Hemophilia A is not a disease just for kids
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Introduction
- Acquired factor VIII inhibitors (also referred as Acquired Hemophilia A) is a rare bleeding diathesis caused by autoantibodies directed against clotting factor VIII.
- It is associated with bleeding involving skin, soft tissues, and muscles (as opposed to hemarthroses in congenital factor VIII def.)
- It is known to cause significant morbidity and mortality with mortality rates reported to be 8-22%.
- We report on a case of a patient with an acquired factor VIII inhibitor.

Case Presentation
- A 79 year old Caucasian man presented to the emergency room with complaints of right hip pain and left elbow pain for 2 weeks after falling. The patient stated he tripped causing him to fall on his right hip and used his left arm to break his fall.
- On physical examination, he had a large edematous area with significant ecchymosis over his right gluteal area and left elbow, which were tender to palpation (figures A and B respectively).
- Initial labs showed he was anemic with a Hgb of 6.2g/dL and MCV of 90fL, with a normal platelet count of 300K/µL.
- Coagulation studies revealed a PT of 11.2s, INR of 1.0, and a PTT significantly elevated at 113s.
- A CT scan of the pelvis showed a large right sided retroperitoneal hematoma and subcutaneous hematoma (figures C and D respectively).
- The patient was unaware of any bleeding disorders in his or his families past medical history.
- D-dimer and fibrinogen both came back elevated at 3.4mg/L and 560mg/dL, so DIC seemed less likely.
- After being transfused with 2 units of pRBCs his Hgb initially responded to 8.9g/dL but a day later went back down to 7.3g/dL.
- He received fresh frozen plasma and prothrombin complex concentrate on two separate occasions, which each improved his PTT for one day.

Hospital Course
- A mixing study was done which did not correct.
- Further coagulation studies were sent and came back showing a significantly low Factor VIII activity level of 8IU/dL and an elevated Factor VIII inhibitor level of 28BU/mL. This confirmed a diagnosis of an acquired factor VIII inhibitor.
- He was started on rituximab and received recombinant factor VIIIa after a bleeding episode.
- His PTT did begin to improve and dropped as low as 71s; however, he became unstable after developing abdominal compartment syndrome from the large pelvic hematoma and went into cardiopulmonary arrest and passed away prior to hospice care placement.

Discussion
- Differential diagnosis of a prolonged PTT with normal PT include deficiencies of factors XII, XI, IX and VIII, inhibitors of these factors, lupus anticoagulant and heparin therapy.
- A mixing study is used to confirm the presence of an inhibitor.
- Confirmation of a factor VIII inhibitor is made with low factor VIII activity levels and elevated factor VIII inhibitor levels.
- Treatment involves attempts at raising factor VIII levels with DDAVP and factor VIII concentrates and/or to bypass factor VIII with activated prothrombin complex concentrates or recombinant factor VIIIa in cases of bleeding.
- Agents used to eliminate the inhibitor include rituximab, cyclophosphamide with prednisone, IVIG and cyclosporine.
- Unfortunately, because of the rarity of this condition, these recommendations for treatment are based on small case series.

References