

## Management of a Retroperitoneal Hematoma in a Hemophilic Patient: A multi-disciplinary approach

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### Abstract

Hemophilia is a bleeding disorder with complicated diagnostic and management strategies which, if overlooked, can lead to greater morbidity and mortality. We present a case of traumatic retroperitoneal hematoma in a known case of Hemophilia A, who presented to us in ED with left sided abdominal pain and swelling. A multi-disciplinary approach was sought. The condition of the patient did not improve; hence a diagnostic laparoscopy was performed under cover of factor VIII which resulted in improvement of the patient. Therefore, a combination of conservative and surgical methods can be effective in managing retroperitoneal bleeding in patients with Hemophilia A.

**Keywords:** Bleeding Disorder; General Surgery; Hemophilia; Hematoma

### Introduction

Retroperitoneal Hemorrhage (or Retroperitoneal Hematoma (RPH) refers to an accumulation of blood found in the retroperitoneal space [1]. Few types of RPH have been described in the published reports: Spontaneous, iatrogenic, idiopathic or traumatic hematoma [2]. Idiopathic retroperitoneal hematoma is a rare but potentially life-threatening pathological cause of an acute surgical abdomen. In the early stages, it typically presents as generalized abdominal pain, nausea, anorexia. The symptoms and signs of hypovolemic shock normally present late [3]. Traumatic retroperitoneal hematoma is the common complication of abdominal or pelvic injuries. Retro peritoneum contains several visceral and vascular structures in the gastrointestinal, genitourinary, vascular, musculoskeletal and nervous systems [4].

Traumatic retroperitoneal hematoma is reported as high as 18% - 60% in English literatures [5]. Haemorrhage within the retroperitoneal area may be massive and may exceed 2000 ml of blood. Experimental data have shown that as much as 4000 ml of fluid can extravasate into the retroperitoneal space under pressure equal to that in the pelvic vessels. The diagnosis of

retroperitoneal haematoma is most difficult following blunt trauma to the abdomen and should be suspected in any patient following trauma who had signs and symptoms of haemorrhagic shock but no obvious source of hemorrhage [6].

## Case Report

A 21 years old male presented in the ER with the complaints of pain and swelling in the abdomen along with fever and vomiting for the past 4 days. According to the patient this was preceded by a fall with trivial trauma. The patient was a known case of Hemophilia.

On general physical examination, the patient was pale and unstable hemodynamically with a pulse of 140/min, BP 90/60, Respiratory rate 26/min and febrile 101F. Abdominal examination showed asymmetrical distension. A hard mass palpable in the left hemi abdomen, tender, not crossing the midline, immobile and dull on percussion. Bowel sounds were not audible.

Lab reports showed Hb 7.1 g/dl, TLC 15000/cmm, Platelets 350/cmm, Bleeding time 4 min, APTT > 2 min, PT 15.2 sec, INR 1.15 sec, Na 127 mmol/l, K 5.1 mmol/l, Serum Creatinine 3.1 mg/dl, Urea 95 mg/dl, LFT within normal limits.

FAST scan showed a large hematoma up to 450 ml in left hemiabdomen within the retroperitoneal space with minimal fluid in abdomen.

Patient was initially managed conservatively with fluid resuscitation, Cryoprecipitate and FFP transfusion along with broad spectrum antibiotics for 14 hours. Opinions were sought from both hematologist and nephrologist. However, the patient's condition did not improve. On examination, Grey turner and Cullen signs (Figure 1) appeared on the abdomen. Lab reports showed Hb 5.3 g/dl and APTT 56 sec.

Repeat Ultrasound Abdomen showed hematoma had increased up to 600ml with hemoperitoneum up to 1000ml. It also showed mild left hydronephrosis.



**Figure 1:** Grey Turner and Cullen signs on the abdomen.

High risk consent was taken and the patient underwent a diagnostic laparoscopy under Factor VIII support. Around 2.5 L blood and clots were drained from the peritoneal cavity. Left colon was dilated and thin walled. Liver and spleen were normal. Retroperitoneal space was opened, all clots were removed, and dissection was done till left kidney. All bleeding veins were ligated

with ligasure. Wide bore drains were placed in the Left paracolic gutter, Right subphrenic space and pelvis.

Postoperatively, the patient's vital signs improved, urine output was adequate, and the drains revealed no active bleed. Factor VIII support continued throughout recovery under hematologist opinion. Lab reports showed, Hb 9.7 g/dl, APTT 28 sec, Serum Creatinine 1.1 mg/dl and K 4 mmol/L.

Ultrasound abdomen showed no intraperitoneal or retroperitoneal bleed. Drains were removed and Factor VIII dose was reduced. Patient was discharged on 9<sup>th</sup> postoperative day. (Figure 2)



**Figure 2:** Patient's abdomen on 9<sup>th</sup> postoperative day.

## Discussion

Hemophilia is an X-linked congenital bleeding disorder caused by a deficiency of coagulation factor VIII (FVIII) (in hemophilia A) or factor IX (FIX) (in hemophilia B). The deficiency is the result of mutations of the respective clotting factor genes.

When a patient of Hemophilia presents with acute hemorrhage, it is a medical emergency and hemostasis is maintained instantaneously. Various studies have shown that the target replacement factor level depends on the severity and site of the hemorrhage [7]. Since the patient was a known case of hemophilia, a hematologist was immediately involved and cryoprecipitates and FFPs were instantly administered. Blood samples were sent to the lab on emergency basis and a FAST scan done to localize the hemorrhage which showed a left-sided retroperitoneal hematoma of 450ml. There was prolonged APTT of >2 mins while PT/INR and platelets were also deranged. The hematologist was consulted to maintain factor activity levels above 50%. Nephrologist was consulted as S/Cr and urea levels were deranged as well. The patient was treated conservatively through fluid resuscitation and factor replacement under cover of antibiotics for approximately 14 hours. Since the patient's condition did not show improvement, re-evaluation was done. APTT prolonged at 56s and ultrasound abdomen showed a retroperitoneal hematoma of about 600ml with a hemoperitoneum of about 1000ml. After consultation with a panel including doctors from multiple specialties, it was decided that patient would undergo a diagnostic laparoscopy.

Up until the 1960s, the only surgeries individuals with hemophilia underwent were emergency procedures, due to the high mortality rates of 25%-50% [8]. After development of plasma fractions and cryoprecipitates, the mortality rate fell to 10% and further to 1% after

development of highly active factor VIII and IX concentrates [9]. A high-risk consent was taken from the patient. A plan of action was laid out by a team of surgeons, hematologists and anesthesiologist. A diagnostic laparoscopy was performed, during which the hematoma was drained, and clots were removed. Multiple bleeders were ligated and several drains placed to ensure no recollection of blood.

The patient showed drastic improvement after surgery. He was treated post-op by fluid and electrolyte replacement, complemented by factor VIII transfusion and adequate pain management after consultation from nephrologist, hematologist and anesthesiologist. Patient was deemed fit for discharge on 9<sup>th</sup> day post-operatively with advice to follow up with the hematologist.

This case report is evidence that managing a patient with hemophilia requires a multidisciplinary approach. Bleeding in hemophilia requires urgent infusion of clotting factor concentrates. However, conservative treatment is not always enough, and emergency surgery might be required to control the hemorrhaging. Participation from experts in the respective specialties is essential for optimal clinical outcomes.

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