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Portal Vein Resection for a Portal Vein Thrombus Caused by Nonfunctioning Islet Cell Carcinoma: Report of a Case

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Abstract
We report a case of nonfunctioning islet cell carcinoma of the pancreas causing a tumor thrombus in the portal vein. The patient was a 60-year-old woman whose presenting symptoms were abdominal pain, vomiting, and weight loss. We performed a subtotal pancreatectomy and splenectomy combined with partial resection of the portal vein. Histopathological studies confirmed the diagnosis of nonfunctioning islet cell carcinoma of the pancreas with a tumor thrombus in the portal vein. The patient's postoperative course was uneventful and she is doing well 25 months after the operation.

Key words Nonfunctioning islet cell tumor · Tumor thrombus · Portal vein resection

Introduction

Tumors of the pancreatic islet cells are rare, with an estimated prevalence of less than 1/100000 of the population.[1] They are classified as functioning or nonfunctioning tumors. Functioning tumors cause the typical symptoms of excessive hormone release, whereas nonfunctioning tumors are either found incidentally at the time of surgery or cause symptoms such as abdominal pain, jaundice, or weight loss, depending on their size and location.[2,3] Nonfunctioning islet cells tumors are clinically important because they have a high rate of malignancy, ranging from 60% to 92%.[4,5] We report an unusual case of a portal vein thrombus occurring in association with a nonfunctioning islet cell carcinoma, which was treated by curative surgery.

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Case Report

A 60-year-old woman was admitted to our hospital for investigation of upper abdominal pain, weight loss, nausea, and vomiting. The serum tumor marker and endocrine hormone levels were within the reference ranges (Table 1). A computed tomography (CT) scan of the abdomen showed a 13 × 8 × 7-cm tumoral mass in the body and tail of the pancreas, and a CT image in the venous phase revealed tumor thrombus in the portal vein (Fig. 1). Using contrast-enhanced Doppler ultrasound, we detected a pulsatile flow in the thrombus. Laparotomy revealed a portal vein tumor thrombus extending from the pancreatic malignancy, and we performed a subtotal pancreatectomy with splenectomy and segmental resection of the portal vein, under temporary portal vein occlusion lasting about 25 min. The length of the resected portal vein was 5.6 cm. The pancreatic stump was closed using a single layer of interrupted 3-0 silk and because an end-to-end anastomosis was impossible, a Gore-Tex (Gore & Associates, Newark, DE, USA) vascular graft was needed for reconstruction. Anastomosis of the portal vein was performed with a running suture of 6-0 Prolene (Ethicon, Norderstedt, Germany). The patient had an uneventful postoperative course and was discharged from hospital 14 days after her operation. A follow-up contrast-enhanced Doppler ultrasound was done 12 months later which confirmed patency of the portal vein. She is well without any signs of recurrence 25 months after her operation.

Histopathologic Findings

The partial pancreatectomy specimen consisted of the body and tail of the pancreas, measuring 13 × 10 × 6 cm. A nodular mass, 7.2 cm in greatest dimension, was found in the pancreas, that protruded into the portal vein (Fig. 2). Microscopically, the tumor consisted of a

large amount of fibrovascular stroma and small, relatively uniform cuboidal cells with centrally located hyperchromatic nuclei and eosinophilic cytoplasm. Immunohistochemical staining with keratin and chromogranin was strongly positive in the tumor mass (Fig. 3A,B).

Table 1. Preoperative serum levels of tumor markers and endocrine hormones

Gastrin	155 pg/ml	CEA	1.5 ng/ml
Glucagon	92 pg/ml	CA 19-9	18 U/ml
Somatostatin	11.6 pg/ml	CA 72-4	4.2 U/ml
Insulin	6.4 μU/ml	CA 125	14 U/ml
VIP	10.2 pg/ml		

VIP, vasoactive intestinal polypeptide; CEA, carcinoembryonic antigen; CA, carbohydrate antigen

Discussion

Nonfunctioning islet cell tumors are groups of islet cell tumors that probably do elaborate polypeptides, but are not hyperfunctioning. They account for about 15%–25% of all islet cell tumors and are the third most common islet cell tumor after insulinoma and gastrinoma.[5,6] The age and sex of the patient, and the histologic features and location of the tumors are similar to those associated with functioning islet cell tumors. The most

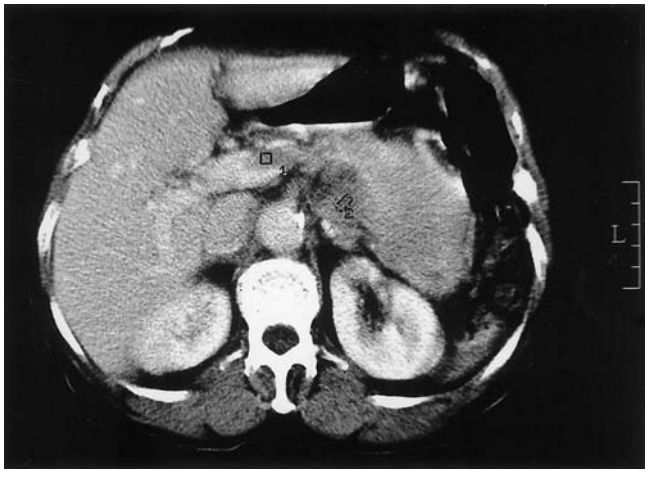

Fig. 1. Abdominal computed tomography (CT) showed a 13 × 8-cm mass in the body and tail of the pancreas. The CT image in the venous phase also showed a portal venous thrombus

Fig. 2. Gross appearance of the nonfunctioning islet cell tumor of the pancreas. There was a large hemorrhagic mass in the corpus and tail of the pancreas, protruding into the portal vein

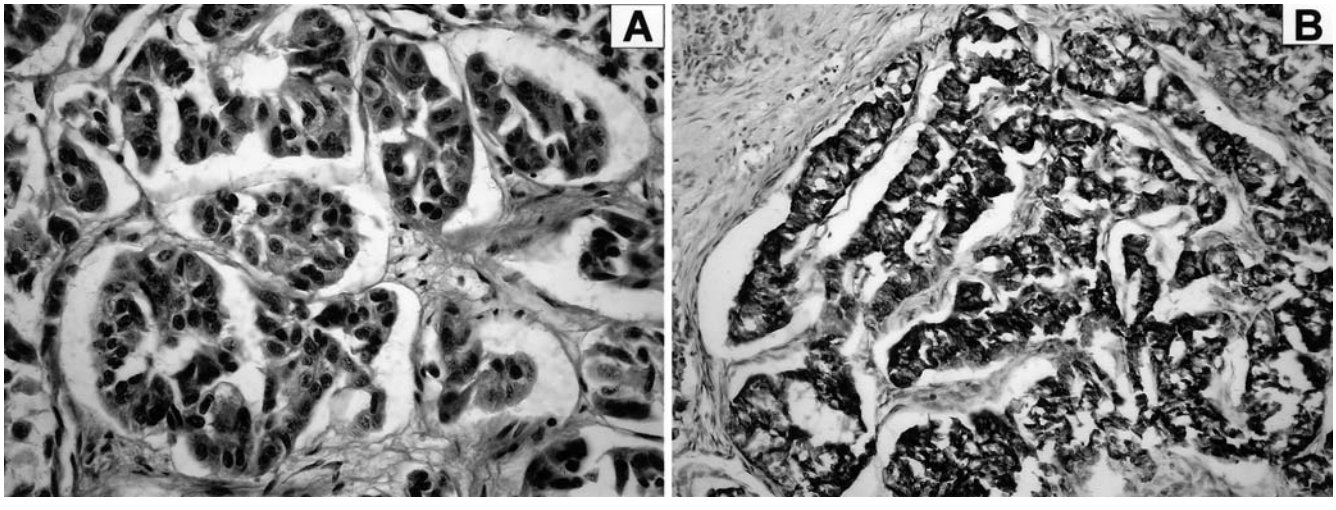

Fig. 3A,B. Microscopic findings. **A** The tumor cells were arranged in a trabecular pattern separated by highly fibrovascular stroma. The tumor cells were cuboidal with hyperchromatic nuclei and eosinophilic cytoplasm (H&E, ×400). **B** Most of the tumor cells showed immunoreactive positivity to chromogranin (×200)

common location of islet cell tumors is the body and tail of the pancreas, correlating with a greater islet concentration in this area.[3] Most nonfunctioning islet cell tumors are malignant and are detected incidentally when they become large enough to compress the adjacent structures.[1,3] Nonfunctioning tumors may also cause obstruction in the biliary tract, which connects the liver to the duodenum and includes the gallbladder, or in the duodenum.[6,7] These tumors may erode and bleed into the stomach or intestines, or both, and may cause an abdominal mass.[8] Whereas venous involvement is a common feature of pancreatic ductal adenocarcinomas, it is a rare feature of nonfunctioning islet cell carcinomas, although cases of venous occlusion, venous encasement, and intraportal tumor growth have been reported.[9] Venous involvement associated with islet cell tumors can be recognized on CT or magnetic resonance images.[10]

Surgical resection is the treatment of choice for pancreatic islet cell tumors, because it can improve the quality of life, prolong survival, and reduce the incidence of metastases, with an acceptable rate of complications and practically zero mortality.[1,2,5,11] In fact, the 5-year survival rate after surgical resection of nonfunctioning islet cell carcinoma of the pancreas was reported to be 77%, whereas that after nonsurgical resection was only 46%.[12] If the tumor is small, curative resection is feasible without a radical approach, but if the tumor is large an aggressive surgical approach is indicated to achieve curative resection without a positive surgical margin.[2] It is well known that palliative resection with a positive margin offers no survival benefit for patients with adenocarcinoma of the pancreas.[13] However, unlike adenocarcinoma of the pancreas, which is frequently considered inoperable in the presence of portal vein invasion, nonfunctioning islet cell carcinoma of the pancreas often warrants aggressive resection in selected cases. If radical tumor resection is combined with portal vein resection, the prognosis is better for patients with nonfunctioning islet cell carcinoma than for those with ductal cell carcinoma.[14] Evans et al.[7] reported that extended resection, including the celiac axis and superior mesenteric-portal venous confluence, may warrant consideration in the subset of patients with locally advanced nonmetastatic islet cell tumors if operative mortality and morbidity is minimized and negative margins are obtained. In conclusion, surgical excision combined with portal vein resection can result in long-term survival with or without residual disease for some patients with islet cell carcinoma and portal tumor thrombus.

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