

Bleeding from isolated gastric varices as complication of a mucinous cystic neoplasm of the pancreas

A case report

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Abstract

Introduction: Bleeding from isolated gastric varices, though uncommon, may be life threatening and may occur as a consequence of splenic vein thrombosis by tumoral compression and subsequent left-sided portal hypertension.

Case overview: We report the case of a 37-year old woman, previously diagnosed with a benign pancreatic cyst, who presented with severe gastric variceal bleeding.

Diagnosis, therapeutics interventions, and outcomes: Abdominal ultrasound (US) and computed-tomography (CT) revealed enlargement and changed morphology of the cystic tumor located on the body of the pancreas. Left-sided portal hypertension was disclosed resulting from splenic vein occlusion. Salvage left spleno-pancreatectomy with lymphadenectomy was undertaken with an excellent postoperative outcome. Histological analysis established the diagnosis of mucinous cystic neoplasm with foci of adenocarcinoma.

Conclusion: Surgical treatment proved to be the election one, leading to a steady hemostasis, removal of the lesion, positive diagnosis, and resolution of the varices. Imaging follow-up of pancreatic cyst should also assess vascular patency to identify the patient at risk for gastric bleeding and to select patients who benefits from surgical resection.

Abbreviations: CEA = carcinoembriogenic antigen, CT = computed tomography, EMA = epithelial membrane antigen, EUS = endoscopic ultrasonography, ICU = intensive care unit, MCA = mucinous cystic adenocarcinoma, MCN = mucinous cystic neoplasia, MIP = maximum intensity projection, MRI = magnetic resonance imaging, US = abdominal ultrasound examination.

Keywords: gastric varices, hematemesis, mucinous cystic neoplasm, pancreas

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1. Introduction

Bleeding from isolated gastric varices is a serious condition. Apart from the endoscopic management of the bleeding, a thorough imaging workup is done to identify the underlying cause of portal hypertension, as well as the inflow and outflow of the varices. We report the case of a young female presenting with upper digestive tract hemorrhage due to huge gastric varices of the fornix.

2. Case report

A 37-year old woman presented to the emergency unit with 4 episodes of hematemesis and melena over the last 4 hours. Her medical history included a pancreatic body thin-walled cyst of 5 x 0.200A;cm, discovered 2 years ago on a routine ultrasound and characterized by CT as a benign lesion (Fig. 1A). As there were no indication of other associated pancreatic or hepatobiliary conditions, the portal system was patent, and the patient refused invasive examinations, a follow-up of the lesion was decided at that time. A follow-up abdominal ultrasound at 1 year showed stability of the lesion. On examination, she was alert and oriented, with a blood pressure of 100/55 mm Hg and heart rate of 110 min⁻¹. She was pale, without any jaundice, lymphadenopathy, or pedal edema. Abdominal examination revealed an enlarged spleen 7 cm below the left costal margin. The digital rectal examination revealed melena and the nasogastric tube

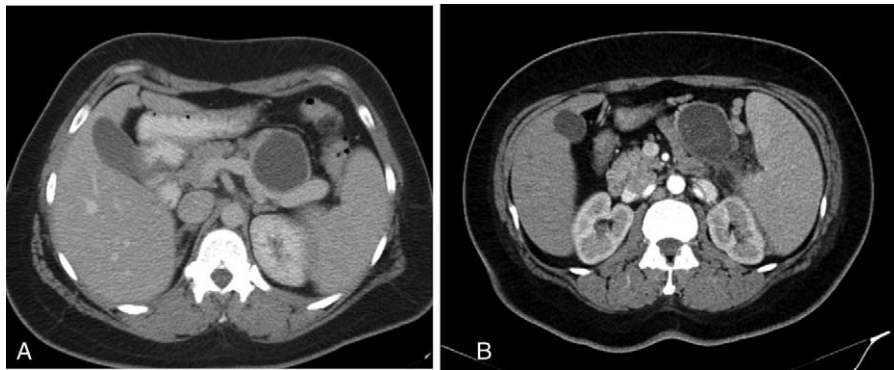


Figure 1. Abdominal CT. January 2015: Incidental finding of a pancreatic cystic lesion located in the tail of the pancreas, well defined, with patent splenic vein and normal-sized spleen (A). January 2017: enlargement of the cystic lesion, with internal enhanced septa and wall (B).

detected a coffee ground aspirate with bright red blood. The rest of the systemic examination was unremarkable and without peripheral stigma of chronic liver disease. She had hemoglobin level of 3.9 g/dL, white blood cell count of $3.35 \times 10^3 \text{ L}^{-1}$, and platelets count of 86.000 mm^{-3} . Her liver function tests, including prothrombin time, were normal, excepting for albumin level of 3.0 g/dL. Hepatitis B and C serology was negative, as well as the seric levels of tumor markers (carcinoembriogenic antigen [CEA] 19–9 antigen). The patient was admitted in the intensive care unit (ICU) for blood volume restitution with blood transfusion, crystalloid, and colloid solutions and started on intravenous omeprazole infusions. The upper tract endoscopy revealed large gastric varices located at the fornix with nipple and red wale signs, indicating the site of bleeding, looking like “varicose veins in varicose veins” (Fig. 2). Abdominal US and CT revealed changes of the size (measuring $7 \times 6 \text{ cm}$ in diameter) and morphology of the pancreatic cyst in the body/tail of the pancreas (still well defined, thin walled, but with enhancing internal septa), associated with segmental occlusion of the splenic vein and homogeneous splenomegaly (Fig. 1B). Dilated veins were identified into the fundus of the stomach (up to 7 mm), without

esophageal varices (Fig. 3A). Maximum intensity projection (MIP) reconstructions identify the inflow at the level of short gastric veins and the outflow to the omental veins and right gastroepiploic veins, draining in the distal part of the dilated superior mesenteric vein and developing a spleno-mesenteric shunt (Fig. 3B). The diagnosis was cystic pancreatic tumor with left-side segmental portal hypertension. As far as the patient was actively bleeding with difficulties in endoscopic management through sclerosis, ligation, or embolization, and 2 high-risk features of the cystic tumor were present (size $>3 \text{ cm}$ and increasing of the solid component represented by septa) a salvage surgery was decided. The peritoneal cavity was entered through a median incision, while the lesser sac was entered by large division of the gastrocolic omentum. Operative findings confirmed nipple gastric aspect, third grade splenomegaly, marked dilation of the gastroepiploic and short gastric veins, and the mass within the body/tail of the pancreas (Fig. 4). Slightly enlarged lymph-nodes along the splenic artery and celiac trunk were also noted. Frozen biopsy from the tumor and several lymph nodes was undertaken. A distal pancreatectomy and splenectomy were performed with lymphadenectomy along the splenic, common hepatic, and celiac

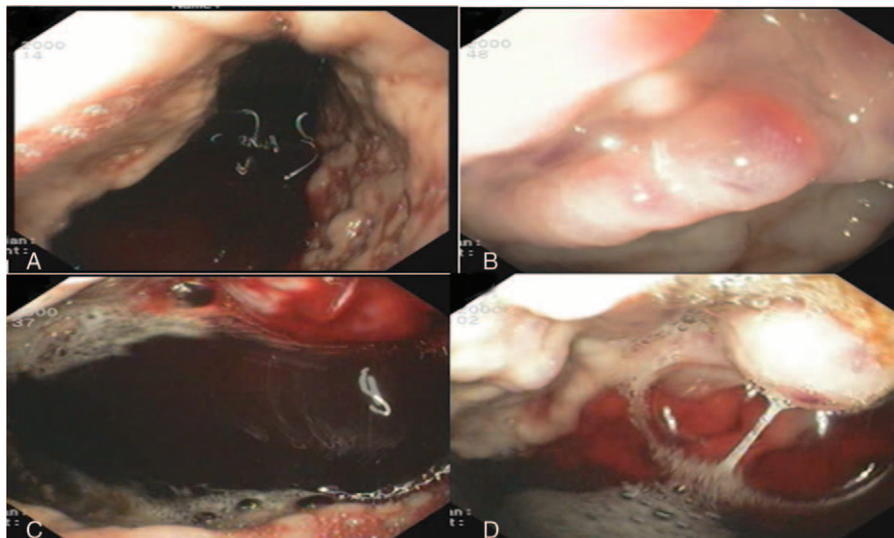


Figure 2. Endoscopy: (A) pale gastric mucosa, varices, and blood into the stomach; (B) congestive gastric varices; (C) gastric varices with active bleeding; (D) gastric varices with signs of recent bleeding.

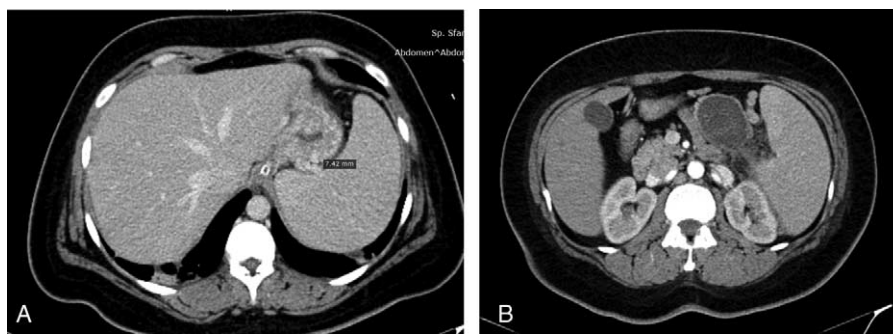


Figure 3. Abdominal CT: January 2017, large gastric varices (A) and gastro-mesenteric shunt (B).

trunk arteries (Fig. 5). The patient had an uneventful postoperative course and was discharged 9 days postoperatively. Histopathology and immunohistochemical marker epithelial membrane antigen (EMA) (MUC1) confirmed the diagnosis of mucinous cystic neoplasia (MCN) with high grade focal dysplasia, foci of proliferation of ovarian stroma inside and invasive component—adenocarcinoma of ductal type with large ducts. She has had no further episodes of upper digestive tract bleeding and a 30th day follow-up endoscopy confirmed complete resolution of the gastric varices (Fig. 6).

3. Discussion

Isolated gastric varices due to splenic vein thrombosis are usually related to the short gastric veins connecting the hilum of the spleen to the greater curvature of the stomach.^[1] The outflow is usually directed to the systemic territory, as a part of a portosystemic shunt, draining into the inferior phrenic vein at the level of gastrophrenic ligament and then forming a gastrenal shunt terminating into the left renal vein, or a gastrocaval shunt terminating into the inferior vena cava.^[2,3] There were also described anastomoses with left pericardiophrenic vein, other peridiaphragmatic and retroperitoneal veins, including the

subcostal and intercostal veins, right inferior phrenic vein, or azygos venous system.^[3,4] In our case the large gastric varices were part of a spleno-mesenteric shunt to overcome a short splenic vein thrombosis.

The 2 commonest clinical features in gastric varices due to splenic vein thrombosis are gastrointestinal blood loss and splenomegaly.^[5] Since isolated gastric varices may be observed in up to 5% of patients with cirrhosis, and up to 10% of patients with non-cirrhotic portal hypertension, further imaging studies are always necessary to evaluate the portal system, as well as the underlying pathology.^[6]

Segmental left-sided portal hypertension represents a clinical entity involving the development of gastric varices in the context of splenic vein thrombosis due to pancreatic pathology. The difference from other forms of portal hypertension resides in the presence of normal liver function and permeable extrahepatic portal vein. Regularly, the reported causes of segmental left-sided portal hypertension include acute and chronic pancreatitis, pancreatic carcinomas, and pancreatic pseudocysts. Less frequently reported are benign pancreatic tumors, although various types of pancreatic cysts, like cystadenoma, choledochal cyst, echinococcal cyst, and also pseudotumors and pancreatic abscesses have all been reported as causing this particular type

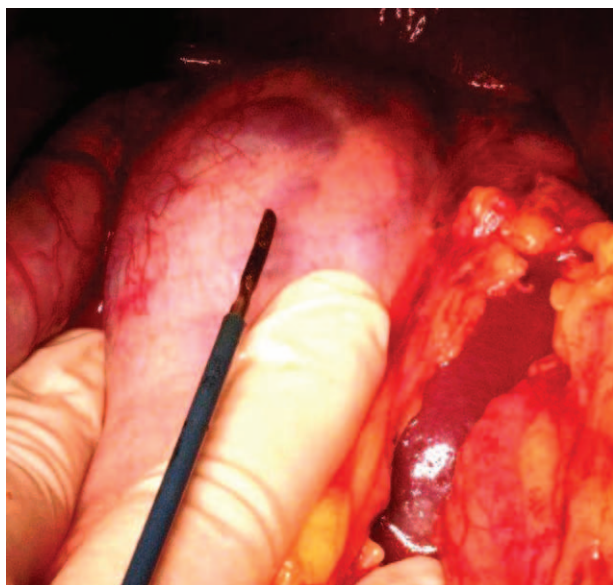


Figure 4. Intraoperative view: large gastric varices.

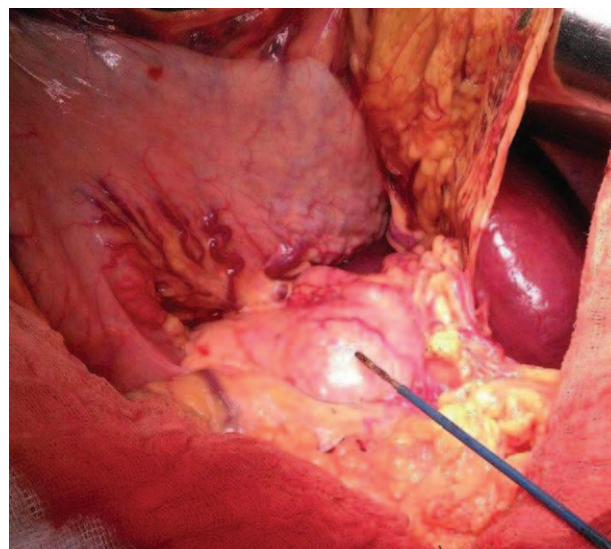


Figure 5. Intraoperative view: pancreatic body cystic tumor, gastric varices, and splenomegaly.

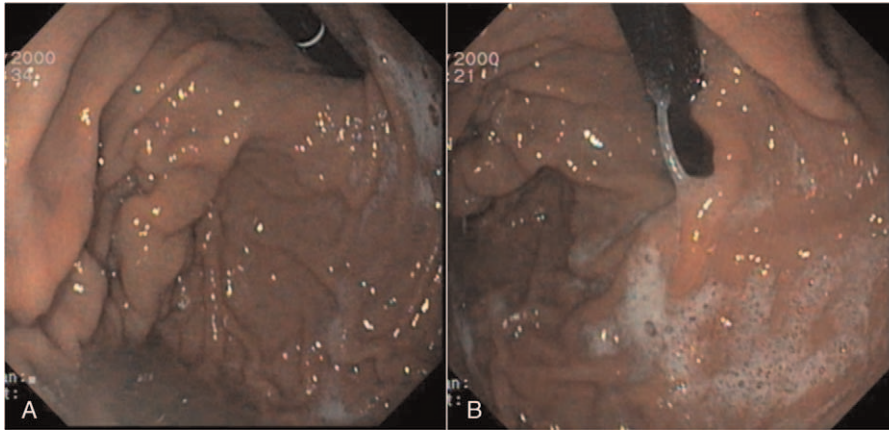


Figure 6. Postoperative follow-up at 1 mo, retroflexed endoscopic view of the gastric fornix: resolution of the gastric varices.

of portal hypertension and subsequent bleeding.^[7–9] Moreover, polycystic disease of the liver and pancreas may determine the development of gastric varices, through compression of the portal vein.^[10]

Management of variceal bleeding in left-side portal hypertension is complex. Endoscopic procedures are largely used to control bleeding, but technical success is related to the size of varices and recurrence of bleeding is frequent.^[11]

Confirming segmental left-sided portal hypertension does not always require surgical intervention in case of benign lesions and the “wait-and-watch” strategy is legitimate in asymptomatic patients.^[11,12] Yet, the presented case required surgical management, due to the presence of 2 high-risk features of the pancreatic cyst and also to the difficulty in performing optimal endoscopic management of bleeding. Splenectomy and surgical resection of the pancreatic lesion can shortly resolve both the varices and the underlying pathology.^[13,14] For cases requiring surgical management, one should always consider that a patient with splenic vein thrombosis may have developed left-side portal hypertension and multiple collateral vessels leading from the spleen to the stomach via the short gastric vessels. Such cases are usually better managed through ligating and dividing the splenic artery early during the procedure.^[14] We opted for an oncologic resection of the lesion, taking into account the inconclusive intraoperative pathology examination of the lymph nodes and cystic wall and the well-known errors due to regressive changes within MCNs.^[15]

MCNs of the pancreas are uncommon (10% of cystic lesions of the pancreas and 1% of pancreatic neoplasms) but with potential to progress to pancreatic adenocarcinoma.^[16–19] There are scarce reports in the literature associating MCNs with left-side portal hypertension.^[20–22] MCNs typically occur in women (F:M ratio 20:1) and are located in the pancreas body and tail (93–95%).^[16,19] The presence of ovarian-type stroma is mandatory to diagnose MCN.^[16] Taking into consideration the highest grade of cytological atypia, different grades of dysplasia are identified: mild, moderate, and severe, the latter considered MCN carcinoma in situ.^[16,19] If invasive malignant disease is confirmed in these lesions, the prognosis is poor with a reported post-surgical 5-year overall survival as low as 17%.^[18] Several epithelial markers have been proposed for mucinous cystic adenocarcinoma (MCA), such as CEA and cytokeratins 7, 8, 18, and 19, as well as the gastric foveolar type markers such as MUC5AC (with MUC1 being present in malignant MCAs). K-

ras, p53, RNF43, and SMAD4 mutations were also identified in dysplastic and invasive MCAs.^[23] Despite the improvement in CT, MRI (magnetic resonance imaging), and EUS (endoscopic ultrasonography) techniques, the ability to differentiate MCN from other types of cystic neoplasms remains poor (25–30%).^[16,19] Therefore, a follow-up protocol of pancreatic cysts is recommended to identify features with high-risk for malignancy such as size >3 cm, solid component, and communication with pancreatic duct and to select patients who benefits from surgical resection.^[12]

4. Conclusion

We have presented a case of MCN particularly by the rapidly developing large size gastric varices as a result of splenic vein thrombosis. Oncological resection of the lesion and splenectomy was the treatment for both MCN and large gastric varices. One has to keep in mind that every pancreatic lesion, even with typical appearance of benignity may produce splenic vein thrombosis and left-side portal hypertension, with life-threatening digestive bleeding; therefore, vascular complications should be included in the follow-up criteria for cystic lesions of the pancreas.

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