01/29/25 Morning Report with @CPSolvers

"One life, so many dreams" Case Presenter: David Serantes (@davserantes) Case Discussants: Sharmin(@sharminzi) & Reza

CC: malaise, myalgia, abdominal pain

HPI: 30 Y/M coming in with malaise, myalgia in shoulders and cervical region abdominal pain with N/V
No rash, no dysphagia.

Admitted to the hospital 1 month ago for subacute course of fever, arthralgia, rash suspected to be Still's started on IV prednisone and later to oral steroids
Negative microbiology & autoimmune Workup. PET CT: Hyper Nodular activity

PMH: None

Meds: Prednisone 10mg
Acetaminophen
ibuprofen

Fam Hx: None

Soc Hx:non smoker, non-alcoholic.
No illicit use of drugs

Health-Related Behaviors: **lives in south spain, contact with horses.**

Allergies:

Vitals: T: afebrile BP: 120/66 mmHg**:** 20**HR**: 88 bpm **Sat:98@RA**
Exam: Gen: not acutely ill.
HEENT: small B/l cervical lymphadenopathy
CV: wnl, **Pulm:** wnl
Abd: tenderness epigastric area, no hepatosplenomegaly, Murphy neg
MSK: folliculitis lesions (mild)- resolved later

Notable Labs & Imaging:
Hematology:
WBC: 8.7, Diff- Nl, Hgb: 16.5Plt:177K
Chemistry
Na: 137K: 4.8Cr:0.8 BUN: 14 Glu:100
CRP: 1.3 LDH: 580 AST:1400 ALT:1900 ALP: 70, Ind Bili:2.75, direct bili: 1.4, alb:4, TP: 7.8 GGT: 115, lipase: 45, CK: 120.
Pro BNP: 40, Trop: Nl, ferritin: 4200-40,000 Procal: Nl
VBG:Ph: 7.38, HC03: 28, lactate: 1
INR: 1.25, PTT: 30, fibrinogen: 240(nl)
UA: trace protein, urobilinogen: Positive
HSV, HBV,HIV: neg
Blood & Urine culture: neg
PCR + for E coli, campylobacter, CMV IgG EBV IgG(+),
Rickettsia, PCR dengue, chikungunya,Brucella, Borrelia, Coxiella: Neg
Two days after admission- Bilirubin: 7, INR: 2.5, Liver enzymes: 2000s, hypoglycemia. **Started on acyclovir, Doxycycline and NAC progressed to acute liver failure.**
Acetaminophen levels: Nl
Imaging:
USG : Nl, GB wall slightly increased, no bile duct dilation. No ascites, doppler:Nl
Copper: Nl, C282 hetergenosity, H63D: Neg. Autoimmune liver workup: Neg
Further complicated by Hepatic encephalopathy.
Liver Biopsy: Unknown.
Received Liver transplantation.
PCR for HHV 6: +
Dx: **Fulminant Hepatic Failure 2/2 HHV 6(possibility due to immunosuppression)**

Problem Representation: Young guy with possible still's disease presenting with Acute Liver Failure Dx as: **Fulminant Hepatic Failure 2/2 HHV 6.**

Teaching Points (Seyma):
Autoinflammatory vs autoimmune (targeting specific antigen)
AOSD: Yamaguchi-criteria (joint pain, neutrophilic leukocytosis, fever, salmon-colored rash etc)
Patients w/ underlying autoimmune/autoinflammatory conditions: Accurate dz ? Flare-up (and what manifestations of flare/complications need acute intervention)? Complication of dz itself (e.g. hypercoag), Drug-related complications (immunocompromised?, Adrenal insufficiency 2/2 steroids), Is it unrelated?, Later: How was the dx made?
Pustular skin lesions: SNAPE (non-infectious mnemonic): **S**APHO, **N**eutrophilic dermatoses (Sweet syndrome, Behcet), **A**utoinflammatory (Pyoderma gangraenosum, Still's), **A**GEP, **P**ustular psoriasis, **E**osinophilic folliculitis (Ofuji-syndrome)
AST or ALT >1000: ***severe acute liver injury*** → prioritize drugs (**p**arenchyma: infection (viral: Herpes-, Hep virus, HIV, Yellow fever), drugs & herbal supplements (Tylenol, Green Tea, Augmentin), autoimmune, Wilson, HLH, infiltrative malignancy; **p**regnancy (HELLP), **p**lumbing: biliary, ischemic) ***+ HE & INR >1.5 → Acute Liver Failure***
AST >ALT: could be extrahepatic e.g. from muscle in absence of alcohol, but be aware that ALT has longer half-life (could be also ALT = AST or even ALT >AST)
Hyperferritinemia: HLH, MAS, AOSD, Histo, PJP, ALF, severe sepsis, hemochromatosis
Final note: AOSD → high dose steroids —> higher risk for fulminant Herpesvirus (here: HHV6) infection