6/19/23 Rafael Medina Subspecialty VMR with @CPSolvers

"One life, so many dreams" Case Presenter: Daniel Motta-Calderon (@dmottacalderon) Case Discussants: Kevin Hageman (@Factor_XII)

CC: 79 yo man with AMS and rectal bleeding

HPI: Transferred to the hospital from an assisted living facility five days ago for urinary retention, AMS, hematuria. Patient also has bruises and epistaxis.
One day prior to admission, patient developed painful blisters in the lower extremities. Also developed painless hematochezia, slurred speech and left facial droop.

PMH:
Dementia
GERD
HTN
HLD
Prostate CA
Sacral Ulcers
Osteomyelitis
Diabetes
Previous aspiration episodes

Meds:
Vancomycin
Memantin
Metformin
Mirtazapine
Allopurinol
Doxazosin
Iron

Soc Hx:
retired veteran

Allergies:
NKDA

Vitals: T: 35.1 HR: 65 **BP**: 133/111 **RR:** 15 **SpO2:** 99% on room air
Weight: 270 pounds
Gen: Only oriented to place;
Pulm: normal resp excursion, speaking full sentences, no accessory muscle use; PEG tube.
Extremities/skin: +1 edema,; fluid tense bullae (w/ serous fluid) about 7cm, no surrounding erythema, 1 bullae already popped; , 7x7cm ulcer on sacrum w/ granulation tissue, no signs of local inflammation or purulence, no oral involvement
Genitals: oozing in urethra. Foley catheter in place
Rectal: oozing blood from the rectum. Anal fissure

Hematology & Chemistry
WBC: 11.7 - Differential: Eosinophils: 3100 Hgb: 6.6 HCT: 33% Plt: 388; Hgb 6.6 -> improved to 7.7 after transfusion.
CMP normal
aPTT: 107 INR: 1.1 Repeat PTT: 95
PT 14.6 Fibrinogen: 383 TSH: 0.9 B12 high
Imaging:
CT/MRI of the head: normal
Six hours after admission: patient was tachycardic and hypotensive. Massive transfusion protocol was started. Patient received fibrinogen and PCC.
CT Abdomen/Pelvis: extensive colonic diverticulosis (no diverticulitis),
Angiogram abdomen/pelvis: no contrast extravasation.
Patient continued to deteriorate and received pressors
Mixing study: Did not correct well, suggestive of inhibitor
Red blood cell tagged scan: extravasation in the LLQ in the sigmoid colon
Colonoscopy: multiple diverticula
IgA/IgG/IgM anticardiolipin & b2-microglobulin: negative Lupus anticoagulant: positive.
Factor VIII activity: 2% (low); Factor VIII inhibitor: positive

Final Dx: Acquired factor VIII inhibitor.

Treatment course: patient was started on high dose steroids and rituximab, but quickly developed pseudomonas infection. Patient was transitioned to comfort measures
Patient received FEIBRA and KCENTRA and his bleeding improved.

Problem Representation: 79yM w/ chronic OM, p/w AMS, diffuse bleeding and tense bullae. Labs notable for eosinophilia, prolonged ptt. Mixing study suggestive of factor inhibitor.

Teaching Points (Bettina):

- **AMS:** Structural pathology in the CNS (hematoma), infection (pneumonia, bacteremia, UTI, PEG tube cellulitis), uremia (lower GI bleed, renal failure), bleeding diathesis (coag disorder, thrombocytopenia, vitamin C deficiency), metabolic, toxins
- **Blisters:** Bullous pemphigoid (below BM on IF) vs. pemphigus vulgaris, vasculitis, edema bullae, IgA bullous dermatosis (vancomycin)
  - Skin biopsy can be done
  - BP can be associated with malignancy
- **Hypothermia:** Infectious process (check for hypotension + tachypnea)
  - Dementia patients on AChE inhibitors (donepezil) can present with bradycardia so check medications
- **Bleeding:** DIC, extrinsic vs. intrinsic vs. terminal pathway, platelet dysfunction, GI bleeding, acquired coagulopathy
  - **Prolonged PTT:** heparin/enoxaparin, lupus anticoagulant, vWF deficiency (vWF carries FVIII), factor VIII inhibitor
    - Do mixing study and see if it corrects
  - Do tagged red cell scan if unsure about bleeding source
  - CT angio can be done for hemodynamically unstable
  - Coagulopathy will manifest as bleeding in a patient with risk factors (ie, diverticular bleed is usually controlled in a patient w/o coagulopathy)
  - Prioritize stabilization of the patient while exploring underlying dx
- **Eosinophilia:** Hypereosinophilic syndrome with multiorgan infiltration, marrow process (myeloproliferative neoplasm with factor deficiencies), hypersensitivity reaction (allopurinol), parasitic
- **Markedly elevated B12** → BM malignancy, hypereosinophilic syndrome
- Eosinophilia + bleeding → think of BM malignancy, autoimmune
- **Acquired FVIII inhibitor:** Rituximab + steroids, extremely rare (1-2 cases per million), can acquire remission in 2 months, associated with underlying malignancy or rheumatologic diseases