ENDOSCOPIC DIAGNOSIS OF INTRAMURAL HEMATOMA IN THE COLON SIGMOIDEUM IN A CHILD WITH HIGH TITER INHIBITORY HEMOPHILIA A

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ABSTRACT
Lower gastrointestinal bleeding is a rare condition in childhood pathology. The incidence of this disorder in the general population of Bulgarian children is unknown. We report a case of a 7-year-old child with diagnosed hemophilia A and high titer of factor VIII inhibitor; the patient was admitted into the Department of Pediatrics and Medical Genetics for rectorrhagia after falling onto his buttocks while playing. Colonoscopy showed submucosal hematoma 25 cm from the anocutaneous line occluding the intestinal lumen with a lesion of the overlying mucosa as long as 20 mm. If a patient presents with rectorrhagia, timely and carefully planned colonoscopy could identify the source of bleeding, determine the severity of bleeding and the size of hematoma, and assess the need for surgical intervention. The reported case supports the modern view that patients with inhibitor hemophilia should not be denied interventional procedure or surgical intervention for fear of uncontrolled bleeding.

Key words: emergency colonoscopy, rectorrhagia, hematoma of the sigmoid colon, inhibitor hemophilia A

REЗЮМЕ
Кровотечение из нижнего гастро-интестинального тракта это редкая патология в детском возрасте. В общей популяции болгарских детей частота заболевания неизвестна. Авторы описывают случай семилетнего ребенка с доказанной гемофилией А и с наличием высокоотитированным ингибитором /фактор VIII/. Поводом настоящей госпитализации является геморрагия, появившаяся через несколько часов после падения на седалище во время игры. Колоноскопия доказала наличие субмукозной гематомы, локализованной в 25 см от ano-кутантной линии и обтурирующей просвет кишки повреждением /лезней/ надлежащей слизистой оболочки по протяжению 20 мм. При проявлении ректоррагии своевременно проведенная после тщательного планирования колоноскопия дала возможность локализировать источник и тяжесть кровотечения, определить величину гематомы и оценить необходимость в хирургическом вмешательстве. Представленный авторами случай подкрепляет современную точку зрения, что пациентам с ингибиторной гемофилией А не следует отказывать в инвазивной процедуре или хирургическом вмешательстве из-за страха от неконтролируемого кровотечения.

Ключевые слова: неотложная колоноскопия, ректоррагия, гематома colon sigmoideum-a, ингибиторная гемофилия A
INTRODUCTION

Bleeding from the lower gastrointestinal tract (GIT) is rare in childhood. Generally, colon bleeding can be categorised into spontaneous and post-traumatic bleeding. The most common cause of nontraumatic rectorrhagia is colon polyposis, followed by chronic ulcerative colitis and intestinal polyposis.\textsuperscript{1,2} GIT hemorrhage is not unusual in congenital coagulopathies, but it is quite rare to have intramural hematomas formed along the colon.\textsuperscript{3} If there are clinically-significant, nontransient and high titer FVIII factor inhibitors (>10 BU), the hemorrhagic episodes are hard to manage and there are even proclivity for spontaneous bleeding.

CASE STUDY

We report a case of a 7-year-old boy (medical record No 5201/2011) diagnosed with mild hemophilia A at 4 years of age and formed factor VIII inhibitors (8 Bethesda Unit), found 10 months prior to the current hospitalisation of the patient. He received a recombinant FVIIa therapy when necessary. The patient was admitted to hospital for rectorrhagia which presented with colicky pains in the suprapubic region with an onset a few hours after the boy fell on his buttocks while playing but without direct trauma in the region of the abdomen or in the small back. Previous to hospitalisation the patient had had fresh bright blood in the stools at three successive instances.

At admission the patient presented with no anemia and no coagulation abnormalities that may indicate disseminated intravascular coagulation. The FVIII inhibitor titer was > 10 BU.

The therapy initially administered to the patient included multiple (3 times) administration of recombinant FVIIa (90 μg/kg every 2 hours), but in spite of this therapy, the hemorrhage started again after a lucid period of 12 hours. This was the reason we started a therapy with a medicinal preparation containing activated prothrombin complex concentrates (aPCC), reaching the maximum 24-hour dose of 200 U/kg in the following days, and still the bright blood rectorrhagia persisted. This made it necessary to locate the lesion site and 36 hours after hospitalisation, along with aPCC replacement therapy the patient was subjected to colonoscopy. Videocolonoscopy was performed after 36 hours of total parenteral feeding without a complementary bowel preparation for fear of aggravating the rectorrhagia. The endoscopic study showed submucosal hematoma occluding the intestinal lumen 25 cm away from the anocutaneous line with a lesion of the overlying mucosa as big as 20 mm (Fig. 1). There were both small free clots and small amount of bright blood in the rectum.

The abdominal ultrasound study and computed tomography showed thickening of the wall of the sigmoid colon without any free moving fluid in the abdominal cavity.

A control study three days after that found that the hematoma had resorbed and the sigmoid colon had restored its patency. In the region of the lesion we found a fibrin coagulum without changes in the rest of the colon (Fig. 2).

Both endoscopic studies found the rectum and sigmoid colon very efficiently cleaned.

Hemodynamically, the patient remained stable.

Figure 1. The lumen of sigmoid colon occluded with a hematoma.

Figure 2. Restored colon patency with a formed fibrin coagulum.
throughout the procedures. Surgical approach was considered twice, but was not realised as there was no clinical evidence of intestinal obstruction. The patient received infusion of Sandostatin, Nexium, wide spectrum antibiotics, and one-time blood transfusion. He was discharged at 18 days after admission with restored colon patency and no pathological impurities in his stools.

DISCUSSION

We performed a meta-analysis in MEDLINE of published cases of sigmoid colon disorders in children with hemophilia. We found 4 reports done over the last 40 years that describe intramural hematomas in this region.4-7

Rectorrhagia in hemophiliac children is considered an emergency condition that requires a precise early diagnosis and therapeutic management. Barnert et al. consider colonoscopy to be the method of choice in diagnosing lower GIT bleeding, while angiography is indicated only if bleeding of unknown origin takes place. Other radiographic methods of study can be considered in intermittent hemorrhages with a bleeding source impossible to identify in a colonoscopy study. Unplanned emergency colonoscopy (performed within 48 hours) in patients with moderate lower gastrointestinal bleeding is a highly informative and complication free procedure.9

The case reported herein is of casuistic interest because of the rare bleeding site in high titer inhibitor hemophilia A and its effective management using conservative approach.

CONCLUSIONS

Patients with high titer inhibitor hemophilia are a therapeutic challenge due to the high severity of the hemorrhagic events and their hard management. In rectorrhagia, a timely colonoscopy performed after thorough planning makes it possible to localise the source of bleeding, determine the severity of bleeding and the size of hematoma, and assess the need for surgical intervention. The reported case supports the modern view that patients with inhibitor hemophilia should not be denied interventional procedure or surgical intervention for fear of uncontrolled bleeding.

REFERENCES