12/30/22 Morning Report with @CPSolvers

Case Presenter: *Kirtan Patolia (@KirtanPatolia)* ***Case Discussants***: *Rabih Geha (@rabihmgeha)*

CC: Persistent fever

HPI: 27yo M w/ no PMH p/w 5 days of persistent fever, HA, n/v, sore throat, neck and shoulder pain and diffuse body aches.

PMH:
-

Meds:
-

Fam Hx:
-

Soc Hx:
-

Health-Related Behaviors:

No travel

Allergies:
-

Vitals: **T**: 38.4 **HR**: 130 **BP**: 126/82 **RR**: 18 **SpO_2**: 98% RA
Exam: **Abd:** mild splenomegaly, diffuse TTP

Notable Labs & Imaging: **Referring hospital**: monospot test positive
Hematology: WBC: low 2.7 (1.8 lowest) Hgb: 14.2 Plt: 30k
Chemistry: AST: 98 (peak 474) ALT: 272 Alk-P: 132 > 187 T. Bili: 2.2 (direct: 0.5), LDH: 1496 > 3k, GGT: 64 > 135
Thyroid: wnl
Imaging: CXR: wnl, CT a/p: wnl except for splenomegaly
Infectious and autoimmune work-up: negative
CSF and blood cultures, serum fungal, urine culture, HIV, CMV, HSV, COVID, influenza, RSV - negative
Clinical course: Abdominal pain went away on it's on. HA became CC. 10/10 HA despite analgesics.
Obtained EBV serologies: capsid antigen IgG >750, IgM negative
Nuclear antigen IgG negative: EBV PCR > suggesstive of acute infection
Further labs: Ferritin: 7500, PT 16.9 INR 1.41 aPTT 40.3, d-dimer: elevated
CT scan to r/o hemorrhage - unrevealing
Started on gancyclovir: within 3 days – improved labs, fever resolved, ferritin came back to 2k

Bone marrow biopsy > increased monocytes and macrophages.

Month later > soluble IL-2 receptor came back positive: 35k

Final Diagnosis: HLH from infectious mononucleosis (HA 2/2 PRES)

Problem Representation: 27yo M with no PMH p/w persistent fever, sore throat found to have infectious mononucleosis, splenomegaly, leukopenia and H-criteria consistent with HLH from infectious mononucleosis and HA 2/2 PRES.

Teaching Points (Debora):
- Fever → infection. pharyngitis w/ systemic features + severe Tachycardia. Check for COVID, virus: influenza and RSV
Complications of pharyngitis: abscess near the tonsils, sinusitis and ear infection.
Management: Check the vitals signs, fluids, swab tests
- Infeccion + splenomegaly → Viral: mononucleosis, bacterial: syphilis, parasitic: malaria. But leukopenia is unusually for mononucleosis, look for a cause. E.g. HLH can be primary or secondary.
- Test: Mono, HIV (acute retroviral infection), TSH (hypothyroidism) + image of pelvis and abdomen.
- Headache refractory: brain abscess, meningoencephalitis. Can be a sctrural lesion in the brain.
Hemophagocytic lymphohistiocytosis: systemic, multi-organ system hyperinflammatory state, caused by inappropriate activation of immune cells. Can be primary (potentially genetic), typically infants and young adults or secondary infection (likely - herpesviruses, parvo, COVID, brucella, TB, histoplasmosis, leishmaniasis, etc), tend to be in old patients.
H-criteria (need 5/8): - Fever - Cytopenias (at a minimal two lineages) - Splenomegaly - Hypertriglyceridemia +/- hypofibrinogenemia - Hemophagocytosis on biopsy - Ferritin > 500 ng/ml - Low or absent NK-cell activity - Elevated sIL2Ra.
- HLH and headache → neurologic involvement or complication like a hemorrhage
Hemophagocytic lymphohistiocytosis in response to **EBV** or otherwise remains a life-threatening problem for patients with genetic disorders that cause HLH. Early recognition and initiation of HLH-directed therapy remain key for patient survival.