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Journal of Gynecology Case Reports

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# Beyond the Uterus: An Unusual Cause of Transfusion-Dependent Vaginal Bleeding

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Received: Oct 11, 2023

Accepted: Nov 27, 2023

Published Online: Nov 31, 2023

Journal: Journal of Gynecology Case Reports

Publisher: MedDocs Publishers LLC

Online edition: http://meddocsonline.org/



**Abstract**

Approximately 1-1.5 people per million will develop the rare coagulopathy of acquired hemophilia A. This autoantibody-mediated destruction of Factor VIII and can lead to severe hemorrhage. Here we describe a first presentation of an acquired hemophilia A in an 89 year old female who had recently undergone hysteroscopic surgery, and presented with transfusion-dependent anemia. We review the work-up, diagnosis, and multidisciplinary management of this rare but potentially life-threatening condition.

## Clinical Presentation

An 89 year old female, presented to the Emergency Department of a regional hospital, from home, accompanied by her husband. She was five days post-operative from a hysteroscopy and polypectomy with endometrial sampling, which was uncomplicated and unremarkable. She was discharged home that same day with scant vaginal bleeding. On post-operative day four, she developed heavy vaginal bleeding, which prompted her to present to a smaller, community centre, where her hemoglobin was noted to be 121 and she was prescribed Tranexamic Acid (TXA). Despite being compliant with medications, the bleeding continued and on post-operative day five, lead to syncope prompting her to phone an ambulance which took her to our regional centre. Upon evaluation, her hemoglobin had fallen from 121 to 74 in 24 hours. Upon examination, we cleared copious blood and clots from the vagina but the cervical os was closed without active bleeding. TXA was started along with intravenous premarin for presumed uterine bleeding. She received two units of blood in the Emergency Department. Several hours later, she passed more large clots per vagina and speculum exam again revealed blood in the vagina but a closed os without active bleeding and no lacerations. One her sacrum, was a large bruise tracking up the backside, which her husband stated had been there for several weeks. She was admitted to Gynecology for observation and kept NPO.

Of note, her medical history is significant for polymyalgia rheumatica however she had been off prednisone for the past year. She is also known for myotonic dystrophy, osteoporosis, and genitourinary syndrome of menopause, for which she takes denosumab, and vagifem, respectively. She denied any used of supplements, anticoagulants, Acetylsalicylic Acid (ASA), Non-Steroidal Anti-Inflammatory Drugs (NSAIDs) and, in private questioning, she denied any intimate partner violence or elder abuse. She does have a history of easy bruising for approximately 18 months but no history of epistaxis, prior transfusion, hemearthroses or family history of hemophilias.

**Cite this article:** Manion R. Beyond the Uterus: An Unusual Cause of Transfusion-Dependent Vaginal Bleeding. J Gynecol Case Rep. 2023; 2(2): 1012.

### Diagnostic process

On the third assessment within six hours, the Gynecology team was called because the patient was again passing large clots and was persistently tachycardic post-transfusion. Hemoglobin was not improving as expected to transfusion (hemoglobin persistently 74 post-transfusion of two units of red blood cells). Speculum exam was unchanged from prior assessments, again with no active bleeding coming from the cervix, but copious blood in the vagina. Ultrasound revealed scant fluid within the endometrial canal and an avascular clot measuring 2.6cm. We continued our transfusion protocol with another two units of red blood cells and ordered coagulation studies, LDH and haptoglobin to rule out hemolysis. In addition, we ordered a CT pelvis with IV contrast to rule out active extravasation from elsewhere in the pelvis. PTT returned significantly elevated at 88 seconds (normal range 19-28 seconds). Hemolysis studies were normal and CT revealed active extravasation from a small vaginal wall vessel on the left vaginal sidewall.

Given the significant elevation in PTT, Hematology was consulted for a possible coagulopathy in the extrinsic pathway, which is depicted in **Figure 1** [1]. Hematology noted that this elevation was almost certainly pathologic, and follow-up studies showed partial correction of her PTT on mixing studies (**Figure 2**). Mixing studies involve mixing the patients plasma in a 1:1 ratio with control plasma. If the PTT corrects after mixing, this points towards a factor deficiency, if the numbers fail to correct, this suggests the presence of inhibiting factors [1]. While this patients labs were indicative of an underlying coagulopathy, factor levels were sent to the tertiary care hospital to confirm a diagnosis of acquired hemophilia and would take 24 hours to return.

### Treatment and management

In the interim, Hematology suggested that surgery could be extremely dangerous as this patient would be unlikely to form clot on her own, should any major procedure be undertaken. We discussed the active extravasation however, and the need for ongoing transfusion, and collectively agreed that an examination under anesthesia with small sutures placed in the vaginal fornix would be acceptable, so long as recombinant factor VIIa was started preoperatively to aid with coagulation. The reasoning behind starting recombinant factor VIIa is because, statistically, most acquired hemophilia cases are due to a deficiency in factor VIII from autoantibody production against factor VIII (2). Factor VIIa is known as a bypassing agent, and can bypass the need for factor VIII in the coagulation cascade (**Figure 1**) and its use in major bleeding in acquired hemophilia cases has been documented [2,3]. Alternatively, activated prothrombin complex concentrate (which contains factor VII and the other vitamin-K dependent clotting factors), is appropriate as well [2,3].

### Outcome

She was taken to the operating room where an examination under anesthesia was performed and showed an extremely small vessel less than 1mm in diameter, in the left vaginal fornix actively bleeding. This vessel would have been compressed by the inferior blade of a speculum during speculum examination, likely why it was not seen upon earlier examination. One figure of eight suture was placed to obtain hemostasis. Vaginal packing and foley catheter were left in situ for twelve hours.

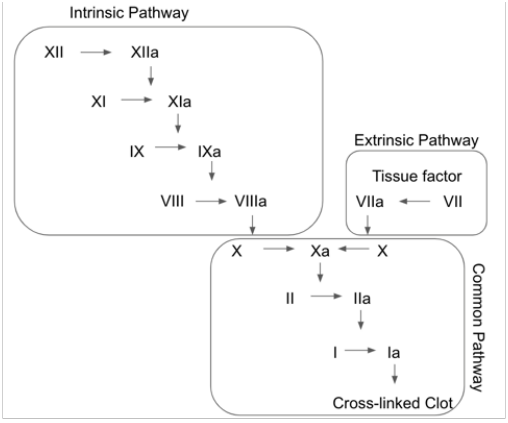


**Figure 1:** Simplification of the coagulation cascade.

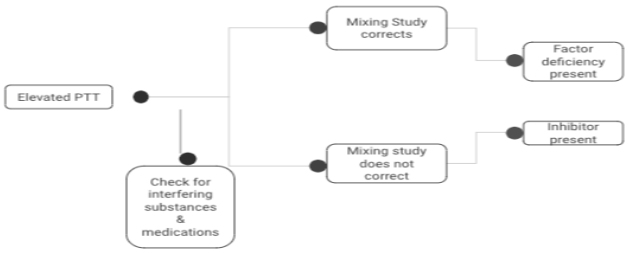


**Figure 2:** Laboratory evaluation of Prlonged PTT and overview of miing study interpretation.

On post-operative day one, factor levels returned from the tertiary care centre confirming that this patient had an acquired Factor VIII deficiency. She was stable from a hemostasis and post-operative perspective and she was transferred to Hematology at the Tertiary Hospital where she received several infusions of recombinant factor VIIa and was discharged home several days later. At follow-up in hematology clinic, she was started on immunosuppressive medication and her bleeding parameters had normalized.

### Concluding thoughts

Acquired hemophilias are a rare group of coagulopathies, with an incidence of 1-1.5 per million per year, not commonly encountered in Gynaecology [2]. Given the rarity of this condition, delays in diagnosis and treatment are common and mortality rates are as high as 20% in acute bleeding [2]. An acute bleeding episode is not an uncommon first presentation, however the majority of first presentation bleeding episodes are subcutaneous bleeds, with vaginal bleeding being far less common, though post-operative and post-partum hemorrhage is reported [3,4]. The pathophysiology of acquired hemophilia A is IgG autoantibody mediated inactivation of factor VIII (4). Therefore treatment focuses on bypassing factor VIII in acute bleeding episodes, and immunosuppression in long-term management [3]. Though diagnosing an acquired hemophilia is outside the scope of gynecology, it is imperative that we consider coagulopathies in patients with an unusual presentation - where the history and physical exam are discordant, or where a clear source of active bleeding cannot be determined. In this particular case, the workup was confounded by the fact that the patient had recently undergone uterine surgery. Furthermore, this case highlights the importance of carefully reviewing baseline coagulation studies in bleeding patients, and considering consultation for a mixing study if the parameters are markedly elevated. Finally, when encountering a patient with active bleeding and a suspected or diagnosed acquired hemophilia, a

multidisciplinary approach is required, as medical management is the cornerstone of reversing the coagulopathy and aggressive surgical management of vaginal bleeding can worsen this rare clinical scenario [5].

## References


1. Palta S, Saroa R, Palta A. Overview of the coagulation system. Indian J Anaesth. 2014; 58: 515-523.

2. Pishko AM, Doshi BS. Acquired Hemophilia A: Current Guidance and Experience from Clinical Prac-tice. Journal of blood medicine. 2022; 13: 255–265. https://doi.org/10.2147/JBM.S284804

3. Mo L, Bao GC. Acquired factor VIII deficiency: Two case reports and a review of literature. Exp Hematol Oncol. 2017; 6: 8.

4. El Demerdash DM, Ayad A, Tawfik N. Acquired Hemophilia A (AHA): underreported, under diagnosed, undertreated medical condition. Egypt J Intern Med. 2022; 34: 12.

5. Scully MF, Shublaq W, Oliver GD. Acquired Hemophilia a Presenting as a Bleeding Diathesis in a Postpar-tum Patient: Diagnosis and Management. J Obstet Gynaecol. 2002; 24: 430-432.