

CASE REPORT

Iliopsoas hemophilic pseudotumor with bowel fistulization

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Abstract: Haemophilic pseudotumors are usually observed in the diaphysis of long bones. Pseudotumors due to psoas muscle hematoma are rare and surgical management is difficult. Surgical treatment of these lesions is usually associated with high morbidity and mortality rate. Here, we present a case with iliopsoas haemophilic pseudotumors with bowel fistulization who underwent three abdominal operations and survived. Based on our experiences in this patient, we recommend to wait for the intraabdominal hematoma and adhesions to resolve and organise, so that the dissection can be kept to a minimum, which decreases the chances of iatrogenic injury and surgical bleeding (Fig. 3, Ref. 15). Full Text (Free, PDF) www.bmj.sk.

Key words: haemophilia, pseudotumor, psoas muscle, hematoma, bowel fistulization, enterocutaneous fistula.

Hemophilic pseudotumors are usually seen in the diaphysis of long bones. Pseudotumors caused by psoas muscle hematoma are rare and their surgical management is quite challenging. This difficulty is primarily due the inherent properties of underlying disease which increases the risk of bleeding and the dense adhesions formed between the pseudotumor and visceral organs, which increases the likelihood of iatrogenic bowel injuries. We present a case with iliopsoas hemophilic pseudotumor with bowel fistulization, and discuss our experience related to medical and surgical management of this rare entity (1–3). Upon recurrence of abscess after percutaneous drainage, the patient required three consecutive operations and recovered well. The patient's initial presentation, imaging workup, and intraoperative and postoperative courses are discussed.

Case

The patient, 61 years old male has been followed with diagnosis of hemophilia A since 1960, in our Hematology Department. Hemophilia was diagnosed in another hospital when he was 2 years of age. He suffered a right iliac fossa hematoma in 1985, and was treated with factor VIII concentrates. In 2001, he underwent bilateral prosthetic total knee replacement with factor VIII concentrates infusion and he was discharged to home without any complication (4–6). His history was remarkable for several factor VIII concentrates infusions from time to time due

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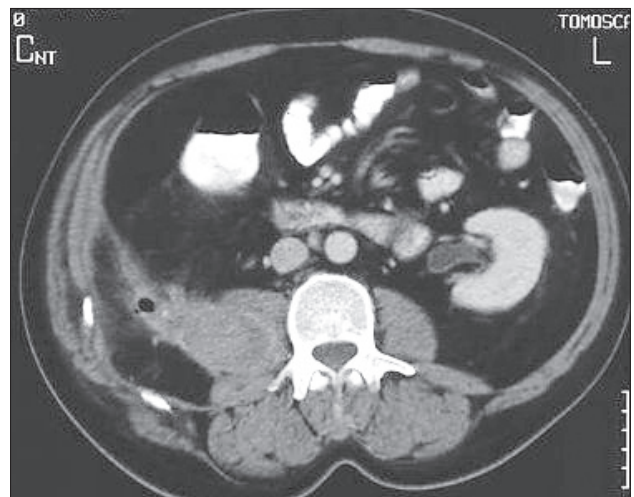


Fig. 1. Contrast enhanced abdominal CT shows the right retroperitoneal abscess.

to bleeding episodes in his various joints. He recovered well from the bleeding episodes with no limitations in his daily activities.

The patient was admitted to another hospital with a history of high fever for 3 days and a painful right lower quadrant mass. Presuming a septic focus parenteral antibiotic treatment was instituted and the patient was referred to our hospital. On admission, his hemoglobin level was 8.2 g/dL. He was transfused with packed red cells and factor VIII concentrates.

Physical examination was unremarkable besides high fever (39 °C) and a painful right lower quadrant mass approximately 10 cm in diameter. The biochemical and urinary examinations were within normal limits.

Contrast enhanced abdominal computed tomography (CT) demonstrated a right retroperitoneal abscess with dimensions of



Fig. 2. After 10 days of drainage, CT image shows no residual collection.

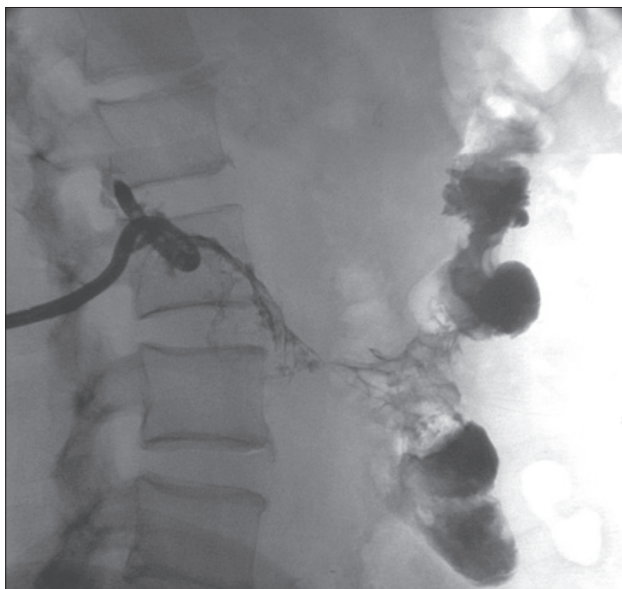


Fig. 3. Abscessogram shows a fistulous communication between the abscess cavity and right colon.

8 by 5 by 5 cm (Fig. 1) This abscess was catheterized percutaneously under ultrasound and fluoroscopy guidance with IV sedation. A 14 Fr pigtail drainage catheter (Flexima APDL, Boston Scientific, USA) was placed with Seldinger technique and a total of 100 ml purulent-hemorrhagic fluid was aspirated. Parenteral sulbactam ampicillin therapy was started. *E. coli* and *Morganella* species were isolated from the samples. After the second day of drainage, the fever subsided and the mass became progressively smaller. On the 10th day control tomography confirmed the absence of residual collection (Fig. 2) but abscessogram obtained with gravity drainage showed a fistulous communication between the abscess cavity and right colon (Fig. 3). Daily drainage de-

creased progressively but the catheter dislodged spontaneously on the 50th day. Total amount of drainage was about 650 ml at that time. Since the daily output was less than 5 ml/day and the patient's clinical course was uneventful, conservative management was decided. His factor VIII level, which was 3 % with no inhibitor at the beginning, was maintained at around 100 % during the hospital stay (5–8). He was discharged on day 61 of admission.

Two weeks later he was admitted to the hospital for the second time with the same clinical and radiological findings plus rectal bleeding. Abdominal tomography confirmed abscess recurrence which was percutaneously drained. There was no communication with the colon. After drainage and factor VIII replacement the patient discharged on the 15th day of his second hospitalization.

Three weeks later the patient presented with the recurrence and Recatheterization and factor VIII replacement were performed. Abscessogram revealed colonic fistula and surgical exploration was planned.

The patient was operated on the 149th day of the first admission. Operation was covered with factor VIII concentrates. The abdomen was entered through a vertical midline incision. Exploration revealed dense fibrous adhesions in the right lower quadrant which has converted distal small bowel and ascending colon into a conglomerated mass. The adhesions were taken down with meticulous dissection to avoid iatrogenic injury to bowel and taking great care for haemostasis. Upon mobilization of right colon, a thick fibrous fistula tract was identified between cecum and lateral abdominal wall. The percutaneous catheter was palpated within the fistula tract, and was removed. The fibrous fistula tract was detached from cecum by taking care to leave a small defect with clear margins. The defect was closed with two seromuscular interrupted sutures. Excision of fistula tract revealed a piece of synthetic material which was thought to be the prolene string that has been detached from the percutaneous catheter. The lateral abdominal wall where the fistula communicates with skin was debrided, and after the control of haemostasis, the abdomen was closed.

The early postoperative course was uneventful. The patient continued to receive factor VIII concentrates. On the sixth postoperative day, his body temperature was elevated to 39 °C, and based on the previous culture results, broad spectrum antibiotic therapy was instituted parenterally. On the seventh postoperative day the patient became tachycardic. His hemoglobin level showed a progressive decrease and his abdomen became distended. On reoperation, a hematoma of approximately one liter was found and evacuated. There were patchy foci of minimal bleeding. Haemostasis was performed and the abdomen was closed. On the 14th day the patient was reoperated due to rebleeding and haemostasis was performed. Before closure of the abdomen all peritoneal surfaces were washed with Tranexamic acid, expecting potential benefit [8]. The following postoperative course was uneventful and the patient was discharged. The patient was followed for 3 years postoperatively and has been doing well.

Discussion

Hemophilic pseudotumors are rare clinical entities. Although a benign process, a hemophilic pseudotumor can be complicated with life threatening complications. They occur in 1–2 % of the patients with severe FVIII or FIX deficiency or in patients with inhibitors (9–13). They arise from subperiosteal or intramuscular bleeding, which forms a blood filled capsulated cystic mass. They are mostly seen in pelvis and lower extremities, orbits, mandible, clavicle, hand and spinal canal being less common sites. Recurrent bleedings cause expansion of the pseudotumor which in turn lead to compression and pressure necrosis in the surrounding tissues. Rarely a pelvic pseudotumor may erode into bowel resulting in fistula formation. Percutaneous aspiration or drainage attempts may predispose to fistulization.

Uncomplicated hemophilic pseudotumors may be treated by factor VIII replacement to keep an activity of 100 % (4–6, 8). For patients with inhibitors, rFIIa or prothrombin complex concentrates can be used (11, 13). If signs of neural or ureteric compression are present, or pseudotumor is expanding despite adequate factor replacement, surgery and/or radiotherapy may be required. But in general, surgery is advised only if conservative measures fail to stop the growth of the pseudotumor.

Fistulization into the bowel causes a more complicated clinical scenario, because of septic complications. The typical place of fistulization is right colon. Fecal contamination of pseudotumor content leads to an intraabdominal abscess, which must be drained. Surgical drainage is both dangerous and difficult, because of bleeding tendency and due to the dense adhesions between bowel and abscess cavity. That's why radiologically guided percutaneous drainage must be attempted first (1–3, 7, 14).

From a surgical standpoint, our case was challenging. In addition to the presence of the enterocutaneous fistula and intraabdominal abscess, the patient had significant bleeding diathesis which significantly increased the risks of operative intervention. Relatively distal localization of the fistula resulted in a low output, and allowed the follow up of patient with percutaneous drainage. This time interval allowed the adhesions to resolve and organise, so that dissection could be kept to a minimum, which is important in patients with bleeding diathesis. We believe that the use of Tranexemic acid contributed to the cessation of the bleeding during the last operation.

To our knowledge, this case presents the first report of a patient with psoas hemophilic pseudotumor who underwent three consecutive operations for recurrent bleeding and recovered well. Based on our experiences in this patient, we recommend waiting for the intraabdominal hematoma and adhesions to resolve and organise, so that the dissection can be kept to a minimum, which decreases the chances of iatrogenic injury and surgical bleeding.

Conclusions

In this study we present a case with mild haemophilia A, in whom bleeding into right pelvic area recurred three times despite factor VIII coverage and drainage and he was operated on.

Because of recurrences, the patient had undergone three abdominal operations and survived. The decision of operation was very difficult; because the patient had a bleeding diathesis and the risk of uncontrolled sepsis was substantial (14, 15).

References

1. Makris PE, Moros J, Foka Z, Kouskouras K, Chourmouzi D, Pithara E, Dimitriadis A. Pseudotumors in haemophiliacs: a complication of major psoas muscle hematoma. *Haema* 2002; 5 (2): 118–124.
2. Heaton DC, Robertson RW, Rothwell AG. Iliopsoas hemophilic pseudotumors with bowel fistulation. *Haemophilia* 2000; 6 (1): 41–43.
3. Rodriguez-Merchan EC. Hemophilic cysts (pseudotumors). *Haemophilia* 2002; 8 (3): 393–401.
4. Marmor L. Total knee replacement in hemophilia. *Clin Orthop Relat Res* 1977; 125: 192–195.
5. Heim M, Horoszowski H. Hemostasis: A practical review of conservative and operative care. *Clin Orthop* 1996; 1 (328): 34–38.
6. Atilla B, Pekmezci M, Tokgözoğlu M, Alpaslan M. Hemofilide Kas-Iskelet Sistemi Problemleri ve Ortopedik Tedavi Girişimleri. *Türk Ortopedi ve Travmatoloji Birliği Dergisi* 2003; 2:102–109.
7. Coon WW, Penner JA. Management of abdominal hemophilic pseudotumor. *Surgery* 1981; 90 (4): 735–740.
8. Villar A, Jimenez-Yuste V, Quintana M, Hernandez-Navarro F. The use of haemostatic drugs in haemophilia: desmopressin and antifibrinolytic agents. *Haemophilia* 2002; 8 (3): 189–193.
9. Rodriguez-Merchan EC, Rocino A, Ewenstein B, Bartha L, Batorova A, Goudemand J et al. Consensus perspectives on surgery in haemophilia patients with inhibitors: summary statement. *Haemophilia* 2004; 10 (Suppl 2): 50–52.
10. Slaoui M, Lambert T, Stieltjes N, Claeysens S, Borel-Derlon A. Study Group. Intestinal surgery with activated recombinant factor VII prophylaxis in patients with haemophilia A and high responding inhibitors: A report of five cases. *Blood Coagul Fibrinolysis* 2004; 15 (8): 687–691.
11. Sheth S, Dimichele D, Lee M, Lamour J et al. Heart transplant in a factor VIII-deficient patient with a high-titre inhibitor: perioperative management using high dose continuous infusion factor VIII and recombinant factor VIIa. *Haemophilia* 2001; 7 (2): 227–237.
12. Ingerslev J. Efficacy and safety of recombinant factor VIIa in the prophylaxis of bleeding in various surgical procedures in hemophilic patients with factor VIII and IX inhibitors. *Semin Throm Hemos* 2000; 26 (4): 425–432.
13. Tjonnfjord GE. Activated prothrombin complex concentrate (FEI-BA) treatment during surgery in patients with inhibitors to FVIII/IX: The updated Norwegian experience. *Haemophilia* 2004; 10 (Suppl 2): 41–45.
14. Ahlberg AK. On the natural history of hemophilic pseudotumor. *J Bone Joint Surg Am* 1975; 57 (8): 1133–6.
15. Bicknell S, Mason A. Hemophilic pseudotumor of the chest. *J Thorac Imaging* 2001; 16 (3): 188–90.

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