



Emergency Splenectomy For A Traumatic Splenic Rupture In An Elderly Patient With Undiagnosed Acquired Hemophilia: A Case Report

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Abstract

Among the rare coagulopathies that can be revealed by trauma or surgery, acquired hemophilia holds a special place. It is an autoimmune pathology linked to the presence of inhibitors directed against coagulation factor VIII, resulting in spontaneous or trauma-induced bleeding. We report the case of a 70-year-old patient admitted to the Yaoundé emergency center, presenting with a traumatic splenic rupture, in whom the persistence of an unusual hemostatic disorder led to the diagnosis of acquired hemophilia.

Emergent general surgery leads to significant morbidity and mortality in older patients. While clinical outcomes after elective operations are similar in younger and older patients, the latter have higher rates of complications and mortality after emergency surgery. Abdominal surgical procedures such as colectomy, small-bowel resection, cholecystectomy, appendectomy, and others constitute ~80% of all emergent surgeries performed in older patients. The risk of surgical bleeding can be minimized by judicious administration of fresh frozen plasma at induction, during, and after surgery under the close supervision of hemophilia specialists.

The successful outcome demonstrates that major emergent abdominal surgery, even highly hemorrhagic procedures like splenectomy, can be safely performed in patients with hemophilia (or severe coagulopathy) provided a continuous and closely monitored factor replacement regimen is implemented.

Background

Traumatic splenic rupture is one of the main causes of hemoperitoneum in emergency surgery. In the elderly, reduced physiological reserve and the frequency of comorbidities increase the severity of hemorrhagic shock and mortality. In this context, rapid hemodynamic stabilization and early surgical control of the bleeding determine the prognosis.

The occurrence of perioperative bleeding disproportionate to the injury mechanism should raise suspicion of a hemostasis abnormality. Among the rare coagulopathies that can be revealed by trauma or surgery, acquired hemophilia holds a special place. It is an autoimmune pathology linked to the presence of inhibitors directed against coagulation factor VIII, resulting in spontaneous or trauma-induced bleeding [1]. It preferentially affects elderly subjects with a median age between 70 and 76 years, with no distinction between sexes [2,3]. Its diagnosis is often delayed due to the absence of a hemorrhagic history and a non-specific

clinical presentation, which complicates surgical management.

The combination of severe abdominal trauma requiring emergency splenectomy with undiagnosed acquired hemophilia is exceptional and poses a real perioperative challenge due to the major risk of uncontrollable hemorrhage and the complexity of the transfusion strategy.

We report the case of a 70-year-old patient admitted to the Yaoundé emergency center, presenting with a traumatic splenic rupture, in whom the persistence of an unusual hemostatic disorder led to the diagnosis of acquired hemophilia. This case highlights the need to suspect an underlying coagulopathy when faced with an atypical bleeding profile in traumatology and discusses the principles of operative management in light of recent data.

Case Presentation

A 70-year-old female patient, with no known hematological history, was admitted to the emergency department for facial swelling

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and facial bleeding following a road traffic accident involving a truck with initial loss of consciousness.

On admission, her upper airways were clear, and a cervical collar was placed. She presented with a respiratory rate of 28 cycles/min, a pulse oximetry of 99%, a blood pressure of 67/42 mmHg with a pulse of 75 beats/min, as well as peripheral signs of hemorrhagic shock. Fluid resuscitation with 1000 mL of normal saline restored her blood pressure to 107/62 mmHg. Neurologically, the patient was conscious but confused with a Glasgow Coma Scale of 13/15 and presented no sensory nor motor function impairment. Blood glucose was 1.94 g/L. The examination noted a bleeding lip wound. An E-FAST revealed intraperitoneal fluid.

The diagnosis included multiple trauma, combining a mild traumatic brain injury and a blunt thoraco-abdominal trauma, complicated by hemorrhagic shock. Initial management included the transfusion of 2 units of whole blood.

A full-body scan revealed a massive hemoperitoneum secondary to a splenic rupture. A CBC showed severe microcytic anemia at 7 g/dL, initial thrombocytopenia at $97,000/\text{mm}^3$ rapidly dropping to $11,000/\text{mm}^3$, and a collapsed prothrombin time. These hemostatic abnormalities, disproportionate to the blood loss alone, raised suspicion of an underlying coagulopathy. The clinical context led to the suspicion of acquired hemophilia.

The patient underwent an emergency laparotomy: a total splenectomy with drainage of approximately 2000 mL of hemoperitoneum. Intraoperatively, she received 3 units of packed red blood cells, 1 pack of fresh frozen plasma, 3 platelet concentrates, as well as vitamin K and tranexamic acid. The coagulopathy made managing the hemorrhage particularly difficult, reinforcing the suspicion of acquired hemophilia.

In the immediate postoperative period, the CBC showed hemoglobin at 12.5 g/dL, thrombocytopenia at $24,500/\text{mm}^3$, and a PT (prothrombin time) at 78.9% after hemostatic correction. The outcome was favorable, allowing discharge on Day 11 with vitamin K for 5 days. One week later, she showed satisfactory clinical progress, with no recurrence of bleeding.

Discussion

Surgery in elderly patients

Emergent general surgery leads to significant morbidity and mortality in older patients. While clinical outcomes after elective operations are similar in younger and older patients, the latter have higher rates of complications and mortality after emergency surgery [4].

Abdominal surgical procedures such as colectomy, small-bowel resection, cholecystectomy, appendectomy, and others constitute ~80% of all emergent surgeries performed in older patients [5,6]. Pre-operative frailty is associated with increased poor clinical outcomes after emergent or non-emergent general surgery in older adults. The outcomes that may be adversely influenced by baseline frailty include post-operative complications and morbidity, hospital length of stay, 30-day mortality, and long-term mortality.

The utility of frailty assessment should extend beyond its role in pre-operative risk stratification. In fact, the positive identification of frailty in an older surgical patient should trigger the initiation of a set of interventions that may reduce morbidity and enhance functional recovery after surgery [7].

Potential types of interventions to be considered include: (1) the Enhanced Recovery After Surgery (ERAS) pathways, an integrated clinical care delivery program recently shown

to improve clinical and functional outcomes in older patients after colorectal surgery [8,9]; (2) prehabilitation programs that aim to optimize physical function and comorbidities pre-operatively [10]; and (3) geriatric interdisciplinary assessment and treatment models that have been demonstrated to improve clinical outcomes of frail older adults [11]. While further investigation is needed, clinical care pathways that integrate frailty assessment, geriatric medicine-focused peri- and post-operative management, and patient-centered interdisciplinary care models should be delivered as a comprehensive intervention in frail older adults undergoing surgery [12].

Surgery in hemophilic patients

Haemostasis planning

After assessment of the patient's underlying factor deficiency, inhibitor status, and associated complications or comorbidities are aligned with the planned surgical procedure, and the haematology team develops an individualized haemostatic management plan, including the preoperative, intraoperative, and rehabilitation periods. Replacement therapy, doses, and intervals are established, including method of administration (bolus/continuous infusion), as well as the need for concomitant antifibrinolytic therapy [13,14,15]. A contingency plan should be in place (e.g., pharmacy and blood bank support) if unexpected bleeding occurs.

The quantity of clotting factor product required for surgery is determined. Patients with mild haemophilia A undergoing surgery may not require clotting factor concentrate and may be treated with non-specific haemostatic agents (e.g., desmopressin acetate). Less-invasive procedures, such as radio- and chemical synovectomy, typically require smaller amounts of clotting factor concentrates. Prior to major elective procedures, an in vivo recovery and half-life study should be considered when the patient is not bleeding, using either a standard or a Bayesian approach to pharmacokinetic analysis [16,17].

Attention to the possibility of inhibitor development during and following surgery requiring replacement therapy is imperative. A factor assay monitoring schedule must be coordinated with the coagulation laboratory to ensure timely and accurate results to permit dosing adjustments. Expertise in inhibitor identification and treatment must be available. Some patients with non-severe haemophilia A may have mutations that increase their risk of inhibitor development after intensive exposure to FVIII [18]. The haemostatic plan must be approved by the haematologist, communicated to all teams involved in the surgery, and a copy filed in the medical records. Patients with existing inhibitors require additional attention, and the quantity and mode of administration of bypassing agents and the need for adjunctive therapy (e.g., antifibrinolytics) agreed upon [14].

Postoperative recovery period

The main goal of an inpatient stay is maintenance of adequate haemostasis and initiation of required rehabilitation to assure a safe discharge. For PWH without inhibitors, rehabilitation begins immediately, as early as the transfer to the postanaesthesia care unit. The role of the MDT coordinator may shift from the surgical service to the medical or haematology service, although the surgeon will remain involved to respond to surgical queries.

Haemostasis and thrombosis prophylaxis

The haemophilia nurse ensures that the patient's haemostatic plan is carried out and that haemostasis is adequately monitored, particularly for patients with severe haemophilia requiring frequent or continuous infusions of clotting factor

concentrate. Patients with inhibitors require careful monitoring to ensure that they receive consistent, timely treatment with the appropriate bypassing agent [19]. The haematologist is available to determine changes to the haemostatic plan as required. The surgical and haematological teams monitor wound healing and drainage. The MDT should be fully aware of concomitant medications the patient receives, their potential impact on coagulation, and the need for continuity throughout the hospital stay. Patients receiving clotting factor concentrate by continuous infusion are monitored to ensure appropriate product delivery and adequate factor levels. Inhibitor and clotting factor monitoring should be performed more frequently in patients with mutations that increase their risk of inhibitor development after intensive exposure to FVIII.

Incidence of symptomatic venous thromboembolism in PWH undergoing major orthopaedic surgery (some of whom received thromboprophylactic interventions) may be similar to that estimated in the general population not receiving thromboprophylaxis [20]. Although there is no consensus in current haemophilia practice, a thrombosis risk assessment should be performed and postoperative prophylaxis considered. Minimally, the entire team should be aware of the potential for thrombosis and remain alert to clinical signs and symptoms [21]. Mechanical prophylaxis with graduated compression stockings or impulse foot compression is commonly used. Chemical prophylaxis is uncommon [22].

Surgery in elderly patients with hemophilia

The risk of surgical bleeding can be minimized by judicious administration of fresh frozen plasma at induction, during, and after surgery under the close supervision of hemophilia specialists. Frequent monitoring to maintain adequate levels of factor VIII makes surgical options safe even when faced with major emergent trauma surgery. In our case, continuous fresh frozen plasma infusion therapy was proven effective in preventing bleeding complications during the laparotomy and during the patient's prolonged hospitalization, especially in elderly people. There are several reports in the literature in favor of surgery in hemophilic patients with continuous or interrupted [23] infusion of clotting factors with minimal risk of hemorrhage [24].

We report a rare case of successful emergency splenectomy on a trauma patient with hemophilia. Abdominal surgery in a hemophilia A patient is feasible without hemorrhagic complications under continuous fresh frozen plasma replacement therapy (factor VIII included). Elective resection of an abdominal hemophilic pseudotumor should be considered prior to the development of major complications. Successful emergency splenectomy was performed on a trauma patient with previously undiagnosed hemophilia, a rare occurrence. This case demonstrates the feasibility of abdominal surgery, such as splenectomy, in a patient with hemophilia A without hemorrhagic complications, provided continuous factor VIII replacement therapy is administered.

Conclusion

The management of perioperative hemostasis in an emergent surgical setting presents unique challenges, particularly in the elderly patient population and those with underlying coagulopathies, such as hemophilia.

The successful outcome demonstrates that major emergent abdominal surgery, even highly hemorrhagic procedures like splenectomy, can be safely performed in patients with

hemophilia (or severe coagulopathy) provided a continuous and closely monitored factor replacement regimen is implemented. This underscores the necessity of a pre-established, comprehensive hemostatic management plan, emphasizing factor monitoring and appropriate bypassing agents (if inhibitors are present) or factor concentrates for optimal surgical safety and prevention of hemorrhagic complications.

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