11/6/20 Morning Report with @CPSolvers

Case Presenter: *Kiara Camacho (@kiaracamacho96)* ***Case Discussants***: *Rabih Geha (@rabihmgeha) and Reza Manesh (@DxRxEdu)*

CC: pleuritic chest pain

HPI: 19M, 3mo ago had pleural CP, c/w pleural effusion. Tx w Abx. WL 15kg, cough with expectoration, N/V, foamy urine, swollen legs. Since then, lost to follow up. Presented 1-2 months later and hospitalized with above Sx.

PMH: Asthma, controlled without meds

Meds:

Fam Hx:

Soc Hx: Lives in Iquiros, jungle part of Peru
Eng student
Health-Related Behaviors:

Allergies:

Vitals: **T**: **HR**: **BP**: **150/80** **RR**: **SpO_2**:
Exam:
Gen: not well appearing
HEENT: retroauricular, jaw, and perioral hyperpigmentation. Alopecia (present 3y prior) , epistaxis, pain in pharynx and erythematous
CV:
Pulm: slightly diminished sounds in lower R lung
Abd: pain on palpation, distended, liver 2-3cm below costal border, tympanic
Neuro:
Extremities/Skin: pale skin, pale stretch marks on abd; LEE

Notable Labs & Imaging:
Hematology:
WBC: **3.8** (N 72, L 20) Hgb: **8.2**, microcytic, hypochromic Plt: **110**
Fe **48**, transferrin **87**, TIBC **110.5** , transferrin sat- **43**, ferritin **2000**
Vit D: **7.5 (deficit <12)**
Chemistry:
Na: **126** K: 3.97 Cl: 97 CO2: BUN: **145** Cr: **4.47** (no baseline) glucose: 116 Ca: **6.3**; iCal 0.8 Phos: **9** (high)
TP: 4.3; Alb **1.89 ;** AST: ALT: Alk-P: **214** T. Bili: Albumin:
UA: **prot 3+, Hgb 2+, Leukocytes, RBC 28, 24h albumin- 7147m/24h (0-30), total prot 10.91g (0-0.15)**
TSH: **9**, T3: **0.82 (low mild)**
Imaging
CXR: R-middle lobe consolidation and slight pleural effusion
Bone marrow Bx: hemophagocytic cells
Neg: IgA-IgE, cANCA, pANCA, VDRL, anti-Rho/La, Coombs, dsDNA, EBV, TB, Histo/parasites, HTLV1, CMV, antiCCP, C1Q, antiPhospholipid, anti-cardiolipin, anti-B2 glycoprotein, varicella, cortisol Red congo stain - negative
Positive:
ANA: 1/160, **ssDNA**>200, **SPEP-** total prot 3.85, **Alb** 1.65→ hypogammaglobulinemia, **Alpha 1/2 increased,** Alb/glob inversion , **fibrinogen increased 517**, **B2 microglobulin 25 (high)**; Serum K/L chains 9.7 (high), Haptoglobin (high), Urine L high, K nl, **complement low C3: 38/C4: 9.6, TG 260 (high)**, CH50: 13 (low)
U/S - HM, SM, large kidneys; **CT**: pancreas increased in size, normalized within 1 mo hospital course
Final Dx per biopsy: lupus nephritis.

Problem Representation: 19M p/w subacute pleuritic CP, wt loss, alopecia, hyperpigmentation w/ labs c/w HLH, nephrotic syn, +ssDNA, ANA and final dx per renal bx as lupus nephritis.

Teaching Points (Sukriti):
Investigating a Sx of pleuritic chest pain
Pleuritic chest pain -- Think of the structures that the lung makes contact with: Chest wall (irritation of the pleura, costochondritis), Mediastinum (pericarditis), Diaphragm (hepatic abscess, splenic infarct)
Collecting clues: Wt loss + volume overload -- suggests a sinister etiology
Wt loss = inflammation - IMADE mnemonic -- **Inflammatory pleural effusion** (Infection, Malignancy, Autoimmune, Others - GI, ovarian)
Volume overload - heart, kidney, lung -- **New onset HTN + volume overload** -- **Kidney** > Heart, liver
Framing a hypothesis: Volume overload + foamy urine + hyperpigmentation + alopecia
Foamy urine - proteinuria
Hyperpigmentation: primary adrenal insufficiency, heavy metals, acanthosis nigricans
Alopecia in a young adult - Base rate of physiologic cause low, pattern impt. To identify pathology -- think autoimmune
Testing the hypothesis: Nephrotic syndrome + HLH + Hepatosplenomegaly + ANA+ ssDNA
Pearl: Nephrotic syndrome is not equal nephrotic range proteinuria -- triad of LE edema, nephrotic range proteinuria, hypoalbuminemia
Complications: Immune deficiency, thrombosis, hyperlipidemia
Nephrotic syndrome -- Histologic types; base rate for primary cause highest in membranous nephropathy PLA2 receptor Ab (80%)
Working hypothesis: Pancytopenia + hemophagocytic cells = HLH - 1 Cause explains all: Peripheral destruction: AI (lupus), Infection (tick borne), Malignancy (CLL), spleen
Hepatomegaly + splenomegaly = cells > water, molecules -- iMADE
CRP: Solid tumors cause nephrotic MC, do not go the spleen