Abstract

Background: Haemophilia A is a hereditary haemorrhagic disorder that can cause bleeding in the intestinal loops and, on rare occasions, simulate an acute surgical abdomen. Careful assessment of coagulation must be performed in these patients, followed by an attempt to correct the dysfunctions. Often, the administration of the deficient factor is sufficient to resolve the problem, avoiding unnecessary surgeries.

Case report: We present a male patient, 15-years-old, of indigenous descent, who was diagnosed with haemophilia A. The young man was admitted with abdominal pain in the right iliac fossa; ultrasonography suggested acute appendicitis. He underwent an exploratory laparotomy that revealed a normal appendix and the presence of a caecal wall haematoma, without other abnormalities.

Conclusion: This case describes an unusual instance of decompensation of a patient with haemophilia A that simulated an acute surgical abdomen. The case suggests the need for further evaluation of carriers of coagulopathies, whether acquired or congenital, when they suffer abdominal pain. Otherwise, clinically treatable dysfunctions are prone to surgical treatment, with a potential for increased morbidity and mortality.

Keywords
Acute abdomen, haemophilia A, factor VIII, TAP block, coagulopathy
Background

Haemophilia A is a hereditary disease linked to the X chromosome and occurs in 1 out of 10,000 men [1]. This coagulopathy is caused by a factor VIII deficiency that manifests as spontaneous bleeding of the joints and tissues, including those forming the digestive and nervous systems [2]. These patients may develop conditions that require surgical treatment, whether they are related to the underlying pathology or to another disorder [3]. Haemorrhagic complications in the small or large intestine may simulate an acute surgical abdomen, causing a potential increase in morbidity and mortality [4]. The case described involves the surgical treatment of an abdominal problem, suggestive of appendicitis, in a patient with haemophilia A [5].

Case presentation

A 15-year-old male of indigenous descent was admitted to hospital with an 8-day history of abdominal pain. The patient had been previously diagnosed with haemophilia A, and was being monitored by the haematology service of another department. His pain began in the epigastric region and remained localized to the right iliac fossa. Over time, he developed vomiting, asthenia, anorexia, and abdominal guarding. As a result, he underwent abdominal ultrasonography, which suggested appendicitis. Abdominal tomography suggested a lesion to the right of the psoas muscle, but the appendix could not be visualized (figure 1 and figure 2). The results of a laboratory exam revealed a haemoglobin level of 9.4 g/dL, a leucocyte count of 16,000/µL, a left-shifted leukogram; an international normalized ratio of 1.21; an activated partial thromboplastin time (APTT/INR) of 2.86; and a platelet count of 326,000/µL. The patient’s renal and liver function tests were within normal limits [6].

The patient received 3000 IU of factor VIII, preoperatively, and was anaesthetised with propofol, fentanyl, cisatracurium, and lidocaine; anaesthesia was maintained with 2.5% sevoflurane. Standard monitoring, using pulse oximetry, electrocardiography, non-invasive blood pressure measurement, and capnographic analysis of inhaled and exhaled gases, was continuously performed. A midline incision was performed and a normal caecal appendix was visualized. After inspection of the loops, extensive haematoma was found in the caecum, near the base of the appendix (figure 3). An appendectomy was performed, with layered suturing, and a drain was placed (figure 4). At the end of the procedure, a bilateral transverse ab-
dominis planeblock was performed with 20 mL of 0.5% ropivacaine. The patient awoke well, without pain or signs of bleeding. Post-surgically, his APTT was again determined and another dose of factor VIII was administered.

Discussion

Hemophilia A and B are hereditary diseases that result from functional deficiencies in specific circulating blood clotting factors termed factor VIII and factor IX [7]. The gene for factor VIII is located on the X chromosome and is relatively large. Changes resulting from deletion or inversion portions of the genome or changing wrong direction, result in factor VIII activity below 1% of normal, causing severe forms of the disease. Point mutations and smaller depletions result in blood levels of Factor VIII larger than 1% and milder disease [8].

The clinical severity is defined by the factor VIII activity, generating losses in the intrinsic coagulation, extending the partial thromboplastin time (APTT) [9].

The disease can present intramural hematoma of the bowel, simulating acute abdomen, or depending on the extension of the hematoma, leading to intestinal muscles paralysis, with rare chance, however, possible to evolve as bowel obstruction and necrosis of bowel segments [10]. In addition, the carrier of hemophilia A is subject to abdominal surgical pathologies common as suspected appendicitis in our case, the clinical symptoms (nausea, vomiting and abdominal pain well located in the right iliac fossa with the presence of positive Blumberg sign). In addition, features high value of leukograma, suggestive of an infectious process (16,000 leukocytes per mcL) which led to a strong suspicion of appendicitis.

It was contemplated the possibility of intramural hematoma handles, however, there was no tomographic confirmation and held Factor VIII replacement at a dose of 30 IU/kg 8/8 h without worsening of symptoms [11].

For any surgical intervention it is important to determine the factor VIII level and make appropriate preoperative replacement to ensure good surgical hemostasis [12]. In our case, the APTT was with INR 2.86. In this sense, well above suitable for surgery, including being absolute not indicated to the central regional block (spinal-anesthesia or epidural anesthesia) for anesthesia or postoperative analgesia [13].

The ideal previous dose of Factor VIII to allow any surgery is 50-60 U/Kg. Nevertheless, even adequate replacement of Factor VIII, is not indicated to the central anesthetic block. For this
reason we chose general anesthesia, followed by TAP Block guided by ultrasound to postoperative analgesia, which was adequate.

The case report suggests that, in frames of acute abdomen, even with clinical and laboratory evidence of mimicking appendicitis, suspected intramural hematoma handles, which, in most cases resolves with appropriate replacement of Factor VIII (deficient) [14]. Clinical treatment avoids surgery, high morbidity and mortality factor in these patients.

**Conclusion**

In patients with coagulopathies, particularly the more severe coagulopathies, who present with abdominal pain, complications of the underlying disease must always be assumed [6, 12]. The complications may include retroperitoneal hematoma, hematoma of the intestinal loops or hematoma of the abdominal musculature [7, 8, 14]. These complications may arise spontaneously or after minor trauma, simulating an acute abdomen [9]. Often these complications can be reversed with appropriate clinical treatment [10, 11, 15].

General anaesthesia is indicated in these patients. Central blocks as spinal anesthesia or epidural anesthesia are not indicated in patients with coagulopathy patients at high risk of progressing to spinal hematoma. In exploratory laparotomy, always related to the possibility of holding the TAP Block guided by ultrasound with local anesthetics long term as bupivacaine or ropivacaine for postoperative analgesia (Figure 5).

**Consent**

Written informed consent was obtained from the patient’s mother for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

**Figure 5:** Displaying ultrasound showing the structures for carrying out the TAP Block. From top to bottom: external oblique, internal oblique muscle and transverse abdominis. The anesthetic is deposited between the internal oblique and transversus abdominis in volume of 20 ml. The innervation of the lower abdomen spend all that space.

**Ethics questions**

The current study was approved by the ethical committee of the Regional Hospital of Cariri, for case presentation.

**List of abbreviations**

- APTT=activated partial thromboplastin time
- TAP= transversusabdominis plane

**Competing interests**

The authors declare no conflicts of interest. All research was conducted with their own resources.
Financial resources
The authors state that they used their own resources and did not receive financial aid from any institution.

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Authors' contributions
HMTB conceived the study. ICPN, MPLN, GBM DE S, WRP, IMPB, JFC, LTA, RDR, LCA analysed the results and helped to draft the document. All authors read and approved the final manuscript.

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References


