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# WHISPERS OF THE ABYSS: UNMASKING A LURKING RETROPERITONEAL HEMORRHAGE IN THE ENIGMA OF UNDIAGNOSED HEMOPHILIA A

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**ABSTRACT** Spontaneous retroperitoneal hematoma (SRH) is an unexpected and life-threatening occurrence, especially in patients with underlying coagulopathies like hemophilia. We present the case of a 32-year-old man who arrived with seemingly nothing more than abdominal discomfort, but lurking beneath was a rapidly unfolding crisis—a spontaneous retroperitoneal hematoma. In a race against time, we faced the formidable challenge of diagnosing and treating a condition that could turn fatal at any moment. The culprit? Undiagnosed Haemophilia A, a silent predator that had been hiding in his blood all along. This case unravels the suspense of a life-threatening bleed, the adrenaline-fueled decisions, and the critical interventions that ultimately saved the patient's life.

**KEYWORDS :** Spontaneous retroperitoneal hematoma, hemophilia A, Factor VIII replacement, abdominal pain

## INTRODUCTION

Imagine embarking on a casual bike ride, only to later find yourself battling a life-threatening condition. His symptoms, though non-specific at first, soon spiraled into a terrifying cascade that would leave the medical team scrambling to uncover the cause. Unbeknownst to the patient and doctors alike, a catastrophic bleed was brewing inside his body—a silent storm about to erupt. This is the story of how we unraveled the mystery of a spontaneous retroperitoneal hematoma in a patient with undiagnosed hemophilia A. SRH presents a clinical challenge with its stealthy onset but potentially devastating complications, including hypovolemic shock and multi-organ failure. This case highlights the unpredictability of hemophilia and the vital importance of early intervention.

## CASE PRESENTATION

### A SUDDEN TURN OF EVENTS

Our patient, a 32-year-old male, presented to the Emergency Department (ED) with two alarming episodes of syncope and severe abdominal discomfort radiating to his hips. He had experienced these symptoms just after completing a routine 20-kilometer bike ride the day before. Importantly, there was no history of trauma, headache, vomiting, seizures, or bowel disturbances. Yet his condition suggested something far more sinister than simple muscle strain.

### INITIAL EXAMINATION

Upon arrival, his airway was patent, and his respiratory rate was stable at 18 breaths/min, with an oxygen saturation of 99% on room air; heart rate 165 beats/min, blood pressure 90/60 mmHg, with noticeable pallor—his vital signs painted a picture of a body on the edge. He was fully conscious and oriented but clearly in discomfort. Abdominal examination revealed generalized tenderness, but abdomen was soft. There were no external signs—no trauma, no visible injury—yet something insidious was lurking just beneath the surface.

### STABILIZATION IN THE ED

The patient's management began immediately upon his presentation to the ED. Fluid resuscitation was initiated with 1 Liter of normal saline, followed by IV administration of 50 mg of tramadol and 4 mg of ondansetron for pain and nausea control. With his low blood pressure, pallor, and tachycardia, we initially suspected internal bleeding.

The arterial blood gas revealed:
pH: 7.508 (alkalotic); Lactate: 10.21 mmol/L (signaling tissue hypoxia); hemoglobin (Hb): 6.22 g/dL—a shocking revelation of internal blood loss But where was the blood? There were no obvious injuries.
POCUS revealed free fluid in the hepatorenal pouch and splenomegaly.

A unit of uncrossed packed red blood cells (PRBC) was transfused, and 10 mg of IV vitamin K was administered.

### PROVISIONAL DIAGNOSIS AND FURTHER EVALUATION

The next step was a computed tomography (CT) scan, which revealed what we feared: a large retroperitoneal hematoma.

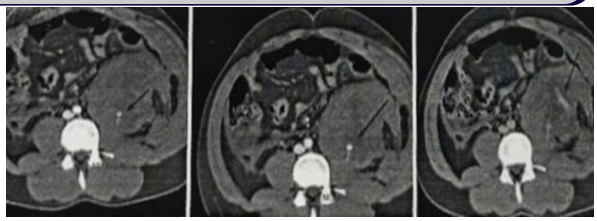

**Figure1:** Large Retroperitoneal Hematoma

Thromboelastography was also sent as we were suspecting underlying coagulopathy as this bleed was not associated with any significant trauma.

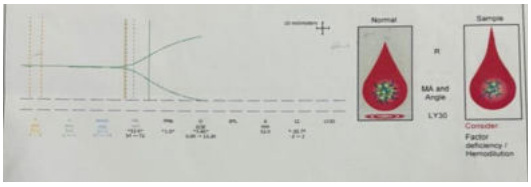


**Figure 2-** Thromboelastography result

### THE LAB RESULTS—MORE THAN MEETS THE EYE

The patient's clotting profile came back, and the pieces finally came together: aPTT: 72.7 seconds (profoundly prolonged); Factor VIII Level: 4% Undiagnosed hemophilia A—a condition that had been lurking in the shadows his entire life revealed as the catalyst for this life-threatening event.

Immediate administration of recombinant Factor VIII was initiated, and the patient was transferred to the intensive care unit (ICU) for close monitoring.

### IMAGING AND SURGICAL CONSULTATION

A digital subtraction angiography (DSA) was performed by the vascular surgery team, ruling out any active arterial bleeding. This meant that while the hematoma was large, there was no ongoing hemorrhage from major vessels—a relief in terms of immediate intervention. However, the hematoma needed to be monitored for potential complications, including compartment syndrome.

### ICU MANAGEMENT AND FACTOR VIII REPLACEMENT

In the ICU, the patient received 8 units of cryoprecipitate, 2 units of fresh frozen plasma (FFP), and continuous Factor VIII replacement. His clinical condition improved steadily, and serial ultrasounds demonstrated a gradual reduction in the size of the hematoma. Over the next few days, Factor VIII levels were carefully monitored, and his therapy was gradually reduced as he stabilized.

### THE ROAD TO RECOVERY

By day four, the patient's Factor VIII levels had risen to 73.9%, and his condition had improved significantly. Repeat imaging showed further reduction in the hematoma, and he remained hemodynamically stable. He was discharged on Factor VIII replacement therapy with instructions for close outpatient follow-up. His recovery was smooth, and there was no recurrence of bleeding.

## DISCUSSION

**CLINICAL PRESENTATION AND DIAGNOSIS OF SRH IN HEMOPHILIAA**: SRH, particularly in individuals with undiagnosed coagulopathies like hemophilia A, presents a diagnostic challenge. Hemophilia A, an X-linked recessive disorder, results from a deficiency of Factor VIII, essential for the coagulation cascade. Without prompt recognition and treatment, SRH can lead to severe complications such as hypovolemic shock and organ failure. [1] Although hemophilia patients may present with spontaneous hemorrhages, the retroperitoneal space poses an additional diagnostic challenge. Due to its large volume capacity, bleeding can accumulate substantially before overt symptoms appear.

The classical signs of SRH—flank pain, hematuria, and shock (Wunderlich syndrome)—were notably absent in this case, underscoring the atypical nature of the presentation. Studies show that SRH in hemophiliacs is a rare complication but can have devastating consequences if not recognized early. [2] A high index of suspicion is essential, particularly in patients with coagulopathies who present with non-specific symptoms like abdominal pain and syncope [3].

The diagnosis hinges on rapid imaging techniques. Point-of-care ultrasound (POCUS), as used in this case, is an invaluable first-line tool for detecting free fluid, though it cannot localize the bleeding source. CT imaging with contrast, which revealed the large retroperitoneal hematoma in this patient, remains the gold standard for a definitive diagnosis. [4]

**PATHOPHYSIOLOGY OF RETROPERITONEAL HEMATOMAS IN HEMOPHILIA**: Hemophilia A patients, due to their profound Factor VIII deficiency, experience spontaneous haemorrhages into deep tissues such as muscles, joints, and retroperitoneal space. SRH occurs when small vessels within the retroperitoneum rupture, leading to blood accumulation. The extent of the hematoma often depends on the degree of coagulopathy and the delay in diagnosis. [5] . As Factor VIII is essential in forming stable fibrin clots, the lack of this coagulation factor means that even minor vessel injuries can result in significant haemorrhage. [6] In the context of physical exertion, such as the 20-kilometer bike ride this patient had undertaken, even mild trauma or muscular strain can precipitate bleeding in hemophilia A patients [7]. This case serves as a reminder of the fragile balance between physical activity and bleeding risk in patients with undiagnosed bleeding disorders.

**MANAGEMENT STRATEGIES IN SRH AND HEMOPHILIA A**: Factor Replacement Therapy is the cornerstone of treatment in hemophilia A patients experiencing major bleeding episodes. The administration of recombinant `Factor VIII in this case rapidly corrected the underlying coagulopathy, halting further bleeding and stabilizing the patient.[1] Guidelines from the World Federation of Hemophilia recommend that for major bleeds, such as SRH, Factor VIII levels should be raised to at least 80-100% of normal levels, with continuous monitoring to ensure haemostasis. [1] In conjunction with Factor VIII therapy, aggressive supportive care is necessary.

Fluid resuscitation and blood transfusion are essential to combat hypovolemic shock resulting from significant blood loss. Close ICU monitoring allows for early detection of complications, such as compartment syndrome, which can occur as the hematoma expands within the confined retroperitoneal space.[8] Surgical Intervention is typically reserved for cases where imaging demonstrates active arterial bleeding. In this case, digital subtraction angiography (DSA) confirmed that no major vessels were involved, thus obviating the need for immediate surgical intervention. However, hematoma expansion and pressure effects need continuous surveillance during the hospital stay[9].

**PROGNOSIS AND LONG-TERM MANAGEMENT**

With prompt diagnosis and Factor VIII replacement, the prognosis for SRH in hemophilia A is generally good. As seen in this case, serial ultrasounds documented a reduction in hematoma size over time, and the patient stabilized without further bleeding complications. Long-term management includes patient education regarding bleeding risks, avoidance of high-risk activities, and regular follow-up with a hematologist. Factor VIII replacement prophylaxis can significantly reduce the risk of spontaneous bleeding episodes in severe hemophilia A patients. [10] In addition, newer therapies, such as gene therapy and extended half-life Factor VIII concentrates, may offer long-term solutions for hemophilia management. [11]

**CONCLUSION**

This case exemplifies the importance of maintaining a high degree of suspicion for coagulopathies in patients with unexplained spontaneous bleeding, particularly when no trauma is evident. Hemophilia A, while often diagnosed in childhood, can remain undetected until a major bleeding event brings it to the forefront, as it did in this patient. Factor VIII replacement therapy, combined with vigilant monitoring, allowed for a successful outcome in this case. Moving forward, patient education and careful management of bleeding risks are key to preventing recurrence. Regular follow-up with hematology and continued Factor VIII therapy will ensure long-term stabilization and quality of life for this patient.

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