07/12/21 Morning Report with @CPSolvers

***Case Presenter**: Travis Smith (@RosenelliEM) **Case Discussants**: Ravi Patel and Dhruv Srinivasachar (@TheRealDSrini)*

CC: Bleeding.

HPI: 54 year old man with a recent diagnosis of bullous pemphigoid (3 months) presents with new bleeding from his blisters. Bleeding started 2 days ago, oozing from wounds, no increase in pain. Diagnosis of pemphigoid was done by biopsy, was given oral steroids and lesions improved. By that time there was no bleeding. No petechiae, bleeding, ulcers. No joint swelling or stiffness.

Vitals: **T**: Afebrile **HR**: 80 **BP**: 120/70 **RR**: **SpO_2**: Normal.
Exam:
Gen: Alert, no acute distress, oriented.
HEENT: Normal.
CV: Normal.
Pulm: Normal.
Abd: Normal.
Neuro: Normal.
Extremities/Skin: Multiple skin lesions on his chest, lower extremities. Some scabs, some unroofed. Oozing on most lesions. No petechiae, bruising. Some of the blisters looked hemorrhagic.

PMH: None

Meds:
None.

Fam Hx: None

Soc Hx:
No alcohol, tobacco or drug use.

Health-Related Behaviors:
None

Allergies: None

Notable Labs & Imaging:
Hematology:
WBC: 6 (Normal differential) Hgb:10 Plt: 240

Chemistry:
Normal metabolic and liver panel.
INR 1.1, PTT 88.
Mixing study did not correct. Lupus anticoagulant negative. Von Willebrand factor negative. Factor VIII 3%

Bethesda Assay: Factor titer.

Final Dx: Factor VIII Acquired Hemophilia A. Treatment with rituximab, bleeding and blisters improved.

Problem Representation: 54M with a 3 month-diagnosis of bullous pemphigoid, presents with new onset bleeding from his blisters. Laboratory were remarkable for a mild anemia and prolonged PTT.

Teaching Points (Vale):

- **Bullous Pemphigoid:** Autoimmune skin condition that causes large fluid-filled blisters, often in flexor areas. More common in older adults. IgG antibodies against hemidesmosomes.
- **Looking for clues:** Autoimmune goes with autoimmune: Rule out other coexisting conditions. Is this bleeding out of proportion to disease progression? (side effects from treatment? complication?)
- **New bleeding:** Damage to vessels (trauma, vitamin-related, vasculitis) vs Coagulation pathway (1ry-Platelets; 2ry Coagulation cascade PT/INR, von Willebrand, Acquired Hemophilia).
  - Bleeding in gums or mucosas and presence of petechiae clues us to platelet disorder.
  - Bleeding in articulations and bruising clues us to coagulation cascade, but the absence of hemarthrosis does not rule out acquired factor inhibitor.
- **PTT Prolongation:** Intrinsic Pathway. Coagulation factor deficiency (VIII is the most common).
  - Factor VIII is not synthesized by the liver.
  - Factor V helps rule out vitamin K deficiency.
- **Mixing studies:** Helps differentiate antibodies (PTT does not correct) from coagulation factor deficiency (corrects).
- Drugs that cause Bullous Pemphigoid-like lesions: Furosemide, spironolactone, NSAIDs, amoxicillin, PD-1/PD-L1 inhibitors, Gliptins and TNF-alpha inhibitors.
- **Acquired Hemophilia:** Associated with pregnancy, LES, medications.
  - APS can present with acquired hemophilia, but rarely bleeding.