7/8/20 Morning Report with @CPSolvers

***Case Presenter**: Imran Nizamuddin (@INizamuddinMD) **Case Discussants**: TJ La, Jr. (@TonLaJr) and Rob Weber (@RobWeberMDPhD)*

CC: fever, lymphadenopathy, pancytopenia
HPI: 27 Male w PMH of Hashimoto disease. He was admitted from clinic due to recent fever, lymphadenopathy and pancytopenia.

He has been feeling unwell for past 2 months. Fever 101 at home w poor intake and fatigue for a month. Was found at a neighboring hospital to have pancytopenia and diffuse lymphadenopathy.

Lymph node biopsy was negative for malignancy. Symptoms attributed to infectious mononucleosis. Also noted burning sensation of bilateral lower extremities and hands, as well as weakness requiring wheelchair use. He moved in with his parents due to inability to do ADL.

PMH: Hashimoto disease during childhood Controlled with diet.

Meds: None

Fam Hx: Father (HTA and DM) and
RA in maternal grandmother
Soc Hx: Now lives in Chicago. Worked and lived in northern California.
Health-Related Behaviors:
No ETOH, smoke. Marijuana vaping daily.
4 months ago camp in desert California. Denies bite.
1 female sex partner with intermittent condom use.
Allergies: None

Vitals: **T**: 100.4 **HR**: 91 **BP**:117/60 **RR**: **SpO_2**: 95% (room air)
Exam:
Gen: Appears chronically ill
HEENT: Moist oral mucosa, no oral lesions. No cervical lymph. Bilateral axillary and inguinal lymphadenopathy. Hard, non-tender
CV, **Pulm**, normal
Abd: Tender hepatomegaly, no splenomegaly
Neuro: CN II-XII Normal, ⅗ proximal strength of bilateral extremities and 5/5 distal extremities. Sensory and reflexes exam normal.
Extremities/Skin: Rash in peri-nasal area, that spared nasal fold, and rash in preauricular area. He had 4-5 erythematous papules and papules on anterior chest.

Notable Labs & Imaging:
Hematology:
WBC: 3.3 (62% neut, 17%lymph and 11% mono) Hgb: 8.6 Plt:74
Chemistry:
Na: 136 Cl:104 CO2: 25 BUN:19 Cr:0.61 glucose:101 Ca: 8.3
AST: 573 ALT: 172 Alk-P:162 T. Bili:5.8 DB:3.9 Albumin: 2.2
CK: 320 UA: Normal no WBC no RBC 2+ protein, ketones
Creatinine: normal Creatinine/ Protein: 2.5
HIV: Positive, but then confirmatory negative CD4: Normal
Peripheral smear: pancytopenia
Ferritin: 6000 TG:355
Bacterial and fungal tests: negative
ANA: 1:1280 dsDNA, anti- Ro: positive
Kidney biopsy: lupus nephropathy
Imaging:
CXR: Hepatomegaly, splenomegaly 18 cm spam, abdominal and bilateral inguinal 2x3 lymphadenopathies
Final dx: HLH secondary to SLE

Problem Representation: Young man with a history of Hashimoto's thyroiditis, presents with a subacute course of fevers, malaise, and weakness, found to have pancytopenia, proximal muscle weakness, and diffuse lymphadenopathy, as well as positive ANA, dsDNA, consistent with SLE c/b HLH.

Teaching Points (Jack):
Fever: An initial approach uses IMADE (infection, malignancy, autoimmune, drugs, DVT, and endocrinopathy).
Fever of Unknown Origin: When a fever persists for multiple weeks, we can classify it as an FUO. Infectious diseases (viral, bacterial, fungi, parasites), autoimmune diseases (most often SLE or a systemic vasculitis), and malignancies are the primary categories that cause FUO.
An approach to pancytopenia: The more cell lines that are depressed, the higher the likelihood of a bone marrow problem (e.g., decreased production of cell lines).
Neuropathy + Systemic Inflammation: Consider autoimmune diseases, such as lupus and a vasculitis, as well as infectious diseases, such as HIV or infectious complications of HIV.
Not all facial rashes = lupus: Seborrheic dermatitis can cause a facial rash and is common in HIV. Dermatomyositis can also have facial rashes (seborrheic dermatitis or heliotrope rash).
Severity of the illness frames the breadth of the workup: When someone has a slowly progressive illness, we have time to stage a workup. However, in rapidly progressive diseases, the urgency of a diagnosis and directed therapy warrants an extensive initial workup and potentially early invasive tests, even if serologic tests are available, due to the time some labs take to come back.
Antibody review: ANA titers can be positive in a number of autoimmune diseases (e.g., SLE, Hashimoto's) or in individuals without any autoimmune disease (titers often lower here). Anti-Ro antibody can be found in SLE as well as Sjogren's Syndrome. dsDNA is most often seen in SLE.
HLH: In children, it may be primary. In adults, often secondary to underlying infection, autoimmune disease, or malignancy.