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Case Report

Acute Intestinal Obstruction due to Intramural Hematoma of Sigmoid Colon in the Five-Years Old Boy with Severe Haemophilia A

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Abstract

Spontaneous intramural hematoma of sigmoid colon is rare clinical condition, presenting usually as bowel obstruction.

5 year old boy, was admitted due to bloody vomiting, severe rectal bleeding, abdominal distension significant abdominal tenderness and anemia. Due to deterioration and hemodynamic instability patient was operated revealing threatening rupture and 10 cm length bloody intramural mass. Colostomy was opened and sigmoid resection according to Hartmann procedure. Postoperative course was followed with acute massive bleeding from wound and stoma site. Additional blood tests revealed severe hemophilia A, with factor VIII level of 0.60%. Substitution therapy of factor VIII started. Histopathologic examination of the specimen revealed an intraluminal submucosal hematoma with no features of malignancy. Stoma closure was performed six months later with preoperative correction of factor VIII level up to 100%.

Emergency Hartman procedures can be performed safely in hemophiliac patients with spontaneous intramural hematoma of the sigmoid colon complicated with acute obstruction.

Keywords: *Haemophilia A; Pseudotumor; Intestinal Obstruction*

Introduction

Intramural hematoma of sigmoid colon caused by haemophilia A is rare clinical condition. Symptoms and clinic signs are not specific, presenting usually as bowel obstruction [1].

Case Presentation

A 5 year old boy weighing 23 kg presented with bloody vomiting and severe rectal bleeding was admitted to Pediatric surgery. Patient had small bruises, abdominal distension followed by suprapubic pain and significant tenderness.

Tests revealed anemia (RBC 2.98 X 10^9/L, HGB 85 g/L, HCT 0.253 L/L), white blood cell count was 18 X 10^9/L. Other laboratory tests were in normal range: bleeding time 3 minutes 15 seconds, clotting time 4 minutes 45 seconds, PLT 322 X 10^9/L, CRP 4.07 mg/L. Plain abdomen X-ray showed dilatation of left colon. Ultrasound of the abdomen showed free fluid in the peritoneal cavity and mass in the left colon. Due to of clinical findings, deterioration and development of hemodynamic instability patient was planned for laparotomy. During

surgery enormous sigmoid dilatation with threatening rupture and 10 cm length bloody intramural mass was found. Stoma was opened and sigmoid resection is done according to Hartmann procedure (Figure 1). Histopathologic examination of the specimen revealed an intraluminal submucosal haematoma with no features of malignancy (Figure 2).

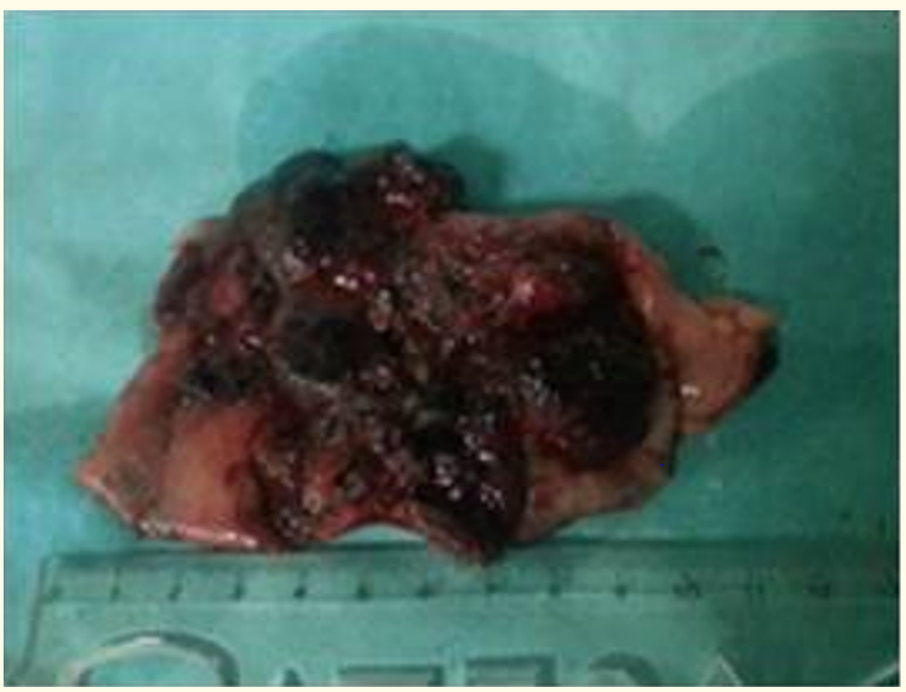

Figure 1: *Giant intramural haemathoma in sigmoid colon.*

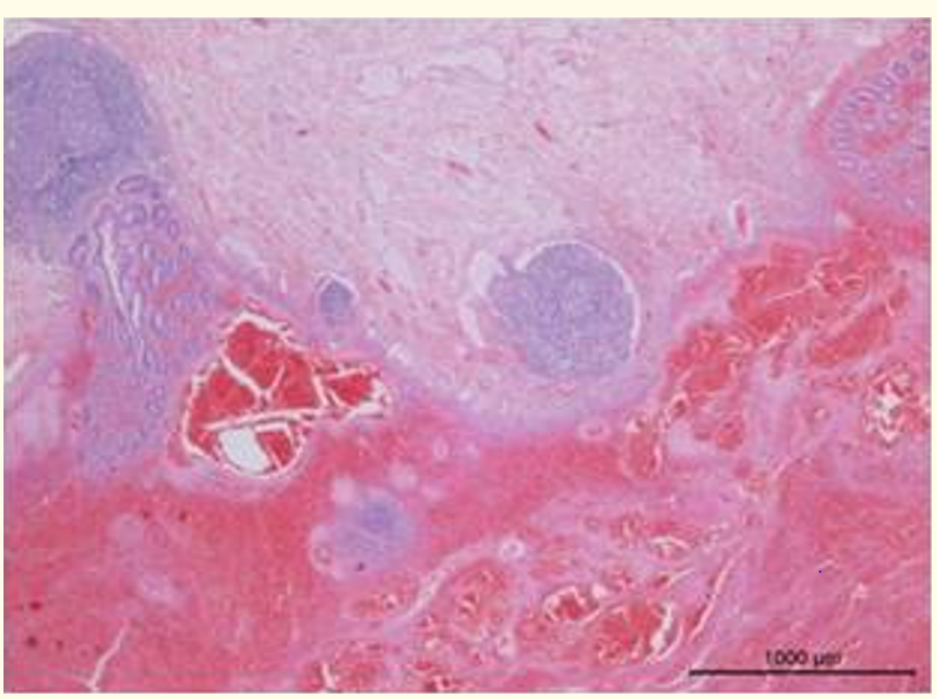

Figure 2: *Histopathologic examination of the specimen revealed an intraluminal submucosal haematoma with no features of malignancy.*

In the next few postoperative hours, acute massive bleeding was noticed from wound and stoma site followed by significant amount of blood in the drainage system and bag. One unit of red blood cells and two fresh frozen plasma units were administered. Additional blood tests revealed hemophilia A, with factor VIII level of 0.60% and prolonged aPTT. Initial substitution begun with a bolus of 1000 IU of factor VIII , and then continued with 500 IU factor VIII units on every 8 h, in decreasing manner during next week, continued twice a week at a dose of 25 IU/kgBW after discharge. Family history did not reveal relatives with haemophilia A.

Stoma closure was performed six months later, with preoperative correction of factor VIII level up to 100%. During the stoma clossure procedure we decided to follow the fast-track management as a safe and effective principle in order to minimise discomfort. Postoperative course was uneventful. The patient is coming to the regular quarterly controls and to this day there were no complications. He is listed in the national register of patients suffering from hemophilia A and parents are educated and informed of all preventive measures in case of potential surgical treatment.

Discussion

Intramural hematomas of colon are extremely rare, having been observed in less than 5% of all intramural hematomas. There are usually the consequence of traumatic lesions, hemophilia and anticoagulants administration. Large hematomas are typical for submucosa layer, because of its vascularity [3]. Duodenum is typical site, while changes to the colon are rarity.

The most probable physiopathology of pseudotumors is caused by stratification of terminal arterial during penetration of the intestinal muscular layer. Consequently, new-formed blood cysts can delaminate intestinal wall between the muscularis mucosa and the muscular layers, with preservation of mucosal viability [2,4].

The diagnosis of haemophilia A is often suspected due to family history and laboratory findings such as very reduced factor VIII and elevated PTT. PT, bleeding time, platelet count and clot retraction will be normal in these patients, since none of these tests are dependent on factor VIII. The most common cause of gastrointestinal problems in hemophilia is caused by arterial erosion that is attributable with massive bleeding, bowel obstruction due to pseudotumor "blood cyst" formation, or palpable abdominal mass [5,6]. Most of pseudotumors can be treated with conservative, non-surgical approach, with spontaneous complete resolution within a few weeks, during factor VIII replacement [7]. This approach is favorable for recent hemorrhage. If there is no response to conservative treatment, surgical intervention is mandatory, especially for complicated cases (progressive growth of hematomas, hemodynamic deterioration, sepsis, peritoneal effusion due the bowel perforation and ileus). Some studies stated that surgery as a primary option for treatment has better outcomes compared to surgery that followed after failure of conservative therapy [8].

MRI is useful tool that is superior to CT scan due to less radiation exposure but has a longer scan time. No comparative study has been done to compare the advantage of one imaging study over the other.

The factor VIII replacement to near normal values (100%), can make surgery feasible in patients with severe hemophilia A, even for major interventions.

During general anesthesia, nasal intubation should be avoided, since traumatic mucosal bleeding can lead to aspiration. In order to prevent intramuscular haemathomas or haemarthrosis, special care is focused in positioning the extremities, and avoiding the high soft tissue pressure.

Fast-track principles, first described by Kehlet Henrik [9] could be efficient model in pediatric patients as recent trials stated [10]. Fast -track can minimize discomfort but also nullifies the risk of lesions in the region of nasal mucosa, urethra and the skin at the site of abdominal drain placement. Patients with hemophilia have increased bleeding tendency, while fast-track management can be considered as the most favorable surgical option, that allows early postoperative feeding and prompt activation of intestinal function and motility.

Conclusion

A high index of suspicion should be maintained in boys with acute abdominal pain who have any bleeding disorders. Early diagnosis and prompt medical treatment are important in order to avoid unnecessary surgical interventions. Surgery should be reserved for complicated cases or when the diagnosis is uncertain. Emergency Hartman procedures can be performed safely in hemophiliac patients with spontaneous intramural hematoma of the sigmoid colon complicated with acute obstruction. The reported case supports the modern concept of treatment of patients with hemophilia. Careful preoperative preparation of the hemophiliacs reduces the risk of uncontrolled bleeding during surgery, and should not be avoided.

Ethical Approval

The patient is involved in project implemented by the Ministry of Health Republic of Serbia No. 175092, and approved by Ethical Committee.

Support

There are no sources of support for this work.

Conflict of Interest

The authors have no conflict of interest to disclosure.

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