9/9/26 Morning Report with @CPSolvers

"One life, so many dreams" **Case Presenter**: Rida **Case Discussants**: Sharmin & Kirtan

https://clinicalproblemsolving.com/present-a-case/

Scribing (Eyron):

CC: 22/M p/w yellow discoloration of sclera and dark colored urine for 3 days

HPI:

He was in usual state of health when until he developed gradual onset of yellowing of sclera and dark colored urine which increased in severity a/w dark urine, pale stools and pruritus as well as decreased appetite and disturbed sleep.
Also with fever - no chills/rigors
1 day prior, he noted pain to large joints which affected his ability to walk

ROS: (+) hair loss, palpitations, blurring of vision, vertigo

PMH: recurrent episodes of yellow discoloration (2010, 2022) s/p PRBC

Meds:
Iron tablets

Fam Hx:

Social Hx: married

Health-Related Behaviors: non-smoker, no EtOH, no drugs

Allergies:

Vitals: T: 98F **HR**: 88 **BP**: 110/70 **RR**: 18 **BMI**: nl
Exam: Gen: jaundiced
HEENT: hyperpigmented cheeks, no palpable lymph nodes
Abd: soft, nt,nd, palpable liver 2 cm below right costal margin, non-tender, liver span 13cm, nonpalpable spleen, no shifting dullness
Extremities/skin: flat nails

Notable Labs & Imaging:

Hematology:
WBC: 15.3 Hgb: 6.2 Plt: 152 MCV: 67
Chemistry:
Electrolytes nl | PT 16.3 INR 1.57
AST: 244 ALT: 129 ALP: 158 T. Bili: 9.5 Direct Bili 6.0 Indirect Bili 3.5
T. protein: 7.0 Alb: 3.3 Glob 4.1 CRP: 88.3
Peripheral Smear: target cells, nucleated rbcs, occasional howell jolly bodies, sickle cells
Reticulocyte count 2.5%, direct coombs negative
Ferritin 20k, LDH 1160
Hb Electrophoresis: HbS 95.6% HbA2 4.4%
Hepatitis panel: Non-reactive
Autoimmune Workup:
ANA (+), speckled pattern, anti-dsDNA (+), Anti-Ro (+), ASMA (+), RF, anti-CCP negative | C3 decreased | IgG 38.8
Imaging:
US Abdomen: liver is normal size, smooth margins, no free fluid, normal spleen

Dx: Sickle Cell and Thalassemia Overlapping Syndrome with SLE and Autoimmune Hepatitis

Problem Representation:
22/M with recurrent episodes of yellow discoloration of sclera, dark colored urine, and pale stools found to have microcytic anemia and direct hyperbilirubinemia. Hb electrophoresis notable for HbA2 and HbS with autoimmune workup revealing (+) ANA, anti-dsDNA, and ASMA with decreased complement consistent with sickle cell and thalassemia overlap with SLE and AIH

Teaching Points (Varsha)

Jaundice: painless vs painful, direct vs indirect
- Acute no miss causes: Acute liver failure (Acute confusion/odd behavior), hemolysis, acute cholangitis
- Something wrong with
  - **hematologica**l (Elevation in bilirubin + change in heme index)
  - **hepatobiliary system (**associated with nausea, vomiting, ab discomfort, severe pain - peritoneal/hepatic capsule involvement)
  - **Extrahepatic** (macroscopic disease) vs **intrahepatic** (microscopic involvement - external: meds/toxins/infections Vs internal: autoimmune/infection/infiltration)

Associated symptoms: infection/autoimmune/toxin exposure
Hair loss + palpitation in young patient → check thyroid profile
Arthralgia in young patient → infection → mostly viral (arthralgia vs arthritis → exam)/ autoimmune
Blurry vision: true neurological change (Optic nerve, ICP) vs subjective symptom; persistent vs intermittent; **Vertigo**: true vertigo (VC nerve) vs non specific
Episodic anemia: most likely not bleeding (no jaundice), could be hemolysis. **Trigger** → oxidative stress/medications or toxins/ no trigger
- G6PD, hereditary spherocytosis, hemoglobinopathies
- Labs: LDH, haptoglobin, direct/indirect bili, PS, PI/INR

Visible jaundice → bilirubin 4-5 . If liver is intact, it should be able to conjugate all the bilirubin from hemolysis unless the liver is also compromised
Hyperpigmented cheeks: increased melanin → sun exposure vs pathologic (vitamin deficiency → alters RBC production as well)
Etiologies that affect both liver and heme
Any hemolytic anemia → gallstones/cholangitis (RUQ US), Wilson's→ can cause hemolysis in end stage liver damage due to free Cu, sickle cell anemia→ affects tiny hepatic sinusoids
low MCV: IDA,sideroblastic anemia, thalassemia → ineffective erythropoiesis → iron overload→ 2nd hemochromatosis→ liver damage (HLH – % sat either normal/low vs 5sat here → high)
PS → **sickle cell +** → has Sickle cell anemia to some extent for sure; **howell jolly bodies**: asplenia + (neither appear physiologically)
Sickle cell anemia and liver complications – acute: sinusoidal obstruction, hepatic ischemia, budd chiari syndrome, chronic: long term iron overload → 2nd hemochromatosis
Severe complication of sickling: bone marrow necrosis due to too much sickling within the marrow