

Diffuse idiopathic colonic varices presenting with lower gastrointestinal bleeding in an elderly patient : a case report and review of the literature

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Abstract

We report a case of lower gastrointestinal bleeding caused by idiopathic colonic varices. A 74-year-old woman presented with rectal bleeding. Colonoscopy revealed numerous varices of the entire colon, which, after an extensive work-up, proved to be idiopathic. No specific therapy or transfusions were required and there has been no further bleeding to date (follow-up 10 months). Review of the literature demonstrates that diffuse idiopathic colonic varices are a rare cause of lower gastrointestinal bleeding, especially as the first presentation in an elderly patient. (*Acta gastroenterol. belg.*, 2006, 69, 15-19).

Introduction

Varices of the colon are a rare clinical entity. Since the original description of colonic varices (C.V.) in 1954 (1) no more than 100 cases have been reported in the literature. C.V. are almost invariably associated with liver cirrhosis and consequent portal hypertension ; other, less common causes, include mesenteric vein thrombosis, pancreatitis with splenic vein thrombosis, adhesions and congestive heart failure (2,3). However, sometimes no specific cause is found, in which case C.V. are called idiopathic. To date only 31 cases of idiopathic C.V. have been reported in the English literature (4-28). We report a case of lower gastrointestinal (GI) bleeding in a patient with C.V. of unknown etiology, involving the entire colon.

Case report

A 74-year-old woman presented with a 3 day history of passing bright red blood per rectum. The patient denied any associated symptoms or similar episodes in the past. She also denied alcohol consumption, tobacco use, intravenous drug abuse or prior abdominal surgical procedures. There was no family history of GI bleeding. Her physical examination was unremarkable and the rectal vault was empty. The initial Hct was 37,5% with microcytic indices (Hgb 11.9 g/dl). The rest of the blood chemistries, including liver function tests, were within normal limits. A full colonoscopy was done on the next day and prominent varices (Fig. 1) were seen throughout the entire length of the colon. No active bleeding was found, but one varix of the sigmoid colon was covered by hemocysts (Fig. 2) and this was presumed to be the bleeding site. Esophagogastroduodenoscopy revealed

neither esophageal nor gastric varices and there was no ulcer. Ultrasonography of the abdomen and echo-Doppler of the portal and splenic veins were normal. A C/T scan of the abdomen with intravenous contrast revealed a normal liver and spleen and no other abnormalities. Superior and inferior mesenteric angiography showed normal arterial and venous anatomy, with no evidence of pooling or early venous filling. Although it could be argued that a liver biopsy or a Hepatic Venous Pressure Gradient (HVPG) measurement should have been performed, in order to exclude rare causes of pre-sinusoidal portal hypertension, the clinical and laboratory data obtained from our patient did not indicate schistosomiasis, early primary biliary cirrhosis, toxins and myeloproliferative syndrome as probable diagnoses. Capsule enteroscopy (M2A, Given Imaging, Yoqneam Israel) did not detect any small bowel varices, however a few dilated and tortuous veins were seen in the ileum (Fig. 3). The patient was treated conservatively with bulk agents and intravenous fluids. She was hospitalized for four days and her bleeding did not recur after a follow-up period of 10 months.

Discussion

Idiopathic C.V. are extremely rare. A review of the literature reveals that only 30 cases have been reported to date (4-28). These cases are shown in Table 1. In our patient, despite the extensive work-up, no specific cause for the development of C.V. was identified ; therefore it is considered a true case of idiopathic C.V.

Idiopathic C.V. usually present with hemorrhage characterized by painless, bright red blood per rectum of varying magnitude (13), which, in most of the cases, ceases spontaneously. As shown in Table 1, hemorrhage from idiopathic C.V. has been reported in all age groups (mean 45.5) and the diagnosis is usually established after a long history of recurrent bleeding episodes. Our case is rather intriguing in that the first episode of bleeding occurred in such an advanced age, in a previously asymptomatic patient.

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Fig. 1. — Colonic varices presenting as tortuous dilated veins with a bluish discoloration, involving the entire colon.

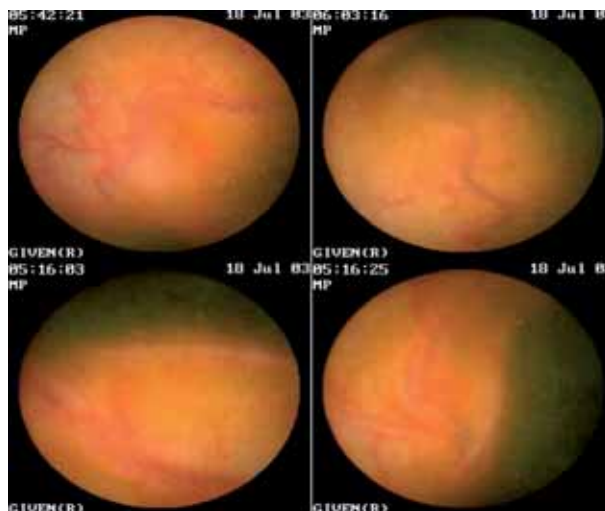
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Fig. 2. — Varix with hematocysts

No firm conclusions about the etiology of idiopathic C.V. have been established. Some investigators have suggested an inborn vascular anomaly (18), while others imply an acquired venous dysplastic disorder (19). Idiopathic C.V. have been reported in two members of six families (8,14,19,20,24,26), suggesting a possible familial tendency, with an autosomal recessive pattern of inheritance. On the other hand C.V. can not be solely a vascular degenerative disease, since it has been reported even in very young patients. In our case there was no evidence for the existence of varices in any member of the patient's family, while the disease's first presentation occurred in the age of 74. This probably suggests that a multi-factorial etiology is responsible for the development of the disease.

What causes the varices to bleed is unknown. Focal erosions into a varix in the rectosigmoid area from hard stools or erosion of a varix into the mucosa have been proposed as probable mechanisms (4,16). Meanwhile the bleeding site is often difficult to identify, since colonic varices are rarely found to actively bleed during endoscopy (13). In most of the cases reported to date, bleeding has been attributed to C.V. just because they were found in colonoscopy, except for two cases in



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Fig. 3. — Capsule enteroscopy images showing dilated veins in the ileum.

which a blood clot (13) and reddish points (11) on the varices were regarded as stigmata of recent hemorrhage. In our case, the presence of hemocysts, although not recognized as a stigma of recent hemorrhage when seen on esophageal varices, was presumed to be indicative of the source of bleeding (Fig. 2).

The most reliable tool for diagnosing C.V. is endoscopy. C.V. appear as tortuous bluish vascular dilations; occasionally they resemble polyps and if biopsied can bleed profusely. Blood flow can be demonstrated by endoscopic Doppler sonography (25) and this modality, if available, can be used in cases where the endoscopic diagnosis is in doubt. Selective mesenteric angiography, with attention to the venous phase, usually demonstrates the varices (13) and failure of the angiogram to do so is attributed to dilution of the radio-opaque agent in the venous phase. Angiography is invasive and expensive, and – as stated earlier – cannot always demonstrate the varices; thus this method is not considered necessary for the diagnosis although it can be helpful for the identification of vascular abnormalities (superior or inferior mesenteric vein obstruction) responsible for the formation of varices. A barium enema is a poor diagnostic tool, since C.V. can be missed (10) or misinterpreted as fecal material, air bubbles, multiple polypoid lesions (14,15,18,19) or even carcinoma (30). Scintigraphic studies (T 99-m labeled RBCs or T 99-m sulfur colloid scan) are not adequate in localizing the segment of colon associated with bleeding varices, but have greater sensitivity than angiography to detect hemorrhage (2).

C.V. are sometimes accompanied by varices of the small bowel, as shown by ileoscopy, enteroclysis, angiography or laparotomy. Endoscopic capsule is able to detect small bowel varices (27,29). To our knowledge, our patient is among the first that received endoscopic capsule for further exploration of idiopathic C.V. (27).

Table 1. — Previous reports of idiopathic colonic varices

Reference	Sex	Age	Presentation	Location	Family history	Therapy	Follow-up
Place, 2000 (4)	M	27	1w tarry stools	Ascending colon	–	Conservative	No rebleeding (4y)
Van Gossum 2000 (5)	M	54	3d hematochezia	Entire colon	–	Conservative	No rebleeding (3y)
Abraham 2002 (6)	M	37	2y intermittent hematochezia	Entire colon	–	Sigmoid colectomy	2 episodes of lower g.i. bleeding (18y)
Vella-Camilleri 1986 (7)	F	44	2d hematochezia	Entire colon	–	Conservative	No rebleeding (3m)
Solis-Herruzo 1977 (8)	F	18	7y intermittent hematochezia	Entire colon	+	–	–
	F	26	15y intermittent hematochezia	Entire colon	+ (aunt)	–	–
Weingart 1982 (9)	M	70	63y intermittent hematochezia	Entire colon	–	Conservative	–
Ibsister 1989 (10)	F	14	8m intermittent hematochezia	Ascending and sigmoid colon	–	Right hemicolectomy	No rebleeding (duration not stated)
Sugiyama 1992 (11)	M	62	1 episode of hematochezia	Ileocecal	–	Right hemicolectomy	–
Wagner 1970 (12)	M	47	3m intermittent hematochezia	Entire colon	–	Subtotal colectomy	No rebleeding. 4m later death from A.M.I.
Villarreal 1995 (13)	M	41	Massive lower g.i bleeding	Descending colon	–	Segmental resection	No rebleeding (6m)
Beerman 1988 (14)	M	65	No bleeding	Entire colon	+	Conservative	–
	F	64	11y intermittent hematochezia	Entire colon	+ (sister)	Conservative	–
Nikolopoulos 1990 (15)	M	22	10d melena	Entire colon, small bowel	–	Subtotal colectomy	No rebleeding (12m)
Shrestha 1994 (16)	M	33	3d hematochezia	Entire colon	–	Conservative	2 episodes of rebleeding (5m)
Vescia 1985 (17)	M	38	20y intermittent hematochezia	Entire colon	–	Total colectomy	No rebleeding (3y)
Iridale 1992 (18)	M	58	10w intermittent hematochezia	Entire colon	–	–	–
Hawkey 1985 (19)	M	66	41y intermittent hematochezia	Entire colon	+	Conservative	–
	F	71	50y intermittent hematochezia	Jejunum, ileum, colon	+ (sister)	Segmental jejunal resection	Multiple episodes of rebleeding (50y)
Atin 1993 (20)	M	23	9y intermittent hematochezia	Rectum, sigmoid, ascending colon, small bowel	+	Resection of terminal ileum, right colon and rectosigmoid	No rebleeding (2y)
	F	17	3y intermittent hematochezia	Rectum, sigmoid colon	+ (sister)	Conservative	–
Detry 1996 (21)	M	32	12y intermittent hematochezia	Rectum, sigmoid colon	–	Resection of sigmoid colon and rectum	No rebleeding (2y)
Pickens 1980 (22)	M	46	6m intermittent hematochezia	Rectosigmoid area	–	Conservative	No rebleeding (2m)
Planas 1988 (23)	M	73	3d melena	Ascending, transverse, descending colon	–	Conservative	No rebleeding (7m)

Table 1. — Continuation

Reference	Sex	Age	Presentation	Location	Family history	Therapy	Follow-up
El-Dosoky 1994 (24)	M	30	20y intermittent melena	Entire colon, small bowel	+	—	—
	M	—	1 episode of melena	Descending colon and rectum	+ (father)	—	—
Schlling 1996 (25)	M	25	2y intermittent hematochezia	Entire colon, terminal ileum	—	Conservative	No rebleeding (14m)
Morini 1993 (26)	W	67	Recent episodes of hematochezia	Entire colon,small bowel	+	—	—
	W	65	6y/4 episodes of massive rectal bleeding	Entire colon,small bowel	+ (sister)	—	—
Tang 2004 (27)	W	62	20y obscure g.i. bleeding	Entire colon,small bowel	—	Conservative	—
Mehta 2004 (28)	M	30	3d melena	Hepatic flexure varices	—	Conservative	No rebleeding (8m)

Whether the dilated and tortuous veins that were detected in the ileum represent an early stage of small bowel varices remains to be verified by subsequent enteroscopies.

The prognosis of hemorrhage from idiopathic C.V. appears to be good and current treatment recommendations include mainly conservative measures (resuscitative measures, selective mesenteric arterial infusion of vasopressin if needed and close follow-up). Younger patients with recurrent hemorrhage or failure of conservative therapy require segmental colonic resection; almost 1/3 of patients with idiopathic C.V. underwent that procedure (6,10,11,12,13,15,17,19,20,21).

In conclusion, we present a case of lower GI bleeding in a 74 year old patient due to idiopathic colonic varices. We, therefore, suggest that the minor possibility of bleeding from idiopathic C.V. should not be overlooked, even in an elderly, previously asymptomatic patient. Family members should be questioned, since a familial tendency has been reported. Small bowel involvement is not infrequent, thus, whenever idiopathic C.V. are demonstrated, we recommend thorough endoscopy of the gastrointestinal tract. Conservative management is the first treatment approach and resection is reserved for cases of continuing or recurrent bleeding.

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