



## Case report

# Isolated non-hemorrhagic cecal varices

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Ectopic varices (those outside of the gastro-esophageal region) are occasionally found on endoscopy in patients with portal hypertension; however they account for a small minority of all variceal bleeds. Cases of isolated cecal varices are quite rare and, when described, often present with acute hemorrhage or evidence of occult bleeding. We present the case of a 29-year-old male with a history of idiopathic portal vein thrombosis and known esophageal varices, who presented for evaluation of abdominal pain. Cecal varices were found on endoscopy, without evidence of bleeding and without varices in the remainder of the colon or rectum. Endoscopic ultrasound and computed tomography were useful in confirming the diagnosis and natural history of these unusual varices.

**Keywords:** endoscopic ultrasound; cecal varices; portal vein thrombosis

## CASE REPORT

A 29-year-old caucasian male, with a history of idiopathic chronic portal vein thrombosis (PVT) with prior esophageal variceal hemorrhage, complained of intermittent right lower quadrant abdominal pain at a visit for routine surveillance esophago-gastroduodenoscopy (EGD).

The patient's PVT was discovered three years earlier during an abdominal ultrasound scan to evaluate right upper quadrant pain. Initial EGD revealed mild portal hypertensive gastropathy without esophageal or gastric varices. There were no indications for colonoscopy. Work-up for auto-immune and hypercoagulable conditions was unremarkable, except for heterozygosity of the prothrombin G20210A mutation, which a hematology consultant did not consider to be causative.

The patient was anticoagulated with warfarin, and repeat right upper quadrant ultrasound with Doppler showed the return of hepatopetal flow. Unfortunately, one year after anticoagulation was discontinued the patient presented with hemorrhage from esophageal varices, which were banded to eradication.

A computed tomography (CT) scan of the abdomen was performed at another hospital, revealing a 6.5 cm cecal mass with increased vascularity and calcifications (Figure 1), stable in size and appearance when compared with a CT scan three years earlier, in which the mass was described as ingested material.

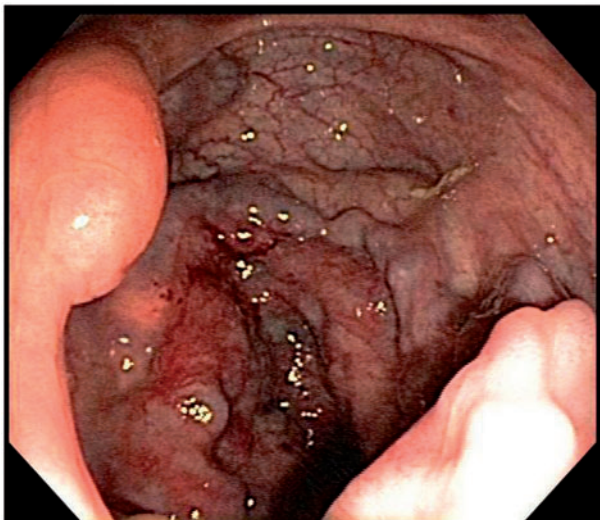
The findings of a physical examination were unremarkable and the patient demonstrated no stigmata of cirrhosis. There was no family history of clotting disorders or liver disease, and he denied significant alcohol consumption.

A colonoscopy revealed an endoscopically normal colon, except for multiple circumferential blue-purple cords from the appendiceal orifice to the ileocecal valve, which appeared consistent with varices, some with overlying vascular ectasias (Figures 2 and 3), without other colonic or rectal varices noted.

Examination with a radial endoscopic ultrasound mini-probe demonstrated the lesions to be anechoic tubular structures, consistent with varices (Figure 4). Superficial biopsies of the mucosa adjacent to the varices showed benign colonic mucosa, with increased sub-epithelial capillaries and focal glandular architectural changes. The absence of



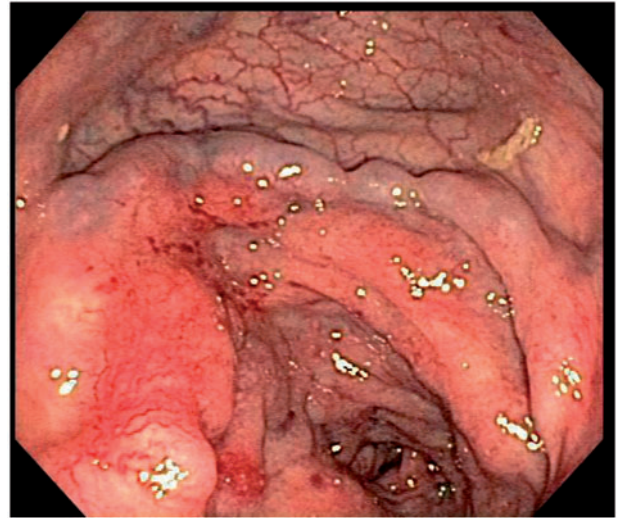
**Figure 1.** Coronal section of a CT scan of the abdomen demonstrating an apparent 6.5 cm cecal mass with calcifications.



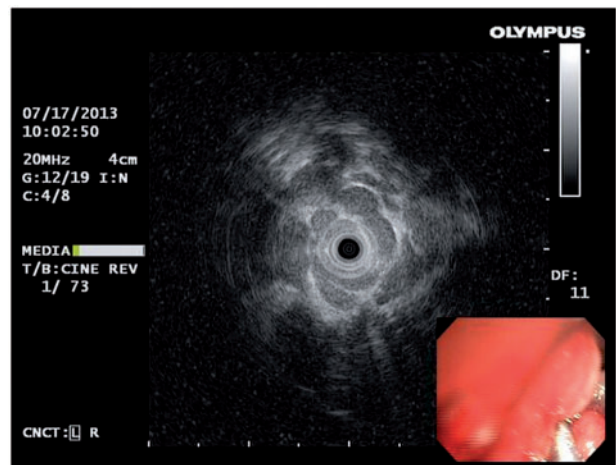
**Figure 2.** Endoscopic view demonstrating cecal varices below the ileocecal valve (left) and above the appendiceal orifice (bottom center).

submucosal vascular ectasias argued against underlying congenital vascular malformation.

The patient's abdominal pain was subsequently diagnosed as irritable bowel syndrome and treated



**Figure 3.** Close-up endoscopic view demonstrating cecal varices (center), overlying vascular ectasia (bottom left), and appendiceal orifice (bottom center).



**Figure 4.** Endoscopic ultrasound of the varices shows thin-walled, anechoic tubular structures.

symptomatically with anti-spasmodics. Propranolol was prescribed, initially 10 mg twice daily, titrated to target heart rate of 55–65 beats per minute.

## DISCUSSION

Ectopic varices are known manifestations of portal hypertension and account for 1–5% of all variceal bleeds [1]. They have been described in the small intestine, biliary system, colon, rectum, and surgical ostomy sites [2]. Colonic varices are less common but it is important to recognize them, as the initial presentation is often massive

hemorrhage. First described in 1954 [3], approximately 100 cases of isolated colonic varices (in the absence of esophageal or other ectopic varices) have been reported [2]. Colonic varices are most often associated with portal hypertension and have been reported in 3.4% of these patients, but are also described in cases of biliary atresia or sclerosis and mesenteric vein thrombosis [4]. Cecal varices are an even rarer entity and are hardly ever found in the absence of pan-colonic varices [5, 6]. In this case, portal hypertension resulting from PVT was believed to be the cause.

Only 17 cases of cecal varices have been reported [2], and all but one presented with massive lower gastrointestinal hemorrhage. Previous cross-sectional imaging of our patient had shown what appeared to be a right lower quadrant mass at the time the PVT was discovered, stable to the time of colonoscopy. This case demonstrates that cecal varices may be present in patients for a significant length of time without hemorrhage. Due to a paucity of literature on the topic [1, 2, 7], the dilemma of how to diagnose and treat such a patient remains.

Methods proposed for detection of colonic varices include routine endoscopy, barium enema, angiography, nuclear scintigraphy, computed tomography and CT angiography, capsule endoscopy, endoscopic ultrasound, and color Doppler-flow imaging [1]. With advances in technology, CT angiography is a promising option [1, 8], and is better able to localize and define colonic varices in a minimally invasive manner, but barium enema, angiography, and scintigraphy have limited utility in evaluating ectopic varices [1, 5, 7, 9, 10].

Endoscopy remains a crucial diagnostic and therapeutic test, but over-insufflation causes the collapse of varices, leading to under-diagnosis, an effect that is compounded during acute hemorrhage with hypotension [5, 11]. Thus, a combination of CT angiography and meticulous endoscopic evaluation of the relevant area (with adjunctive use of endoscopic ultrasound [11]) may yield the best results, although further research is necessary.

Color Doppler imaging has also been used to evaluate gastro-intestinal varices [12]. It may be useful for stratifying bleeding risk in gastro-esophageal varices by demonstrating reversal of flow in the left gastric vein, but is unproven in colonic varices. However, as it can reliably detect direct communication with the inferior vena cava [12], color Doppler may be employed prior to non-emergent sclerotherapy or embolization of colonic varices.

Another promising study for ectopic varices is capsule endoscopy, given its high overall concordance with endoscopy (96.9% for detection of esophageal varices [1, 13]). Because the vast majority of ectopic varices have a distinct medical etiology (e.g. portal hypertension), and capsule endoscopy has demonstrated that at least 8.1% of patients with portal hypertension have small intestinal varices [14],

it is an attractive diagnostic modality that can be followed by EGD, double-balloon enteroscopy, or colonoscopy for therapeutic interventions. Its usefulness in evaluating colonic varices will probably be limited due to the limits of visualization of the colon in routine practice.

Multiple approaches to the management of acute hemorrhage from colonic varices have been reported: from conservative management with resuscitation and observation, to partial or total colectomy. The management of non-bleeding colonic (and especially cecal) varices is not currently directed by a consensus treatment algorithm.

The use of vaso-active compounds such as octreotide, terlipressin, beta-blockers, and nitrates has not been well studied in the treatment of colonic varices. In one case the use of propranolol was described as resulting in the reduction of the size of colonic varices, but the patient did not tolerate the side-effects of therapy [11].

Endoscopic therapies are limited, as band ligation proximal to the rectum is often not safe or even feasible. Injection sclerotherapy and embolization therapy can be applied but have not been well studied, and care must be taken (especially with cecal varices) to avoid vessels that communicate directly with the inferior vena cava. Finally, sclerotherapy and embolization should be considered as bridges to definitive therapy, since there is no decompression of the portal venous system, and collaterals or progression of other varices are likely to develop [5].

In patients without significant contra-indications, transjugular intrahepatic portosystemic shunt (TIPS) may be considered as a bridge to definitive therapy (i.e. liver transplant). This may also be a valid option in patients unfit for surgery. TIPS may be considered therapeutic with or without adjunctive variceal embolization [1].

## SUMMARY

Isolated cecal varices are a recently described, probably under-reported, and potentially distinct pathophysiological entity. There is no 'gold standard' for diagnosis, imaging or treatment, especially in the absence of acute hemorrhage. This case demonstrates that cecal varices may be present in patients for a significant length of time without hemorrhage. Given the natural history of the condition and the difficulties with pre-emptive treatment, close watchful waiting is reasonable with the understanding that variceal hemorrhage can be life-threatening.

**Conflict of interest:** The authors hereby certify that they have no affiliations with or involvement in any organization or entity with any financial or non-financial interest in the subject matter, devices, or materials discussed in this manuscript.

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