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Resected Acinar Cell Carcinoma of the Pancreas with Tumor Thrombus Extending into the Main Portal Vein: Report of a Case

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Abstract: The incidence of acinar cell carcinoma has been reported to be about 1% of all pancreatic neoplasms, and pancreatic cancer combined with tumor growth extending into the portal vein is a rare condition. We herein report a case of acinar cell carcinoma of the pancreas with a tumor thrombus extending into the main portal trunk. Preoperative imaging of the portal vein, consisting of computed tomography (CT), magnetic resonance imaging (MRI), and angiography, revealed an oval shadow defect in the main portal trunk along with an irregular mass in the pancreatic head. At operation, we confirmed a tumor thrombus extending from a tumor in the pancreatic head into the main portal trunk via the pancreatoduodenal veins. A pancreatoduodenectomy combined with partial resection of the portal vein was thus performed under a temporary portal vein shunt from the ileocecal vein to the umbilical vein. Immunohistochemical examination for α_1-antichimotrypsin and electron microscopic examination confirmed the diagnosis of acinar cell carcinoma of the pancreas with a tumor thrombus in the portal vein. Surgical excision combined with portal vein resection may therefore improve the prognosis of selected patients with portal tumor thrombus.

Key Words: acinar cell carcinoma, tumor thrombus, portal vein

Introduction

Recent advances of imaging modalities such as ultrasonography (US), computed tomography (CT), and magnetic resonance imaging (MRI) have greatly contributed to the increased detection of vascular involvement in malignant tumors.[1–5] Tumor thrombus in the portal vein most commonly occurs in advanced hepatocellular carcinoma.[6] Recently, however, it has been reported in association with gastric adenocarcinoma.[7,8] Less frequently, intraportal tumor growth has also been demonstrated in patients with islet cell tumors of the pancreas.[9,10] In pancreatic cancer, although extrinsic obstruction is frequent, tumor growth extending into the portal vein is a rare condition.[11–13] There has only been one previous case report of acinar cell carcinoma accompanying tumor thrombus in the superior mesenteric vein in the Japanese literature.[14] As far as we could ascertain in the English literature, this is the first report of acinar cell carcinoma of the pancreas with a tumor thrombus extending into the main portal vein.

Case Report

A 61-year-old woman presented with symptoms of epigastric pain accompanied by general fatigue and loss of appetite of 1 month's duration. Physical examination showed no remarkable findings. The laboratory data on admission were as follows: serum lactate dehydrogenase (LDH), 612 IU/l (normal value, 227–416); amylase, 170 IU/l (50–158); and lipase, 260 IU/l (0–129). All other laboratory data were within normal limits. Serum carbohydrate antigen 19-9 (CA 19-9), carcinoembryonic antigen (CEA), alpha-fetoprotein (AFP), and other tumor markers (SPAN-1, DUPAN-2, and KMO-1) were in the normal ranges.

Endoscopic examination disclosed an ulcerated tumor in the posterior wall of the superior duodenal flexure. A biopsy of the tumor demonstrated poorly differentiated adenocarcinoma cells mainly located in the submucosal layer of the duodenum. Hypotonic duodenography revealed a tumor of the superior duodenal flexure, but no abnormal finding was noted in the second and third portions of the duodenum. On endoscopic retrograde cholangiopancreatography

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(ERCP), the main pancreatic duct was smooth, and not dilated over its entire course. ERCP also showed a slight dilatation of the upper part of the common bile duct accompanied by narrowing and displacement of its lower half to the left. The ampulla of Vater was intact. Abdominal US revealed a tumor in the pancreatic head and a hypoechoic mass in the main trunk of the portal vein. An abdominal CT scan with contrast enhancement demonstrated a mass lesion in the pancreatic head and a filling defect with a crescent enhancement of the peripherally patent lumen in the portal vein, which thus suggested the presence of a tumor thrombus. No metastatic lesion was detected in any hepatic lobes. The abdominal MRI also showed a mass lesion in the pancreatic head and tumor thrombus in the main portal vein. Celiac arteriography revealed an encasement of the posterior superior pancreatoduodenal artery and anterior superior pancreatoduodenal artery accompanied by hypervascularity in the feeding areas of these arteries. A superior mesenteric arterial portogram showed an egg-shaped shadow defect in the main portal vein, thus indicating the tumor thrombus (Fig. 1).

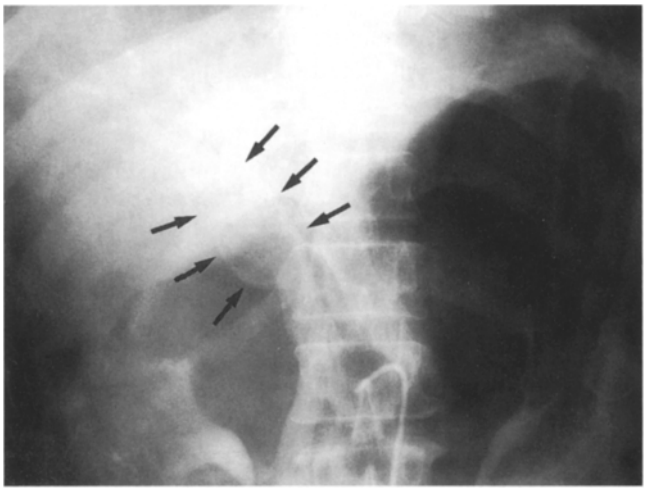

Fig. 1. Venous phase of a superior mesenteric arteriogram shows tumor thrombus (*arrows*) in the main portal vein

At surgery, neither liver metastasis nor peritoneal dissemination was noted. The tumor measured 5.0 × 3.8 cm and was located in the posterior wall of the superior duodenal flexure with good mobility. A floating tumor thrombus extending into the main portal vein via the pancreatoduodenal vein was found during the operative procedures. Except for the above-mentioned site, the portal venous wall was free from carcinoma invasion. A pancreatoduodenectomy accompanied by partial resection and reconstruction of the portal vein was performed with a temporary pump-assisted shunt from a branch of the superior mesenteric vein to the intrahepatic portal vein through the umbilical vein in the hepatic round ligament. The tumor thrombus was removed through a spindle-shaped resection of the portal vein which was reconstructed with a saphenous vein patch. Gastrointestinal reconstruction was performed by Child's procedure.

In the resected specimen, macroscopically, the tumor was located close to the posterior wall of the duodenum. However, the tumor existed below the submucosal layer of the duodenum, and the duodenal mucosa was intact. The tumor had no fibrous capsule and its cut surface showed no areas of cystic degeneration (Fig. 2A). No accessory pancreatic duct could be identified. Fig. 2B shows the tumor thrombus removed from the portal vein, which measured 4.0 × 2.0 cm and was egg-shaped.

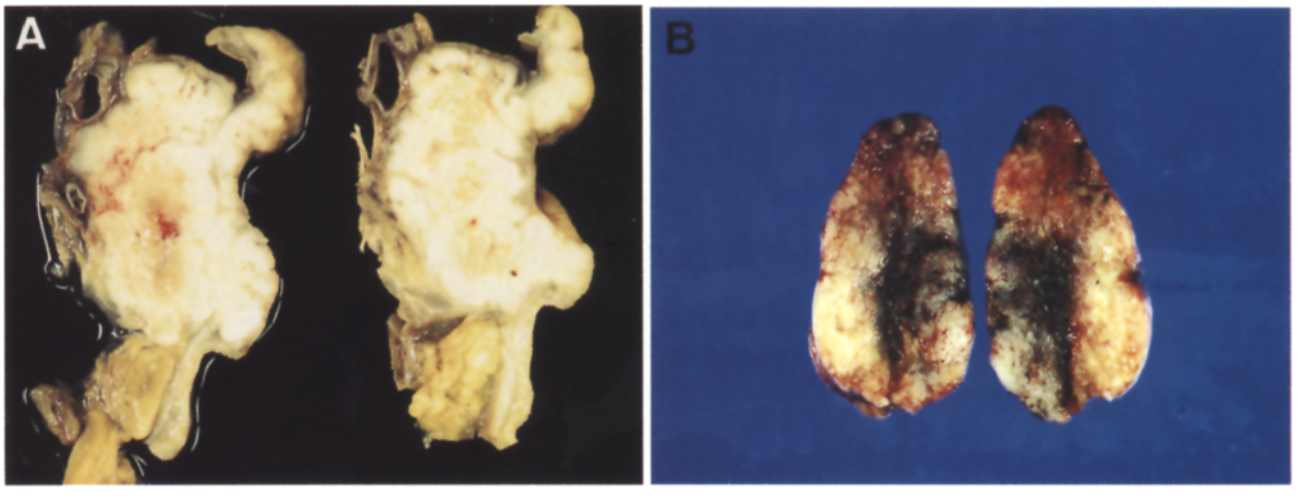

Fig. 2. **A** Macroscopic appearance of the main tumor of the pancreas. Neither a fibrous capsule around the tumor nor cystic degeneration in its cut surface is observed. **B** Tumor thrombus removed from the portal vein

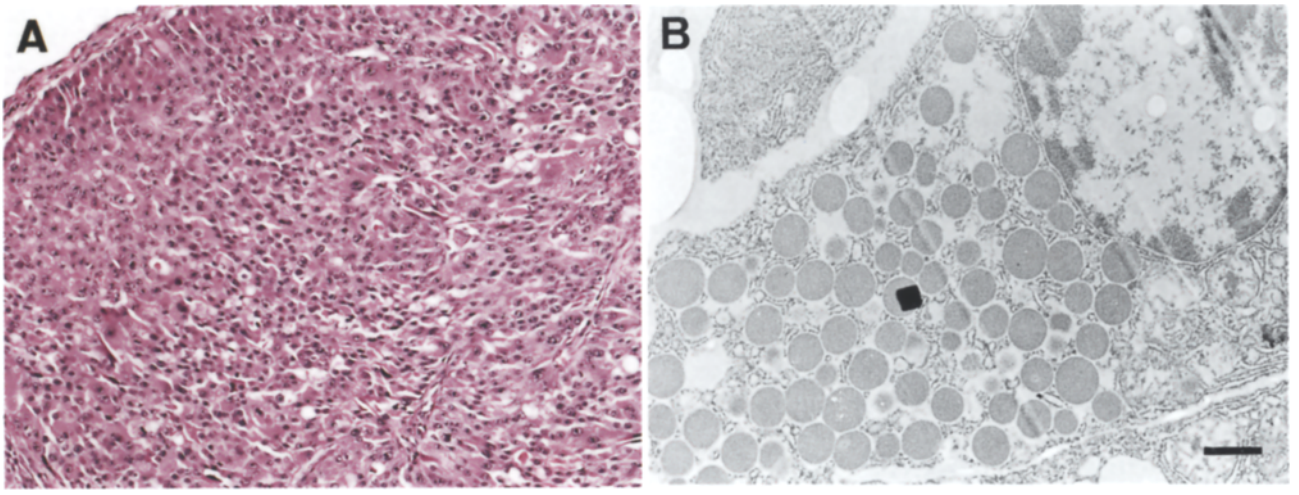

Fig. 3. **A** Light microscopic appearance of the tumor (staining with H&E). Microscopically, it is difficult to differentiate acinar cell carcinoma from undifferentiated islet cell tumor. **B** Electron microscopic appearance of the tumor cell. The tumor cell is observed to contain abundant electron-dense zymogen granules (500–1,000 nm in diameter). Bar: 1,000 nm

Light microscopically, hematoxylin and eosin (H&E) staining of the main tumor (Fig. 3A) revealed atypical cells with pleomorphic nuclei. Mitotic figures were sparse and the cytoplasm was eosinophilic. The amount of stroma was scanty (medullary type) and the extent of venous invasion was severe. Because of these histological findings, it was difficult to differentiate acinar cell carcinoma from undifferentiated islet cell tumor of the pancreas. Therefore, an immunohistochemical examination was carried out; it revealed the tumor cells to be positive for α_1-antichimotrypsin, but negative for both neuron-specific enolase (NSE) and CA 19-9. Furthermore, electron microscopy demonstrated abundant zymogen granules 500–1,000 nm in diameter in the tumor cells (Fig. 3B). Based on these findings, we concluded the diagnosis to be acinar cell carcinoma originating either from the pancreas near the accessory pancreatic duct or the ectopic pancreas. The patient had an uneventful recovery and is still doing well at 15 months after surgery without any sign of recurrence.

Discussion

An obstruction of the portal vein secondary to malignant disease can result from a variety of conditions such as direct invasion, compression, and tumor thrombus. Pancreatic head carcinoma often accompanies a direct invasion of the portal vein, but rarely accompanies portal tumor thrombus. However, as far as we could ascertain in the English literature, acinar cell carcinoma of the pancreas accompanying an intravenous tumor extension into the portal vein has not been previously described. Webb reported 11 cases of acinar cell carcinomas, 9 of which had liver metastases, but there were no cases accompanying tumor thrombus in the portal vein.[15] It is well known that portal tumor thrombus is frequently associated with liver metastases. It has also been reported that gastric cancer with a medullary growth pattern has a high incidence of venous invasion, portal tumor thrombus, and liver metastases.[16] In our case, no intrahepatic metastases were observed, and the tumor thrombus was solitary, which thus indicated that this tumor thrombus most probably developed from the main tumor of the pancreas via the pancreatoduodenal vein. It is thus speculated that the unique mode of tumor extension found in this case may be associated with the peculiar location of the tumor in addition to its medullary growth pattern.

The majority of pancreatic carcinomas are adenocarcinomas of ductal origin. Acinar cell carcinoma is rare and its incidence is only about 1% of all pancreatic neoplasms. A histological diagnosis of acinar cell carcinoma is usually very difficult; therefore, the diagnosis should be based on its distinct histologic pattern and the immunoreactivity of the tumor cells to a number of pancreatic enzymes such as lipase, trypsin, and chymotrypsin. An electron microscopic demonstration of zymogen-like granules in the tumor cells is considered to confirm the diagnosis.[17–19] First of all, in this case, the appearance of solid and cystic tumors (SCT) was negligible based on the gross features of the tumor, because SCT is macroscopically characterized by the fact that the tumor is encapsulated by a well-defined fibrous capsule and the cut surface shows solid areas and cystic degeneration areas due to hemorrhagic necrosis.[20] Secondly, H&E staining did not permit differentiating acinar cell carcinoma from undifferentiated

islet cell tumors; however, an immunohistochemical examination for α_1-antichimotrypsin and an electron microscopic examination did provide strong evidence that this tumor represented acinar cell carcinoma.

Pancreatic head carcinoma accompanied by portal invasion is often unresectable, and the prognosis of such patients is generally poor. However, recent advances in various imaging techniques now allow for the preoperative detection of portal vein thrombus in these patients. In addition, thanks to the incorporation of bypass techniques to prevent splanchnic blood pooling, pancreatoduodenectomy accompanied by portal vein resection can now be safely and easily performed.[21,22] In this case, no liver metastasis was found and the tumor thrombus was solitary. A surgical resection was successfully done. Fifteen months after the surgery, the patient is alive and doing well without any sign of recurrence. Based on the above findings, we believe that surgical excision combined with portal vein resection may therefore improve the prognosis of selected patients with portal tumor thrombus.

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