

Solitary Variceal Rupture in the Small Intestine

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Abstract

A 70-year-old man was referred to our hospital to undergo treatment for hepatocellular carcinoma. In hospital, he complained of hematochezia and a laboratory analysis revealed a decreased level of hemoglobin. Abdominal computed tomography revealed a tumor in the small intestine, with slow enhancement of the dorsal region. Double-balloon enteroscopy revealed a submucosal tumor with a depression in the jejunum. Partial enterectomy was performed and a pathological examination demonstrated the presence of a solitary varix. Solitary varix in the small intestine has not been reported previously. We herein report an extremely rare case of solitary varix in the jejunum.

Key words: solitary varix, small intestine, rupture

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Introduction

Varices other than those of the esophagus and stomach were classified as ectopic varices by Leberc et al. in 1985 (1). Ectopic varices located in the small intestine are uncommon and are usually caused by portal hypertension secondary to hepatic cirrhosis (2). A survey conducted by the Clinical Research Committee of the Japan Society for Portal Hypertension (3) reported that varices of the small intestine accounted for 6.4% (11/173) of ectopic varices, with almost all cases involving portal hypertension associated with liver cirrhosis and portosystemic shunt. Conversely, solitary varices do not involve portosystemic shunt and are considered to differ from the common varices related to portal hypertension.

We herein report a case involving the resection of a solitary varix in the small intestine and discuss the findings, with reference to the pertinent literature.

Case Report

A 70-year-old man was admitted to our hospital for a fourth attempt at transcatheter arterial chemoembolization (TACE) against hepatocellular carcinoma (HCC). His medical history included the following conditions: hepatic cirrhosis due to hepatitis C virus; gastric varices, which were treated with balloon-occluded retrograde transvenous obliteration (B-RTO); esophageal varices, which were treated with endoscopic injection sclerotherapy (EIS); and the three previous attempts at TACE for HCC. The patient's Child-Pugh score was class A and a laboratory evaluation on admission showed elevated levels of liver enzymes and tumor markers: alanine aminotransferase, 76 IU/L; aspartate aminotransferase, 91 IU/L; alkaline phosphatase, 662 IU/L; γ -glutamyl transpeptidase, 127 IU/L; α -fetoprotein, 300.8 ng/mL; and protein induced by vitamin K absence/antagonist II, 831.6 mAU/mL. However, his hemoglobin level was normal (13.8 g/dL).

He complained of hematochezia at 2 days after admission, the day before TACE had been scheduled. His vital signs

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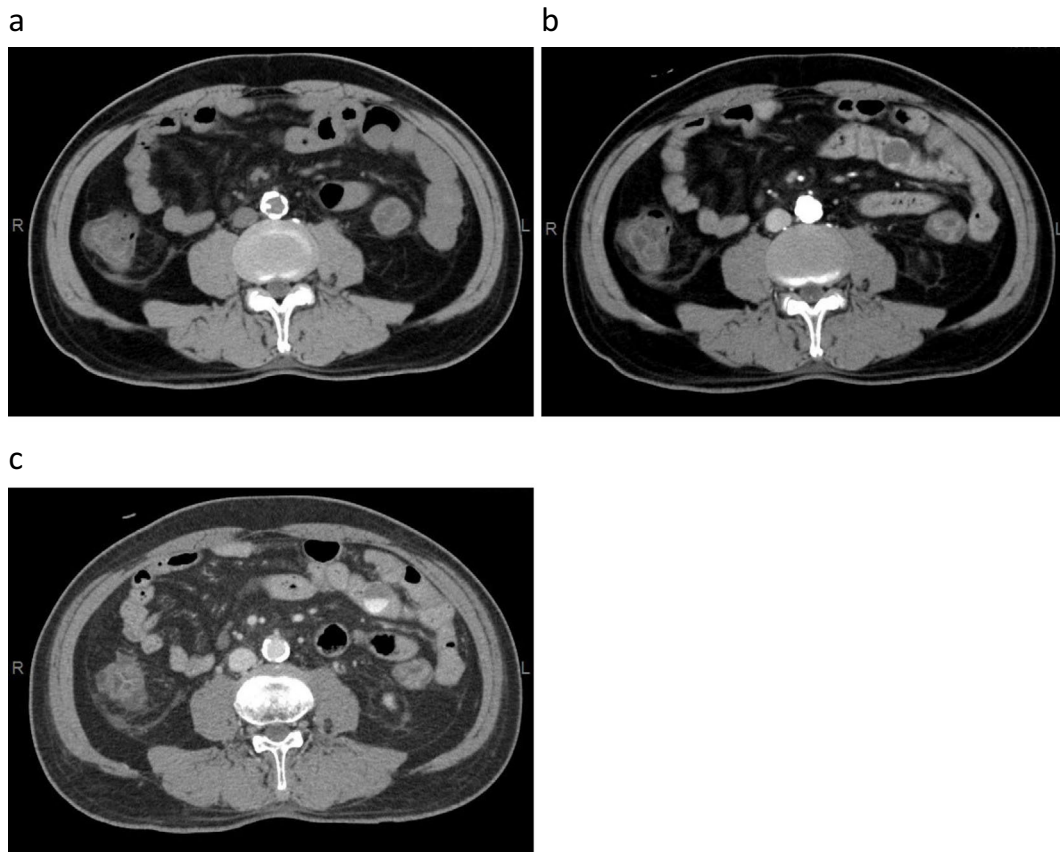


Figure 1. Plain computed tomography (a) demonstrated a low-density tumor of 22 mm in diameter, in the small intestine. The gradual retention of contrast agent was observed in the dorsal area of the tumor from the arterial phase (b) to equilibrium phase (c).

were as follows: heart rate, 109 beats/min; blood pressure, 110/80 mmHg; temperature, 37.0°C; and respiratory rate, 24 breaths/min. A physical examination showed pale palpebral conjunctivae but no abdominal tenderness. Laboratory testing revealed that the hemoglobin level had rapidly decreased to 8.4 mg/dL, and emergency examinations were performed. Dynamic computed tomography of the abdomen demonstrated a low-density tumor of 22 mm in diameter in the small intestine. The dorsal area of the tumor showed the gradual retention of contrast agent from the arterial phase to the equilibrium phase. No meandering or dilated vessels were seen around the tumor (Fig. 1). Esophagogastroduodenoscopy showed esophageal varices (Lm, F1, CW, RC0) but no evidence of bleeding. Colonoscopy showed no source of bleeding, but fresh blood was seen on the oral side of the intestinal tract in the ileocecal region. As the possibility of active bleeding from the small intestine was considered, emergent abdominal angiography was performed. Celiac angiography, superior mesenteric angiography and aortography revealed no extravasation, aneurysm or tumor staining. These results suggested the absence of active bleeding, thus conservative medical treatment was selected at that time. Double-balloon enteroscopy was performed at 8 days after admission to study the small intestinal tumor, revealing a submucosal tumor measuring about 20 mm in diameter in the jejunum. A depression attached to a clot was observed

on the top of the tumor (Fig. 2). Biopsy was not performed out of concern over the risk of re-bleeding. The tumor appeared to be localized to the submucosa on gastrografin (Schering, Berlin, Germany) fluoroscopy (Fig. 3). Abdominal ultrasonography showed a well-circumscribed, round tumor in the small intestine, measuring 17 mm in diameter and arising from the third and fourth layers of the intestinal wall. The internal echogenicity of the tumor was almost anechoic, but part of the tumor was hyperechoic (Fig. 4). On Doppler ultrasonography, no blood flow signal was detected inside or around the tumor. Contrast-enhanced ultrasonography (CEUS), using Sonazoid as a contrast agent (Daiichi-Sankyo, Tokyo, Japan), also showed the absence of blood flow in the tumor and no abnormal veins around the tumor.

Based on all of these results, we diagnosed bleeding from the submucosal tumor in the small intestine. Partial enterectomy revealed a submucosal tumor located 100 cm on the anal side of the ligament of Treitz and measuring 18×15×10 mm. The tumor showed a unilocular cystic lesion, which contained a clot inside, with an ostium on the mucosal surface (Fig. 5).

A histological examination (Fig. 6a and b) revealed a cystic lesion extending from the submucosa to the muscularis propria. The cystic lesion contained a blood clot and was surrounded by the thinning of the smooth muscle. Laceration of the muscularis mucosa and mucosal layer was ob-

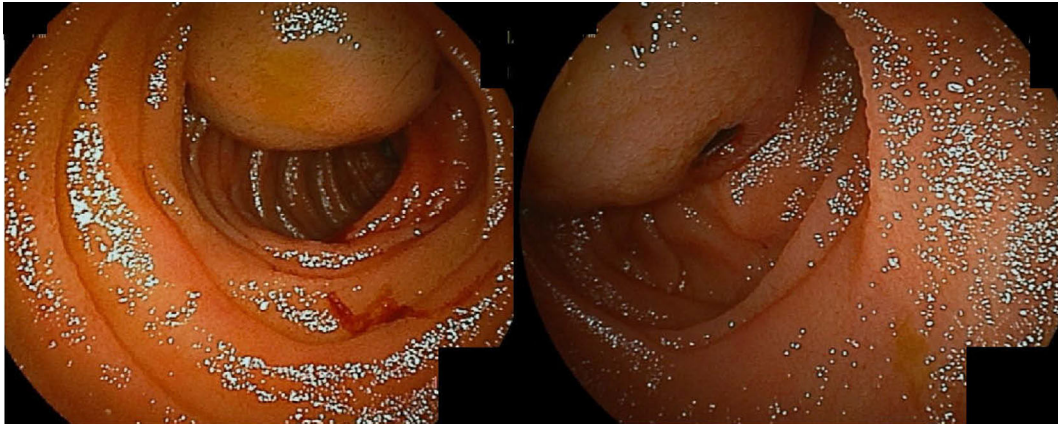


Figure 2. Double-balloon enteroscopy showed a submucosal tumor of approximately 20 mm in diameter in the jejunum. A depression attached to the clot is shown at the top of the tumor.

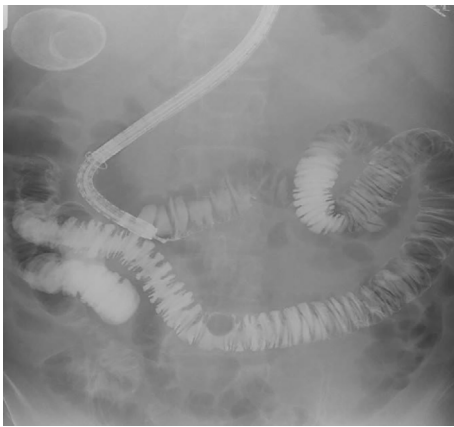


Figure 3. Gastrografin fluoroscopy showed a mass with a smooth margin forming obtuse angles with the jejunal wall.



Figure 4. Abdominal ultrasonography showed a well-circumscribed, round tumor in the small intestine, arising in the third and fourth layers of the intestinal wall.

served at the top of the cystic lesion. An immunohistochemical examination (Fig. 6c and d) revealed that the flat cells on the luminal surface of the cystic lesion were positive for factor VIII and CD34 and negative for D2-40 and keratin, indicating vascular endothelial cells. The thinned smooth muscle in the cyst wall was positive for muscle actin (HHF35) and was considered to be equivalent to vascular media. No migration of organization from the artery to vein (which would imply arteriovenous malformation), was seen, and no dilated vessels in the submucosa were evident (as would be present in typical varices). Based on the pathological and imaging findings, we finally diagnosed a solitary varix in the jejunum. The patient's postoperative course was uneventful and he was discharged from hospital on postoperative day 12.

Discussion

The origin and clinical significance of solitary varices have not yet been sufficiently investigated. Solitary varices in the upper and middle esophagus are considered to differ from both the common esophageal varices related to portal

hypertension and 'downhill varices' in the upper esophagus related to elevated pressure in the superior vena cava (4-9). A solitary varix in the esophagus rarely causes morphological changes or hemorrhage (10-12). However, rupture cases have been reported, and Garret et al. reported a case in which the patient died of hemorrhage (4). Although the etiologies of esophageal solitary varix have been clarified, Fukuda et al. reported that age-related focal fragility of the submucosal vein was more frequently the main cause of solitary varix in elderly patients (10). The patient in our case was also elderly and we suspect that age-related focal fragility of the submucosal vein was a cause. Although the possibility that it was caused by portal hypertension cannot be ruled out, we do not consider this to have been a direct cause.

In the present study, all varices other than those of the esophagus and stomach were defined as ectopic varices. A nationwide questionnaire survey conducted in 1990 revealed that ectopic varices were diagnosed at an extremely low frequency (129 of 18,540 cases, 0.7%) (11). Small intestinal varices, which were initially reported by Blackburn. in 1956 (12), are reported to account for only 6.4% (11/173)

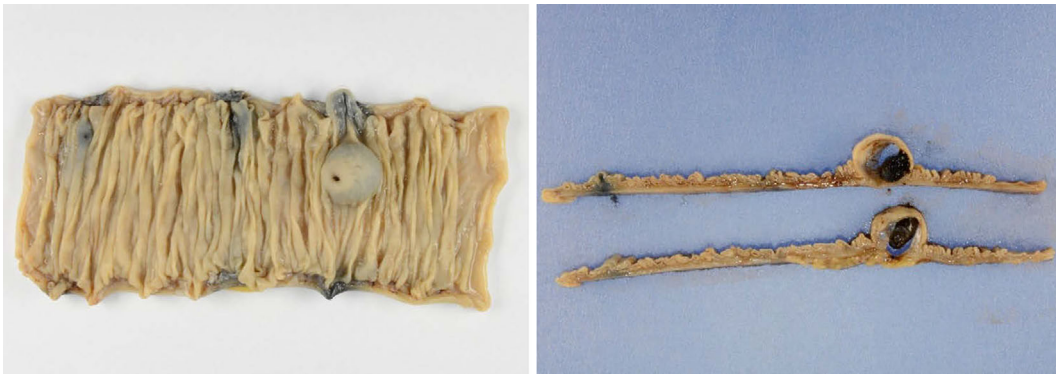


Figure 5. A macroscopic examination revealed a unilocular, cystic lesion, measuring 18×15×10 mm in size and containing a clot. An ostium was apparent on the mucosal surface.

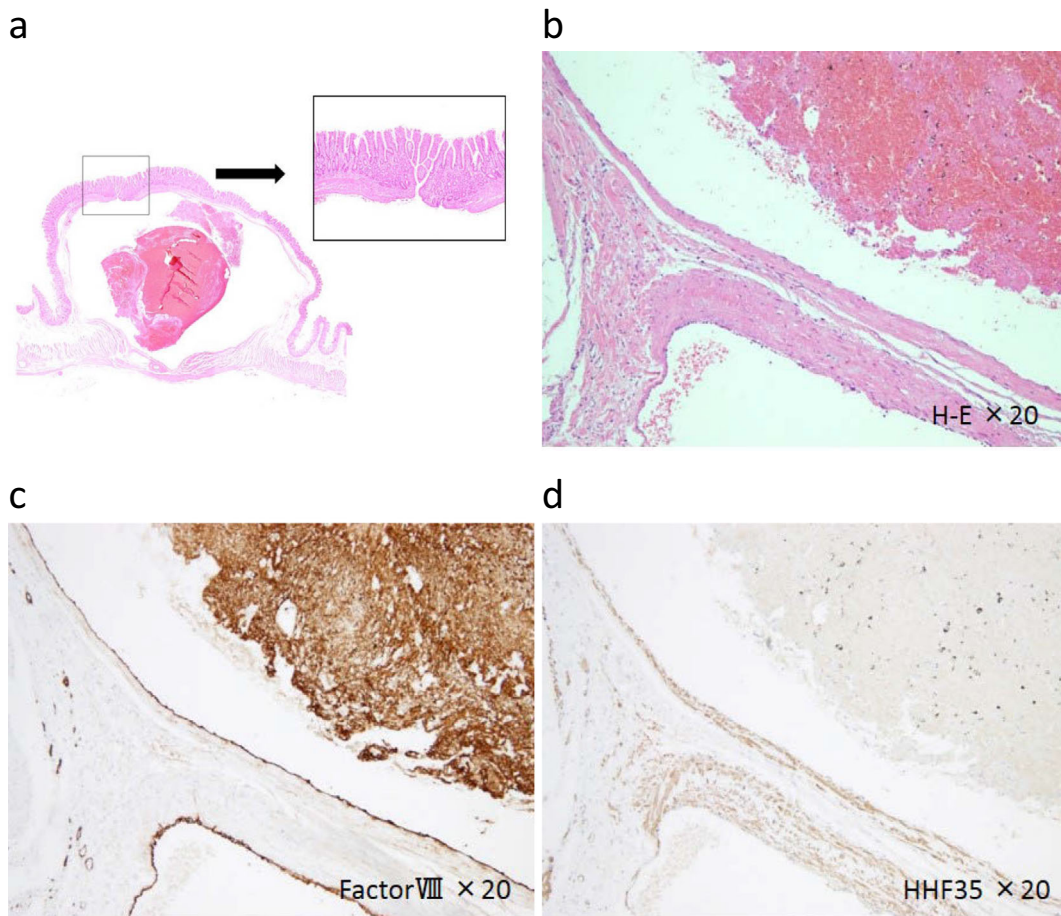


Figure 6. A histological examination (a, b) showed a cystic lesion extending from the submucosa to the muscularis propria. A blood clot was present inside, with a laceration of the muscularis mucosa and the mucosal layer at the top. An immunohistochemical examination showed that the cells covering the surface of the cystic lesion were positive for factor VIII (c) and that the wall was positive for HHF35 (d).

of ectopic varices (3) and are therefore regarded as relatively rare. The majority of patients with small intestinal varices have portal hypertension in the context of cirrhosis or portal vein thrombosis and a history of previous abdominal surgery, and adhesion of the intestinal tract due to surgery and/or chronic intraperitoneal inflammation is etiologically important. Vascularization is considered to occur at sites of ad-

hesion and the pressure overload resulting from portal hypertension added to the fragile vessels causes portosystemic shunting (13). In our case, the patient had portal hypertension as a result of hepatic cirrhosis due to hepatitis C virus; thus, it was important to differentiate from ectopic varices related to portal hypertension. The crucial difference from existing reports is that the patient's varix showed no inflow

or outflow vessels on imaging, operative, or pathological examinations. Moreover, a histological examination revealed the absence of dilated vessels in the submucosa and the varix displayed a solitary form, like a submucosal tumor. Such findings are not present in typical varices, thus this varix was considered to be a truly solitary varix, and the mechanisms that led to its development differed from those that are usually involved in the formation of varices. Arteriovenous malformation should be considered as a differential diagnosis of this case. However, a superior mesenteric angiogram did not reveal any evidence of arteriovenous malformation of the jejunum and the pathological findings showed no migration of the organization from artery to vein; thus, arteriovenous malformation could be excluded. Our investigation of the literature found many reports about solitary varices in the esophagus, but no reports of solitary varices in the small intestine, as occurred in the present case. Thus, the present report represents the first description of such a case.

This disease represents one of the differential diagnoses of gastrointestinal hemorrhage. The imaging findings presented in this case are therefore important for reaching a correct diagnosis. The distinctive feature of the imaging findings in the present case was the gradual retention of the contrast agent. This finding was considered to reflect slow, weak internal blood flow. On ultrasonography, part of the interior of the varix was hyperechoic, probably indicating a blood clot; this finding may be useful in the differential diagnosis.

The treatment methods for small intestinal varices are limited and surgery has been the main treatment option for bleeding small intestinal varices (14-16). Some recent reports have described the application of interventional radiology (IVR) treatment methods, such as transjugular intrahepatic portosystemic shunt (TIPS) and B-RTO (17, 18). Our patient did not show any portosystemic shunting, IVR was not considered to be feasible and surgery was judged to be the most appropriate treatment method. Although the necessity of preventive resection in cases in which solitary varix is found incidentally (similar to the present case) remains controversial, it is important to determine a treatment plan after obtaining informed consent from the patient. However, in cases in which a varix shows a tendency to increase in size, resection is preferable because of the risk of rupture.

In conclusion, we encountered a case of a solitary varix in the small intestine. We reported this as an extremely rare case in which integrative diagnostic imaging and a histopathological examination were performed.

The authors state that they have no Conflict of Interest (COI).

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