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An intra-abdominal pseudotumor is a rare complication of hemophilia. Surgical treatment is associated with high morbidity and mortality rates and reported cases are scarce. We present a 66-year-old Caucasian male suffering from severe hemophilia type A treated for 10 years with Factor VIII. Major complications from the disease were chronic hepatitis B and C, cerebral hemorrhage and disabling arthropathy. Twenty-three years ago, retro-peritoneal bleeding led to the development of a large intra-abdominal pseudotumor, which was followed-up clinically due to the high surgical risk and the lack of clinical indication. The patient presented to the emergency department with severe sepsis and umbilical discharge that had appeared over the past two days. Abdominal computed tomography images were highly suggestive of a bowel fistula. The patient was taken to the operating room under continuous infusion of factor VIII. Surgical exploration revealed a large infected pseudotumor with severe intra...

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Successful emergency resection of a massive intra-abdominal hemophilic pseudotumor

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Author contributions: Coubeau L, Hubert C and Frezin J performed the surgery; Frezin J and Marique L reviewed current literature and wrote the paper; Lambert C, Hermans C and Jabbour N coordinated the paper elaboration and revised the article.

Ethics approval: The study was reviewed and approved by the Institutional Review Board of the Université catholique de Louvain.

Informed consent: The patient gave his verbal consent for reporting this case.

Conflict of interest: None.

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Abstract
An intra-abdominal pseudotumor is a rare complication of hemophilia. Surgical treatment is associated with high morbidity and mortality rates and reported cases are scarce. We present a 66-year-old Caucasian male suffering from severe hemophilia type A treated for 10 years with Factor VIII. Major complications from the disease were chronic hepatitis B and C, cerebral hemorrhage and disabling arthropathy. Twenty-three years ago, retro-peritoneal bleeding led to the development of a large intra-abdominal pseudotumor, which was followed-up clinically due to the high surgical risk and the lack of clinical indication. The patient presented to the emergency department with severe sepsis and umbilical discharge that had appeared over the past two days. Abdominal computed tomography images were highly suggestive of a bowel fistula. The patient was taken to the operating room under continuous infusion of factor VIII. Surgical exploration revealed a large infected pseudotumor with severe intra-abdominal adhesions and a left colonic fistula. The pseudotumor was partially resected en bloc with the left colon leaving the posterior wall intact. The postoperative period was complicated by septic shock and a small bowel fistula that required reoperation. He was discharged on the 73rd hospital day and is well 8 mo after surgery. No bleeding complications were encountered and we consider surgery safe under factor VIII replacement therapy.

Key words: Hemophilia A; Hemophilic pseudotumor; Colonic fistula; Factor VIII replacement therapy; Surgery in hemophilic patient

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Core tip: We present a patient suffering from hemophilia A complicated by a large intra-abdominal hemophilic pseudotumor. This condition is rare and there is no consensus for treatment. Emergency resection was required because of bowel complications and septic shock. Based on our experience, we recommend elective surgery prior to complications under appropriate factor
The laboratory tests were significant for C spontaneous fecal discharge from the umbilicus distension without guarding, no bowel sounds and distension. Physical examination showed abdominal recent onset of diarrhea, vomiting and abdominal was admitted to the emergency room for sepsis, responded to conservative treatment.

Intra-tumoral bleeding with hemorrhagic shock that to the current episode, the patient was hospitalized for the past 10 years, spironolactone 25 mg once a concentrate 2000 IU (Refacto AF®, Pfizer) twice a week

Hemophilia A is a congenital disease with an estimated incidence of 10 to 20 cases for 100000 males¹. Spontaneous bleeding and hemorrhaxis are the most frequent complications. Hemophilic pseudotumors are less common, found in severe cases of hemophilia (1%-2%) and mainly located in the limbs². Pseudo-tumors consist of encapsulated, chronic, slowly expanding hematomas. Abdominal pseudotumors are rare and their management is still controversial. Replacement therapy is often the first therapeutic approach. However, surgery is the most effective and the only definitive treatment² even though it is associated to high morbidity and mortality rates³. Therapeutic alternatives include radiotherapy, percutaneous drainage, embolization and external radiation². We describe our experience in the surgical management of a large intra-abdominal hemophilic pseudotumor.

Our patient is a 66-year-old Caucasian male with severe hemophilia A type C complicated by arterial hypertension, chronic hepatitis B and C, a cerebral hemorrhage 20 years ago and many spontaneous bleeding episodes causing disabling diffuse arthropathy. His medical treatment includes recombinant factor VIII concentrate 2000 IU (Refacto AF®, Pfizer) twice a week for the past 10 years, spironolactone 25 mg once a day and painkillers.

At the age of 44, an intra-abdominal bleeding episode led to the development of an intra-abdominal hemophilic pseudotumor, which was treated conservatively as it was asymptomatic. Follow-up by annual abdominal computed tomography (CT) showed stability of the tumor with no sign of complication. A year prior to the current episode, the patient was hospitalized for intra-tumoral bleeding with hemorrhagic shock that responded to conservative treatment.

This most recent event started when the patient was admitted to the emergency room for sepsis, recent onset of diarrhea, vomiting and abdominal distension. Physical examination showed abdominal distension without guarding, no bowel sounds and spontaneous fecal discharge from the umbilicus (Figure 1). The laboratory tests were significant for C reactive protein 300.9 mg/L, white blood cell count 17490/L, hemoglobin 10.8 g/dL, platelets 429000/µL, prothrombin time 15.7 s (normal 8.6-13.8 s), factor VIII 59% (after factor VIII replacement). Abdominal CT showed air within the tumor (Figure 2). The patient was brought to the operating room. A bolus of 2000 IU factor VIII infusion (Refacto AF®, Pfizer) was given before incision followed by continuous infusion of factor VIII (6000 U per day). Abdominal exploration through a midline incision revealed severe adhesions between the pseudotumor and the small and large bowel. There was no peritonitis. The cyst was filled with stools and old clots and a fistula between the left colon and the tumor was found. Intra-operative management consisted of left hemicolectomy en bloc with the cyst. The posterior cyst wall was left in situ because of involvement of the aorta, inferior vena cava and right ureter. A terminal colostomy was performed, including a Hartmann pouch. Hemostasis was achieved easily. The abdomen was closed after saline irrigation, and 3 drains were left in place. The total operative time was 353 min and the patient was transfused 6 units of red blood cells.

In the immediate postoperative period, the patient was given a continuous infusion of 6000 U/d of factor VIII in order to maintain factor VIII levels between 30% and 40%. On the 3rd postoperative day, he developed a small bowel fistula complicated by intra-abdominal sepsis that required surgical re-exploration. A T-tube was inserted in the fistula site and later served as a feeding jejunostomy.

On the 55th postoperative day, the patient underwent a negative re-exploration because of an inflammatory syndrome with abdominal CT scan findings of portal venous gas and intestinal pneumatosis. He recovered well postoperatively, and quickly resumed enteral feeding. He was discharged to a rehab facility on the 73rd postoperative day.

The patient had no bleeding complication during his hospital stay. Eight months after surgery, he is well.

Hemophilia is a group of inherited blood coagulation disorders. The mutation is located on the X chromosome, therefore only men are afflicted and women are carriers. Depending on the remaining level of factor VIII activity, hemophilia A is classified as mild (> 5%), moderate (1%-5%) or severe (< 1%)¹. Most common complications are spontaneous bleeding into joints and muscles⁴. Hemophilic pseudotumors are rare complications of hemophilia occurring in approximately 1% to 2% of patients suffering from severe hemophilia¹,². The pseudotumor is caused by recurrent bleeding episodes into bone or soft tissue leading to the formation of an encapsulated mass of clotted blood and necrotic tissue. In children, pseudotumors are most likely to occur in the limbs or jaw bone, whilst in adults they...
previously reported in the literature in favor of surgery in hemophilic patients with continuous or interrupted infusion of clotting factors with minimal risk of hemorrhage. The particularly large size of the cyst (over 30 cm in diameter) and the duration of its evolution made the resection hazardous in the present patient. The fact that surgical long-term outcome was favorable despite the patient’s complications and emergency context should encourage elective resection.

We report a rare case of successful emergency resection of a large abdominal hemophilic pseudotumor. Abdominal surgery in a hemophilic A patient is feasible without hemorrhagic complications under continuous factor VIII replacement therapy. Elective resection of an abdominal hemophilic pseudotumor should be considered prior to the development of major complications.

COMMENTS
Case characteristics
A hemophilic patient known for a large intra-abdominal hemophilic pseudotumor presenting with abdominal distension, umbilical fecal discharge and vomiting.

Clinical diagnosis
Physical examination showed signs of severe sepsis and abdominal distension without guarding.

Differential diagnosis
Peritonitis, intestinal obstruction, pseudotumor infection, bowel fistula.

Laboratory diagnosis
C-reactive-protein (300.9 mg/L), white count (17490/L), hemoglobin (10.8 g/dL), platelets (429000/µL), prothrombin time (15.7 s), factor VIII (59%).

Imaging diagnosis
Abdominal computed tomography showed air within the pseudotumor.

Pathological diagnosis
Pathological analysis of the surgical specimen confirmed the diagnosis of large bowel fistula into the hemophilic pseudotumor.

Treatment
Emergency exploratory laparotomy was performed. The pseudotumor was resected en bloc with the left colon because of the presence of a large bowel fistula.

Related reports
Few cases of successful resection of intra-abdominal hemophilic pseudotumors have been reported. The treatment of those tumors is still controversial.

Term explanation
A hemophilic pseudotumor is a chronic slowly expanding hematoma. An intra-abdominal hemophilic pseudotumor can lead to severe complications such as massive bleeding or bowel fistulas.
Experiences and lessons
Our case describes the management of a complicated intra-abdominal hemophilic pseudotumor requiring an emergency resection. The patient was discharged after 73 d and 2 further exploratory laparotomies due to complications. The authors recommend resection before complication, under appropriate factor VIII replacement therapy.

Peer-review
In this report, the authors described a patient with a hemophilic intraabdominal pseudotumor.

REFERENCES