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【 CASE REPORT 】

Unusual Gastrointestinal Hemorrhaging Mimicking a Rupture of Solitary Gastric Varices Due to a Gastric Gastrointestinal Stromal Tumor with Exogenous Growth

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Abstract:
Gastric gastrointestinal stromal tumors can lead to upper gastrointestinal hemorrhaging, which is usually caused by dimpling or ulceration on the tumor surface. While rare, pedunculated gastric gastrointestinal stromal tumors outside the stomach can present as a huge mass with delayed complaints. We herein report an unusual hemorrhaging mimicking a rupture of solitary gastric varices due to a pedunculated gastric gastrointestinal stromal tumor. In this case, contrast-enhanced computed tomography (CECT) was essential for tumor detection. An endoscopic investigation revealed dilated, aberrant veins and arteries in the submucosa of this tumor, recognized as solitary gastric varices.

Key words: gastrointestinal stromal tumor, GIST, pedunculated gastrointestinal stromal tumor, pedunculated GIST, upper gastrointestinal hemorrhaging, UGIH

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Introduction

Gastrointestinal stromal tumors (GISTs) show highly variable growth features (1). The gross appearance includes protrusion into the luminal side (type I), a wide connection with the proper muscle and protrusion into the luminal side (type II), an intra- or transmural location (type III), and protrusion mainly into the serosal side of the gastric wall (type IV) (1, 2). Endogenous growth of GISTs often leads to upper gastrointestinal hemorrhaging (UGIH); however, the presentation of clinical symptoms of exogenous growth often occurs later (3, 4). UGIH is the most common symptom of GIST, leading to 52.0-54.5% of diagnoses (5, 6). Particularly in cases of GIST-related emergencies, UGIH is the most common cause of admission (48.9%) (7, 8). The presence of dimpling on the GIST surface and mucosal break or ulceration after tumor growth >5 cm in size are independent risk factors for GIST-related UGIH (9, 10).

We herein report a case of an unusual UGIH derived from vascular-enriched gastric submucosa that mimicked solitary gastric varices, caused by a pedunculated gastric GIST.

Case Report

A 64-year-old man was admitted to our emergency department because of hematemesis. He presented with a normal blood pressure (120/90 mmHg) and tachycardia (102 beats/min). His medical history included neither peptic ulcers nor liver diseases. No anticoagulants, antithrombotic agents, or non-steroidal anti-inflammatory drugs were prescribed. The patient was a non-smoker and social drinker.

Blood test results upon admission were as follows: white blood cell count of 11,100/μL; hemoglobin level of 4.6 g/dL; platelet count of 20.8×10^4/μL; C-reactive protein level of 0.06 mg/dL; blood urea nitrogen level of 25.4 mg/dL [upper normal limit (ULN): 20.0 ng/mL]; creatinine level of 0.70 mg/dL (ULN: 1.07 U/mL); prothrombin time-international normalized ratio of 1.12; carcinoembryonic antigen level < 0.5 ng/mL (ULN: 5.0 ng/mL); and carbohydrate antigen 19-

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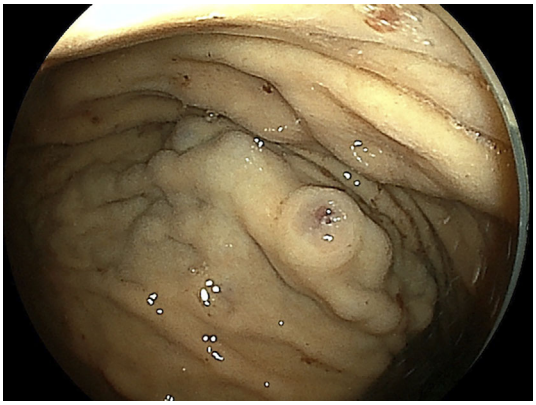

Figure 1. **Upper endoscopic findings on admission. Bluish-bloated gastric mucosal folds of the greater curvature with a string-of-beads aspect and a red spot. This finding mimicked the post-ruptured status of solitary gastric varices.**

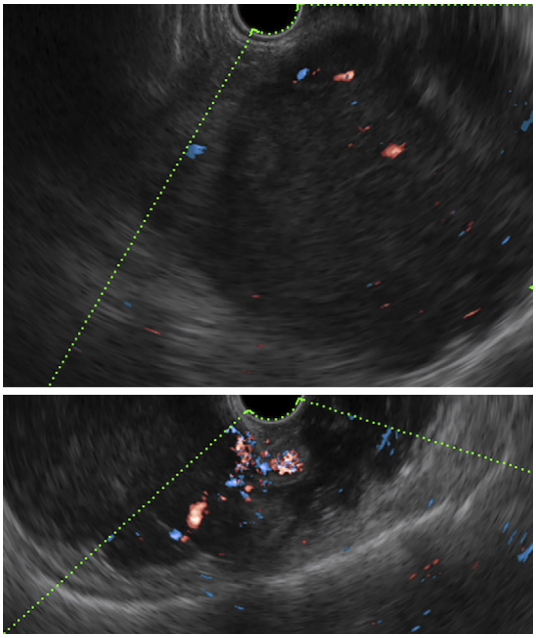

Figure 3. **Endoscopic ultrasonography (EUS) at 7.5 MHz. Doppler-EUS at 7.5 MHz depicted hypervascular mass with vascular-enriched submucosa.**

9 level of 2.0 U/mL (ULN: 37.0 U/mL). Since the patient had a Glasgow Blatchford score of 13 (11-13), he underwent urgent upper endoscopy with prompt intravascular volume replacement using crystalloid fluids.

Upper endoscopy revealed bluish-bloated gastric mucosal folds of the greater curvature with a string-of-beads aspect, and a red spot was suspected as the bleeding point. The endoscopic findings mimicked the post-ruptured status of 'solitary gastric varices'(Fig. 1). No other lesion was detected in this examination. A small amount of brownish-colored remnant remained in the stomach. Endoscopic hemostasis was not performed following spontaneous hemostasis.

Contrast-enhanced computed tomography (CE-CT) performed following blood transfusion revealed an irregular-

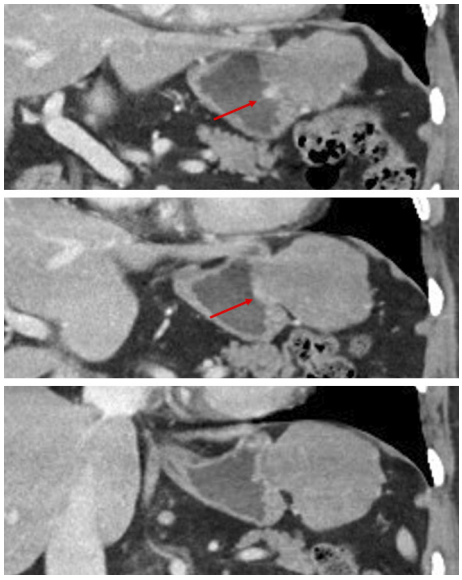

Figure 2. **Coronal contrast-enhanced computed tomography. An irregularly shaped tumor 60 mm in diameter on the serosal side of the stomach. The enhanced mucosal side of the tumor suspected of being vascular-enriched gastric submucosa.**

shaped tumor 60 mm in diameter on the serosal side of the gastric greater curvature initially. Hyper-vascular gastric submucosa at the connecting point was revealed on coronal CT (Fig. 2). There were no signs of liver cirrhosis, splenomegaly, or collateral vessels. We carried out further examinations using endoscopic ultrasonography (EUS) at frequencies of 7.5 and 12.0 MHz. These EUS investigations depicted a hyper-vascular submucosal tumor with vascular-enriched submucosa and previous crater-like mucosal collapsed point connected to the developed gastric submucosal vessels (Fig. 3, 4). The specimen obtained by an EUS-guided fine-needle aspiration biopsy (EUS-FNAB) revealed GISTs derived from the stomach, and surgical intervention was scheduled.

Wedge resection under laparoscopic surgery during a single admission was performed with an uneventful perioperative course. The intraoperative findings showed an irregular-shaped tumor pedunculated from the gastric greater curvature, with a narrow contact base. The developed vessels were confirmed on the tumor surface (Fig. 5). Peritoneal dissemination was also identified.

The resected tumor was 80 mm in diameter. A microscopic examination revealed that the muscular layer-derived tumor grew exogenously, but the luminal side of the muscular layer was maintained. This vascular-rich submucosa contained dilated, aberrant veins and arteries, similar to 'angiodysplasia'(Fig. 6). This angiodysplasia was recognized as solitary gastric varices on an endoscopic examination. The diagnosis was gastric GIST based on the immunohistochemical staining findings of c-KIT (+) and CD34 (+) (Fig. 7) with a mitotic index <5/50 high-power fields. Gastric GISTs of intermediate risk were also confirmed (14).

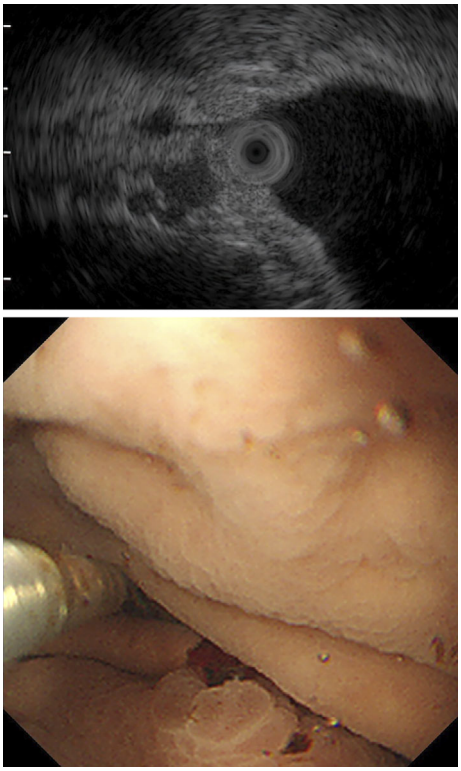

Figure 4. **Endoscopic ultrasonography at 12.0 MHz from an endoscopic view. Crater-like, hypoechoic mucosal collapsed point. This was connected to the gastric submucosal vessels.**

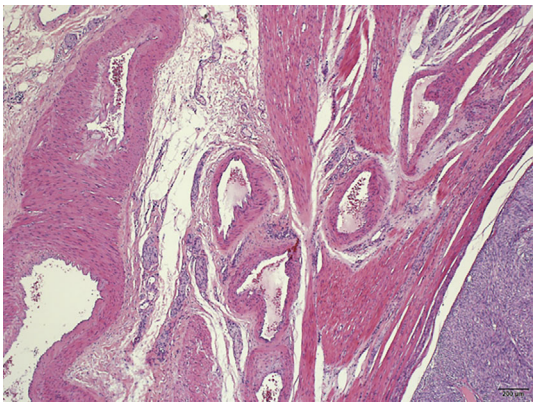

Figure 6. **Representative microscopic findings of submucosal layers on the tumor. Tumor derived from the muscular layer. Dilated, aberrant veins and arteries resembling angiodysplasia were observed in the submucosal layer.**

Discussion

We encountered a rare case of a pedunculated gastric GIST that caused UGIH mimicking a rupture of solitary gastric varices.

UGIH is the most common manifestation of GIST, leading to 52.0-54.5% of diagnoses (5, 6) and 48.9% of admissions (7, 8). In general, GIST-related UGIH is caused by dimpling on the GIST surface (9) or a mucosal break or ulceration caused by a tumor diameter >5 cm (10). GIST-related UGIH was caused by mucosal and submucosal destruction triggered by tumor growth, vascular rupture caused by invasion of nutrient vessels, and tumor necrosis (15).

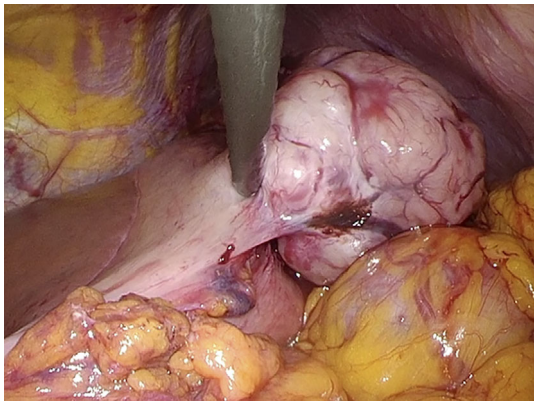

Figure 5. **Intraoperative gross appearance of the tumor. An irregular-shaped tumor pedunculated from the gastric greater curvature, with a narrow contact base.**

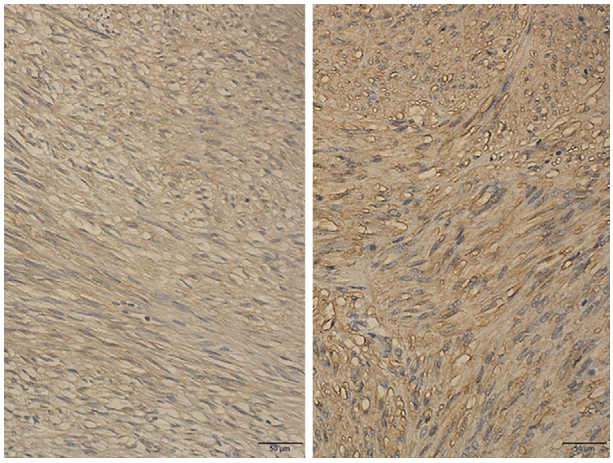

Figure 7. **Immunohistochemistry staining for c-KIT and CD34 in the tumor. Staining was positive for both c-KIT (left side) and CD34 (right side) in this tumor.**

GIST protrudes not only from the mucosal side of the gastric wall (type I and II) but also the serosal side (type III and IV) (1, 2). As many as 79% of GISTs show exogenous growth with clinical symptoms occurring later (16), and most exogenously pedunculated gastric GISTs are huge masses (12-15 cm) with these patients presenting with delayed complaints of abdominal masses (17-19). Therefore, in some studies, these GISTs were misdiagnosed as pancreatic (17) or liver (20) tumors. In gastric GIST-related emergencies, manifestations other than UGIH include spontaneous rupture, massive intra-abdominal bleeding, and peritonitis (7). In addition to the extra-gastric features, tumor size, and mitotic index, GIST-related UGIH has been reported as an independent risk factor for a poor prognosis. If GIST-related UGIH develops, it should be equated with tumor rupture and considered to have malignant potential (16).

In the present case, unusual UGIH from submucosal ves-

sels was suspected to have been caused by a pedunculated gastric submucosal tumor. Submucosal dilated, aberrant veins and arteries, similar to angiodysplasia, were recognized as solitary gastric varices on endoscopic examinations. Unitarily, these vessels developed as a result of tumor growth. However, even after a discussion with pathologists, it was difficult to confirm whether this angiodysplasia coexisted with GIST incidentally or had developed secondary to GIST.

CT, ideally with contrast enhancement, is clinically useful in cases of UGIH to identify the cause of bleeding, such as peptic ulcers, esophageal and gastric varices, carcinomas, pseudoaneurysms, and other tumors, including GISTs.

Laparoscopic resection is feasible for gastric GISTs <50 mm but is challenging for those >50 mm (21). The present GIST was 60 mm in diameter; however, it was located in the gastric greater curvature and showed extra-gastric pedunculated growth. Therefore, wedge gastrectomy under laparoscopic surgery was considered reasonably safe and feasible.

To our knowledge, there have been no reports of solitary gastric pseudo-variceal rupture caused by pedunculated GISTs. At present, there are no standard hemostasis guidelines for GIST-related UGIH, such as adrenaline injection, argon plasma coagulation, sclerotherapy, hemoclips, and hemostatic graspers (10). In the present case, we were fully prepared to perform hemostasis, including sclerotherapy with histoacryl, however, spontaneous hemostasis had been already achieved and therefore hemostasis no longer needed to be performed.

In conclusion, we described a rare case of pedunculated GIST that caused UGIH, mimicking rupture of solitary gastric varices.

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

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