Idiopathic colonic varices : an unusual cause of massive lower gastrointestinal hemorrhage

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Abstract

Varices of the entire colon are very rare. This rare cause of massive lower gastrointestinal hemorrhage is almost invariably associated with cirrhosis of the liver and consequent hypertension or portal venous obstruction.

We report about a patient with massive lower gastrointestinal bleeding from extensive colonic varices. Despite extensive investigation and a follow-up of 3 years, the etiology of the colonic varices could not be determined. Only a few cases of apparent idiopathic (familial or non - familial) colonic varices have been described.

Recognition of this abnormality is important, however, because colonic varices may be the cause of recurrent, frequently massive lower gastrointestinal hemorrhage.

A misleading endoscopic diagnosis can lead to inappropriate biopsies, resulting in major bleeding. (Acta gastroenterol. belg., **2000**, 63, **397-399**).

Key words : colonic varices, hemorrhage, idiopathic.

Case report

A 54-yr-old, otherwise healthy man was admitted to the hospital with the history of bright red blood per rectum for 2-3 days. He had no previous gastrointestinal complaints and there was no family history of gastrointestinal bleeding.

On examination he was moderately obese and looked well.There were no stigmata of chronic liver disease.

Laboratory studies showed a hemoglobin value of 14.6 gr/ dl and hematocrit of 43 %.

GGT were moderately increased.

Colonoscopy revealed large, tortuous bluish veins throughout the entire colon. There was no evidence of active bleeding and the site of bleeding was not identified. There were no ileal varices detected .Additionaly, a sessile polyp more than 10 mm in size was seen in the sigmoid colon ; biopsy caused a sligh bleeding that stopped spontaneously without any endoscopic treatment. Histologic examination revealed a tubulo-villous adenoma with mild dysplasia.

Upper GI endoscopy revealed neither esophageal nor gastric varices, and there was no ulcer and no vascular estasies.

The liver was bright on sonographic scan, consistent with fatty charge, the spleen was normal and there was no evidence of an abnormal portal vein or variceal change around the cardia and splenic hilum.

The 99 Tc-sulphur colloid liver and spleen scans and the aminopyrine breath-test were normal.

The patient's bleeding did not recur and he was discharged to clinic follow-up. Four days later, he was readmitted for recurrent bleeding and his hematocrit dropped to 34.8%

Superior and inferior mesenteric arteriography was undertaken. This showed a normal arterial anatomy with no evidence of early venous filling ; in general the venous anatomy was normal although the veins of the rectum and sigmoid were prominent.

Echodoppler of the pelvic and abdominal vessels was normal. A CT scan of the abdomen with intravenous contrast was normal.

The Hepatic Venous Pressure Gradient (4 mmHg) excluded portal hypertension.

A liver biopsy undertaken at the time of portography showed the histological features of fatty change with no fibrosis or other specific or significant pathological features.

After a follow-up of 3 years, there was no recurrent rectal bleeding ; colonoscopic examination confirmed multiples dilated veins protruding into the lumen without active bleeding and a upper GI endoscopy was always normal.

Based on the results of these investigations a diagnosis of diffuse intestinal varices was made wtih no obvious etiology and most probably of congenital origin.

Discussion

Colonic varices are a rare cause of lower gastrointestinal bleeding. Although the exact incidence of colonic varices remains unknown, one study of 2,912 adult patients with gastrointestinal bleeding undergoing autopsy identified colonic varices as the source of bleeding in only 2 patients (0.07 percent) (1).

Bleeding from colonic varices has been reported to occur from 2.5% of patients attending for sclerotherapy to oesophageal varices (2), and between 3.6% and 56% of cirrhotics patients have been shown to have rectal varices (3).

Males and females appear to be equally affected. The age at diagnosis ranges from 5 to 80 (mean, 50 years) (4). However, nearly 75 percent of all patients

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with colonic varices manifest symptoms between 36 and 65 years of age (5).

Colonic varices are more frequently located on the left side of the colon in the inferior mesenteric venous distribution (66 percent).

Less commonly, they present in the superior mesenteric venous distribution (26 percent). The remainder are distributed diffusely throughout the colon (4,6).

It has been suggested that both primary and secondary varices of the colon are due to congenital abnormalities. Moreover, the familial occurence in the absence of any sign of portal hypertension and the involvement even of those areas lacking porto-systemic anastomoses (transverse colon and small bowell between duodenum and ileocecal area) suggest an inherited vascular alteration on which portal hypertension may be superimposed, although not mandatorily (7).

Portal hypertension is by far the most common cause of colonic varices, which almost also occurs with rectal varices. They are more frequent in alcoholic cirrhotic patients with oesophageal varices than in those without oesophageal involvement.

Segmental portal hypertension with colonic varices has been described as a complication of local mesenteric or splenic vein obstruction after pancreatitis or ligation of the inferior mesenteric vein during left hemicolectomy (8).

Other causes include crowding and kinking of splancnic arterioles and capillaries due to adhesions, compression of the superior mesenteric vein by anormal ileocaecal anatomy and congenital failure of the ileocaecal vein (9).

Our case, with colonic varices throughout the entire colon without detectable cause, is exceptionnaly rare. We only found 25 cases in the literature(8,10,11). Of interest is the fact that in 14 of these patients another family member with the same condition could be detected.This strongly suggests the importance of genetic factors in the etiology.

Thus, some authors have used the term "familial colonic varices" (12).

Clinical presentation of colonic varices is characterized by painless, bright red blood per rectum of varying quantity. More than one-half of all patients with colonic varices have had multiples episodes of bleeding over a period of more than one year (5,13).

Colonic varices can also be asymptomatic (10).

Colonoscopy and mesenteric angiography are considered to be the most helpful diagnostic methods. Endoscopy allows direct visualization of the varices. However, the sensitivity of colonoscopy is greatly reduced during period of active bleeding and when adequate mechanical bowel preparation has not been achieved (4).

Colonic varices appears as tortuous vascular dilatations with a bluish discoloration.

Collapses of the varices, during periods of hypotension secondary to decreased intravascular volume or because of increase intraluminal pressure secondary to the insufflation of air reduce the sensitivity of colonoscopy and may result in the failure to identify these lesions.

Occasionally, colonic varices may resemble polyps and, if biopsied, can bleed profusely (5).

The management of an associated polyp requires the highest care, since polypectomy or even a simple biopsy of the polyp may lead to bleeding (7). Splanchnic angiography is poor contributive and moreover it is invasive and expensive (12).

Barium enema, particularly without double contrast, may not lead to the correct diagnosis, since it may be interpreted as normal or varices may be described to inflammatory bowel disease, polyposis, air bubbles, fecal material... (13).

A technetium-99m-labeled red blood cell or, more easily, a technetium-99-sulfur colloid scan may localize bleeding to the left or right side of the colon (4).

Esophagogastroduodenoscopy should be performed to exclude an upper gastrointestinal source of bleeding and a wedged hepatic vein pressure to exclude portal hypertension. The prognosis of hemorrhage from idiopathic varices appears to be good, especially compared with varices secondary to portal hypertension and cirrhosis.This may be related to low pressure in the varices as well as the absence of significant hepatocellular disease.

Recurrent hemorrhage in a young patient often necessitates segmental or subtotal colectomy.

On the 25 published cases, 9 patients had been treated by segmental colectomy.

For patients presenting in later life, it may be appropriate to make every effort at conservative medical support (5,8).

The therapeutic approach of bleeding from colonic varices due to portal hypertension is controversial (14). Conservative treatment is usually successful in minor bleeding episodes with only hemochezia. The effect of beta-blocking agents and nitrates on the colonic varices of hypertension portal is unknown. If bleeding from colonic varices is massive and not stopped following correction of coagulopathy and replacement therapy, emergency surgery is indicated. Once the bleeding site is located, resection of the involved colonic segment seems to be the procedure of choice. With a good liver function, selective shunting (portal anastomosis or a transjugular intrahepatic portal systemic stent) seems to be the safer procedure concerning the short-term outcome, but long-term follow-up results are not yet available (4,15).

In conclusion we report an unusual case of idiopathic colonic varices responsible for massive bleeding. A congenital venous anomaly remains the sole explanation.

The prognosis of hemorrhage from idiopathic varices appears to be good, especially compared with varices secondary to portal hypertension and cirrhosis. However, approximatively 30% of the published patients with idiopathic varices have to be treated by surgery.

Recognition of this abnormality is important to avoid improper biopsy that may cause severe hemorrhage.

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