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Case Report

Spontaneous retroperitoneal hematoma, an uncommon case of acute surgical abdomen in a patient with hemophilia: case report

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ABSTRACT

Hemophilia A is an X-linked hereditary disorder with a predilection for men. A worldwide incidence of 1 in 5 000 inhabitants is estimated. Currently, there is no curative treatment, which can lead to the development of multiple complications. However, abdominal presentations related to acute abdomen are uncommon and require surgical treatment. Similar cases are related to psoas muscle hematoma and hemophilic pseudotumors. We presented a 25-year-old male diagnosed with hemophilia A with acute abdominal pain and peritoneal irritation signs. After evaluation, we decided to perform a laparotomy, which reported a hematoma in the right mesocolon from the ileocecal valve to the hepatic flexure with right colic artery disruption. After seven days, he was discharged due to clinical improvement, and we indicated an outpatient general surgery follow-up. Acute abdomen caused by a retroperitoneal hematoma in patients with hemophilia is uncommon. Therefore, its management is still controversial. We concluded that a surgical approach might reduce the risk of complications in patients with poor clinical courses.

Keywords: Hemophilia A, Hematoma, Mesocolon, Acute abdomen, Case report

INTRODUCTION

Hemophilia A is an X-linked hereditary disorder with a predilection for men. A worldwide incidence of 1: 5000 males is estimated and there are approximately 6022 patients reported in Mexico, according to the Mexican Hemophilia Federation.[1] Currently, there is no curative treatment.

The most common complications include neurological, musculoskeletal and infectious complications. However, abdominal presentation which provokes an acute abdomen and requires surgical intervention is infrequent. Among the reported cases are Iliopsoas muscle hematomas.[2]

CASE REPORT

We presented a 25-year-old male who was diagnosed with hemophilia A two years ago and under treatment with factor VIII primary prophylactic, with no other medical history. He was admitted because of a generalized progressive abdominal pain developed for 24 hours with a pain intensity of 9 and associated to adrenergic symptomatology, no aggravating or provoking factors identified. During clinical examination we described a flat abdomen with absent bowel sounds and painfully palpation predominantly on the right hemiabdomen, positive Blumber's sign and peritoneal irritation signs.

At presentation, the complete blood count reported hemoglobin of 14.3 g/dl, a white blood cell count of 16.6×103, platelet count of 2,86,000 and hematocrit of 40.5%. The coagulation results revealed a prothrombin time of 11, partial thromboplastin time of 62.4 with an INR of 0.96 and a fibrinogen of 341. Laboratory findings are shown in Table 1.

Table 1: Laboratory findings.

Laboratory tests	Values	Normal range
Hemoglobin (g/dl)	14.3	12.50-16.80
Hematocrit (%)	40.5	36-47
Platelets (×10^3 µl)	286	142-424
Leukocytes (×10^3 µl)	16.6	4.07-11.23
Neutrophils (%)	87.7	37-80
Glucosa (mg/dl)	117.5	74-106
BUN (mg/dl)	8.6	7-25
Urea (mg/dl)	18.4	17-43
Creatinina (mg/dl)	0.7	0.66-1.09
Uric acid (mg/dl)	7.7	2.60-6
Total cholesterol (mg/dl)	211.11	0-200
Triglycerides (mg/dl)	397	0-150
Total bilirubin (mg/dl)	0.5	0.30-1.20
AST (u/l)	36	0-35
ALT (u/l)	54	0-35
LDH (u/l)	259	140-271
Alkaline phosphatase (u/l)	92	30-120
GGT (u/l)	89	9-64
Albumin (g/dl)	5	3.50-5.20
Total protein (g/dl)	8.1	6.60-8.30
Globulin (g/dl)	3.1	2.20-3.90
Albumin/globulin ratio	1.6	1.20-2.20
Sodium (meq/l)	137.8	136-145
Potassium (meq/l)	3.4	3.50-5.10
Chloride (meq/l)	104	98-107
Calcium (mg/dl)	8.9	8.60-10.30
Phosphate (mg/dl)	3.2	2.50-4.50
Magnesium (mg/dl)	2.0	1.90-2.70
Amylase (u/l)	65	22-80
Lipase (u/l)	25	0-67
Prothrombin time	11	9.10-12.10
INR	0.96	0.80-1.10
Partial thromboplastin time	62.4	<40
Fibrinogen (mg/dl)	341	180-350

Abdominal ultrasound reported free fluid in the right lower quadrant. Pelvic computed tomography during single-phase and with contrast medium showed a collection of approximately 64×35 mm with free fluid (Figure 1 and 2). We administered factor VIII before the exploratory laparotomy suspecting a bleeding hemophilic pseudotumor. We found an hemoperitoneum of approximately 1200 with a hematoma extended to right mesocolon, cecum, ascending colon and the splenic flexure. We also discovered infiltration through the zone II, according to Kuds and Sheldon's retroperitoneal hematoma classification, and right hemicolon changes suggestive of ischemia related to the disruption and thrombosis of the middle colic artery (Figure 3). Therefore, we performed a right hemicolectomy with manual ileotransverse end-to-side anastomosis in two planes without any 'never events''.

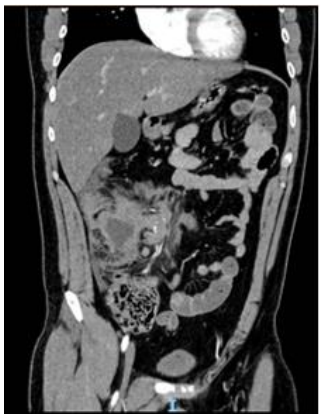

Figure 1: Single-phase coronal computed tomography up to 25 UH with a reinforced heterogeneous collection.

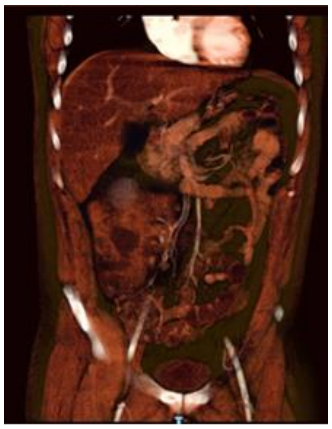

Figure 2: Coronal computed tomography with an heterogeneous collection and middle colic artery sudden course interruption.

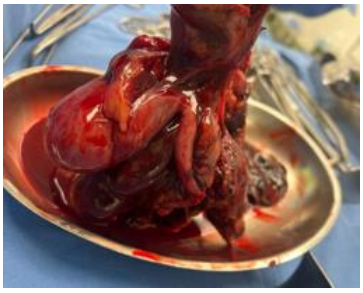

Figure 3: Surgical specimen with infiltrative hematoma and multiple areas of ischemia obtained from right hemicolectomy.

The patient was hospitalized for further observation and we administered factor VIII at a dose of 50 UI/kg alongside antibiotics and analgesics. We kept the patient fasting for 72 hours and began enteral alimentation. We discharged him on the seventh day due to clinical improvement and we indicated an outpatient general surgery follow-up.

Histopathological examinations showed multiple hematomas in mesocolon, which the biggest measured 5.5×4.5 cm in diameter and the smallest measured 3×2 cm, non-specific chronic colitis with thrombi in mesocolon and necrosis areas without malignancy signs.

DISCUSSION

Abdominal injuries leading to massive bleeding in patients with hemophilia A are infrequent, although some cases have been reported with perforation without signs of trauma. Low-energy trauma transmitted to any solid viscera might lead to a pericapsular hemorrhage, intestinal occlusion or perforation.[3]

Identifying the opportune moment for surgical management is still in discussion, since its evolution may be limited or compromise the hemodynamic status despite initial resuscitation with crystalloid solutions and/or hemoderivatives. In these cases, in which there is no defined management, when evolution is unfavorable and the patient's clinical condition meets the criteria, immediate surgical management by exploratory laparotomy may be indicated to remove or repair the involved organ.[3] Due to the lack of experience in these situations, the risk of an expansive hematoma and imminent rupture may appear despite the absence of evidence of previous trauma.[4] Although infrequent, its possibility should not be ruled out.

The most similar cases to an abdominal bleeding in patients with hemophilia A are hematomas in the psoas and obturator muscle, which are usually self-limited and rarely require surgical management.[2] The usual clinical presentation of bleeding is usually intra-articular. Instead, spontaneous retroperitoneal bleeding associated with hemophilia A is infrequent. They usually require conservative management for three to four weeks, in addition to an individualized dose of factor VIII. Some cases have been reported with a surgical approach, such as in duodenal hematomas associated with some degree of trauma as it might occur at endoscopy.[6] The incidence has been reported to be as high as 0.1% and as low as 0.00008% (1:1250 patients).[6]

It is important to determine surgical intervention in patients with clinical and laboratory data with a torpid evolution alongside peritoneal irritation signs. The clinical presentation is usually nonspecific with abdominal pain of sudden or gradual onset, generalized and intense associated with adrenergic discharge and hemodynamic instability. The imaging study of choice when intra-abdominal bleeding is suspected is computed tomography, which is comparable to conventional angiography that allows detecting bleeding and identifying as arterial or venous.[4,7] Currently there is no consensus to determine the specific surgical approach according to the degree of injury and the affected organ.[8] Therefore, it is important to determine the degree of damage to the affected organ in order to decide the management, which can range from drainage of the hematoma to intestinal resection with a primary or secondary anastomosis.

CONCLUSION

Spontaneous retroperitoneal hematomas in patients with a medical history of hemophilia are infrequent. However, in patients with a poor clinical course, peritoneal irritation signs and evidence of free fluid or indirect signs of an expensive hematoma are absolute criteria for surgical approach which reduces the risk of complications.

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