinophil, any stage	2
Parasites	1
Microcytic	1
Blast, undifferentiated	1
Total Population:	333
Intended result was	
Basophil, any stage	

No.
293
11
10
6
3
3
2
1
1
1
333



*To see the original full-sized images, please refer to the original CD or sign on to your data entry sheet at <http://www.aab-pts.org/>

Specimen #2: History: *A 62-year-old woman is brought to the Emergency Department after being found unresponsive at home by her neighbor. On physical examination, the patient is now alert, but lethargic, has pale oral mucosa and conjunctivae, petechiae on her lower extremities, and splenomegaly. Pressure on her sternum elicits a painful response. CBC results: WBC 48,000/ μ L, Hgb 5.6 g/dL, Hct 16.8%, Plts 31,000/ μ L. Identify the indicated cells.*

The automated CBC results show marked anemia, thrombocytopenia, and leukocytosis. Review of the peripheral smear confirms the thrombocytopenia and elevation of WBCs. While mature granulocytes are seen, the majority of cells are blasts with dysplastic nuclei, prominent nucleoli, and minimal cytoplasm. The cells to be identified are basophils, which are increased in number. A basophil should be similar in size to a neutrophil. The cytoplasm contains numerous, fairly large blue-black granules that partially obscure the nucleus. In contrast, although a promyelocyte (answer chosen by a small percentage of participants) also has granules scattered throughout the cytoplasm, the granules do not obscure the nucleus and the cell is larger than a neutrophil.

This patient has chronic myelogenous leukemia (CML) with blast transformation. CML is a clonal stem cell disorder characterized by detection of the fusion protein BCR/ABL that occurs as a result of a translocation between chromosomes 9 and 22. The t(9;22) translocation is also called the Philadelphia chromosome. The incidence of CML increases with age and men are affected more than women. CML begins with a chronic phase during which time patients may be asymptomatic or complain of fatigue, lethargy, or pain due to splenic enlargement. Patients can then transform to the accelerated phase, characterized by increasing extramedullary hematopoiesis with splenomegaly and bone pain, particularly sternal tenderness, and increasing numbers of WBCs blasts in the peripheral blood accompanied by basophils and thrombocytopenia. The blast phase is characterized by the presence of at least 20% blasts in the peripheral blood and is consistent with transformation of CML to acute leukemia. The acute leukemia is myeloid in 70% of cases and lymphoid in 30% of cases (generally B-cell type). With transformation to the blast phase, the prognosis is grim with death occurring within 3 to 6 months due to infection and hemorrhage.