ISSN: 1607-8322, e-ISSN: 2220-5799 Anaesthesia, Pain & Intensive Care
Vol 28(5); October 2024 DOI: 10.35975/apic.v28i5.2556

CASE REPORT PERIOPERATIVE MEDICINE

# Acute abdomen: an atypical presentation of acquired hemophilia

**Muhamad Rafiqi Hehsan** [1,2,3]

**Author's affiliations:**

1. Department of Anesthesiology & Critical Care, School of Medical Sciences, Health Campus, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan
2. Faculty of Medicine & Health Sciences, Universiti Sains Islam Malaysia, Bandar Baru Nilai, 71800, Nilai, Negeri Sembilan, Malaysia.
3. Faculty of Medicine, Universiti Sultan Zainal Abidin, 20400 Kuala Terengganu, Terengganu Darul Iman, Malaysia; E-mail: muhamad_rafiqi@usim.edu.my

**Correspondence:** Muhamad Rafiqi Hehsan; E-mail: muhamad_rafiqi@usim.edu.my

## ABSTRACT

Acquired hemophilia is a rare but potentially life-threatening disorder characterized by the development of acquired hemophilia A, leading to bleeding complications. Spontaneous, mild-to-life-threatening bleeding in typically elderly individuals without a personal or family history of bleeding is the hallmark of acquired hemophilia A (AHA), a rare autoimmune bleeding illness caused by the formation of auto-antibodies against endogenous factor VIII (FVIII). This hallmark represents our present case, with no obvious history related to the bleeding issue. Prompt detection and treatment of bleeding are essential for the best outcomes.

We present a case of a 66-year-old male who had acute-onset severe abdominal pain and with constipation for five days, associated with nausea and loss of appetite. On physical examination, the patient appeared pale, and abdominal examination revealed diffuse tenderness with guarding. Initial evaluation revealed tachycardia and hypotension. Initial abdomen X-rays revealed features suggestive of small bowel obstruction. The patient was stabilized with intravenous fluids, referred to a surgical team, and admitted to the acute bed in the general ward for further management.

**Keywords:** Acquired Hemophilia; Auto-Antibodies; Factor VIII; Intraabdominal Bleeding

**Citation:** Hehsan MR. Acute abdomen: an atypical presentation of acquired hemophilia. Anaesth. pain intensive care 2024;28(5):974–976; **DOI:** 10.35975/apic.v28i5.2556

**Received:** March 19, 2024; **Reviewed & Accepted:** September 08, 2024

## 1. INTRODUCTION

Acquired hemophilia is a rare but potentially life-threatening disorder characterized by the development of acquired hemophilia A (AHA), which leads to bleeding from different sites of the body. There is spontaneous, mild-to-severe bleeding in typically elderly individuals. There may not be any personal or family history of bleeding. AHA, a rare autoimmune bleeding illness is caused by the formation of auto-antibodies against endogenous factor VIII (FVIII). Prompt detection and treatment of bleeding are essential for the best outcomes. Nonetheless, a lack of awareness of the condition's urgency, its clinical manifestations, and the laboratory diagnosis of acquired hemophilia limits the ability to provide effective management.[1]

We present a case of an old man who presented with acute abdomen. He was anemic, tachypneic and restless. Radiological examination revealed small bowel obstruction, which needed surgical treatment.

## 2. CASE REPORT

A 66-year-old male presented with acute-onset episodic abdominal pain and constipation for five days, associated with nausea and loss of appetite. On physical examination, the patient appeared pale, and his abdominal examination revealed diffuse tenderness with

 Open access attribution (CC BY-NC 4.0)

guarding. Initial evaluation revealed tachycardia and hypotension. Abdomen X-rays revealed features suggestive of small bowel obstruction (Figure 1).

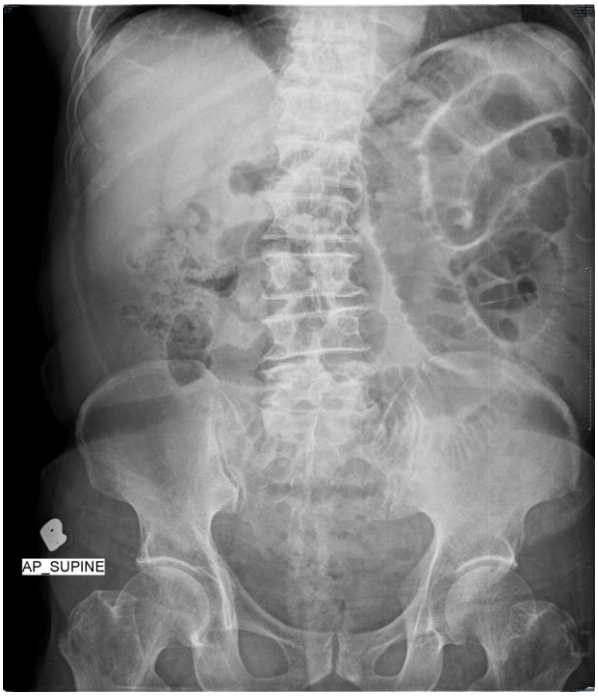


**Figure 1: Initial abdomen X-rays revealed features suggestive of small bowel obstruction.**

However, on the next day, he complained of severe, persistent abdominal pain. Laboratory investigations revealed a significant drop in hemoglobin levels from 8.9 g/dL to 5.5 g/dL, prolonged activated partial thromboplastin time (aPTT) 73.9 sec (normal 21-35 sec), prothrombin time (PT) 10 sec, and an International Normalized Ratio (INR) 0.9. A complete blood count (CBC) revealed macrocytic anemia, and reactive lymphocytes, which may be associated with a viral infection or autoimmune disorder. There was also thrombocytosis with a platelet count of 492 K/µL. The computed tomography (CT) angiography of the mesentery was performed urgently and revealed a huge right retroperitoneal hematoma with evidence of active contrast extravasation associated with the right iliacus and possible right psoas and lateral abdominal wall muscle involvement (Figure 2).

Further investigation into the etiology of the intraabdominal bleeding with severe anemia and prolonged activated partial thromboplastin time (aPTT) led to the measurement of the factor VIII level to rule out hemophilia. A zero percent level of factor VIII in this patient established the diagnosis of acquired hemophilia. The patient had no history of prior exposure to factor

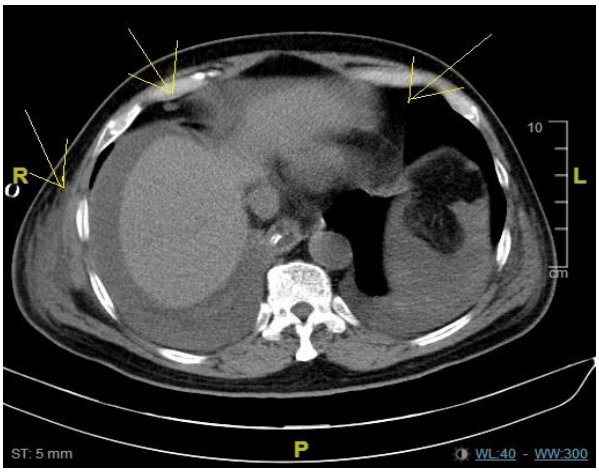


**Figure 2: Huge right retroperitoneal hematoma with evidence of active contrast extravasation associated with right iliacus and possible right psoas and lateral abdominal wall muscle involvement (yellow arrows).**

replacement therapy or other potential triggers for acquired hemophilia. Other investigations of exclusion,

including carcinoembryonic antigen (CEA), antinuclear antibody (ANA), and Lupus anticoagulant screening, only showed low and weak results. Given the high risk of exsanguination, emergent surgical intervention was warranted. He was then admitted to the ICU for close monitoring and optimization.

The patient's management involved a multidisciplinary approach, including hematologists, intensivists, and general surgeons. The decision was made to initiate immunosuppressive therapy to suppress the production of autoantibodies against factor VIII. Treatment included corticosteroids (prednisone) and bypassing agents, including recombinant activated factor VII (rFVIIa), FEIBA (Anti-Inhibitor Coagulant Complex), and activated prothrombin complex concentrate (aPCC). The massive transfusion protocol was activated, and the patient received blood products and packed red blood cells (pRBCs) including 6 pints of cryoprecipitate, 4 pints of fresh frozen plasma (FFP), and 5 pints of (pRBCs), with close monitoring of hemostatic parameters and clinical response.

Despite aggressive medical management, the patient's clinical condition deteriorated, with persistent hemodynamic instability and ongoing hemorrhage. While surgery had been scheduled, the patient developed respiratory distress that necessitated intubation in the ICU for an impending respiratory collapse. Hemodialysis was needed due to worsening renal function caused by an acute kidney injury. Unfortunately, the patient's hemodynamics deteriorated

 Open access attribution (CC BY-NC 4.0)

**Box 1: Ten principles of AHA care (Dolan, G. et al.[1, 2])**

| | |
|---|---|
| **Principle 1** | **Improving initial diagnosis of AHA in all settings** |
| Principle 2 | Differential diagnosis of AHA: Laboratory assessment of patients with unusual bleeding |
| Principle 3 | Effective communication between laboratories, physicians, and specialists |
| Principle 4 | Improving clinical care: networking between HCPs in the treating hospital and specialist hemophilia treatment centers |
| Principle 5 | Comprehensive assessment of bleeding is required |
| Principle 6 | Appropriate use of bypassing agents for prompt treatment |
| Principle 7 | Long-term follow-up and monitoring for efficacy and safety of immunosuppressive treatment |
| Principle 8 | Promotion of patient care and follow-up in the home, inpatient and outpatient settings |
| Principle 9 | Access to innovative and disruptive treatments will be important |
| Principle 10 | Promotion of collaborative research to improve patient outcomes |

Abbreviations: AHA, acquired hemophilia A; HCP, healthcare professionals.

during dialysis, requiring double inotropic support until pulseless ventricular tachycardia (VT) and asystole developed. CPR commenced for 30 min; however, he succumbed to death.

# 3. DISCUSSION

Acquired hemophilia is a rare but potentially life-threatening disorder characterized by the development of autoantibodies against clotting factors, leading to bleeding complications.[1] Spontaneous, mild to life-threatening bleeding in typically elderly individuals without a personal or family history of bleeding is the hallmark of acquired hemophilia A (AHA), a rare autoimmune bleeding illness caused by the formation of auto-antibodies against endogenous factor VIII (FVIII).[2] This hallmark represents our present case, with no obvious history related to the bleeding issue.

Although hemarthrosis and spontaneous mucocutaneous bleeding are frequent presentations, intraabdominal bleeding resulting in an acute abdomen is rare and offers a difficult scenario for diagnosis as well as management.[3] Individuals with acquired hemophilia can present with anemia or occult bleeding, as presented in our case. In contrast to patients with congenital hemophilia, those with acquired hemophilia may experience more severe bleeding symptoms that occur in a distinct pattern.[1,2] Clinically, acute abdomen manifests as intense abdominal pain accompanied by various associated symptoms such as nausea, vomiting, and abdominal distension.[3] In our case, the patient presented with acute abdomen due to intraabdominal bleeding secondary to acquired hemophilia. The diagnosis was established based on prolonged aPTT and the zero percent factor VIII activity. The management involved a multidisciplinary approach with close collaboration between hematologists, intensivists, and the surgeons for optimizing patient outcomes.

There are 10 principles of AHA care proposed by Dolan, G. et al.[1, 2] (Box1).

# 4. CONCLUSION

Intraabdominal bleeding in acquired hemophilia is a rare and challenging clinical presentation. Prompt recognition, accurate diagnosis, and aggressive treatment with bypassing agents and immunosuppressants are crucial for managing this life-threatening condition. This case underscores the importance of considering acquired hemophilia in the differential diagnosis of unexplained bleeding, even in individuals with no prior history of bleeding disorders. Early intervention and a multidisciplinary approach are essential to achieving favorable outcomes in these complex cases.

## 5. Conflict of interests

The author declares no conflict of interest. Ethical committee clearance was not required.

## 6. Author's contribution

MRH was the sole author of this case report.

# 7. REFERENCES


1. Dolan G, Benson G, Bowyer A, Eichler H, Hermans C, Jiménez-Yuste V, et al. Principles of care for acquired hemophilia. Eur J Haematol. 2021;106(6):762–73. [Pubmed] DOI: 10.1111/ejh.13592
2. Bitting RL, Bent S, Li Y, Kohlwes J. The prognosis and treatment of acquired hemophilia: a systematic review and meta-analysis. Blood Coagul Fibrinolysis. 2009;20(7):517-23. [Pubmed] DOI: 10.1097/MBC.0b013e32832ca388
3. Gans SL, Pols MA, Stoker J, Boermeester MA, Expert Steering Group. Guideline for the diagnostic pathway in patients with acute abdominal pain. Dig Surg. 2015;32(1):23-31. [Pubmed] DOI: 10.1159/000371583


 Open access attribution (CC BY-NC 4.0)