Painless Hematuria: A Case of Acquired Hemophilia A

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Introduction

- Acquired hemophilia A (AHA) is a rare disease with 1.3 to 1.5 cases per million per year.

Case Presentation

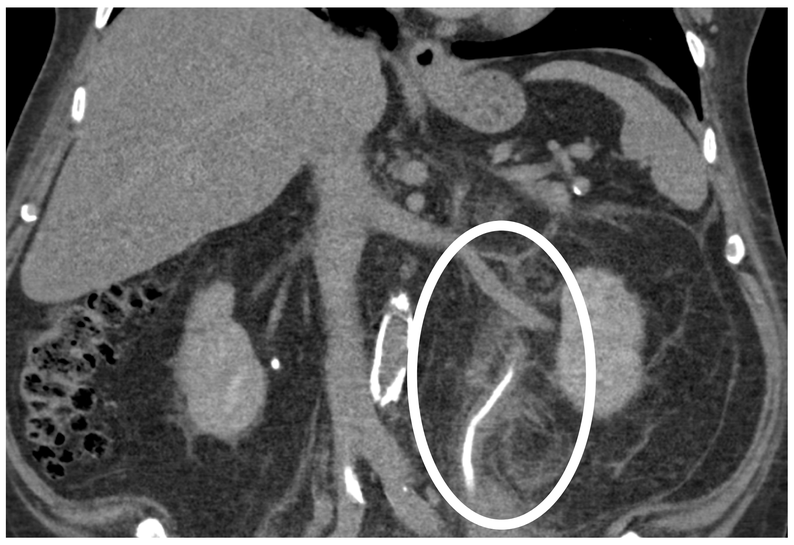
HPI:

- A 74 year-old man with prior psoriasis (not on immunosuppression) presents with gross hematuria, malaise, lethargy, left flank ecchymoses, and left groin pain.

Labs / Imaging:

- **Hgb 8.7 g/dL (L)** ← 13.4 prior.
- **aPTT 90.5 seconds (H)**
- INR 1.2
- ESR 94 mm/hr (H)
- CRP 162.8 mg/dL (H)
- Fibrinogen 594 mg/dL
- Haptoglobin 282 mg/dL (H)
- LDH 124 IU/L (L).
- Urinalysis: >180 RBC (H)
- **Factor VIII Activity: 3% (L)**
  - Ref: 0.6-1.5%
- **Factor VIII Inhibitor: 5.8 Bethesda Units (H)**
  - Ref: <0.6 Bethesda Units
- Von Willebrand Disease Panel: negative.

Figure 1. CT Abdomen/Pelvis showing edematous stranding of left ureter and collecting system — infiltrative process.



Clinical Course

- Initial Treatment Course:
  - **Daily high-dose prednisone.**
  - Received **rituximab** 1.0 grams IV x2 doses.
  - Blood transfusions as needed.
- Complications:
  - Left lower extremity weakness developed on hospital day #6.
  - Asymmetric **left retroperitoneal iliopsoas** and **medial adductor compartment hemorrhage**.
  - Treatment:
    - **Activated Factor VII**
    - Tranexamic Acid

Figure 2. Trend of Factor VIII activity level and Factor VIII inhibitor level before and after treatment. * = normal inhibitor level / ** = normal activity level.

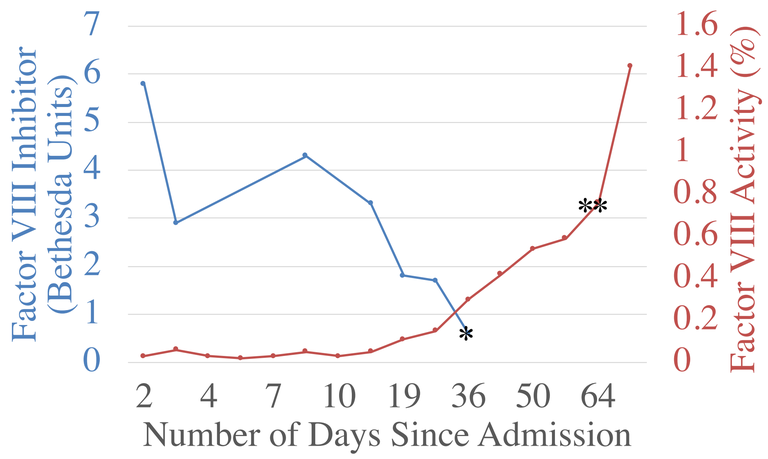
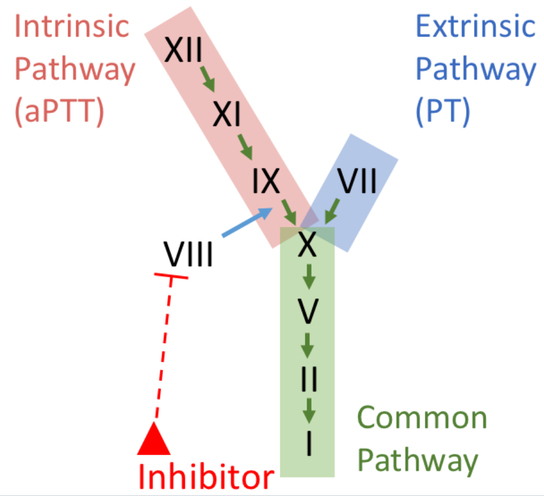


Figure 3. Simplified coagulation cascade with depicted Factor VIII inhibitor.



Discussion

- AHA Etiologies:
  - **Idiopathic** (51.9%), autoimmune diseases, prior/current **malignancy** (solid > hematologic), medications (antibiotics, clopidogrel, interferon, NSAIDs, amiodarone, heparin), monoclonal gammopathy of unknown significance, dermatologic disorders (i.e., **psoriasis**).
- AHA Presentation:
  - **Spontaneous bleeding**, often subcutaneous, deep muscle, or retroperitoneal bleeding. **Rare hemarthroses**.
- Treatment of Active Bleeding in AHA:
  - DDAVP
  - Factor VIII concentrates
  - Activated recombinant human Factor VII.
- Long-Term Treatment of AHA:
  - Glucocorticoids + Rituximab or Cyclophosphamide
- Relapse in 20% even after normalization.

Teaching Points

- Work up elevated aPTT with fibrinogen, LDH, D-dimer, mixing study, von Willebrand Disease panel, factor VIII inhibitor and activity levels.
- Monitor for signs of acute thrombosis at bleeding sites when using activated human recombinant factor VII to treat active bleeding.

References

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