Hemepath Case 53: 4-Year-Old Girl

HISTORY

A 4-year-old girl presents with oliguria, hematuria, and increasing irritability. The child has a 1-week history of crampy abdominal pain, vomiting, and bloody diarrhea which were just beginning to clear up when these new symptoms appeared. The child first became sick after drinking unpasteurized apple juice.

On physical examination, the girl appears pale and restless. She is feverish and mildly hypertensive. Her ankles are slightly swollen, and a petechial rash is noted on her legs and arms.

CBC

Hgb (g/L)	Low
MCV	N
Reticulocyte Count	High
WBC	High with left shift
Plt	Low

OTHER LABORATORY FINDINGS

BUN	High
Serum Creatinine	High
Urinalysis	Cellular casts with dysmorphic RBCs

DESCRIPTION OF SLIDE

Peripheral Blood Smear

Anemia and thrombocytopenia are evident on the peripheral smear, as well as leukocytosis demonstrating a left shift (see circles). RBCs show echinocytosis (see rectangles) and severe schistocytosis (see arrows). Many large platelets are also present.

*** To see the slide annotations in Imagescope, click on VIEW, then ANNOTATIONS, and then on the "eye" icon adjacent to the word "Layers". In the “Layer Attributes” box, a brief description of the annotations is provided. You may also click on individual layer region (e.g. region 1) in the “Layer Regions” box to locate each annotation – this is especially helpful in identifying annotations when the slide is not zoomed in. ***

MORPHOLOGICAL DIAGNOSIS

Hemolytic uremic syndrome

DISCUSSION

Hemolytic uremic syndrome (HUS) classically presents with a triad of thrombotic microangiopathy, thrombocytopenia, and acute renal failure. It is in fact the most frequent cause of acute kidney failure in children.

In many patients, there is a history of infection by *E. coli* 0157:H7, which may be found in undercooked hamburger meat, unpasteurized apple juice or cider, and in other foods as well. Patients usually present with symptoms of colitis including bloody diarrhea. The *E. coli* bacterium produces a Shiga toxin which is absorbed from the gastrointestinal tract and travels via the bloodstream to capillaries in the renal glomeruli. The toxin binds to and damages the endothelial lining of the blood vessels, activating the body's inflammatory response and leading to deposition of fibrin in the vasculature wall. RBCs that traverse these vessels are "cut" by the fibrin strands; this is why we see erythrocyte fragments (schistocytes) in the peripheral blood. Reduced blood flow to the renal parenchyma results in organ failure. Renal failure and dehydration may together cause RBC echinocytosis (burr cell formation).

Platelet number is also reduced in patients with HUS. Increased consumption of platelets in the pathological intravascular coagulation, as well as sequestration of platelets in the liver and spleen, contribute to this phenomenon.