

Case Report

Ilio-psoas Bleed mimicking Acute Appendicitis in a Newly Diagnosed Haemophilic; A Case Report of Near Miss Surgical Catastrophe

Dalha Gwarzo Haliru, Musa Aminu, Fatima Usman Zangina, Halima Isma'il.

Department of Haematology, Bayero University, Kano/Aminu Kano Teaching Hospital, Kano

Abstract

Ilio-psoas hemorrhage is a serious complication in patients with Haemophilia. Bleeding into the ilio-psoas muscle can mimic acute appendicitis clinically. Ilio-psoas hematoma is commoner in patients with Haemophilia than the general public and unlike the general public where bleeding is secondary; it is usually spontaneous in haemophiliacs. We present a case of a 55-year-old man referred to the emergency room with a history of abdominal pain and distention. We took a detailed history which led to a suspicion of Ilio-psoas hematoma likely due to a bleeding disorder. Clotting profile and mixing studies supported the diagnosis of Haemophilia A. The diagnosis of Ilio-psoas hematoma in Haemophilia is still a challenge and requires a high index of suspicion. It also requires careful history taking, a detailed clinical examination and performing basic investigations to distinguish ilio-psoas bleeding from other causes of acute abdomen like acute appendicitis.

Keywords: Haemophilia, Ilio-psoas, Appendicitis, Clotting Factors

INTRODUCTION

Haemophilia is the second most common inherited bleeding disorder after Von Willebrand's disease.¹ About 1 in 5,000 and 1 in 20,000 male live births are expected to have Haemophilia A (HA) and B (HB) respectively.² It is an X-linked recessive disorder; divided majorly into HA (Factor VIII deficiency) and HB (Factor IX deficiency) which are clinically indistinguishable.^{1,3} The pathology lies in having a decreased and/or a defective clotting factor (CF).¹ The usual presentation is prolonged bleeding following injury and deep seated bleeds into muscles and joints, and the absence of muco-cutaneous bleeds.⁴ Patients are categorized depending on the residual CF in their plasma into mild, moderate or severe.^{1,2} The lower the level of the residual CF, the more severe the clinical features.⁴ Therefore, those with mild disease bleed only following trauma, while those with severe disease bleed spontaneously. Ilio-psoas haematoma in patients with Haemophilia is a well-known complication that limits the functional capacity of the patient.

The aim of this case report is to emphasize the need for a proper history taking, physical examination and investigations prior to any invasive procedure.

Correspondence:

Dalha Gwarzo Haliru. Department of Haematology, Bayero University, Kano/Aminu Kano Teaching Hospital, Kano. ORCID-0000-0002-8082-352X dalhatugwarzo@yahoo.com +2348036241535

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CASE REPORT

We present a 55-year-old man referred to emergency room of our hospital with 11-day

history of abdominal pain and distension. There was associated history of initial passage of melena, haematemesis and haematochezia. No history of bleeding from other orifices, no history of peptic ulcer disease nor history of chronic liver disease. He received 4 units of whole blood at the referring hospital. He was initially reviewed by the surgical team; he was acutely ill, in painful distress with severe pallor. Vital signs were stable with a pulse rate of 100bpm and blood pressure of 100/80mmHg. Abdomen was uniformly distended with generalised tenderness and guarding marked around the right iliac fossa (RIF) and the right lumbar region. A palpable mass of about 8cm x 8cm in the right iliac fossa was noted.

The abdominal tenderness prevented further examination. Digital rectal examination revealed dark coloured stool. Abdomino-pelvic Ultrasound Scan (USS) showed intra-peritoneal fluid collection in the RIF suggestive of a ruptured appendix. Full blood count result showed haemoglobin of 5.9g/dl, mild neutrophilic leucocytosis with WBC count of $13 \times 10^9/L$ and absolute neutrophil count of $11.5 \times 10^9/L$. Platelet count and renal function test were essentially normal.

A diagnosis of generalized peritonitis due to a ruptured appendix in a patient at risk of intra-operative bleeding was made. He was prepared and counseled for emergency surgery. Patient then volunteered a history of prolonged bleeding following injury. Consequently, haematologists were invited to review. We documented additional history of recurrent haematomas, haemarthroses and a family history of prolonged bleeding. His younger sibling died following prolonged post-circumcision bleeding.

Furthermore, we established history of repeated use of non-steroidal anti-inflammatory drugs (NSAIDs) due to the recurrent haemarthroses and its use just before this presentation. On examination, there was evidence of bilateral haemarthropathy; right knee affected more than the left. We requested for a clotting profile which showed isolated prolongation of APTT. Mixing studies showed correction with both normal and Factor IX deficient plasmas and a failure to correct with Factor VIII (FVIII) deficient plasma. We made an impression of Ilio-psoas bleed in a newly diagnosed haemophilia A patient. We did not have facilities to do CF assay. We ordered for a repeat Abdomino-pelvic USS, providing a more detailed clinical history which confirmed the diagnosis.

Patient was managed with recombinant FVIII. Initially, his FVIII level was raised to 100% 12hourly for 3 days and then subsequently to 50% 12hourly for additional 4 days. Patient got better

and was later discharged to outpatient haematology clinic.

DISCUSSION

Haematomas are a common presentation of patients with haemophilia accounting for about 25% of bleeds.⁵ Ilio-psoas bleed has been reported in this group of patients. Due to the fact that it can mimic acute appendicitis; it is critical to distinguish the two conditions.⁶ Failure to separate them can lead to a surgical catastrophe with the patient bleeding uncontrollably and a possibility of death on table. History taking and the use of USS can easily delineate the diagnosis.⁷ Although diagnosis of haemophilia can be made using simple tests, it is still a challenge in Nigeria, as well as in several other developing countries.

This case classically depicts the diagnosis of Haemophilia for the first time at the age of 55 years. The decision to invite haematologists to review this patient was really important. This stresses the benefit of multi-disciplinary approach in patient's management. This patient could have ended up having a surgical procedure he did not need with all the possible dreaded complications. Use of NSAIDs in Haemophiliacs should be avoided as much as possible, as it predisposes them to peptic ulceration and a higher propensity to haemorrhage.⁸

We strongly believe that this index presentation was provoked by the use of Ibuprofen. Furthermore, the more specific USS diagnosis of Ilio-psoas haemorrhage following the repeat investigation was obtained mainly because of the detailed clinical history provided to the sonologist. This justifies the call for provision of adequate history by clinicians while requesting for investigations.

CONCLUSION

Diagnosis of Haemophilia is still a challenge in our setting. This case highlights the importance of a high index of suspicion to make the diagnosis of ilio-psoas bleeding. In a patient with bleeding tendency, the history of RIF pain and difficulty in walking with presence of a mass in the RIF gives a clue to the diagnosis of ilio-psoas bleeding. Detailed history taking, physical examination and basic investigations will help in making the correct diagnosis, thus preventing any untoward effects of a rushed surgical procedure.

The authors have declared no conflict of interest

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