10/19/26 Mainstream Monday with @CPSolvers

"One life, so many dreams" ***Case Presenter****: Syeda(@)* ***Case Discussants****: Youssef (@saklawiMD) & Ann Marie (@annkumfer)*

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

Scribing (Valentina and friends):

CC: 22-year-old presents to the ED with scleral icterus and confusion

HPI: Patient is previously healthy with no prior hospitalizations. She now has one week of fatigue, decreased appetite, nausea, intermittent abdominal discomfort and non-bloody vomit

Darker urine, yellowing of the eyes x 3 days

ROS: No significant trauma.

PMH: never hospitalized, no anemia or jaundice

Meds: occasional acetaminophen

Fam Hx:

none

Social Hx:

Unremarkable

Health-Related Behaviors:

Denies supplements, extra medications, recreational drugs, travel

Allergies:

Vitals: T: 98,8 F (37.1 C) **HR**: 106 **BP**: 104/66 **RR**: 18 **Sat**: 99 on room air

Exam: Gen: Scleral icterus and diffuse jaundice

Abd: mild discomfort with palpation RUQ, no rebound or guarding

Neuro: awake, follows commands, oriented to self, place, but slow, following commands, slow to respond. No asterixis.

MSK/skin: small bruises over arms and legs

Notable Labs & Imaging:

Hematology:

WBC: 11.8 Hgb: 8.7 MCV: 91 Plt: 118

Chemistry:

Na: 136 K: 3.6 Cl: HCO3: 19 Ca: Mg: BUN: Cr: 1.1 Glu:

AST: 186 ALT: 72 ALP: 38 T. Bili: 26.4 Direct Bili: 10.1 T. protein: Alb: 2.9

Coagulation panel: PT: 24.6 aPTT: 41 INR: 3.1

Pregnancy test negative

Acetaminophen: undetectable

Acute Hep A, B, C: negative

Imaging:

Echo: no biliary ductal dilation; patent hepatic and portal vasculature

worsening acute liver failure and hepatic encephalopathy with increasing INR unresponsive to vit K administration - ICU transfer

Hematology: Hgb: 6.9; Plt: 92

Coagulation panel: PT: 35.2; aPTT: 53;

AST: 228; ALT: 81

No obvious bleeding

Hemolysis labs: Reticulocyte: 9.8%; LDH: 1420; Haptoglobin: <10; Direct Coombs: negative

Peripheral smear: polychromasia without significant schistocytosis

Iron studies: Serum iron: 168; Ferritin: 684; TSAT: 74%

Fibrinogen: 165

D-dimer: 3.8

New Liver function tests: Marked hyperbilirubinemia, ALP 36; AST>ALT

Autoimmunity panel: ANA: nl; SMA: nl; serum IgG: nl

Serum ceruloplasmin: 11

24h urine collection: increase in copper excretion

Slit-lamp examination positive for Kayser-Fleischer rings

Unresponsive to supportive management the patient underwent transplantation. The explanted liver showed elevated copper concentration

Final dx: Wilson disease

Problem Representation: 22-year-old F with acute hepatic failure, progressive encephalopathy, Coombs negative hemolysis

Teaching Points (Gillian)

In a young person without past medical history:

- May have medical problem not yet recognized
- Large physiologic reserve, takes a lot to bring patient to care
- Schema: bad genes, risky behavior, bad luck; *In assessment of risky behavior, safe space for questioning is imperative*

Nausea, abdominal pain, anorexia, fatigue: -itis inside the GI tract, GU, Diffuse: toxin, CNS

Jaundice: Hemolysis vs hepatobiliary

- To see jaundice, you need massive hemolysis
- Hepatobiliary can still have mixed direct/indirect bilirubin
- Massive bilirubin elevation makes the liver very likely involved, can have concomitant hemolysis

Bilirubin + abdominal pain/nausea syndrome

- Can localize to hepatobiliary system
- Inflammatory response to initial syndrome

Petechiae: blood (platelets, coagulation cascade) vs vessel.

- When you add encephalopathy, worry about TMA (TTP, DIC)
- Stat CBC, CMP, Liver labs, hemolysis labs

Bicytopenia: bone marrow, spleen, consumption (thrombocytopenia, anemia)

- MCV can be helpful: TTP <110, in B12 def MCV very high
- Intermediate thrombocytopenia (50-150) can be early consumption, but makes concern slightly less

Liver enzymes:

- Low Alk Phos compared to bilirubin: zinc deficiency, wilson's disease
- AST, ALT lower than expected may suggest sub acuity
- Can check AST/LDH ratio

Coagulopathy related to liver disease: can check factor VIII, vWF

Acute liver failure: Substance, Ischemia, Acetaminophen, Hepatitis, HELLP, Systemic syndrome: (autoimmune hepatitis, HLH, wilson's disease)

Hemolytic anemia + liver disease: burr cell hemolytic anemia, zieve syndrome, AIHA + autoimmune hepatitis, sickle cell disease, HELLP, Wilson's disease

Wilson's Disease: Bilirubin/Alk Phos ratio >4, AST/ALT ratio >2 , coombs negative hemolytic anemia