Case Report

Acute Lower Gastrointestinal Bleeding Caused by Congenital Portosystemic Shunt:A Case Report and Review of the Literature

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ABSTRACT

Lower gastrointestinal bleeding refers to bleeding within the lumen of the gastrointestinal tract located below the ligament of Treitz. In this study, we present a case with bleeding from massive anorectal varices, caused by a congenital high pressure Porto-systemic shunt.
A 31-year-old man with a prolonged history of painless rectal bleeding was referred to our center. The onset of symptoms was at the age of two years. Finally, he underwent semi-elective surgery due to severe bleeding and a diagnosis of portosystemic shunt was made intraoperatively.

Keywords: Gastrointestinal Bleeding, Congenital, Portosystemic Shunt.

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INTRODUCTION

We presented a rare case of bleeding from the lower GI system, which might be classified into the category of anorectal varices. Anorectal varices are large and dilated veins originating from a portosystemic shunt. In most cases, increased blood flow between the upper hemorrhoidal vein (portal) and lower and middle hemorrhoidal veins (systemic) is established due to portal hypertension.

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CASE REPORT

A 31-year-old man with a prolonged history of painless rectal bleeding was referred to our center. The onset of symptoms was at the age of two years. Since then he had continuously suffered from bleeding at different times with different volume. He reported no signs of bowel habits changes, constipation, mucus discharge, or problems such as rectal evacuation disorder. There were several episodes of hospital admission since childhood in his history, and diagnostic studies such as upper gastrointestinal (GI) endoscopy and colonoscopy had been conducted, which resulted in various diagnoses such as FAP (Familial Adenomatous Polyposis), angiodysplasia, hemorrhoids, and telangiectasia, but none of them was definitely confirmed. Therefore, he was continuously under surveillance. Considering his chronic symptoms and multiple blood transfusions in his history, he had numerous referrals to medical clinics causing him to develop a personality disorder such that he had an aggressive mood and it was hard to communicate with him.

Physical examination of the patient revealed nothing

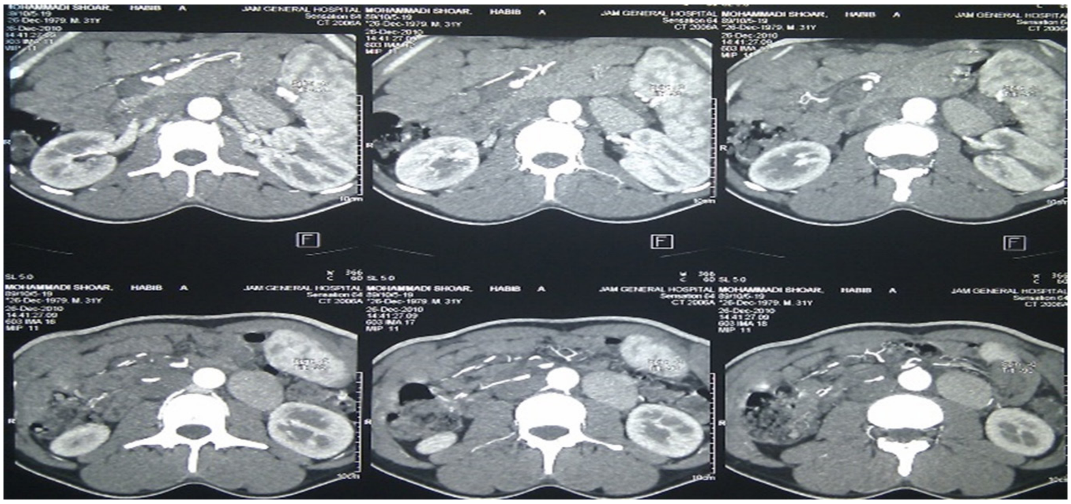

Fig. 1: **Abnormal vein structure in the abdominal cavity (55 mm in diameter)**

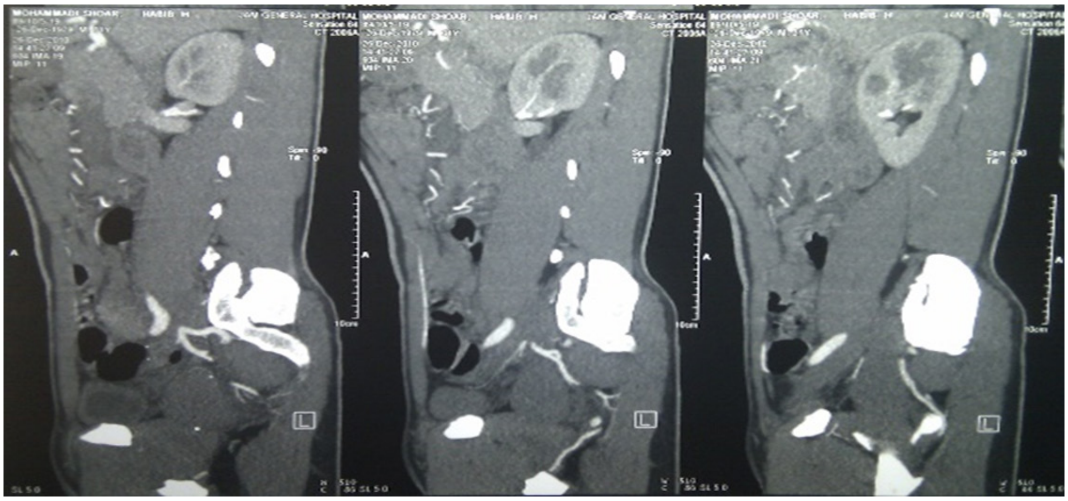

Fig. 2: **Abnormal vein structure in the abdominal cavity**

notable except mild scoliosis and mild splenomegaly. Moreover, external and internal hemorrhoidal cushions were observed in perineal and rectal examinations. There were no signs of prolapse, painless descent, fissure, or hemorrhoidal thrombosis. In digital rectal exam, tone and length of sphincter were normal.

Total colonoscopy revealed three quite large submucosal veins on the posterior wall of the rectum at the rectosigmoid junction and a large submucosal vein in the cecum. The terminal ileum was normal. There was no history of liver or cirrhosis. All liver tests and necessary investigations in term of liver function were normal.

Computed tomographic angiography (CTA) (figures 1,2) demonstrated a very large venous structure in the abdominal cavity (55 mm in diameter) coursing between the bladder and rectum and connecting to the splenic and portal vein; possibly a very large inferior mesenteric vein (IVC). Moderate dilatation of the superior mesenteric vein (SMV) and portal vein was also observed. The IVC was somewhat dilated and mild splenomegaly was also observed. The left lobe of the liver was large and evidence of chronic parenchymal damage could be seen. The arterial system had a normal appearance. According to the CTA report, a primary diagnosis of

Fig. 3: **Abnormal vein structure in the abdominal cavity**

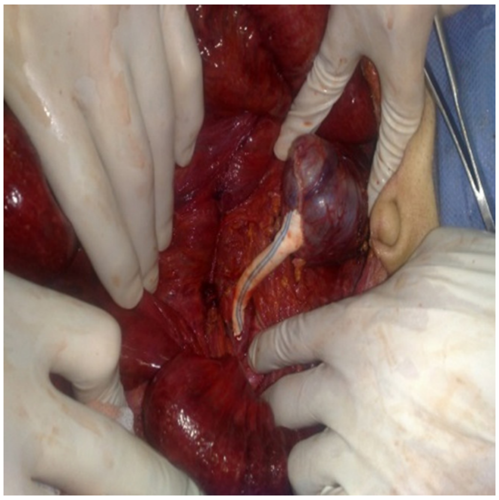

Fig. 4: **Venous bypass between the inferior mesenteric vein and IMV (Inferior Mesenteric Vein) stump**

portosystemic shunt between IMV (Inferior Mesenteric Vein) and internal iliac vein was considered. The patient then underwent digital subtraction angiography (DSA) as well as angioembolization (figure 3) to decrease the arterial inflow to the venous structures. 24 hours after angioembolization, the patient developed generalized severe abdominal pain, fever, tachycardia, and slight abdominal distension. In physical exam, there was no tenderness in the abdomen. White blood cell and hemoglobin were $12000\times1000/mm^3$ and 9.9 gr/dl, respectively, and other tests were normal. Chest radiography was also normal. Ultrasonography revealed an interloop collection (21×27 mm) along with free fluid in the abdomen and pelvis. Due to the patient's condition, he was transferred to operating room for laparotomy.

There was a large venous network in the pelvis with a fully engorged vein diameter (8 cm) adjacent to the mesorectum, sigmoid, and descending colon. The IMV was much larger than normal (about 5 cm in diameter) and even larger than the adjacent colon. Moreover, fully dilated veins along the mesorectum, cecum, and even appendix and terminal ileum were seen, although the sigmoid and the mesosigmoid were normal. The diameter of the inferior mesenteric artery was normal and there was mild splenomegaly.

Firstly, classic dissection of the rectum and sigmoid were done. Then, the dilated pelvic vein was occluded by the use of a vascular stapler. Next, the IMV was ligated superiorly with a vascular stapler. However, this caused a sudden and severe congestion and bowel edema. Because of this impaired venous drainage, a decision was made to perform a venous bypass between the IVC and the IMV stump. This was done with an 8-mm PTFE (Polytetrafluoroethylene) graft, end to side in fashion (figure 4). Fortunately, intestinal edema and congestion resolved quickly. The graft was covered with omentum. Due to the short mesocolon and dilated veins throughout the colon and rectum, total proctocolectomy was done along with a part of the terminal ileum just adhere to the colon and terminal ileum for preservation of collateral vessels. After that, end to end anastomosis between the terminal ileum and anal canal was performed with the use of a 28-mm circular stapler. Finally, a protective ileostomy was fashioned. The bowel edema resolved completely while the abdominal wall was being closed, so the

Table 1: Different types of portosystemic shunt

I	There is no flow in the liver (there is no portal vein as congenital).
II	Partial shunt with remaining portal vein flow toward liver aII: Derived from the right or left portal vein (including patent dactusvenule) bII: Derived from the main portal vein (including bifurcation site or splenomesenteric junction) cII: Derived from the mesenteric, gastric, or splenic vein

abdomen was closed without any tension. There was no significant bleeding during surgery and the patient had a good urinary output. After surgery, the patient was transferred to the intensive care unit (ICU). He was tachycardic during the first 24 hours, but his blood pressure was stable. There was a large amount of ascetic fluid drainage, which led to acute tubular necrosis (ATN), as the patient's BUN rose to 120 mg/dl. Fortunately, all of these problems resolved after intensive medical care.

The patient was discharged from the hospital 10 days after surgery. The ileostomy was closed 6 months later. Three years later, the patient is in the full recovery, has a better mental status, has married, and has a child.

DISCUSSION

In this report, we presented a rare case of bleeding from the lower GI system, which might be classified into the category of anorectal varices(1,2). Anorectal varices are large and dilated veins originating from a portosystemic shunt. In most cases, increased blood flow between the upper hemorrhoidal vein (portal) and lower and middle hemorrhoidal veins (systemic) is established due to portal hypertension. Ectopic varicose veins can pass beyond the anorectal area and involve other regions. A widespread study conducted on the bleeding from ectopic varicose veins by Norton and colleagues revealed that stomal varices are the most common (26%), followed by 17% in the duodenum, 17% in the jejunum or ileum, 4% in the colon, 9% in the peritoneum, and 8% in the rectum (3,4).

Our patient had varicose veins in the rectum and the cecum as well, due to a congenital portosystemic shunt that has not been diagnosed during childhood without apparent portal hypertension.

Congenital absence of the portal vein with end-to-side portocaval shunt was defined for the first time by John Abernethy (5). Different types and subtypes of portosystemic shunt have been reported since then. Morgan and Superina (6), defined the classification based on the status of the portal vein in 1994.

Currently, there is a common categorization for portosystemic shunts considering the anatomic variants, which is summarized in table 1 (7,8).

According to the table 1, our patient had type cII shunt. A variety of types II portosystemic shunts have been reported. This diversity is seen in the presentation of clinical symptoms. In one article, extrahepatic shunt was reported in 61 cases in 2007 (9). Although the symptoms usually begin during the childhood, some cases have remained asymptomatic even to the middle age (10).

In type II, no superiority is seen compared with type I. There is a great diversity of type cII concerning the extrahepatic portosystemic shunt. It can be derived from mesenteric, gastric, or splenic veins drain into the renal vein, azygos, or iliac veins and branches.

Depending on the type of the shunt, signs and symptoms can vary. Portosystemic shunt may cause symptoms of hyperammonemia and hypergalactosemia. These conditions increase the risk of hepatic encephalopathy. In fact, metabolic changes can lead to psychological symptoms in such patients. Cardiovascular malformations, skeletal system, and the urinary tract system anomalies may co-exist (7,8). In one study, a shunt between IMV and a branch of the iliac vein caused frequent vaginal bleeding in a patient, whose clinical symptoms resolved with the ligation of the shunt (7).

In another study conducted by Shapiro and colleagues, two patients with very large paraesophageal and paraumbilical varices were described (11). Akgul and co-workers also presented a disease discovered in a patient's follow-up during sonography and CT scanning in which the patient had a varicose IMV that was due to the increase in portal vein flow (12).

Since the disease is uncommon, different treatments have been recommended. Transjugular intrahepatic portosystemic shunt (TIPS) and endovascular methods can be effective (13). Coil embolization has been also associated with successful results (14). Open surgery and laparoscopy can be used for ligation of the shunt (15). In most cases, clinical symptoms have resolved after surgery, and even psychological symptoms have improved.

Summary of experiences, and recommendations:

When a patient's bleeding history goes back to his childhood, a possibility of vascular anomalies should be considered and the patient should be completely evaluated in this regard. Moreover, during the surgery it is crucial to attend and guide a vascular surgeon in addition to a general or colorectal surgeon and be prepared for using necessary instruments such as stapler and graft. However, the cooperation between gastroenterologists and radiologists, nephrologists, and intensive care specialists, is so important in the management of the patient before and after the surgery.

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CONFLICT OF INTEREST

The authors declare no conflict of interests related to this work.

REFERENCES

1. David E. Beck, Patricia L. Roberts, John L. Rombeau& other .The ASCRS Manual of Colon and Rectal Surgery (2009).427- 439.
2. Vernava AM 3rd, Moore BA, Longo WE, Johnson FE. Lower Gastrointestinal Bleeding. *Dis Colon Rectum* 1997;40:846-58.
3. Park CM, Shin WG, Hong KW, Park JW, Jung JC, Kwon MJ, et al. Arteriovenous Malformation Causing IleocecalVariceal Bleeding in Liver Cirrhosis: Case Report and Review of the Literature. *Gut Liver* 2008;2:54-9.
4. Norton ID, Andrews JC, Kamath PS. Management of ectopic varices. *Hepatology* 1998;28:1154-8.
5. Abernethy J. Account of two instances of uncommon formation in the viscera of the human body. *Phil Trans R Soc* 1793;83:59-66.
6. Morgan G, Superina R. Congenital absence of the portal vein: two cases and a proposed classification system for portasystemic vascular anomalies. *J Pediatr Surg* 1994;29:1239- 41.
7. Lautz TB, Tantemsapya N, Rowell E, Superina RA. Management and classification of type II congenital portosystemic shunts. *J Pediatr Surg* 2011;46:308-14.
8. Stringer MD. The clinical anatomy of congenital portosystemic venous shunts. *Clin Anat* 2008;21:147-57.
9. Kandpal H, Sharma R, Arora NK, Gupta SD. Congenital extrahepatic portosystemic venous shunt: imaging features. *Singapore Med J* 2007;48:e258-61.
10. Bazzocchi G, Pastorelli D, Laviani F, Simonetti G. New type of asymptomatic congenital portosystemic shunt . *Clin J Gastroenterol* 2009;2:43-6.
11. Shapiro RS, Mendelson DS, Silvers AR, Gray CE, Halton KP, Miller CM. Giant varices in portal hypertension: CT Findings. *Comput Med Imaging Graph* 1990;14:269-72.
12. Akgul E, InalM, Aikimbaev K, Celiktas M. Giant varicose inferior mesenteric vein. *Eur J Radiol Extra* 2003;45:122-5.
13. Vangeli M, Patch D, Terreni N, Tibballs J, Watkinson A, Davies N, et al. Bleeding ectopic varices- treatment with transjugular intrahepatic porto-systemic shunt (TIPS) and embolisation . *J Hepatol* 2004;41:560-6.
14. Gorini P, Johansen K. Portal Decompression Using the Inferior Mesenteric Vein. *HPB Surg* 1998;10:365-9; discussion 369-70.
15. Nii A, Takehara HO, Kuyama H, Shimada M. Successful preemptive surgical division of type 2 – congenital extrahepaticportosystemic shunt in children. *J Med Invest* 2009;56:49-54.