12/02/24 Rafael Medina Subspecialty VMR with @CPSolvers

"One life, so many dreams" Case Presenter: Franco Murillo () Case Discussants: Brendan Denvir (@), Dr. Michael Cammarata (@)

CC: periorbital swelling and joint pain

HPI:
77 year old presented with 2 weeks of left periorbital swelling. He reports that 1 month ago has URI(self-limited productive cough, hoarseness and intermittent fever). At that time, he went to his PCP and got a prescription for azithromycin and prednisone. His symptoms resolved shortly after however 2 weeks before admission he reported left orbital swelling and bilateral wrist and right knee pain and recurrence of fevers

PMH:
Pemphigus vulgaris
rituximab x4 , remission

Meds:
Atorvastatin
tylenol

Fam Hx:
no
Soc Hx:
no
Health-Related Behaviors: **no**

Allergies: none

Vitals: T: 39 BP: 130/70 **RR:**18 **Sat: 96 HR:**95
Exam: Gen: not ill appearing
HEENT: left periorbital edema, no oral ulcers, redness around eyelid
CV, Pulm: non significant
Abd: nontender, nondistended
Neuro: AOX4, left foot drop, no paresthesias
Skin:no rash, no plaques
MSK: no synovitis, pain with flexion and extension, no effusion

Notable Labs & Imaging:
Hematology:
WBC: 13K Hgb: 7 Plt:187k Hct: MCV:101
Neut 88% eosinophils 0%, Lymph 4.3%
Chemistry
Cr 3.4 (0.8), BUN 67 AST, ALT NL
Bili normal; ALP normal
ESR 104 CRP 8
Troponin :8
TIBC:160, transferrin 128 ferritin 400 Iron 23
b12>2000
Total protein 7.7 Alb 3
UA: trace protein on dipstick, no rbc, no wbc
ANA 1:320 dsDNA:neg
Anti CCP, Rheum Factor - neg
c ANCA 1/40 P-ANCA : neg
ELISA anti PR3: neg/Anti-MPO: neg
Imaging:
CT chest: bilateral lung nodules with bilateral pleural effusions
CT Abd/plv:unremarkable
MRI orbit: soft tissue swelling around left periorbital area
Kidney bx: glomerulosclerosis, tubulointerstitial scarring, negative IF, no evidence of vasculitis

Dx: VEXAS syndrome

Problem Representation:
77 year old male presented with 2 weeks of left periorbital swelling who was found to have VEXAS

Teaching Points (Dan):
-Initial approach to periorbital swelling: Focus on eye. Redness? Intact EOM? CN intact? Vision? Consider involving Optho colleagues. Initial ddx: Infection? GPA vs IGG4-related disease?
-Arthralgia: joint pain (e.g. viral illness w/ joint ache) vs. Arthritis: pathology of joint
- Serum sickness associated with rituximab (chimeric mAb)
- Foot Drop ddx: consider Cryoglobulinemia vs ANCA-associated vasculitides vs SLE/RA. Consider EMG/NCS w/ muscle bx.
- Anemia + Elevated MCV: consider liver disease, B12, Folate, & consider VEXAS
- Urine protein:creatinine ratio to better characterize proteinuria
- ANCA: test of indirect immunofluorescence
- Rituximab: anti-B cell therapy. Measuring anti-PR3 and c-ANCA may be less helpful (more likely to have lower levels of Abs)
- Generally look for concordance between c-ANCA and anti-PR3 vs p-ANCA and anti-MPO
- GPA in Kidneys: crescentic, necrotizing glomerulonephritis which can be focal & segmental
- Diffuse glomerulosclerosis seen on kidney bx suggests chronic process
- Final ddx: infection vs GPA vs VEXAS vs underlying malignancy
- VEXAS: 10-20% of population with mutation can develop vasculitis
- Cytopenias in vasculitis are less common. Typically see normocytic anemia w/ thrombocytosis.
- Macrocytosis or thrombocytopenia w/ relapsing polychondritis: increased pre-test probability for VEXAS (happens in old males)