# 03/15/24 Morning Report with @CPSolvers

*"One life, so many dreams" **Case Presenter**: Jas Bajwa(@JasBajwa18) **Case Discussants**: Rabih Geha (@rabihmgeha)*

**CC**: 49yM w/ PMH of MI 2013 s/p PCI RCA and LCx, w/ evaluation for flu like symptoms **(November 2023)**

**HPI**: He had been in his usual state of health 8 days ago, at that time he developed unspecific flu like symptoms. Cough, nasal congestion, post-nasal drip and 3 non-bloody movements per day. Influenza, RSV, Covid test neg. New AKI (Crea 2.7 (bl 1.1), BUN 55, HypoNa 131, Plt 33k, Hb 6.6, Hct 18, Lactic acid 2.5). CXR: clear; acute knee pain R, XRay neg, no effusion; shoulder+ankle hurt, wrist, no sign of inflammation; LDH 271, high Haptoglobin. On Day 3: new fever 102°F, hypoxia (3l O2), Cef+Doxy, CT chest: minimal bibasilar opacity, (R>L) CT abd/pelvis: hepatosplenomegaly, mild sigmoid diverticulitis, intermittent fever, later encephalopathy, brain MRI: subacute lacunar infarct, Vanc+Cef+Flagyl+Acic started; HIV, RPR, EBV, CMV, Lyme, Ehrlichiosis, Babesia are negative.
UA: some RBC, no cast. Progressive dyspnea + peripheral edema. Echo: large pericardial effusion w/o tamponade physiology, prior EF 45% (now EF 25%)

**PMH**:
MI 2013 s/p PCI RCA and LCx

**Meds**: /

**Soc Hx**:
Works in health-care, gets routine Tb testing

**Health-Related Behaviors**:
No travel, no bird or bat exposure, no Tb exposure, MSM, protected intercourse, last time 3 months ago

**Notable Labs & Imaging**:

**Hematology**:
WBC: 5.1 (lymphopenia; Neutros predom) Hgb: multiple blood transfusions needed Plt: 30-50k

**Chemistry**:
B12+Folate wnl; Ferritin: 1154, D-dimer: >7000, Fibrinogen: 889, PTT: 60 and PT/INR: 16/1.4.
Pericardial effusion: 550 cc's of serous fluid drained
Pericardium: acute on chronic inflammation, cytology neg;
**PBS:** abnormal mononuclear cells, approximately 4% favoring blasts, abnormally segmented neutrophils, giant and hypogranular platelets and nucleated RBC's
ANA+, ANCA neg, Anti-cardiolipin neg, Anti-SSA/SSB neg, C4+C3 nl, RF + ACCPA nl, ESR 61, CRP 32; Anti-Jo nl , RNA Polymerase 3 nl,
WNV neg, IgG4 low, total IgG high, k/L low, kappa and lambda high, SPEP nl (no poly-or monoclonal gammopathy)
**BM biopsy: tp53** mutation, hypercellular marrow with multilineage dysplasia, increased blast forms (7% of marrow cellularity), some ring sideroblasts. **Flow cytometry of the bone marrow aspirate:** no monoclonal B cell population, no aberrant T cell population
Patient was unfortunately readmitted to the hospital in the beginning of **January 2024** due to N/V, non bloody diarrhea, abdominal pain and fever for the last two days. No sick contacts. **CT abdomen:** thickening of urinary bladder consistent w/ cystitis; mild proctitis, pancreatitis. PCT high, leukocytosis, hypotension, high Lactate (2.8). Zosyn started, IV fluids. CRP 294, ESR 91, IL2 10.000, Ferritin 5400 → 5600. CT images reviewed: Cystitis, Proctitis, myocarditis, Pancreatitis & hepatosplenomegaly that resolved during the next CT. Possible AML transformation? UBA1 for Vexas negative. BM biopsy: no evidence of HLH. Abdominal pain (some resolving organs + some new inflamed organs. **Repeat BM biopsy:** increased blasts from 8-10% and Increased histiocytes noted showing hemophagocytosis. PET scan 1 month prior: no uptake on lymph nodes. Genetic panel for 1° HLH neg.
**Final dx: HLH 2/2 high grade MDS**

**Problem Representation**: A 49yM w/ flu like symptoms, and later on intermittent fever, bicytopenia, lymphopenia, hypoxia, AKI, hepatosplenomegaly, high Fibrinogen, high PTT, hypercellular BM w/ increased blast forms (7%) and p53 mutation. Ferritin >5k, high sIL2R, high inflammation markers and migratory -itis of several organs. Repeat BM biopsy w/ increased blasts and histiocytes compatible w/ HLH.

**Teaching Points (Anmolpreet)**:

**I] Respiratory infections and diarrhea are interlinked,** mostly seen together; a cytokine like process is responsible for the bowel abnormalities;
Resp tract: problem; Gi symptoms: consequence

**II] Severity of labs with minimal symptoms** makes us think that the lab changes are subacute; eg: Hb of 6 acutely will make the patient really sick!

**III] 5Ps for AKI:**

1. **Pigment nephropathy** (CK),
2. **Paraprotein** (SPEP),
3. **Pharmacy** (drug induced),
4. **Pointy -** crystals (uric acid, ethylene glycol),
5. **Platelets**

**IV] To find out the core problem in so many symptoms is important; other could be just the manifestations of the body;s reaction** eg : encephalopathy as a result of multisystem ds; we should get a bone marrow biopsy given pancytopenias and pericardial tap to see if infected.

**V] Systemic imprint:** stroke history makes us think of vascular issue
**febrile, cytopenias with hepatosplenomegaly: CORE PROBLEM**

- Mononucleosis (EBV, CMV)
- Tick borne ds (Rickettsia)
- Granulomatous infection: more resp involvement (TB, Histo)

**VI] Cytopenia, abnormal marrow, coagulopathy : APML**→ can lead to intracerebral haemorrhage

**VII] Diagnosing a patient with HLH is like diagnosing a patient with neutrophilia!**
It just tells us that MAS is a part of the phenotype of the actual ds of the patient.
**Pt has biphasic HLH signaling autoimmune ds** - VEXAS (if we consider BM dysplasia → UBA1 gene testing)

**VIII]** Our differentials seeing the recurrent nature of the ds**: Cancer, lymphoma, multifocal Castleman's ds(absence of LAD reduces its possibility)**

**IX] MDS: 2 forms: 1. indolent (low grade) , 2. aggressive (high grade)**
HLH was secondary to MDS ; HLH can be primary or secondary(acquired)