

## CASE REPORT

# A Patient of Pancreatic Acinar Cell Carcinoma with Dilated Esophagogastric Vessels

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### ABSTRACT

Left portal hypertension and splenic vein occlusion commonly occur with pancreatic tumors, however these signs are rarely observed in patients with acinar cell carcinoma. This report describes a rare left portal hypertension in a patient who presented with a dilated esophagogastric vein upon esophagogastroduodenoscopic examination of a gastric polyp. A contrast-enhanced computed tomography scan revealed a pancreatic tumor, with obstruction of the splenic vein and portal-systemic shunt. The patient was diagnosed with an acinar cell carcinoma of the pancreatic tail. This patient highlights that pancreatic acinar cell carcinoma should be considered as a differential diagnosis in patients with a dilated esophagogastric vein and without signs of liver disease.

### INTRODUCTION

Acinar cell carcinoma of the pancreas is rare, accounting for approximately 1% of pancreatic exocrine tumors [1]. This tumor most commonly occurs in the head or tail of the pancreas of male Caucasians in their seventh decade [2]. The major clinical symptoms of acinar cell carcinoma include weight loss, abdominal pain, nausea, and vomiting [1]. However, the majority of patients are asymptomatic, and the disease is often only discovered at an advanced stage when metastases to the liver or other organs have occurred [1, 3, 4]. The report describes a rare pancreatic acinar cell carcinoma that presented in a patient with dilated esophagogastric vessels upon upper gastrointestinal endoscopy.

### CASE REPORT

The patient was an 84-year-old man, who was referred to our hospital for treatment of an adenoma of the cardia and lesser curvature posterior wall of the stomach that was detected upon upper gastrointestinal endoscopy. The patient had a 20-year history of high blood pressure, megaloblastic anemia that was diagnosed eight years prior, and a 4-year history of asbestosis. The patient reported no alcohol or tobacco use. Initial routine blood tests were normal. Upper gastrointestinal endoscopy of the esophagus and stomach revealed dilated esophagogastric

vessels (**Figure 1**). A mass shadow (32 × 44 mm) at the tail of the pancreas was detected by computed tomography (CT) scan (**Figure 2**). A contrast-enhanced CT scan of the portal vein showed occlusion the splenic vein by the mass at the pancreatic tail, and well-developed collateral veins. Ultrasonography also revealed a low echoic mass in the pancreas tail. Magnetic resonance imaging (MRI) indicated a T1-high, T2-low, and diffusion-high mass. Endoscopic ultrasonography showed low echoic, uniform 40 mm mass with a relatively clear border, indicating there was no cystic portion within the mass. Endoscopic retrograde cholangiopancreatography showed that the pancreatic duct was slightly displaced by the tumor, and cytologic evaluation of the pancreatic juice indicated no malignancy. However, positron emission tomography-CT scan detected a high accumulation (standardized uptake value maximum=13.54) of fluorodeoxyglucose, which indicated that the pancreatic mass was malignant. As the imaging results were also indicative of a neuroendocrine tumor, and the mass was obstructing the splenic vein and causing hyperplasia of the collateral pathways, the decision was made to operate.

A laparoscopic distal pancreatectomy and splenectomy were performed, and pathology of the tumor specimen revealed an acinar architecture (**Figure 3**). The biopsy was immunopositive for trypsin, and a final histologic diagnosis of acinar cell carcinoma was made. There was no vascular invasion and the cause of splenic vein obstruction was determined to be due to a thrombus.

The patient had a liver metastasis five months postoperatively, and ultrasound-guided percutaneous radiofrequency ablation, which was repeated three times. Chemotherapy with S1 was initiated 18 months after the operation. The patient remains alive 35 months postoperatively.

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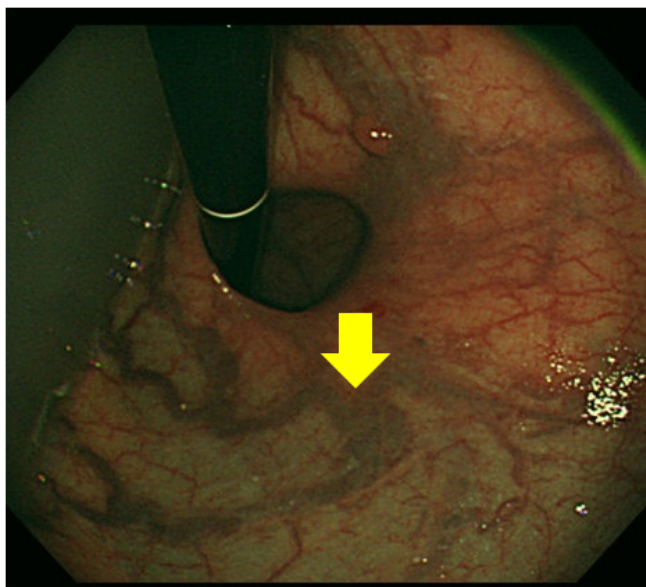
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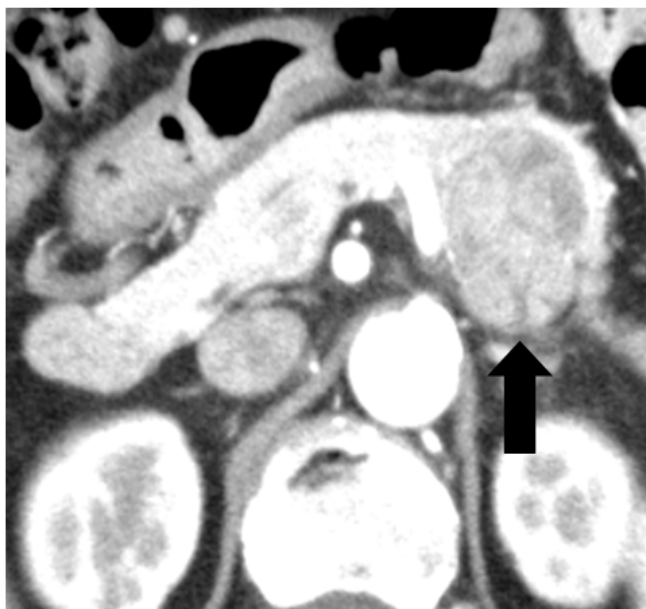
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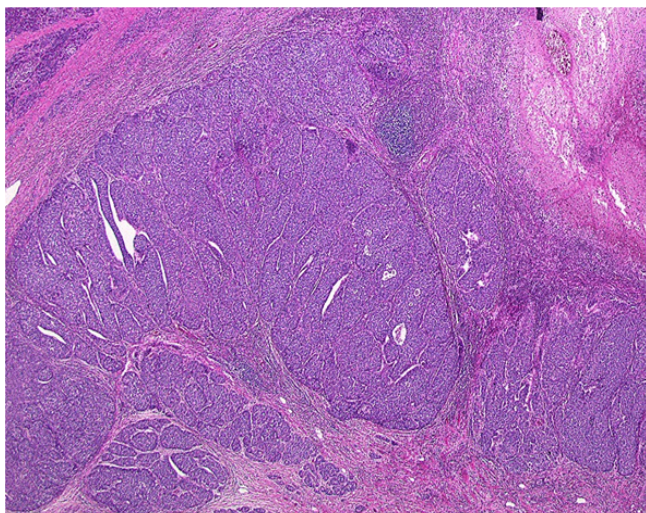
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**Figure 1.** Upper gastrointestinal endoscopy revealed dilated esophagogastric vessels in the wide area of esophagus and stomach.



**Figure 2.** Computed tomography revealed a mass shadow at the tail of the pancreas. The mass (32 × 44 mm) showed a contrast enhancement.



**Figure 3.** Histologic pathology revealed that the tumor had an acinar architecture (hematoxylin and eosin: 500 μm).

## DISCUSSION

The median overall survival for patients diagnosed with acinar cell carcinoma is 19 months, which is longer than reported for ductal adenocarcinoma of the pancreas (9 months) but substantially shorter than for pancreatic neuroendocrine neoplasm (40–60 months) [2]. Once diagnosed, however, median survival in patients with acinar cell carcinoma is extended to 36–41 months following resection [5]. Unfortunately, there is a high recurrence rate (72%) after resection [2], and there are currently no curative treatments. As the majority of patients are detected at an advanced stage, early detection of acinar cell carcinoma is of utmost importance.

Several different imaging modalities have described the appearance of acinar cell carcinoma. On CT scan, the tumor appears as a solitary, heterogeneous, hypodense mass within a well-defined capsule [6, 7]. For the patient described herein, contrast-enhanced CT scan revealed obstruction of splenic vein. However, MRI techniques, such as mangafodipir trisodium-enhanced MRI [5, 8], can also be used to reveal splenic or portal vein thrombosis. Although endoscopic ultrasonography can be used to visualize the pancreas, the features of acinar cell carcinoma have not been clearly defined with this modality [5, 8]. For example, Aql *et al.* [5] reported the appearance of an acinar cell carcinoma as a large mass with both cystic and solid components with endoscopic ultrasonography.

For the patient described herein, the diagnosis of acinar cell carcinoma was confirmed upon pathologic examination. These tumors show a characteristic clear border, with an expansive, tuberous growth pattern comprised of large and small nodules and an acinar structure. Immunohistochemical staining for trypsin and chymotrypsins, digestive exocrine enzymes can also be used [9], as demonstrated in the present patient.

There are only a few similar reports describing pancreatic neuroendocrine tumors. In a case series, pancreatic cancer was found to be the cause of esophagogastric varices, with all patients showing invasion of portal venous systems [10]. The splenic vein was involved only in the patients with pancreatic body and/or tail cancer, similar to the present patient but not to patients with cancer of the pancreatic head. Previous patients have also presented with isolated gastric varices [11–13], as well as a dilated esophagogastric vein, as observed in a patient with a solid pseudopapillary neoplasm [13]. Our patient showed a dilated esophagogastric vein. We thought that some pancreatic tumors, like acinar cell carcinoma, grow expansively and cause dilatation of the esophagogastric vein and not varices because they are less invasive than pancreatic cancer. The expansive growth of the tumor caused a pressing on the splenic vein and slowly formed a thrombus. As a result, it was thought that the similar pattern of growth was likely involved in the esophagogastric vein dilation observed in the present patient.

The detection of acinar cell carcinoma in the present patient occurred as a result of gastrointestinal endoscopy. This method may become useful for future similar diagnoses with continual clarification of esophagus and intragastric vein expansion views.

This patient highlights that pancreatic acinar cell carcinoma should be considered as a differential diagnosis in patients with a dilated esophagogastric vein and without signs of liver disease.

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## Conflicting Interest

The authors had no conflicts of interest

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