8/28/23 Rafael Medina Subspecialty VMR with @CPSolvers

"One life, so many dreams" Case Presenter: Dr. Mike Arcieri (@ArcieriMichael) ***Case Discussants****: Dr. John Chang*

CC: 50M w/ 3w of sleepiness and falls

HPI: 3w ago became slowly but progressively more sleepy. Complicated by frequent falls and increased need for help with ADL's, including showering and toileting (Previously fully independent in ADLs). 2w ago noticed to have "tremors" when holding shopping cart. **ROS:** becomes intermittently more lucid during the history and states his chronic abdominal pain is "the same as always." Diffuse swelling of the extremities and around the eyes. Chronic weight loss. Negatives: No dyspnea, orthopnea, PND, jaundice, melena, hematemesis, weakness, paresthesias, or medication misuse.

PMH:

1) GI: Ileitis and Colitis s/p colostomy 40y ago. Gastric perforation ex- lap 1y ago w/ 30+ GI ulcers. IBD suspected. 2) STEMI 6m ago stenting to RCA and LAD 3) Unprovoked PE 6m prior. 4) Remote history of AMS w/ Tracheostomy and PEG placement years prior.

Meds:

Pantoprazole
Apixaban
Buprenorphine
Aspirin
L-Thyroxine
Trazodone
Gabapentin
Acetaminophen
Metoprolol
Aripiprazole

Soc Hx: Previously unhoused, did not have steady access to food (months in duration)Recently did move in with a friend where food is now consistently available. Out of work for 5 years, prior mechanic. 20 pack-year smoking history. Remote EtOH and Opioid use disorder. Active cocaine user

Vitals: T: 98.1 **HR:** 65 **BP:** 130/70 **RR:** 14 **SpO2 92%** on Fi02 0.21
Gen: somnolent, hard to awaken
HEENT: no jaundice or scleral icterus
CV: No JVD.
Pulm: crackles in R lung base
Abd: Diffuse tender and distended abdomen, no peritoneal signs.
Neuro: challenging; eyes open symmetrically, pupils 3mm reactive, speech mumbled, 4-/5 strength in 4 extremities, asterixis present
Extremities: Diffuse anasarca. 3+ LE, 2+ UE, 2+ sacral, 1+ periorbital
Skin: no stigmata of liver disease, no jaundice.

Notable Labs & Imaging:
Hematology: WBC: 8.7 Hgb: 10 (baseline 12.5) Plt: 474 (high)
Chemistry: Na: 140 K: 3.9 Cl: 108 CO2: 26 BUN: 11 Cr:0.77 glucose: 93 Ca: 9 (corrected)
Phos: 2.5 Mag: 2.2. AST: 13 ALT: 20 Alk-P: 146 T. Bili:0 0.34 **Albumin: 0.9** GGT 52. INR 1.5.
Ammonium: 110.
BNP 133 (during prior STEMI: 500).
Acetaminophen, Gabapentin, Alcohol negative. UDS: + cocaine, - opioids
Urine: trace protein, rest negative. UACR 0.5g . 24h urinary prots: 1.51g
TSH 17, T4 0.9 (low) , T3 1.1 (low)
VBG: ph 7.38. Pc02 43
ANA, C3/C4, HIV/ HCV/ HBV, anti-SM, Ceruloplasmin, AMA, TTG IgA (-). BCx negative.
Folate 4.7 (low) , Vit D 8.9 (very low), VitA 9 (very low), B1/B12, VitE/ VitK nl
CXR: RLL opacity w/ small R pleural effusion. Rest normal. **Abd US:** decompressed gallbladder. No ascites, steatosis vs chronic liver disease. CT head non-con: normal. MRI brain: small vessel disease. **TTE:** EF 60%, I wall motion ab (same as 6m prior). **CT abd/pelvis w/c:** liver and spleen normal. Fat stranding at splenic flexure. Known 4.5 x4cm fluid collection posterior aspect gastric fundus. No lymphadenopathy.
Amino acid analysis: arginine, citrulline, orotic acid, ornithine, homocitrulline normal. Citrullinemia (SLC25A15), HHH syndrome (SLC25A13) wild type. NO urea cycle disorder.
Liver biopsy: steatosis w/o fibrosis: NAFLD. HVPG 5 mmHg (normal)
CTA for trans-yugular liver Bx: splenorenal shunt identified with portal to- systemic shunt. Should improve w/ embolization. Partial improvement with lactulose. **Embolization performed: dramatic reduction in ammonia.**

Final Dx: Hyperammonemia 2/2 splenorenal shunt exacerbated by protein- wasting enteropathy w/intermittent protein loads from intermittent GI bleeds + DAPT/AC

PR: 50M w/ 3w of sleepiness presented diffuse anasarca, hypoalbuminemia and hyperammonemia without cirrhosis found to have a splenorenal portal to systemic shunt which resolved after shunt embolization.

Teaching Points (Seyma):
AMS: MIST-P → Metabolic (Hypo/HyperNa, BUN/Crea, Hypercalcemia, Ammonium, Glucose, Vit B1 & Vit B12), Infection, Structural (high ICP), Toxins/Meds (anticholinergics, opioids, benzos, gabapentin), Psychiatric
Gabapentin toxicity can p/w tremor and AMS; dosage as high as 300 mg/d w/ normal renal function; 100 mg/d on dialysis
CAVE: AMS in AKI most commonly medication induced!
Asterixis: hypercapnia, hyperammonemia
Edema: Heart, **Liver,** Kidney, (Thyroid)
Hepatopulmonary syndrome could explain the low SpO2 (Hypoxemia due to dilated intrapulmonary vasculature in the presence of portal HTN); A-aO2 >20 mm Hg; clinical signs: platypnea/orthodeoxia
Thrombocytosis: reactive, chronic iron deficiency anemia
Low albumin: Renal loss (nephrotic syndrome), Liver failure, Protein losing enteropathy, Inflammation, Malnutrition, Clarkson syndrome (systemic capillary leak)
Ammonium: produced in small intestine & colon due to protein catabolism → transport to liver, then converted into urea
High ammonium:
Liver dz (impaired urea cycle), Urea cycle disorder (most common: Ornithine Transcarbamylase deficiency →excess Carbamoylphosphate gets funneled to orotic acid↑), intake of Depakote (Divalproex sodium), Renal dz, Shock, GI bleeding, Smoking, Urease-producing bugs in UTI (Proteus, Ureaplasma, Klebsiella, Staph. saprophyticus), malnutrition, **portosystemic shunt,** multiple myeloma, neobladder (reservoir function; made of ileum)