



7/8/25 Morning Report with @CPSolvers

"One life, so many dreams" Case Presenter: Zakariyya Case Discussants: Dr. Daniel Restrepo



Scribing (Sam B)
CC: 59F, hx Sjogren's on hydroxychloroquine with sudden onset vision loss
HPI: 3 weeks ago sister noticed she was breathing harder than usual, mild wheezing, similar to asthma exacerbations
 2 weeks ago saw PCP, given 5 day prednisone taper and azithromycin, mild nonproductive cough.
 3 days ago had bloody sputum, hit her head and developed subsequent headache and dizziness
Sudden onset of blurry vision
ROS: Diarrhea, nausea, arthralgia, fatigue, myalgia, night sweats, hemoptysis
 Pain in shoulders, arms, thighs, improved on prednisone. No fevers, chills, weight loss, Raynaud, dry mouth, eye pain, eye redness, sore throat

PMH:
 Asthma dx childhood
 Sjogren's
 Migraines: nausea + dizziness, no aura
 Dry eyes
 +ANA
 Hypothyroidism
 HTN
Meds:
 Hydroxychloroquine started 1 month ago
 Hydralazine
 Lisinopril
 Sumatriptan
 Celecoxib
 Levothyroxine
 Restasis eye drops

Fam Hx:
 Mom hypothyroid
 S/p hysterectomy

Social Hx:

Health-Related Behaviors:
 No alcohol, cigarette, drug use

Allergies: Denied

Vitals: T: 102.6 BP:128/78 HR:118 RR: Sat: 84% RA
Exam: HEENT: No redness, ulcers; decreased central vision, palpable non tender cervical lymphadenopathy.
Ophtho: macular edema, cotton wool spots, retinal hemorrhages, flecken consistent with Purtscher-like retinopathy bilaterally
CV: tachycardic, no murmur
Pulm: diffuse fine crackles throughout
Neuro: nl **Skin:** nl **MSK:** Synovitis of wrists L > R, axillary lymphadenopathy

Notable Labs & Imaging: **Hematology:** WBC:7 Hgb: 9 HCT: 29 Plt:122 MCV: 94
Chemistry: Na: 135 K: 3.7 Cr: 2.4 BUN: 56 Glu: 104 Cl: 103 LDH: 520 AST: 73 ALT: 92 ALP: 190 Total pr: 7.5 Alb: 2.2 CK 156 Haptoglobin undetectable
 TSH 4.0 T4 0.65 (L) +schistocytes, +rhinovirus, +ANA 1:2560, SSA, SSB, IgG4H 866, Ferritin 866, ESR 86, CRP 25
 UA: 1+ protein, 1+ blood, Pr/Cr ratio 2.2, granular casts consistent with ATN
 Negative: HIV, hep ABC, EBV, CMV, parvo, lyme, mycoplasma, RMSF, syphilis, quant gold, histoplasma, AFB culture, fungal culture
 Negative: dsDNA, Sm, RNP, SCL70, histone, Jo1, C3/4, RF, CCP, lupus anticoagulant, anticardiolipin, anti beta-2-glycoprotein, RNA pol3, ANCA, MPO, PR3
 SPEP: polyclonal hypergammaglobulinemia FLC ratio nl IL6 undetectably high
 VEGF very high
Imaging: TTE negative
 CT chest: bilateral ground glass opacities and patchy consolidation with peribronchovascular thickening, mediastinal+hilar lymphadenopathy, small/moderate bilateral pleural effusions
 CT abd: bulky abdominal lymphadenopathy, mild hepatosplenomegaly
Tissue: FNA: Nonspecific inflammation, no atypical or large lymphocytes. Some nodes completely effaced by inflammatory infiltrate, mostly plasma cells, nonspecific. Stains negative for infectious cause including viral, bacteria, AFB, fungal.
 BAL: Negative viral, bacterial, AFB, fungal cultures, negative cytology and flow
 Axillary LN excisional biopsy: marked sheet like interfollicular plasmacytosis, increased vascularity and follicular dendritic cell prominence, nonspecific. Stains for HIV, HSV, EBV, HHV8, AFB, fungal negative. Plasma cells and lymphocytes are polyclonal. No kappa/lambda restriction. Increased IgG4 cells, ratio of IgG4/IgG lower than 40%. No aberrant t-cell population.
 Bone marrow bx: Mildly hypercellular marrow, increased dysmorphic megakaryocytes with hyperplasia of megakaryocytes, mild signs of myelofibrosis, no HLH, no Kappa/lambda restriction, no clonal B/T /plasma on flow
 Renal bx: glomerular microthrombi consistent with TMA, immunofluorescence negative for immune complex deposition of immunoglobulin or complement
Course: Early empiric high dose steroids for ?GCA. Given tocilizumab with complete resolution of symptoms
Dx: Idiopathic multicentric Castleman's disease, NOS subtype (autoimmune overlap)
 Mixed features of plasma cell and TAFRO variants (thrombocytopenia, anasarca, reticulin fibrosis, renal dysfxn, organomegaly)

Problem Representation: 59yoF with history of autoimmune disease with fever, B symptoms, diffuse lymphadenopathy, bicytopenia, elevated inflammatory markers, negative infectious and malignant workup

Teaching Points (Saketh V):

- History of autoimmune disease should trigger thoughts about other autoimmune disorders/vasculitis/medication history (Steroids/Hcq) for the primary disorder that would lead to the current presentation.
 Response to steroids --> signal for steroid response disorders - PMR
- New onset asthma in adulthood - should also rule out CHF/eosinophilic disorders/vocal cord disorders
- Drugs:
 - Hcq --> Can cause retinopathy
 - Hydralazine --> Associated with drug induced lupus and ANCA associated vasculitis.
- Labs: Hemolysis (Intra/Extravascular, Immune/Non-immune) With many extra articular manifestations and Generalised LAD: prioritize CTD, Vasculitis, Malignancies, Lymphoproliferative dz, Chronic Inflammatory Dx
 Elevated gamma globulins: consider HIV,HBV,HCV, IgG4 dz
- Can use PLASMIC Score before the labs come back to predict ADAMTS13 deficiency in suspected TTP
- Idiopathic multicentric Castleman's Dz: NOS Subtype (autoimmune overlap):**
When should you suspect it :
 Disease looks like a Rheumatologic Dz (Positive Serologies)

- High Fever and B Symptoms
- Diffuse LAD
- Cytopenias

Labs: High IL 6 Levels, CRP
 TAFRO Syndrome: TCP,Anemia, Reticulin Fibrosis, Renal Dysfunction and organomegaly -> clue to Castleman Dz