



# 8/8/24 Morning Report with @CPSolvers



“One life, so many dreams” Case Presenter: Mukund Raguram Case Discussants: Lea (@xLea\_B) and Rabih (@rabihmgeha)

**CC:** R lower extremity pain

**HPI:** 84 yo male p/w fever, chills and weakness; 1 week ago: diverticulitis c/p abscess treated w/ ceftriaxone & metronidazol; this morning: fall resulted in R lower extremity pain, denied any recent trauma

Outside hospital  
Ceftriaxone for cellulitis w/n improvement  
Biopsy of the leg: no evidence for vasculitis  
Current Hospital:  
Skin lesion progressed to necrotic lesion w/ blisters -> surgical debridement, treated w/ Vancomycin + Zosyn with no improvement

**PMH:** HT, DM II, CKD, PAD w/o revascularisation, unprovoked DVT, HFpEF, knee replacement 2/2 severe osteoarthritis, chronic inflammatory arthritis, VEXAS

**Meds:**  
Apixaban  
Ruxolitinib  
Prednison  
Bactrim + Acyclovir  
Insulin  
Tocilizumab (IL-6 ab)

**Fam Hx:**  
nil  
**Soc Hx:**  
Lives independently

**Health-Related Behaviors:**  
smokes cigarettes 36 pack years, 2 glasses wine daily

**Allergies:**

**Vitals:** T: 38.1C HR: 106 BP:154/67 RR: 22

**Exam:** Gen:  
HEENT, CV, Pulm, Abd, Neuro: wnl  
**Extremities/skin:** redness and swelling from the upper calf all the way to the ankle - tender to palpation, no open wounds

**Notable Labs & Imaging:**  
**Hematology:**  
**Chronic Pancytopenia** at the outside hospital  
WBC: 2.6, neutrophilic predominance, Hgb: 9.4 Plt: 45

**Chemistry:**  
Cr: 1.53 CRP 15.3 ESR 25  
BNP nl, iron 18, Transferrin 152, TIBC 152, TSAT 12%, ferritin 861  
Bilirubin 0.8 , LDH 265 (nl<243), B12 1002, folate nl  
Coag nl Ret 1.8 % inappropriate nl  
**Blood smear:** bands, bilobed neutrophils, elliptocytes, dacrocytes, nl platelet  
**Infectious panel:** HIV, Hep, Covid, res. Viral panel: nl  
MRSA neg,  
BK neg (4d)

**Imaging:**  
**MRI RL extremity:** nonspecific edema w/o osteomyelitis, fluid collection

**Derm biopsy:** inflam. Infiltration w/ rare neutrophils with variable plasma cells, mycobacteria neg. ; Bactrim resistant MRSA

**Final Dx:** VEXAS associated neutrophilic dermatosis

**Problem Representation:** 84 y/o male p/w chronic hematological condition 2/2 VEXAS syndrome w/systemic features (pancytopenia + neutropenic fever) cb LE swelling w/ necrotic skin lesion.

**Teaching Points (Elena):**

- **Unilateral leg pain:** vascular (DVT, arterial), trauma, skin/soft tissue (cellulitis), neuropathy -> let's look at it and let's find out how deep it is (is the bone, the muscle, the fascia involved)
- no miss: invasive fungal infections (esp. in the context of neutropenia) limb ischemia - is not ruled out by present pulses, sensation or motor function
- Recent hospitalisation + prominent systemic features (is the limb finding a signature of a systemic disease or is an independent finding)
- **VEXAS:** rheum-heme autoinflammatory disease (somatic, X linked UBA1 mutation), male>women
  - **Heme:** macrocytosis (95%), paraproteinemia (25% vs 1%), some have eosinophilia or a myeloproliferative phenotype, MDS (transformation to AML)
  - **Rheum:** surface (centered around the face- oral ulcerations, ear, eyes, urticaria esp. in the context of eosinophilia vs visceral (variable vessel vasculitis, PNA, ultra hypercoagulability - like Ruxolitinib; ILD)
  - **The same concept** of visceral vs surface can be used in Behcet - another inflammatory condition that typically entails folliculitis, acneiform rash instead of neutrophilic dermatosis; hypercoagulability
- Restriction to the calf (w/o foot involvement) decreases the likelihood of limb ischemia
- **Pancytopenia:** destruction < production (meds, nutrition, infiltration) → Abx failure category (microbiology) - labs: inflammation, low retics (production), left shift
- **Space occupying BM processes:** Fibrosis, Granulomatous diseases, cancer
- **Empiric Tx w/o diagnosis:** sepsis, decomp. cirrhosis, rigoring (60-70% likelihood of bacteremia), febrile neutropenia (watch out for angioinvasive fungal infections, think about the molds)
- Necrotic progressive ulcerative lesion in the context of chronic pancytopenia that is unresponsive to treatment and worsens due to manipulation increases the likelihood of a neutrophilic dermatosis (“pathergy”) - r/o a hiding angioinvasive fungal Infx before you give steroids