Rapid Onset Acquired Hemophilia A Causing Spontaneous Retroperitoneal Bleed in a Patient Newly Diagnosed with Bullous Pemphigoid

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Introduction

- Acquired hemophilia A (AHA) is a rare bleeding disorder caused by spontaneous production of circulating autoantibodies against coagulation factor VIII and can present with a potentially life-threatening hemorrhage
- AHA is associated with autoimmune diseases, malignancy, pregnancy and certain medications, however, it is idiopathic in approximately 50% of cases(1)

Case Presentation

- A 77-year-old female presented with a two-day history of rapidly progressing severe tense bullous lesions extending over bilateral extremities, trunk and vaginal mucosa
- Skin biopsy revealed a clear linear pattern with deposition of IgG, C3 and C5B-9 on direct immunofluorescence suggestive of bullous pemphigoid. This was later confirmed by elevated Anti-BP180 and Anti-BPAG230 antibodies.
- She was started on doxycycline and intravenous steroids, which led to significant improvement in lesions
- However, the next day she developed sudden flank pain. After a few hours, she became hemodynamically unstable and required vasopressors.
- Serial labs revealed down trending hemoglobin, CT abdomen showed retroperitoneal hematoma (a) and hemoperitoneum. She was given aggressive fluid resuscitation as per massive transfusion protocol and multiple units of fresh, frozen plasma.
- Despite these measures, she remained hemodynamically unstable, and a repeat CT scan showed interval increase in the size of a hematoma (b)
- She eventually needed IR embolization and was found to have a bleeding right lumbar artery
- Remaining lab work showed decreased factor VIII assay 22% and elevated partial thromboplastin time at 63 seconds
- The mixing study failed to correct aPTT, indicating the patient had a factor inhibitor in the intrinsic pathway
- Patient's hospital course was further complicated by bacterial sepsis, ultimately resulting in her death

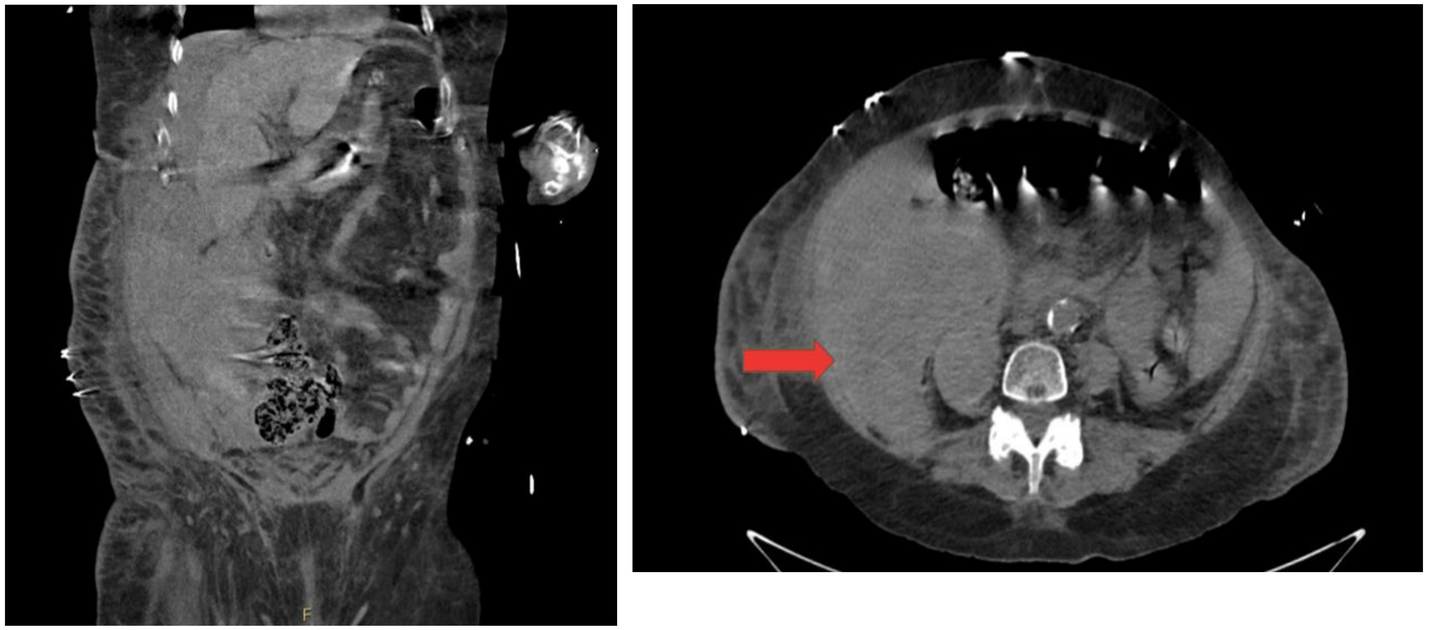

Figure a and Figure b – CT Abdomen showing massive retroperitoneal hemorrhage

Discussion

- Bullous pemphigoid (BP) is an autoimmune bullous disease characterized by autoantibodies against hemidesmosome proteins of skin and mucous membranes
- AHA is a rare complication of BP, postulated to develop due to cross reactivity of autoantibodies
- Patients developing AHA can have rapid clinical deterioration due to life threatening complications like massive hemorrhage or airway obstruction, and the mortality rate of AHA resulting from severe bleeding is estimated to be 7.9%-22.2%(2)
- Typically, AHA occurs a few months after the diagnosis of BP, but there have been rare instances when it developed rapidly within days (as in this case) or even concomitantly with BP. Hence, AHA should be suspected in BP patients who develop signs of bleeding at any point, as early diagnosis and initiation of treatment are important.
- Bleeding control and elimination of autoantibodies with immunosuppressive drugs are the mainstay of treatment, however, prognosis depends on the severity of hemorrhagic complications and patient's response to immunosuppression therapy

Conclusion

- AHA should be high in the differential in patients with BP who develop signs of bleeding and they should be promptly evaluated in a timely manner in order to prevent life-threatening complications

References

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This project is supported by the Health Resources and Services Administration (HRSA) of the U.S. Department of Health and Human Services (HHS) as part of awards totaling $17,038,990 with 63% percent financed with non-governmental sources. The contents are those of the author(s) and do not necessarily represent the official views of, nor an endorsement, by HRSA, HHS, or the U.S. Government. For more information, please visit HRSA.gov.