Spontaneous Intra-abdominal Hemorrhage in a Hemophilia A Patient Masquerading as Intra-abdominal Pseudotumor

Divij Jayant[1] Yashwant Sakaray[1] Uttam Thakur[1] Lileshwar Kaman[1] Arihant Jain[2]

[1] Department of General Surgery, Post Graduate Institute of Medical Education and Research, Chandigarh, India
[2] Department of Internal Medicine, Post Graduate Institute of Medical Education and Research, Chandigarh, India

Address for correspondence Yashwant Sakaray, Department of General Surgery, Post Graduate Institute of Medical Education and Research, Room No. 6, Fifth Floor, F Block, Nehru Hospital, Chandigarh 160012, India (e-mail: sakaraypgi@gmail.com).

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Abstract

Spontaneous intra-abdominal hemorrhage is very rare in hemophilia A patients. High index of suspicion is necessary for successful management. Surgical intervention may be required in cases of diagnostic uncertainty and for adequate hemostasis. Here we report a case of spontaneous hemorrhage in the peritoneal cavity in hemophilia A patient, who was managed with surgery and factor VIII supplementation. Patient did well in the postoperative periods.

Keywords

- factor VIII
- hemophilia A/B
- transfusion medicine
- surgery

Introduction

Inherited hemophilia is caused by a deficiency in clotting factor. Hemophilia A is caused by factor VIII deficiency and B is caused by factor IX deficiency. The incidence of hemophilia A is 10 to 20 cases per 100,000 people.[1] Pseudotumor is defined as an encapsulated, chronic, slowly expanding hematoma. This is a rare complication seen in 1 to 2% of patients with a severe disease, usually seen in extremities.[2] Here, we report our experience of surgical management of giant intra-abdominal pseudotumor in hemophilia A.

Case Report

A 32-year-old male patient presented with left-side upper abdominal pain for 7 days. The patient underwent laparotomy 7 years back at some other center, requiring multiple blood transfusions, but the rest of the details were not available. No family history of hemophilia or any hematological disorder could be elicited in the preoperative period due to the absence of close family members. Patient was conscious, grossly pale, and blood pressure was 108/60 mm Hg with a pulse rate of 120 per min. A firm, nontender lump was present in the left hypochondrium. Hemoglobin was 4.3 g/dL, total leucocyte count was 7,600/mm^3, and the platelet count was 150,000/mm^3. The coagulation profile showed activated partial thromboplastin time (aPTT) of 70 seconds (28–-34 seconds). The patient was initially resuscitated with transfusion of four units of packed red blood cell and fresh frozen plasma transfusion at 15 mL/kg. Posttransfusion hemoglobin was 7.9 g/dL and aPTT was 40 seconds. Ultrasonography (USG) of the abdomen was suggestive of gross ascites with a large heterogeneous mass of 15 × 9 cm in the left upper quadrant. Contrast-enhanced computed tomography of the abdomen (►**Fig. 1a–d**) showed 15 × 7.8 × 14 cm heterogeneously enhancing mass lesion with multiple hypodense areas suggestive of internal necrotic lesions, lying in close relation to the splenic flexure of colon with loss of fat planes. An initial differential diagnosis of retroperitoneal tumor, sudden-onset bleed in liposarcoma, and necrotic leiomyosarcoma with bleed was suspected. Clinically, with the ongoing need for transfusion in the background of suspected tumor bleed the patient was taken up for surgery. Intraoperatively 2 L of hemoperitoneum was present with large clots in lesser sac and left paracolic gutter near splenic flexure of the colon. The solid organs were normal and no source of active bleed was found (►**Fig. 1e, f**). Clot evacuation and lavage was done, hemostasis was achieved, and clots were sent for histopathology report. With the intraoperative findings, a diagnosis of pseudotumor was considered. The patient was shifted to surgical ICU, monitored postoperatively, and consulted with a hemophilia physician.

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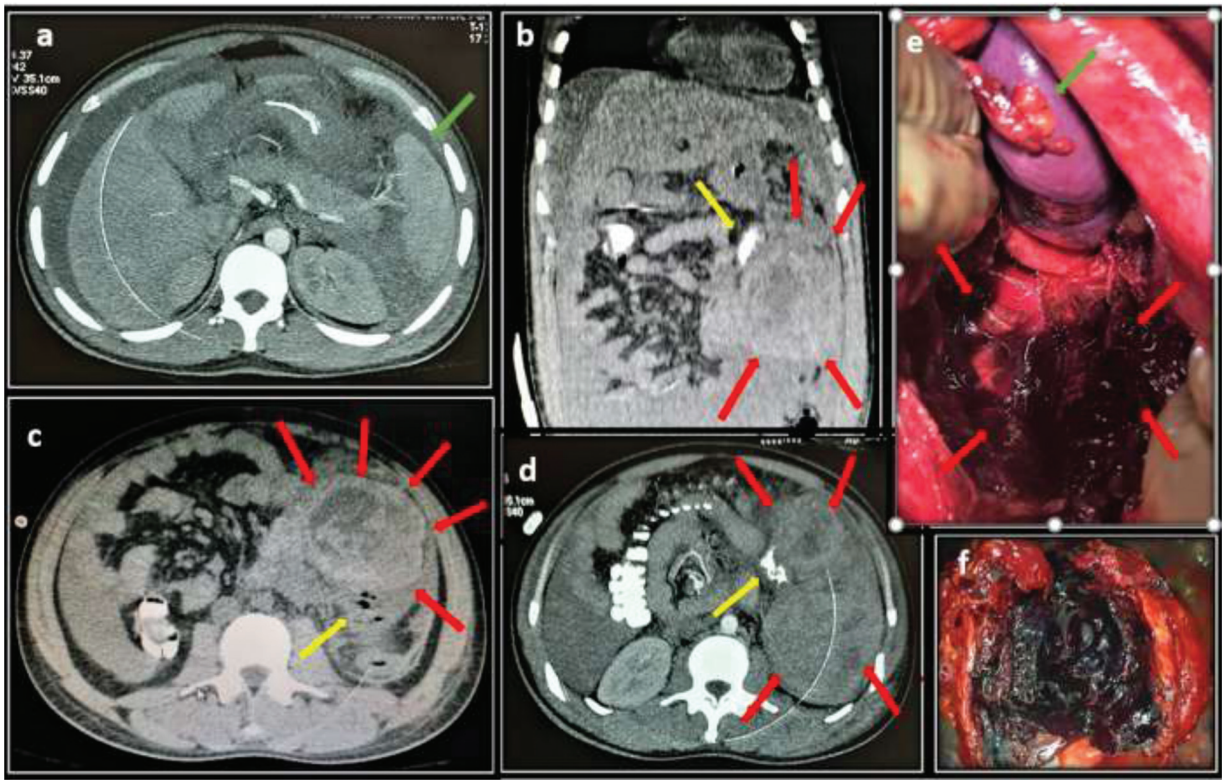

Fig. 1 (**A**) Gross hemoperitoneum (green arrow). (**B–D**) Heterogeneously enhancing mass lesion (red arrows) in close relation to splenic flexure with loss of fat planes and pushing splenic flexure medially (yellow arrow). (**E**) Intraoperative photographs showing intact spleen (green arrow) with hematoma present just superior to splenic flexure (red arrows). (**F**) Evacuated hematoma.

Follow-up

Postoperatively, clinical history was reelicited which was suggestive of swelling of joints since childhood and a history of similar bleeding episodes in maternal male members (►Fig. 2). Subsequent detailed hematological workup confirmed the diagnosis of severe hemophilia A (factor VIII deficiency) for which factor VIII correction was given 100% for 2 days, 70% for 3 days, 40% for 7 days, and 20% for 7 days. The patient was discharged subsequently in satisfactory condition. The patient is being continued on on-demand prophylaxis up to 25 units/kg two to three times per week.

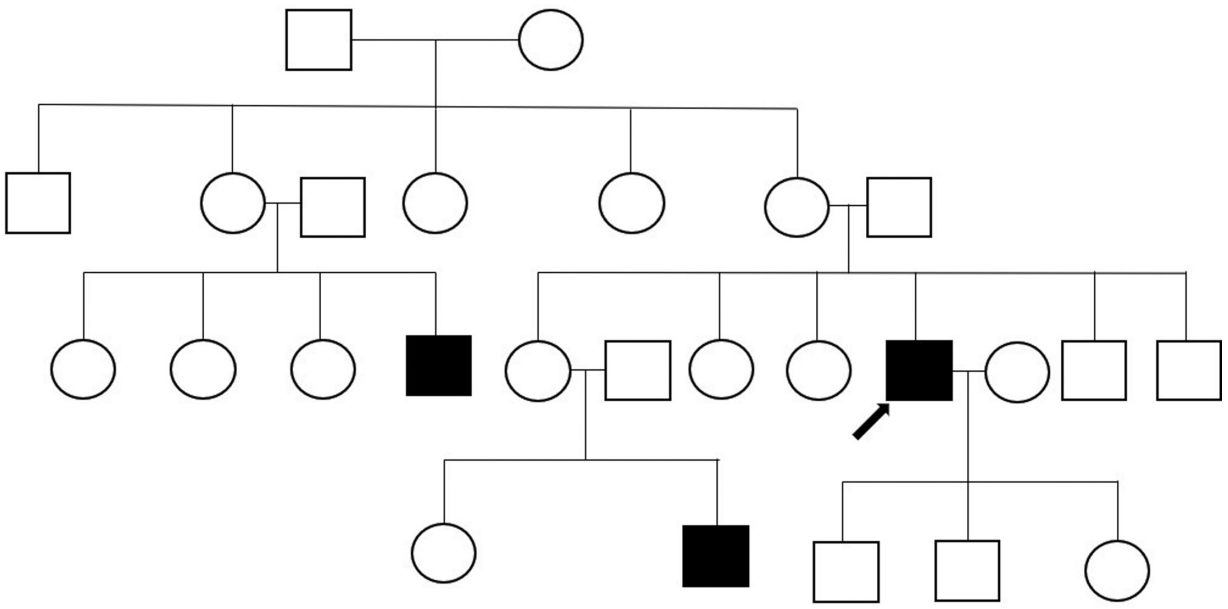

Fig. 2 ■Affected individual. →■ Index patient. □ Male sex. ◦ Female sex.

Discussion

Mortality in patients with intra-abdominal hemorrhage due to arterial bleeding is 8.6%; however, failure in the identification of bleeding site may increase the mortality rate up to 50%.[3] Prompt recognition, resuscitation, and adequate treatment with factor VIII replacement are essential for managing such patients.

In conclusion, spontaneous intra-abdominal hemorrhage in hemophilia A patient is a rare and life-threatening emergency. Surgical intervention may be required under the cover of clotting factor concentrate in cases of diagnostic uncertainty and for adequate hemostasis.

Conflict of Interest

The authors declare that they have no conflict of interest.

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