



Endocrine Carcinoma of the Pancreatic Tail Exhibiting Gastric Variceal Bleeding

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Nonfunctional endocrine carcinoma of the pancreas is uncommon. Without excess hormone secretion, it is clinically silent until the enlarging or metastatic tumor causes compressive symptoms. Epigastric pain, dyspepsia, jaundice, and abdominal mass are the usual symptoms, whereas upper gastrointestinal (GI) bleeding is rare. Here, we describe the case of a 24-year-old man with the chief complaint of hematemesis. Upper GI panendoscopy revealed isolated gastric varices at the fundus and upper body. Ultrasonography and computed tomography showed a tumor mass at the pancreatic tail causing a splenic vein obstruction, engorged vessels near the fundus of the stomach, and splenomegaly. After distal pancreatectomy and splenectomy, the bleeding did not recur. The final pathologic diagnosis was endocrine carcinoma of the pancreas. Gastric variceal bleeding is a possible manifestation of nonfunctional endocrine carcinoma of the pancreas if the splenic vein is affected by a tumor. In non-cirrhotic patients with isolated gastric variceal bleeding, the differential diagnosis should include pancreatic disorders.

Key words: Isolated gastric varices, pancreatic tumor, hematemesis, endocrine carcinoma

INTRODUCTION

Gastroesophageal variceal bleeding is often secondary to liver cirrhosis. In non-cirrhotic patients, isolated gastric varices at the fundus could result from splenic vein obstruction, which causes venous hypertension and the formation of splenoportal collaterals.¹ As such, pancreatic disorders that induce splenic vein obstruction can present with upper gastrointestinal (GI) bleeding.² Here, we describe a young man who presented with gastric variceal bleeding, and the final diagnosis was endocrine carcinoma of the pancreas. Its cause may be segmental portal hypertension secondary to splenic vein involvement by a pancreatic neoplasm.

CASE REPORT

A 24-year-old Chinese man presented with the chief complaint of hematemesis. Physical examination in the emergency room showed no signs of acute abdomen. A blood

test revealed anemia (hemoglobin, 10.6 mg/dL) and a normal platelet count. Liver function test results were normal, and hepatitis B and C serologies were negative. The serum glucose level was also normal (99 mg/dL). Upper GI endoscopy showed an ulcerative lesion with hemorrhagic stigmata at the greater curvature of the gastric fundus suggestive of bleeding gastric varices. The bleeding stopped temporally after endoscopic hemostasis by epinephrine injection and clipping [Figure 1a]. No esophageal varices were seen [Figure 1b].

Subsequent abdominal ultrasonography showed splenomegaly and a pancreatic tumor, while the contour, size, and echogenicity of the liver were normal. Computed tomography (CT) scans of the abdomen revealed an ill-defined lobulated mass measuring 6.8 cm × 4.4 cm × 4.2 cm at the pancreatic tail with heterogeneous contrast enhancement during a dynamic study. The neoplasm completely encased the splenic vein, within which thrombus formation was detected [Figure 2]. Multiple engorged short gastric vessels were noted at the fundus of the stomach.

One week later, the patient experienced massive hematemesis despite repeat endoscopic hemostasis for which an emergent laparotomy was performed. One 4 × 4 × 3 cm indurated mass [Figure 3] at the pancreatic tail constricted the splenic vein and was considered to be associated with splenomegaly and torturous engorgement of the short gastric and gastroepiploic veins. As such, gastric devascularization, distal pancreatectomy, and splenectomy were performed. Microscopic examination showed a low-grade, well-

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differentiated endocrine carcinoma. The tumor had invaded the peripancreatic fatty tissue and metastasized to the peripancreatic lymph nodes. All of the resection margins were free of tumor invasion. Immunohistochemical staining showed tumor cells positive for chromogranin-A [Figure 4], synaptophysin, and neuron-specific enolase. The final diagnosis was nonfunctional endocrine carcinoma of the pancreas (T2N1M0, stage IIB). The gastric variceal bleeding secondary to splenic vein stenosis by the tumor was confirmed and cured surgically.

The patient's postoperative recovery was uneventful, and he was discharged 14 days after the operation. He underwent adjuvant chemoradiotherapy with gemcitabine and cisplatin without target therapy like sunitinib. No recurrence and no GI bleeding were noted during the 3 years of follow-up at our hospital.

DISCUSSION

Endocrine tumors of the pancreas account for approximately 1-2% of pancreatic tumors. Almost half of

these tumors secrete hormones (insulinoma, gastrinoma, glucagonomas, somatostatinomas, vasoactive intestinal polypeptide-secreting tumors [VIPomas]) and are associated with clinical metabolic or GI disorders.³ Instead, nonfunctional endocrine tumors cause symptoms after the enlarging or metastatic tumor compresses the adjacent organ, which manifests as epigastric pain, anorexia, back pain, and jaundice. The increasing use of cross-sectional imaging has resulted in the identification of tumors in more asymptomatic patients.⁴ However, the vague symptoms may also result in a diagnosis that is delayed until metastasis occurs, which worsens the prognosis.

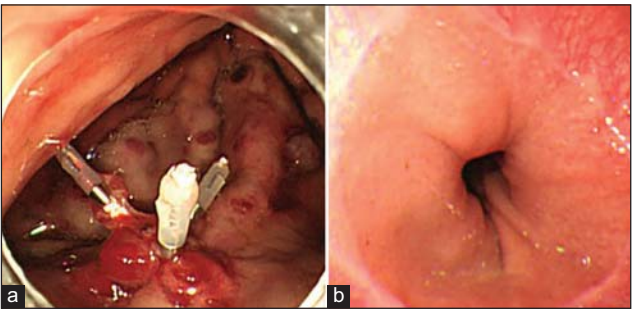


Figure 1. Upper gastrointestinal endoscopy image showing bleeding from varices in the gastric fundus. (a) Emergent hemostasis using epinephrine injection and clipping was performed. (b) EG junction: No esophageal varices or ulcers are seen



Figure 3. One grey-white solid tumor arising from the pancreatic tail (black arrow) with encasement of the splenic vessels (white arrow)

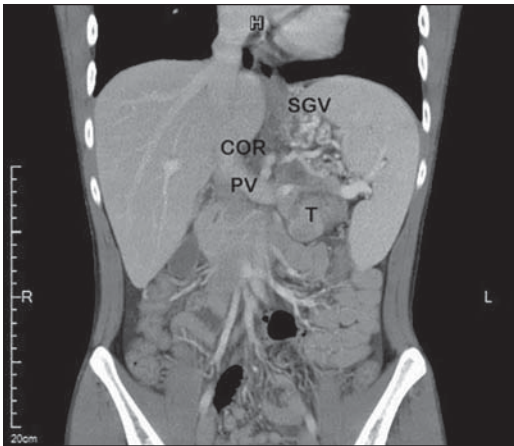


Figure 2. Coronal view of a computed tomography image showing that the tumor (T) occupies the pancreatic tail and involves the splenic vein. Without cirrhosis of the liver or portal vein thrombosis, isolated engorgement of the short gastric veins and coronary vein are remarkable, showing the sequence of splenic vein obstruction by the pancreatic tumor

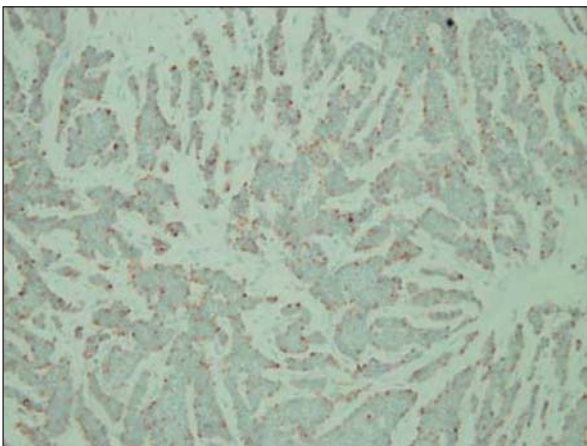


Figure 4. Histopathology image demonstrating the organoid architecture, hyalinized stroma, and coarsely clumped chromatin but few mitotic cells. This finding corresponds to well-differentiated endocrine carcinoma of the pancreas. (Brown spots: Positive immunohistochemical staining for chromogranin, ×200)

Endocrine tumor of the pancreas that presents with acute upper GI bleeding is uncommon. In fact, there are only 22 cases reported in the English literature since 1970. Among them, most were nonfunctional tumor but one was a VIPoma.⁵ Gastric varices were frequently found in the fundus of the stomachs of these patients. The tumors disturbed the splenic venous return in all patients as well, causing segmental portal hypertension (or left-sided [sinistral] portal hypertension). After tumor resection and splenectomy, no recurrence of variceal bleeding was reported.

Isolated gastric varices are defined as gastric varices without esophageal varices.⁶ Portal hypertension secondary to cirrhosis of the liver remains the most common etiology. However, patency of the splenic vein in the presence of pancreatic disorders needs to be checked, especially when fundal varices are involved.¹ Since short gastric veins (SGVs) at the greater curvature of the gastric fundus are confluent with the splenic, gastroepiploic, and coronary veins (CORs), stenosis or occlusion of the splenic vein results in segmental portal hypertension. In this patient, the splenomegaly, engorged SGVs, and CORs noted on CT [Figure 1] could have resulted from the splenic vein obstruction by the neoplasm at the pancreatic tail.

Bleeding gastric varices should be treated with octreotide and balloon tamponade followed with either a transjugular intrahepatic portosystemic shunt or surgery. Endoscopic therapy including variceal sclerotherapy, variceal obturation with cyanoacrylate, and variceal ligation with nylon or snares or rubber bands could be attempted.⁷ Our patient did not receive standard endoscopic therapy during the first acute bleeding episode due to the uncertainty of his endoscopic diagnosis and the fair initial response to the epinephrine injection and clipping. When a tumor of the pancreas is discovered on CT, a subsequent episode of gastric variceal bleeding demands emergent surgery.

Distal pancreatectomy and splenectomy are the standard treatment for neoplasms at the pancreatic tail. Splenectomy is used as curative surgery and releases the segmental portal hypertension since it cuts off venous return from the splenic hilum and a portion of the SGV. In patients with prior bleeding episodes, it is reasonable to perform a splenectomy alone with distal pancreatectomy.⁸

Nonfunctional endocrine tumor of the pancreas is difficult to diagnose owing to its vague symptoms. Upper GI bleeding is a rare but possible manifestation in such patients. In non-cirrhotic patients with isolated gastric variceal bleeding, attention should be paid to segmental portal hypertension related to the pancreatic neoplasm. Gastric varices along the greater curvature of the fundus also require cross-sectional imaging to verify splenic vein patency.

DISCLOSURE

The authors report no conflicts of interest in this work.

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