Case3 Print

Location: office

Presenting complaint: A 25-year-old African-American male presents with jaundice.

Vitals: Pulse:95/min, B.P:110/75 mm Hg, Temp:98.8 F, R.R:16/min, Height:72 inches (180 cm), Weight:72 Kg (158.4 lbs)

HPI: A 25-year-old African-American male presents to the outpatient clinic with the sudden onset of jaundice and dark colored urine. He complains of back pain and fatigue. The patient is afebrile and denies recent travel. He does not smoke, drink alcohol or use recreational drugs and uses condoms whenever he engages in sexual activity. On further questioning, it is revealed that he took TMP-SMZ for diarrhea a few days ago. An uncle has a history of some type of blood disorder.

Hospitalization/Procedures	None
Other Medical Problems	None
Allergies	NKDA
Current Medications	None
Vaccinations	Up to date
Family History	Mother died at the age of 60 yrs due to MI. Father is alive and healthy at the age of 65 yrs. No sibling.
Social History	He is single but has a girl friend. Denies tobacco, alcohol and drug use.
Sexual History	He is sexually active with his girl friend.
Occupational History	Restaurant owner.
Recreational	He plays basketball and enjoys traveling.

Review of Systems:

General	see HPI
Skin	pallor, itch, no rashes
HEENT	Icterus
Musculoskeletal	No muscle aches or joint stiffness
Cardio respiratory	Without complaint
Genitourinary	Dark colored urine, denies dysuria
Abdominal	see HPI

How to approach this case:
This is a presentation of jaundice. Jaundice can be due to hemolytic causes and disorders involving the liver or biliary tracts.

First perform a physical examination:
General examination
Skin
Lymph nodes
HEENT/Neck
Heart exam
Lung exam
Abdominal examination
Extremities
Neuro

Results of PE:
General: Icterus is noted on sclera; He also appears pallor. Abdominal examination: no masses, tenderness, or organomegaly. Normal bowel sounds. Rest of the exam is WNL

Routine Orders:
CBC with differential, routine

Basic metabolic panel, routine
LFTs, routine
Prothrombin time, routine

Results of Labs:

WBC count:	8200/mm3
RBC count:	1.8 million/mm3
Hemoglobin:	9 g/dL
Hematocrit:	33 %
Platelet count:	200,000/mm3
MCV:	98 cu microns
MCH:	28pg/RBC
MCHC:	35 g Hb/dL

WBC differential:

Segmented neutrophils:	72%
Juvenile neutrophils:	2%
Lymphocytes:	18%
Monocytes:	5%
Eosinophils:	2%
Basophils:	1%

Peripheral blood smear:
Normochromic normocytic erythrocytes, bite cells are also present. Leukocytes and platelets are normal in number and morphology.

LFTs

Bilirubin, serum, total	5 mg/dL
Bilirubin, serum, direct	0.5 mg/dL
Aspartate transaminase, serum	25U/L
Alanine transaminase, serum	20U/L
Alkaline phosphatase, serum	182U/L
Protein, serum, total	7.2 g/dL
Prothrombin time	11 sec

Discussion:
This patient has no history of fever or abdominal pain and also no risk factors for hepatitis. Thus, acute cholangitis or hepatitis is unlikely as a cause of his jaundice. Normal abdominal examination with absence of tenderness further excludes liver or biliary tract pathology. The presence of pallor, and dark colored urine suggests an intravascular hemolytic cause of the jaundice. Other points to elicit are positive family history and exposure to sulpha drugs.

Normal LFTs rule out liver or biliary tract disease. Elevated levels of indirect bilirubin are a clue towards hemolysis. CBC shows anemia and presence of bite cells on peripheral smear. This suggests that the jaundice is due to hemolysis. Next do the following tests to confirm the presence of hemolysis and to determine if it is intravascular or extravascular.

Routine Order review:
Admit the patient in floor/ward
IV access, stat
Normal saline, IV, continuous
Diet: Regular diet (avoid fava beans)
Activity - ambulation at will
Reticulocyte count, stat (to confirm that jaundice is hemolytic as reticulocyte count is elevated in cases of hemolysis)
Calculate reticulocyte production index (to correct reticulocyte count for the degree of anemia)
Serum haptoglobin
LDH, serum, stat (it is elevated in intravascular hemolysis)
Urinalysis, stat (to detect hemoglobin or hemosiderinuria)
Type and cross match, stat (for 2 units of blood)

PRBC, transfuse, stat (packed RBC transfusion)

*Repeat Hb and hematocrit (H& H) in 12 hours

Results:

BUN	12 mg/dL
Serum creatinine	0.6 mg/dL
Rest of the BMP	WNL
Serum LDH	400 IU/L
Serum haptoglobin	20 mg/dL
Urinalysis	normal

Elevated reticulocyte count confirms the presence of hemolytic anemia and elevated LDH with low haptoglobin indicate that the hemolysis is intravascular. A positive family history, history of exposure to sulpha drugs and presence of bite cells on peripheral smear are all suggestive of G6PD deficiency anemia. G6PD deficiency is confirmed by G6PD assay. Other hereditary causes of hemolytic anemia are sickle cell anemia, thalassemias and hereditary spherocytosis.

In sickle cell anemia, peripheral smear shows sickle shaped RBCs and in hereditary spherocytosis RBCs exhibit a loss of central pallor. Thalassemias produce a microcytic picture with target cells. For thalassemias and sickle cell anemia, hemoglobin electrophoresis provides useful diagnostic information. Autoimmune hemolytic anemia is an important non-hereditary cause and in such cases coomb's test is an important diagnostic tool. Intravascular hemolysis can also be a part of thrombotic thrombocytopenic purpura, but in such cases fragmented RBCs are found on peripheral smear and platelet counts are also low along with renal impairment. Regarding treatment of G6PD deficiency anemia, all affected individuals should avoid exposure to drugs with oxidant potential. Heterozygous females should also avoid exposure to such drugs during pregnancy and lactation as they may trigger hemolysis in the fetus or neonate. Transfusions are needed when anemia is very severe due to impaired compensatory erythropoiesis.

Routine Order:
G6PD blood, quantitative, stat
Coomb's test, direct, stat (optional-depending on the scenario)

Result:
G6PD levels are low (levels can be normal during or immediately after the acute hemolytic episode, therefore you can repeat the test to confirm the diagnosis)

Order review:
Schedule an appointment after 2 months and at that time re-evaluate G6PD assay
Reassurance
Patient counseling
Limit alcohol use
Regular exercise
Safe sex counseling

Primary Diagnosis:
G6PD deficiency anemia